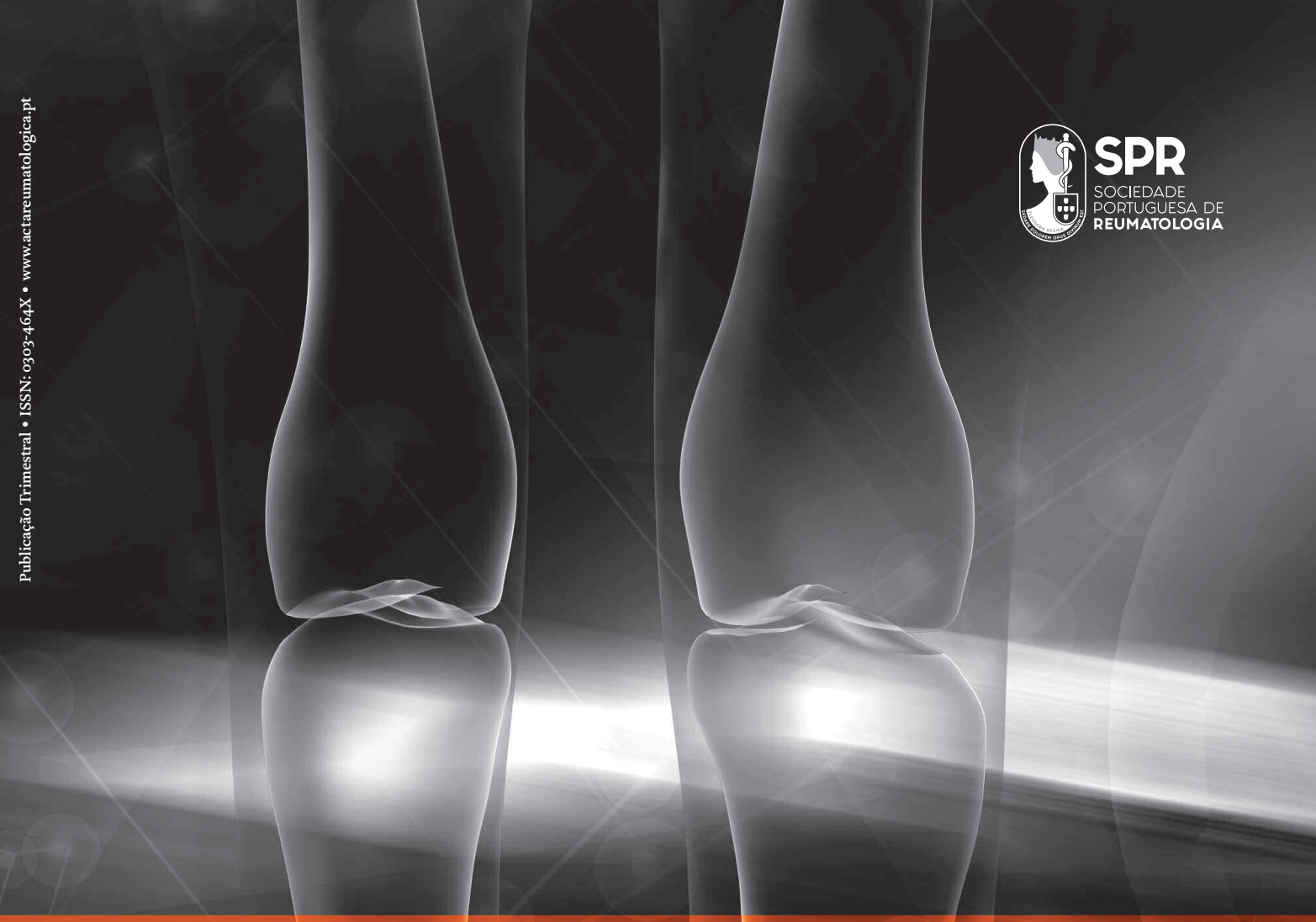
Publicação Trimestral • ISSN: 0303-464X • www.actareumatologica.pt



SPR
SOCIEDADE PORTUGUESA DE REUMATOLOGIA

ACTA REUMATOLÓGICA PORTUGUESA

VOL 46 - Nº 4 | OCTOBER/DECEMBER 2021 (XXIII CPR SPECIAL EDITION)

CPR 2021

XXIII CONGRESSO PORTUGUÊS DE REUMATOLOGIA

ACTA REUMATOLÓGICA PORTUGUESA

ÓRGÃOS SOCIAIS DA SPR BIÉNIO 2020-2022

MESA DA ASSEMBLEIA GERAL

Presidente
Dr. Luís Miguel da Cunha Cristóvão Botelho de Miranda

Secretário
Dr.ª Ana Cristina Van Cauteren Cordeiro

SECRETÁRIO
Dr.ª Inês Sofia Fonseca Cordeiro

DIRECÇÃO

Presidente
Professora Doutora Helena Canhão

Vice-Presidente
Professora Dr.ª Ana Maria Rodrigues

Vice-Presidente
Professora Dr.ª Ana Filipa Mourão

Secretário-Geral
Dr. Tiago Meirinhos

Secretário Adjunto
Dr.ª Anabela Barcelos

Tesoureira
Dr.ª Margarida Oliveira

Vogal Norte
Dr.ª Filipa Teixeira

Vogal Centro
Dr.ª Inês Cunha

Vogal Sul
Dr. Pedro Carvalho

Vogal Ilhas
Dr.ª Lídia Teixeira

Presidente-Eleito
Dr. António Vilar

CONSELHO FISCAL

Presidente
Dr.ª Cátia Duarte

Relator
Dr.ª Cláudia Vaz

Vogal
Dr. Filipe Araújo

A ACTA REUMATOLÓGICA PORTUGUESA É O ÓRGÃO OFICIAL DA SOCIEDADE PORTUGUESA DE REUMATOLOGIA

CPR 2021
XXIII CONGRESSO PORTUGUÊS DE REUMATOLOGIA

SPR SOCIEDADE PORTUGUESA DE REUMATOLOGIA

BOAS-VINDAS

Caros Colegas

Sejam bem-vindos ao XXIII Congresso Português de Reumatologia (CPR).

Neste ano, em que se comemoram os 49 anos da Sociedade Portuguesa de Reumatologia, temos a honra e felicidade de voltar a ter um Congresso Nacional presencial!

Daqui a uns anos vamos olhar para trás e sabemos que a Pandemia que temos estado a viver, mudou para sempre as nossas vidas.

Passámos a utilizar mais meios digitais para comunicar, para ensinar e para aprender. Mas o contacto humano, partilharmos pessoalmente experiências, estarmos uns com os outros e discutir presencialmente os assuntos, é um privilégio que devemos e vamos aproveitar ao máximo.

O CPR decorre de 13 a 16 de Outubro nos Salgados, Albufeira, Algarve.

Começamos com o Curso Pré-Congresso Reuma.pt, seguido da Cerimónia de Abertura e atribuição dos Reumeritus. Temos depois 3 dias repletos de palestras, mesas redondas e simpósios satélites, que abordam os temas mais relevantes e atuais da Reumatologia. As comunicações das mesas redondas são complementadas por apresentações orais dos trabalhos selecionados. Outros trabalhos submetidos serão apresentados sob a forma de poster.

Para que este CPR fosse uma realidade, a direção da SPR contou com a colaboração de muitos colegas que constituem a Comissão Científica, os Júris dos Abstracts, Júris dos melhores posters, comunicações orais e imagens, palestrantes e moderadores. A todos eles agradecemos a dedicação, excelência e generosidade.

A indústria farmacêutica, como habitualmente, foi imprescindível e entusiasta no seu apoio ao CPR.

Esperamos que este CPR do reencontro, seja um momento de excelência científica, aprendizagem e partilha que todos desejamos.

Queremos também reiterar, a bem dos doentes e da população em geral, a necessidade de existir em Portugal um Programa Prioritário para a Saúde Músculo esquelética, que congregue as especialidades de Medicina Geral e Familiar, Reumatologia, Ortopedia, Medicina Física e Reabilitação, outras especialidades associadas, associações de outros profissionais de saúde que trabalham com doentes do foro músculo-esquelético e associações de doentes.

Todos juntos somos melhores.

Desejamos a todos um Excelente e Muito Participado CPR!

Muito obrigada!!

Helena Canhao
Presidente da Sociedade Portuguesa de Reumatologia

Tiago Meirinhos
Secretário Geral da Sociedade Portuguesa de Reumatologia

CPR 2021
XXIII CONGRESSO PORTUGUÊS DE REUMATOLOGIA

SPR SOCIEDADE PORTUGUESA DE REUMATOLOGIA

COMISSÃO CIENTÍFICA

Presidente
Helena Canhão

Anabela Barcelos
Augusto Faustino
Carmo Afonso
Daniela Faria
Fernando Pimentel-Santos
Helena Santos
Jaime Branco
João Eurico Fonseca
José António Pereira da Silva
José Costa
Lídia Teixeira
Lúcia Costa
Maria José Santos
Pedro Carvalho
Raquel Marques
Sandra Falcão
Tânia Santiago
Tiago Meirinhos

COMISSÃO ORGANIZADORA

Presidente
Tiago Meirinhos

Ana Filipa Mourão
Ana Maria Rodrigues
Anabela Barcelos
António Vilar
Filipa Teixeira
Inês Cunha
Lídia Teixeira
Margarida Oliveira
Pedro Carvalho

JÚRIS

JÚRI CASO CLÍNICO
Paulo Monteiro
Margarida Cruz
Joaquim Pereira

JÚRI IMAGEM
Fernando Saraiva
Miguel Matias Sousa
Inês Silva

JÚRI COMUNICAÇÃO ORAL
Augusto Faustino
Cátia Duarte
Filipe Araújo

JÚRI TRABALHO ORIGINAL
Maria João Salvador
Eugénia Simões
Cláudia Vaz

Programa

DIA 13 OUTUBRO (4ª FEIRA)

CURSO REUMA.PT

08:20-08:30h **Abertura**
Presidente da SPR: Helena Canhão
Provedor dos internos: João Eurico Fonseca
Representante dos Internos: Diogo Fonseca

08:30-08:50h **Reuma.pt: 13 anos de evolução**
João Eurico Fonseca

08:50-09:10h **Porquê usar o Reuma.pt?**
Maria José Santos

09:10-09:30h **Como navegar no Reuma.pt (dicas práticas)**
Helena Santos

09:30-09:50h **Projetos de iniciativa do investigador no Reuma.pt - Documentos e prazos**
Diana Carmona Fernandes

09:50-11:00h **MÓDULO PRÁTICO I: Introdução de dados no Reuma.pt – Casos práticos**
- **AR com RTX (2 consultas)**
- **SpA com secucinumab (2 consultas)**
- **OA**

Tutores: Ana Maria Rodrigues, Diana Carmona Fernandes, Fernando Martins, Vítor Chão

11:00-11:15h *Coffee-break*

11:15-11:45h **MÓDULO PRÁTICO I: Introdução de dados no Reuma.Pt - Discussão**
Fernando Martins

11:45-13:15h **MÓDULO PRÁTICO II: Desenvolver um projeto: Desenvolvimento de uma pergunta de investigação e protocolo de estudo com dados Reuma.pt (Trabalho de Grupo)**
Tutores: Ana Maria Rodrigues, Diana Carmona Fernandes, Fernando Martins, Vítor Chão

13:15-13:45h Almoço

13:45-14:15h **SIMPÓSIO JANSSEN**
Algoritmo de investigação na Hipertensão pulmonar - papel do reumatologista
Moderador: Catarina Resende
Palestrante: Tânia Santiago

14:15-15:30h **MÓDULO PRÁTICO II: Desenvolver um projeto (Cont.): Apresentação de uma pergunta de investigação e protocolo de estudo (5 slides com introdução, pergunta de investigação, metodologia, resultados esperados)**
Tutores: Ana Maria Rodrigues, Diana Carmona Fernandes, Fernando Martins, Vítor Chão

15:30-15:45h **ENCERRAMENTO DO CURSO REUMA.PT - Take-home Message**
Coordenadora Reuma.pt: Ana Maria Rodrigues
Presidente da SPR: Helena Canhão
Presidente do Colégio da Especialidade: Augusto Faustino

15:45-16:00h *Coffee-break*

16:00-17:30h **SESSÃO DE ABERTURA E ENTREGA PRÉMIO REUMÉRITUS**
- **Reumatologia e a revolução digital: o que perdemos e o que ganhamos**
  Luís Cunha Miranda

- **A Covid e os doentes reumáticos: onde estamos?**
  Pedro Machado

17:30-18:30h **SIMPÓSIO PFIZER**
The added value of Tofacitinib in Rheumatology
Chair: José António Pereira da Silva

17:30h **Welcome and introduction**
17:35h **Tofacitinib in current treatment strategies –** João Eurico da Fonseca
18:00h **Tofacitinib - Real-world evidence and clinical experience –** Axel Finckh
18:20h **Panel discussion**
18:30h **Meeting Close**

18:30-20:00h **SESSÃO I – DOENÇAS REUMÁTICAS EM IDADE PEDIÁTRICA**
Moderadores: Margarida Guedes; Raquel Marques

- **Seguimento e manutenção de cuidados nas Doenças Reumáticas em idade pediátrica em tempo de pandemia**
  Filipa Oliveira Ramos
- **Doenças Reumáticas em idade pediátrica: o que aprendemos e o que esquecemos nos últimos 20 anos**
  José Melo Gomes
- **COMUNICAÇÕES ORAIS**
  - **Health-related quality of life and function in adults with Juvenile Idiopathic Arthritis – comparison with adult-onset Rheumatic Diseases (92)**
    Filipa Oliveira Ramos
  - **Clinical Course and Risk Factors for Severe/Critical COVID-19 in Patients with Rheumatic Diseases – a Multicenter, Nationwide Study (125)**
    Sofia Barreira

DIA 14 OUTUBRO (QUINTA-FEIRA)

08:00-09:00h Reunião Comissão Nacional Reuma.pt
09:00-10:30h **SESSÃO II – OSTEOPOROSE E SARCOPENIA**
Moderadores: Lúcia Costa, José Romeu

- **Confinamento e Sarcopenia: preço a pagar?**
  Fernando Pimentel-Santos
- **Impacto global da osteoporose: Conseguimos mudar o futuro?**
  José António Pereira da Silva
- **COMUNICAÇÕES ORAIS**
  - **How are we treating patients with fragility fractures? - Assessment of Patients Observed at Emergency Department for Fractures (112)**
    Catarina Dantas Soares
  - **A Fracture Liaison Service implementation aer two years: a retrospective cohort study (264)**
    Susana P. Silva

10:30-11:30h *Coffee-break*
11:30-12:30h **SIMPÓSIO AMGEN**
2 Visões, 1 Outcome: Fraturas de Fragilidade
Moderador: Jaime Branco
Palestrantes: Anabela Barcelos, Céu Mateus

12:30-14:00h Almoço

14:00-15:00h **SIMPÓSIO ABBVIE**

RA 2021 - JAKi for the treatment of Rheumatoid Arthritis

- **Optimizing patient care and clinical outcomes**
- **Growing evidence on the safety profile**

Moderador: Fernando Pimentel-Santos

Palestrantes: José Miguel Bernardes, João Eurico Fonseca

15:00-16:30h **SESSÃO III – NOVOS HORIZONTES NAS ESPONDILARTRITES AXIAIS**

Moderadores: Jaime Branco; Pedro David Carvalho

- **Diferenças de género nas Espondilartrites: impacto clínico e profissional**
  Helena Santos
- **Treat-to-target nas espondilartrites: realidade ou utopia**
  Alexandre Sepriano
- **COMUNICAÇÕES ORAIS**
  - **Molecular profiling of radiographic axial spondyloarthritis patients reveals an association between innate and adaptive cell populations and therapeutic response to adalimumab (122)**
    Rita Pinheiro Torres
  - **Efectiveness of a biologic tapering protocol for rheumatoid arthritis, psoriatic arthritis and axial spondyloarthritis (258)**
    Pedro Ávila Ribeiro

16:30-17:30h *Cofee-break*

17:30-18:30h **SIMPÓSIO JANSSEN**

Guselkumab – Redefinir o tratamento da Doença Psoriática

- **Guselkumag: estaremos mais próximos da remissão sustentada?**
  Elsa Vieira Sousa
- **Experiência clínica com Guselkumab**
  - **Perspetiva do Dermatologista**
    Paulo Ferreira
  - **Perspetiva do Reumatologista**
    Anabela Barcelos

18:30-19:30h **SIMPÓSIO BIOGEN**

As oportunidades e Desafios dos Biossimilares numa era em que a redução de custos é imperiosa

Moderador: José António Pereira da Silva

- **Construir o futuro do SNS: O papel fundamental da sustentabilidade**
  João Pereira
- **O paradigma dos biossimilares em Portugal**
  - **Quais as oportunidades presentes e futuras?**
    José António Pereira da Silva
  - **Quais os desafios enfrentados atualmente?**
    Tiago Meirinhos
- **Q & A**

DIA 15 DE OUTUBRO (SEXTA-FEIRA)

09:00-10:30h **SESSÃO IV – ARTRITE REUMATÓIDE**
Moderadores: Miguel Bernardes, José Costa
- **Integração de cuidados na AR: parceria APPSReuma – The boomerang Effect apresentação dinâmica com 4 preletores (Reuma, Enfermagem, MGF, Doentes)**
  João Eurico Fonseca, Ricardo Ferreira, Elsa Frazão Mateus, José Carlos Marinho
- **COMUNICAÇÕES ORAIS**
  - **Rheumatic Disease Comorbidity Index as a predictor of clinical response to the first biological agent in Portuguese patients with rheumatoid arthritis: a multicenter cohort study (42)**
    Salomé Garcia
  - **Determinants of quality of life in rheumatoid arthritis and spondyloarthritis: how do they interact from a hierarchical perspective? (86)**
    Pedro David Carvalho

10:30-11:30h *Coffee-break*

11:30-12:30h **SIMPÓSIO MSD**
Persisting for the Future

11:30h **Abertura**
Paula Martins de Jesus

11:40h **Persisting for the future – Confiança através de experiência de vida real**
Filipe Araújo

12:20h Conclusões & Encerramento

12:30-14:00h Almoço

14:00-15:00h **SIMPÓSIO NOVARTIS**
The uniqueness of the psoriatic arthritis patient
- **Opening**
- **Every psoriatic arthritis patient is unique**
  Philip Mease
- **The impact of IL17A inhibition in a unique patient**
  Patrícia Nero
- **Clinical case presentation with live pull**
- **Q&A**
- **Wrap-up**

15:00-16:30h **SESSÃO V – ENVOLVIMENTO EXTRA-ARTICULAR NA ARTRITE PSORIÁTICA: O QUE ESTAMOS A PERDER?**
Moderadores: Sandra Falcão; Patrícia Pinto
- **Manifestações extra-articulares na Artrite Psoriática: fisiopatologia e cuidados a ter na prática clínica**
  Maria José Santos
- **Sexualidade e artrite psoriática**
  Manuel Esteves
- **COMUNICAÇÕES ORAIS**
  - **Biologic agents for rheumatic diseases in the outbreak of COVID-19: friend or foe? (4)**
    Cristiana Sieiro Santos
  - **Antibody response aer SARS-COV-2 infection in patients with rheumatic diseases: a multicenter, nationwide study (132)**
    Rita C. Machado

16:30-17:30h *Cofee break*
17:30-18:30h **SIMPÓSIO LILLY**
Olumiant – uma solução real na Artrite Reumatoide
O conhecimento acumulado que sustenta a utilização.
Moderador: Augusto Faustino
- **Baricitinib – Update de Eficácia e Segurança**
  João Eurico Fonseca
- **Baricitinib na Vida Real – RWE na Europa**
  Maria José Santos

20:30-22:00h Jantar do Congresso
Oferta recém especialistas e novos internos
Lançamento do livro da APPSReuma
Apresentação novos consultores

DIA 16 DE OUTUBRO (SÁBADO)

09:00-10:30h **SESSÃO VI – LÚPUS ERITEMATOSO SISTÉMICO E OUTRAS DOENÇAS IMUNOMEDIADAS SISTÉMICAS – Sessão patrocinada GSK**
Moderadores: Tânia Santiago, Ana Cordeiro
- **Hipertensão pulmonar na Esclerose sistémica**
  Mário Santos
- **SLE-DAS: Novo score de atividade "Made in Portugal"**
  Diogo Jesus
- **COMUNICAÇÕES ORAIS**
  - **Predictors of flare in SLE patients attaining Lupus Low Disease Activity State: A real-life cohort study of 292 patients with 36-month follow-up (106)**
    Rita Novais Cunha
  - **Prevalence and clinical associations of dierent autoantibodies in the Reuma.pt systemic sclerosis cohort: is it all really set in stone? (84)**
    Eduardo Dourado

10:30-11:00h *Coffee-break*
11:00-12:00h Assembleia Geral da SPR
12:00-13:00h **ENCERRAMENTO E ENTREGA DE PRÉMIOS**
13:00-14:30h Almoço

VOL 46 • Nº 4 • OCTOBER/DECEMBER 2021 (SUPLEMENTO)

SUMÁRIO/CONTENTS

VOL 46 • Nº 4 • OCTOBER/DECEMBER 2021 (SUPLEMENTO)

SUMÁRIO/CONTENTS

POSTERS

TRABALHO ORIGINAL (COM APRESENTAÇÃO ORAL)

VOL 46 • Nº 4 • OCTOBER/DECEMBER 2021 (SUPLEMENTO)

SUMÁRIO/CONTENTS

TRABALHO ORIGINAL (COM EXPOSIÇÃO)

VOL 46 • Nº 4 • OCTOBER/DECEMBER 2021 (SUPLEMENTO)

SUMÁRIO/CONTENTS

VOL 46 • Nº 4 • OCTOBER/DECEMBER 2021 (SUPLEMENTO)

SUMÁRIO/CONTENTS

VOL 46 • Nº 4 • OCTOBER/DECEMBER 2021 (SUPLEMENTO)

SUMÁRIO/CONTENTS

POSTERS

CASOS CLÍNICOS (COM APRESENTAÇÃO ORAL)

VOL 46 • Nº 4 • OCTOBER/DECEMBER 2021 (SUPLEMENTO)

SUMÁRIO/CONTENTS

CASOS CLÍNICOS (COM EXPOSIÇÃO)

VOL 46 • Nº 4 • OCTOBER/DECEMBER 2021 (SUPLEMENTO)

SUMÁRIO/CONTENTS

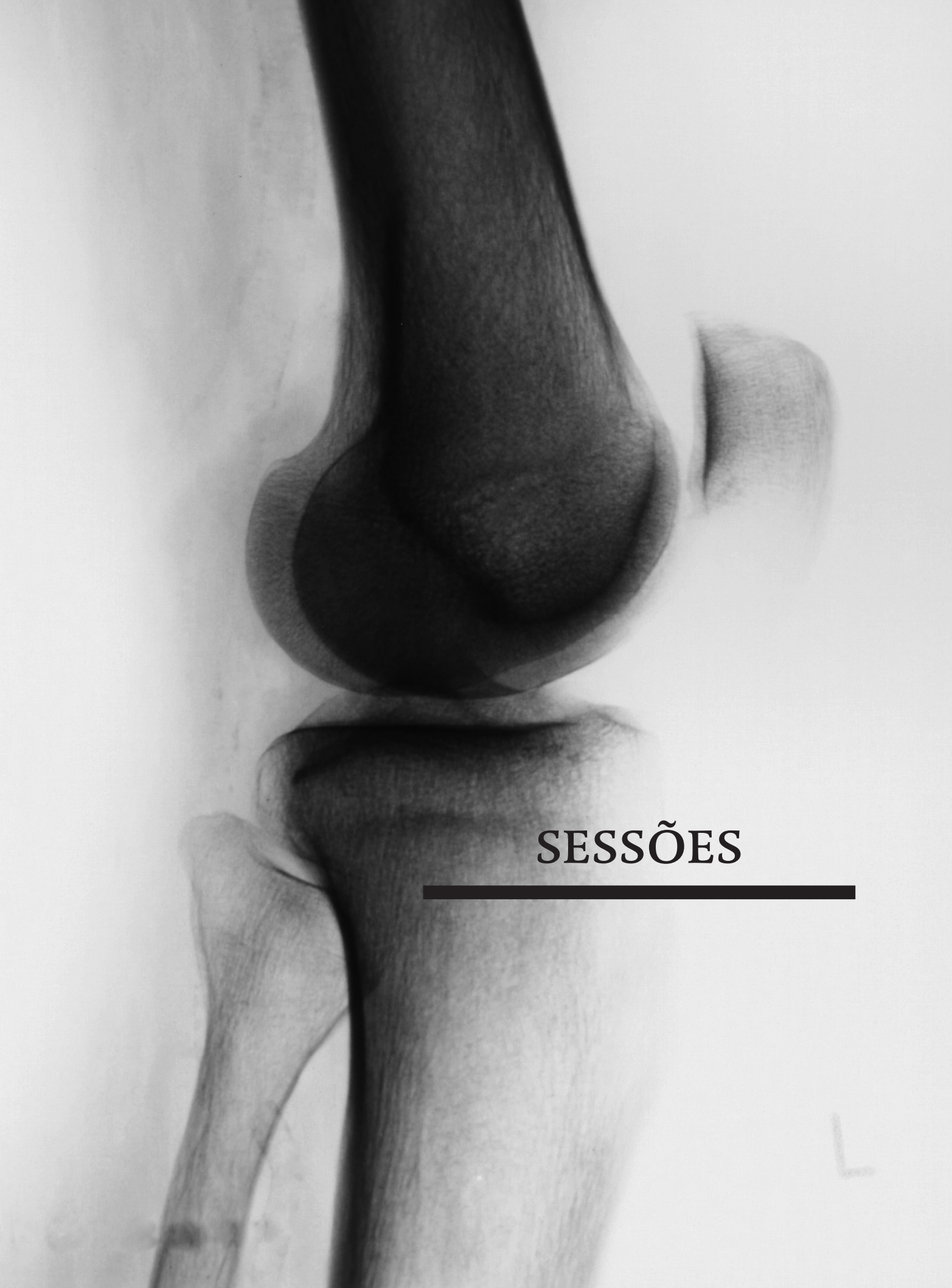
VOL 46 • Nº 4 • OCTOBER/DECEMBER 2021 (SUPLEMENTO)

SUMÁRIO/CONTENTS

IMAGENS

VOL 46 • Nº 4 • OCTOBER/DECEMBER 2021 (SUPLEMENTO)

SUMÁRIO/CONTENTS



SESSÕES

Sessões

ACTA REUMATOL PORT. 2021:46:??-?? (SUP)

SESSÃO I - DOENÇAS REUMÁTICAS EM IDADE PEDIÁTRICA

SEGUIMENTO E MANUTENÇÃO DE CUIDADOS NAS DOENÇAS REUMÁTICAS EM IDADE PEDIÁTRICA EM TEMPO DE PANDEMIA

Filipa Ramos[1]

[1] Serviço de Reumatologia – Centro Hospitalar Lisboa Norte – Hospital de Santa Maria, EPE

A pandemia por COVID-19 trouxe muitos desafios para os sistemas de saúde em geral e em particular para os cuidados dos doentes com doenças crónicas, em que os doentes em idade pediátrica não foram exceção. Houve uma adaptação estrutural importante dos serviços no sentido de reorganizar abordagens dos cuidados dos doentes em várias dimensões, tentando nunca descurar os cuidados e a comunicação entre o doente e as equipas que o seguem. As consultas não presenciais tiveram um papel importante sempre que não foram possíveis presencialmente durante os períodos mais restritivos de confinamento ou para maior comodidade e segurança dos doentes e das suas famílias. Num tempo de incertezas e de mudanças constantes muitas outras alterações tiveram que ser instituídas, de adaptação às necessidades impostas pela pandemia, mas sem nunca comprometer os cuidados de excelência e as boas práticas clínicas no seguimento das crianças e jovens com doenças reumáticas.

DOENÇAS REUMÁTICAS EM IDADE PEDIÁTRICA: O QUE APRENDEMOS E O QUE ESQUECEMOS NOS ÚLTIMOS 25 ANOS

José António de Melo Gomes[1]

[1] Médico Reumatologista e Reumatologista Pediátrico. Responsável pela Consulta de Reumatologia Infantil do Adolescente e do Adulto Jovem do Instituto Português de Reumatologia, Lisboa

A Reumatologia Pediátrica existe em Portugal há cerca de 1 ano, mas alguns de nós têm vindo a dedicar-se a este ramo da Medicina clínica desde há mais de 40 anos. Muito se aprende e, felizmente às vezes, muito se esquece ao longo de mais de 2 décadas de trabalho clínico, mas tal tem sempre aspetos positivos – aprendemos pelos erros e melhoramos quando deixamos de cometer os mesmos erros - e negativos – quem não tem a experiência de erros prévios e desconhece o passado histórico (da sociedade ou da Medicina) pode sempre cair em erros evitáveis.

Nos últimos 25 anos muita coisa se alterou na avaliação, classificação e tratamento das principais doenças reumáticas juvenis. Aprendemos a levar em consideração, duma forma estruturada e sistematizada, a opinião dos pais e dos doentes quanto às alternativas terapêuticas e quanto à avaliação da eficácia das terapêuticas efetuadas – e estes aspetos foram integrados na clínica diária e na avaliação da eficácia e dos efeitos adversos dos novos fármacos que foram surgindo. Aprendemos também a avaliar a atividade clínica das doenças de uma forma mais objetiva e uniformizada, o que teve uma repercussão direta na eficácia dos tratamentos e no reconhecimento mais precoce de eventuais efeitos adversos ou ausências de eficácia, permitindo também a comparação de tratamentos feitos com os mesmos fármacos em países distintos. Aprendemos a criar bases de dados nacionais para as doenças reumáticas mais graves, o que constituiu um marcado avanço no reconhecimento precoce de iatrogenia rara dos fármacos utilizados.

Esquecemos, por vezes, que as doenças reumáticas neste grupo etário exigem também capacidades reforçadas de jogo (crianças mais pequenas) e de diálogo empático (pais e adolescentes adultos jovens), pois o tratamento só é eficaz se for feito e a colaboração dos doentes/família é indispensável para que tal aconteça. Esquecemos que a amiloidose secundária existia nestas crianças e que há 30 anos estávamos a trata-las com muita paciência e empatia, mas com fármacos muito pouco eficazes ou com efeitos adversos muito graves.

SESSÃO II - OSTEOPOROSE E SARCOPENIA

CONFINAMENTO E SARCOPENIA: PREÇO A PAGAR?

Fernando Pimentel-Santos[1]

[1] Serviço de Reumatologia – Centro Hospitalar de Lisboa Ocidental, EPE, Hospital Egas Moniz EPE

A pandemia COVID-19 para além do maior risco de morte foi também responsável por quadros de sarcopenia aguda, em particular nos adultos mais velhos e nos indivíduos com doenças crónicas. A sarcopenia aguda é um quadro pouco conhecido traduzido por insuficiência muscular, ou seja, por declínios na função e / ou quantidade muscular, que se instala em menos de 6 meses, habitualmente associado a uma situação de stress.

Nesta comunicação, serão revistos os mecanismos fisiopatológicos subjacentes (nomeadamente o papel da enzima conversora de angiotensina 2- receptor para o coronavírus e que se expressa no músculo esquelético, associado aos efeitos do repouso, ventilação e modificação de hábitos alimentares) e discutidas as consequências a longo prazo da sarcopenia aguda, no contexto da infeção a COVID-19. Pretende-se alertar para a importância do diagnóstico e para a necessidade de implementação na prática clínica, de medidas que visem manter/desenvolver a força muscular, quantidade e desempenho físico, incluindo exercício físico, reforço proteico e suplementação vitamínica. Adicionalmente alertar para a avaliação do risco/benefício de cada intervenção terapêutica (corticoides, por exemplo) e promover a abordagem multidisciplinar.

Este tipo de estratégias, podem servir como medidas preventivas úteis para reduzir a probabilidade de sarcopenia em geral e no caso de futuros períodos de isolamento bem como das suas consequências em termos individuais e nos sistemas de saúde.

IMPACTO GLOBAL DA OSTEOPOROSE. CONSEGUIMOS MUDAR O FUTURO?

José António Pereira da Silva[1]

[1] Serviço de Reumatologia – Hospital da Universidade de Coimbra

A resposta concreta à pergunta colocada depende de tantas variáveis e de tantos protagonistas que dificilmente pode ser dada com um razoável grau de confiança. A humildade impõe-se ainda mais quando se pede uma perspetiva do impacto global da osteoporose, o que ultrapassa largamente o âmbito em que temos trabalhado e refletido.

Certo é que, se não fizermos mais do que temos feito, o futuro se encarregará de mudar a realidade que atualmente vivemos para uma outra ainda mais pesada em termos sociais e económicos. Basta ter em conta o envelhecimento progressivo da população esperado para os próximos anos. Certo é, por outro lado, que dispomos de meios suficientes para alterarmos de forma positiva essa evolução previsível: temos formas acessíveis de estimar o risco de fratura e de decidir quem deve ser tratado, dispomos de meios eficazes e acessíveis de tratamento, temos, no nosso país como em muitos outros, estruturas de saúde suficientemente organizadas para abordar com eficácia este problema.

Parece faltar apenas que nos mobilizemos, a nós e a esses recursos, para estruturar e efetivar as estratégias mais adequadas. Existem muitas iniciativas nacionais internacionais apelando a essa mobilização. Enquanto isso, o volume de prescrições de medicamentos anti-osteoporóticos diminui progressivamente entre nós...

SESSÃO III – NOVOS HORIZONTES NAS ESPONDILARTRITES AXIAIS

DIFERENÇAS DE GÉNERO NAS ESPONDILARTRITES: IMPACTO CLÍNICO E PROFISSIONAL

Helena Santos[1]

[1] Instituto Português de Reumatologia

São conhecidas as diferenças entre homens e mulheres em muitas doenças reumáticas, em relação à forma de apresentação, manifestações clínicas, impacto da doença e resposta à terapêutica. Nas espondilartrites (SpA), classicamente consideradas doenças do sexo masculino, essas diferenças estão sobretudo estudadas nas formas de espondilartrite axial (axSpA). Sabemos hoje que embora exista uma maior prevalência no sexo masculino da forma radiográfica de axSpA (3:1), a forma não-radiográfica parece afetar de igual modo ambos os sexos. Também o prognóstico, ao contrário do que se pensava há alguns anos, não é melhor no sexo feminino.

Diferentes perfis de expressão génica em homens e mulheres com axSpA, para além da maior prevalência do HLA-B27 nos homens, poderão ter um papel importante na diferente progressão radiográfica observada nos 2 sexos. Os homens apresentam maior progressão radiográfica a nível da coluna vertebral e sacroilíacas, mas as mulheres parecem ter um envolvimento preferencial da coluna cervical. No entanto, as diferenças não estão restritas à progressão radiográfica, já que em diferentes coortes, a mulheres apresentaram maior atividade de doença, mais entesite, mais dactilite, mais dor difusa e pior função, para um mesmo grau de dano radiográfico. Por outro lado,

os homens apresentam mais lesões inflamatórias na ressonância magnética nuclear, tanto a nível da coluna vertebral como das sacroilíacas. As manifestações extra-articulares como a psoríase e a doença inflamatória intestinal são mais frequentes nas mulheres, mas a uveíte aguda anterior parece ser mais prevalente no sexo masculino. À semelhança do que se passa noutras doenças reumáticas inflamatórias, as mulheres com axSpA apresentam pior qualidade de vida e maior impacto na produtividade laboral.

O início insidioso dos sintomas, a prevalência elevada da lombalgia na população e a falta de marcadores de diagnóstico, fazem com que continue a existir um atraso no diagnóstico desta patologia, que se estima ser atualmente de cerca de 8 anos. Nas mulheres com axSpA, esse atraso diagnóstico é ainda maior, por várias razões: o conceito enraizado de se tratar de uma doença do sexo masculino; pelas particularidades da apresentação clínica (mais entesite, mais dor difusa e menos sacroileíte radiográfica); pela maior prevalência de fibromialgia associada.

Também em relação ao tratamento parecem existir diferenças entre os dois sexos. Vários estudos têm demonstrado uma pior resposta aos inibidores do TNF (TNFi) e uma menor persistência em tratamento, o que poderá ser explicado não só pela existência de diferentes perfis de citoquinas, mas também pela diferente apresentação clínica, com manifestações que classicamente têm menor resposta aos TNFi.

Na axSpA é assim importante compreender as diferenças no sexo masculino e feminino, com implicações na abordagem diagnóstica e terapêutica destes doentes.

REFERÊNCIAS:

1. Chandran V. Psoriatic spondylitis or ankylosing spondylitis with psoriasis: same or different? Curr Opin Rheumatol 2019;31(4):329–34.
2. Rusman T, Van Bentum RE, Van Der Horst-Bruinsma IE. Sex and gender differences in axial spondyloarthritis: Myths and truths. Revmatol 2020;59:IV38–46.
3. Wright GC, Kaine J, Deodhar A. Understanding differences between men and women with axial spondyloarthritis. Semin Arthritis Rheum 2020;50(4):687–94.

TREAT-TO-TARGET NAS ESPONDILARTRITES: REALIDADE OU UTOPIA

Alexandre Sepriano[1]

[1] Serviço de Reumatologia – Centro Hospitalar de Lisboa Ocidental, EPE, Hospital Egas Moniz EPE

Treat-to-target (T2T) is a strategy in which clinicians tailor interventions to achieve a predefined level of a relevant outcome measure in each individual patient. In theory, T2T should be better in symptomatic relief, prevention of (further) irreversible damage, decreasing the likelihood of comorbidities and in leading to better health related quality of life and social participation, as compared to the standard of care. International organizations have issued recommendations prescribing that ASDAS should be used to define the target in the management of patients with spondyloarthritis (SpA). Experts have agreed on Inactive disease or, in alternative, low disease activity according to the ASDAS has the target. ASDAS has, recently, be shown to associate with axial structural damage further strengthening its use on T2T strategies in SpA. The benefit of T2T in SpA remains, however, to be formally demonstrated in a randomized experiment. In this lecture, the evidence supporting or refuting the practice of T2T over standard of care in SpA will be critically reviewed. In light of this evidence, we will discuss the likelihood that T2T will ever become a reality in the management of SpA in the daily clinical practice.

SESSÃO IV – ARTRITE REUMATÓIDE

INTEGRAÇÃO DE CUIDADOS NA AR: PARCERIA APPSREUMA – THE BOOMERANG EFFECT APRESENTAÇÃO DINÂMICA COM 4 PRELETORES (REUMA, ENFERMAGEM, MGF, DOENTES)

João Eurico Fonseca, Ricardo Ferreira, Elsa Frazão Mateus, José Carlos Marinho

SESSÃO V – ENVOLVIMENTO EXTRA-ARTICULAR NA ARTRITE PSORIÁTICA: O QUE ESTAMOS A PERDER?

MANIFESTAÇÕES EXTRA-ARTICULARES NA ARTRITE PSORIÁTICA: FISIOPATOLOGIA E CUIDADOS A TER NA PRÁTICA CLÍNICA

Maria José Santos[1]

[1]Serviço de Reumatologia, Hospital Garcia de Orta, Almada

A artrite psoriática (AP) é uma doença heterogénea quer no que respeita ao padrão de envolvimento articular, quer às manifestações extra-articulares. A prevalência estimada da AP na população portuguesa adulta é de 0,3%, ou seja, afeta cerca de 26 000 adul-

tos, os dois sexos numa proporção semelhante, sendo mais frequente em indivíduos caucasianos. Existe uma forte predisposição genética para desenvolver AP, embora vários fatores ambientais também tenham sido identificados como desencadeantes desta artropatia inflamatória em doentes com psoríase.

Para além da psoríase, outras manifestações extra-articulares como a entesite, dactilite ou envolvimento ungueal são bastante comuns, estimando-se a sua prevalência em 30-60%. Já o envolvimento ocular ou gastrointestinal característico das espondilartrites é menos frequente. Outras comorbilidades, tais como a doença cardiovascular, síndrome metabólica, obesidade, diabetes mellitus, dislipidémia, esteato-hepatite não alcoólica, gota úrica ou depressão, estão aumentadas nos doentes com AP e a sua presença associa-se a maior morbi-mortalidade e a uma pior qualidade de vida.

A identificação atempada e abordagem correta das comorbilidades é essencial na gestão do doente com AP.

SEXUALIDADE E ARTRITE PSORIÁTICA

Manuel Esteves

SESSÃO VI – LÚPUS ERITEMATOSO SISTÉMICO E OUTRAS DOENÇAS IMUNOMEDIADAS SISTÉMICAS

HIPERTENSÃO PULMONAR NA ESCLEROSE SISTÉMICA

Mário Santos[1,2]

[1] Unidade de Doença Vascular Pulmonar do Hospital Santo António, Centro Hospitalar Universitário do Porto.

[2] Unidade Multidisciplinar de Investigação Biomédica, Instituto de Ciências Biomédicas Abel Salazar, Universidade do Porto.

A hipertensão pulmonar associada a doença do tecido conectivo é uma complicação frequente da Esclerose Sistémica. Este é um tipo de hipertensão arterial pulmonar muito agressivo e que constitui uma das principais causas de morte destes doentes. Atualmente, estão disponíveis fármacos vasodilatadores pulmonares que demonstraram eficácia neste grupo de doentes com ganhos significativos de sobrevida e qualidade de vida. Contudo, a sua eficácia é tanto maior, quanto mais precoce for o diagnóstico. Nesta apresentação será discutido a evidência científica que suporta a recomendação do rastreio de hipertensão arterial pulmonar em doentes com Esclerose Sistémica, assim como as suas implicações na prática clínica.

SLE-DAS: NOVO SCORE DE ATIVIDADE "MADE IN PORTUGAL"

Diogo Jesus[1,2], Luís Inês[2,3]

[1] Serviço de Reumatologia, Centro Hospitalar de Leiria

[2] Faculdade de Ciências da Saúde, Universidade da Beira Interior

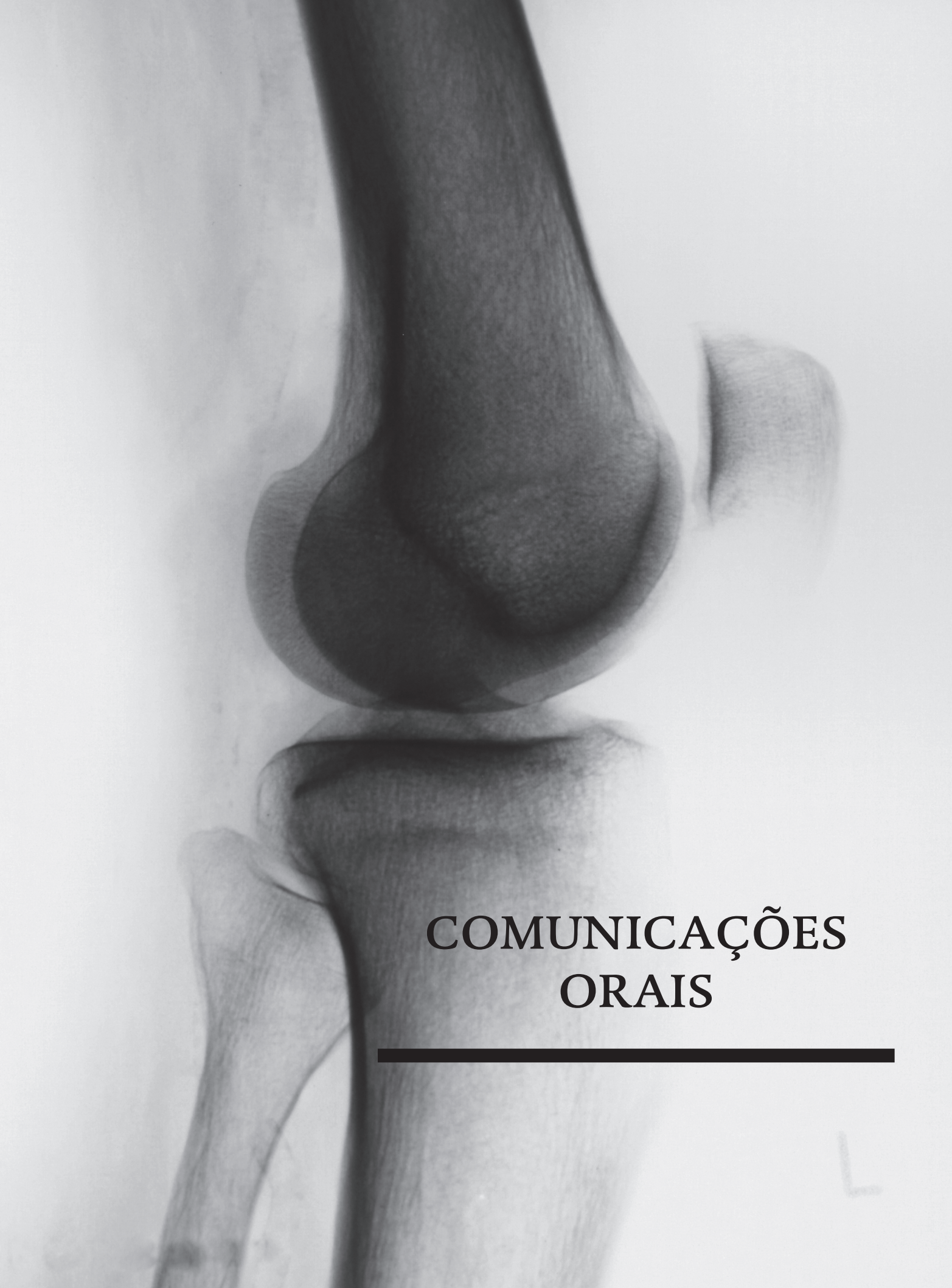
[3] CHUC Lupus Clinic, Serviço de Reumatologia, Centro Hospitalar e Universitário de Coimbra

O *Systemic Lupus Erytemathosus Disease Activity Score* (SLE-DAS) é um índice de atividade clínica composto para medir a atividade de doença do Lúpus Eritematoso Sistémico (LES) que foi recentemente validado e publicado. É constituído por 17 parâmetros clínicos e laboratoriais ponderados, incluindo parâmetros contínuos para a artrite, proteinúria, trombocitopenia e leucopenia, e parâmetros dicotómicos. Ao contrário do *SLE Disease Activity Index* (SLEDAI), o SLE-DAS distingue vasculite mucocutânea de vasculite sistémica e exantema cutâneo limitado de exantema cutâneo generalizado, melhorando assim o sistema de pontuação. Para além disso, inclui manifestações que apesar de raras são clinicamente muito relevantes, como o envolvimento oftalmológico, cardiopulmonar, gastrointestinal e a anemia hemolítica.

Nos estudos de validação já realizados o SLE-DAS demonstrou ter maior precisão na mensuração da atividade do LES, maior sensibilidade à mudança e maior valor preditivo para dano que o SLEDAI. Para além disso, apresentou elevada performance para classificar categorias de atividade de doença de acordo com a opinião de *expert* e com o *British Isles Lupus Assessment Group index* (BILAG) e para definir remissão clínica e baixa atividade de doença de acordo com os critérios DORIS (*Definition of Remission in SLE*) e LLDAS (*Lupus Low Disease Activity*), respetivamente.

Calcular o SLE-DAS utilizando a calculadora online (http://sle-das.eu/) é intuitivo, fácil e rápido, demorando apenas 1 a 2 minutos.

Em suma, o SLE-DAS é um índice de atividade preciso e fácil de utilizar na prática clínica diária que pode facilitar a gestão dos doentes com lúpus utilizando a estratégia *treat-to-target*, a seleção de doentes para ensaios clínicos e que poderá melhorar as medidas de *outcome* dos ensaios clínicos.



COMUNICAÇÕES ORAIS

Comunicações orais

ACTA REUMATOL PORT. 2021:46:??-?? (SUP)

004 - BIOLOGIC AGENTS FOR RHEUMATIC DISEASES IN THE OUTBREAK OF COVID-19: FRIEND OR FOE?

Cristiana Sieiro Santos[1], Clara Moriano[1], Carolina Álvarez[1], Elvira Díez[1], Trinidad Sandoval[1]
[1]Reumatología, Complejo Asistencial Universitario de León, León, Spain

Background: The recent outbreak of COVID-19 has raised concerns in the rheumatology community about the management of immunosuppressed patients diagnosed with inflammatory rheumatic diseases. It is not clear whether the use of biologic agents may suppose a risk or protection against SARS-CoV2 infection however, it has been suggested that severe respiratory forms of COVID-19 occur as result of exacerbated inflammation status and cytokine production.
Objectives: To estimate COVID-19 infection rate in patients treated with biologic disease-modifying antirheumatic drugs (bDMARDs) for rheumatic inflammatory diseases, determine the influence of biologic agents treatment as a risk or protective factor and studying the prognosis of rheumatic patients receiving biologic agents compared to general population in a third level Hospital setting in León, Spain.
Methods: We performed a retrospective observational study including patients seen at Rheumatology department who received bDMARDs for rheumatic diseases between December 1st 2019 and December 1st 2020 and analysed COVID-19 infection rate. Main variable was the hospital admission related to COVID-19. The covariates were age, sex, comorbidities, biologic agent, duration of treatment, mean dose of glucocorticoids and need for ICU care. We performed a multivariate logistic regression model to assess risk factors of COVID-19 infection.
Results: There was a total of 4464 patients with COVID-19 requiring hospitalization. 40 patients out of a total of 820 patients with rheumatic diseases (4.8%) receiving bDMARDs contracted COVID-19 and four required hospital care. Crude incidence rate of COVID-19 requiring hospital care among the general population was 3.6%, and it was 0.89% among the group with underlying rheumatic diseases. Out of the 4464 patients, 869 patients died, 2 of which received treatment with biologic agents. Patients with rheumatic diseases who tested positive for COVID-19 were older (female: median age 60.8 IQR 46-74; male: median age 61.9 IQR 52-70.3) than those who were negative for COVID-19 (female: median age 58.3 IQR 48-69; male: median age 56.2 IQR 47-66), more likely to have hypertension (45% vs 26%, OR 2.25 (CI 1.14-4.27), p 0.02), cardiovascular disease (23 % vs 9.6%, OR 2.73 (1.25-5.95), p 0.02), be smokers (13% vs 4.6%, OR 2.95 (CI 1.09-7.98), p 0.04), receiving treatment with rituximab (20% vs 8%, 2.28 (CI 1.24-6.32), p 0.02) and a higher dose of glucocorticoids (OR 2.5 (1.3-10.33, p 0.02) and were less likely to be receiving treatment with IL-6 inhibitors (0.03% vs 14%, OR 0.16, (CI 0.10-0.97, p 0.03). Patients who tested negative for COVID-19 were more likely to be treated with bDMARDs for a longer period (in months) than patients with a positive result (OR 0.44 (0.29-0.78), p 0.04). When exploring the effect of the rest of the therapies between groups (affected patients vs unaffected), we found no significant differences in bsDMARD proportions. IL-1 inhibitors, IL6 inhibitors, JAK inhibitors and belimumab treated patients showed the lowest incidence of COVID-19 among adult rheumatic patients. We found no differences in sex or rheumatological disease between patients who tested positive for COVID-19 and patients who tested negative.
Conclusions: Overall, the use of bDMARDs does not associate with severe manifestations of COVID-19. Patients with rheumatic disease diagnosed with COVID-19 were more likely to be receiving a higher dose of glucocorticoids and treatment with rituximab. IL-6 inhibitors may have a protective effect.

042 - RHEUMATIC DISEASE COMORBIDITY INDEX AS A PREDICTOR OF CLINICAL RESPONSE TO THE FIRST BIOLOGICAL AGENT IN PORTUGUESE PATIENTS WITH RHEUMATOID ARTHRITIS: A MULTICENTER COHORT STUDY.

Salomé Garcia[1], Augusto Silva[2], Carolina Zinterl[2],

Joaquim Polido Pereira[2], Ana Isabel Maduro[3], Cátia Duarte[3], Catarina Soares[4], Daniela Santos-Faria[4], Susana P. Silva[5], Teresa Novoa[6], Sara Paiva Dinis [7], Filipe Araújo[8], José Tavares-Costa[4], Tiago Meirinhos[9], Margarida Cruz[10], Miguel Bernardes[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal,
[2]Rheumatology and Bone Metabolic Diseases Department, Centro Hospitalar Lisboa Norte, EPE - Hospital de Santa Maria, Lisbon Academic Medical Centre, Lisboa, Portugal,
[3]Rheumatology Department, Centro Hospitalar Universitário de Coimbra, Coimbra, Portugal,
[4]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal,
[5]Rheumatology Department, Centro Hospitalar do Baixo Vouga, EPE, Aveiro, Aveiro, Portugal,
[6]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal,
[7]Rheumatology Department, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal,
[8]Rheumatology and Osteoporosis Unit, Hospital de Sant Ana, SCML, Cascais, Portugal,
[9]Rheumatology Unit, Centro Hospitalar do Tâmega e Sousa, Penafiel, Portugal,
[10]Rheumatology Department, Consultórios Médicos de Caldas da Rainha, Caldas da Rainha, Portugal

Background: Rheumatoid arthritis (RA) is often complicated by medical conditions that coexist with or result from RA. Several comorbidity indices have been developed and validated for use in clinical studies and, more recently, Rheumatic Disease Comorbidity Index (RDCI) was validated in patients with rheumatic and musculoskeletal diseases. The objective of this work was to determine the main predictors of the 12-month clinical response to the first biological disease-modifying antirheumatic drugs (bDMARD) among Portuguese patients with RA, seeking to clarify a possible role of RDCI in this context.
Methods: This is a multicenter, observational study, using data from Reuma.pt. We included patients registered in 11 rheumatology centers, between 2008 until 31st October 2020. Patients meeting the 2010 ACR/EULAR criteria for RA and starting a first biological agent were retrospectively analyzed. The prevalence of comorbidities was computed at baseline and patients were stratified according to RDCI score. Disease activity parameters, physical function and clinical characteristics were evaluated at baseline and follow-up (6 and 12 months). A multivariate logistic regression model was developed to examine the role of RDCI along with other baseline factors as potential predictors of achieving remission, low disease activity (LDA), and EULAR good/moderate response at 12 months of follow-up.
Results: A total of 2001 patients were included in the analysis; 85.4% were women (n=1708). Mean age was 54.1 (± 12.3) years old, with a mean disease duration of 8.9 years (± 2.5). Their mean RDCI score was 1.82 ± 0.51. Arterial hypertension (n=545, 41.0%), dyslipidemia (n= 401, 30.2%), osteoporosis (n=296, 22.3%) and diabetes mellitus (n=272, 20.5%) were the most reported comorbidities, followed by depression (n=247, 18.6%) and Sjögren Syndrome (n=224, 16.9%). Regarding bDMARD, 83.1% (n= 1662) of patients received a tumor necrosis factor alpha inhibitor, with a minority initiating tocilizumab (N=190, 9.5%), rituximab (n=118, 5.9%), anakinra (N=20, 1.0%) and abatacept (n=11, 0.6%). Patients were stratified in 4 subgroups according to RDCI score (RDCI=0; RDCI=1; RDCI=2 and RDCI>=3) and differed only in age and selection of rituximab as the first bDMARD (p<0.001). Patients with higher RDCI scores were significantly older and were more frequently on rituximab as first-line biological agent. No individual comorbidity was identified as a predictor of response to a bDMARD. According to the multivariate logistic regression analysis, lower RDCI score was a predictor of achieving 12-month EULAR good-moderate response (OR 0.780, 95% CI 0.641-0.842, p< 0.001) and DAS28-ESR remission (OR 0.792, 95% CI 0.615-0.950, p = 0.01). Higher baseline DAS28-ESR and combination therapy with MTX were also associated with achieving the same two outcomes (Table 1).
Discussion/Conclusions: The results of this study corroborate the usefulness of this score for screening the comorbidity status in patients who are about to initiate a bDMARD. To the best of the authors' knowledge, this study represents the first attempt to characterize comorbidities and test the hypothesis of a predictive effect of RDCI in a multicentric RA Portuguese cohort. Underreporting of comorbidities is probably the main limitation of this work, as data entry in the Reuma.pt database is voluntary. However, we evaluated a large and homogeneous sample that provided robust data on the impact of comorbidities in RA patients.

TABLE I. RDCI AND OTHER BASELINE FACTORS AS PREDICTORS OF 12-MONTH EULAR RESPONSE AND DAS28-ESR REMISSION

	EULAR good/moderate response		DAS28-ESR LDA		DAS28-ESR Remission	
	OR (95% CI)	*p*	OR (95% CI)	*p*	OR (95% CI)	*p*
Age, years	0.871 (0.780 - 1.072)	0.250	0.974 (0.921 - 1.134)	0.360	0.965 (0.922 - 1.081)	0.352
Sex	0. 978 (0.923 - 1.222)	0.450	0.956 (0.870 - 1.321)	0.683	0.988 (0.907 - 1.255)	0.667
Baseline DAS 28 ESR	1.450 (1.130 - 1.520)	**0.010**	1.201 (0.980 - 1.256)	0.340	1.461 (1.212 - 1.497)	**<0.001**
RF positivity	1.232 (0.990 - 1.371)	0.370	1.302 (0.794 - 1.400)	0.770	1.304 (0.681 - 1.402)	0.768
ACPA positivity	1.871 (0.494 - 1.955)	0.850	0.974 (0.722 - 1.138)	0.780	1.965 (0.560- 1.280)	0.862
Concomitant MTX	1.202 (1.191-1.360)	**<0.001**	0.964 (0.922 - 1.170)	0.673	1.320 (1.161 - 1.623)	**0.020**
RDCI score	0.780 (0.641 - 0.842)	**<0.001**	0.811 (0.570-1.042)	0.090	0.792 (0.615 - 0.950)	**0.010**

ACPAs: Anti-citrullinated protein antibodies; CI: confidence intervals; DAS28-ESR: Disease Activity Score 28-Erythrocyte Sedimentation Rate; EULAR: European league against rheumatism; LDA: Low disease activity; MTX: Methotrexate; OR: odds ratio; RDCI: Rheumatic Disease Comorbidity Index; RF: Rheumatoid Factor.
Multivariate logistic regression analysis. Data expressed in bold presented statistical significancy.

084 - PREVALENCE AND CLINICAL ASSOCIATIONS OF DIFFERENT AUTOANTIBODIES IN THE REUMA.PT SYSTEMIC SCLEROSIS COHORT: IS IT ALL REALLY SET IN STONE?

Eduardo Dourado[1, 2], Raquel Freitas[3], Patrícia Martins[1, 2], Liliana Saraiva[4], Tânia Santiago[4, 5], Francisca Guimarães[6], Emanuel Costa[7], Diogo Esperança Almeida[7], Sara Paiva Dinis [8], Ana Sofia Pinto[8], Alexandra Daniel[9], Marília Rodrigues[9], Maria João Salvador[4, 5], Ana Catarina Duarte[3], Ana Cordeiro[3], Maria José Santos[2, 3], JE Fonseca[1, 2], Catarina Resende[1, 2], Inês Cordeiro[1, 2]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal,
[2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal,
[3]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal,
[4]Serviço de Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal,
[5]Coimbra Institute for Clinical and Biomedical Research (iCBR) - Faculty of Medicine, University of Coimbra, Coimbra, Portugal,
[6]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal,
[7]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal,
[8]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal,
[9]Serviço de Reumatologia, Hospital Distrital de Leiria, Leiria, Portugal

Background: Different autoantibodies (Ab) have been associated with distinct systemic sclerosis (SSc) phenotypes. Most of these associations have not been confirmed in Portuguese patients.
Objective: To evaluate SSc immuno-clinical associations in the Rheumatic Diseases Portuguese Register (Reuma.pt) cohort.
Methods: Multicentre open cohort study including adult SSc patients registered in Reuma.pt up to February 2021. The associations between Ab expression and clinical data were established using Chi-Square, Fischer's Exact or Mann-Whitney U tests. The Bonferroni correction for multiple comparisons was applied to get ≤0.05. Definite associations were defined by p≤0.002, and likely associations by p≤0.05.
Results: 1080 patients were included, with a mean age and disease duration of 60.2±14.6 and 12.4±10.0 years, respectively (Table 1). Most were females (87.5%) and had white European ancestry (WEA, 93.2%). The most common disease subtypes were limited cutaneous (lcSSc, 57.4%), diffuse cutaneous (dcSSc, 17.7%), and very early diagnosis of SSc (VEDOSS, 12.3%). Most patients expressed antinuclear Ab (ANA, 93.4%), and the most frequent were anti-centromere (ACA, 54.6%), anti-topoisomerase I (Scl70, 21.8%), and anti-Pm/Scl Ab (PmScl, 4.7%).

ACA had definite positive associations with female sex, older age at diagnosis, lcSSc, lower modified Rodnan skin score (mRSS, median 0 vs 4), and isolated sclerodactyly, and likely associations with a higher diagnosis delay, WEA and VEDOSS. ACA had definite inverse associations with flexion contractures (FC), myositis, digital ulcers (DU), and interstitial lung disease (ILD), and likely inverse associations with pitting scars (PS) and oesophageal involvement (OI).

Scl70 had definite positive associations with male sex, dcSSc, higher mRSS, FC, DU, PS, ILD, and OI, and likely associations with younger age at diagnosis, ten-

don friction rubs, active scleroderma pattern in capillaroscopy, and heart involvement.

PmScl had a definite association with myositis and likely associations with male sex, calcinosis, joints involvement, and ILD. Anti-U1RNP Ab had definite associations with younger age at diagnosis, MCTD and myositis, and likely associations with a lower diagnosis delay, African ancestry and joint involvement. Anti-RNA polymerase III Ab (RP3) had likely associations with higher mRSS and renal involvement. Anti-U3RNP Ab had a definite association with dcSSc and likely associations with calcinosis and renal involvement. Anti-Th/To Ab had likely associations with male sex and myositis. Anti-Ku Ab had likely associations with systemic lupus erythematosus and mixed connective tissue disease (MCTD) overlap syndromes.

Discussion: There was a higher prevalence of ACA and PmScl compared to other cohorts, most likely due to the high proportion of WEA patients. Most immunoclinical associations described in the literature apply (Figure 1), including ACA with lcSSc and Scl70 with dcSSc, DU, PS and ILD. However, Scl70+ patients did not have an increased risk of renal involvement, and ACA+ patients did not have an increased risk for calcinosis, PAH or OI, contrary to what was described in the literature. New findings included the association of PmScl with ILD and Scl70 with an active pattern in capillaroscopy. Also, anti-U3RNP+ and Th/To+ patients

TABLE I. DEMOGRAPHIC AND CLINICAL DATA FROM THE 1080 PATIENTS INCLUDED IN THE STUDY

Variable	NPAD	Total
Demographic data		
Females, N (%)	1080	945 (87.5)
Ancestry		
European white, N (%)	643	599 (93.2)
African, N (%)	643	30 (4.7)
Non-European white, N (%)	643	7 (1.1)
Mixed African and European, N (%)	643	5 (0.8)
Romani, N (%)	643	1 (0.2)
Asian, N (%)	643	1 (0.2)
Age at the symptom onset, mean (SD)	824	47.3 (15.5)
Age at diagnosis, mean (SD)	927	52.3 (14.7)
Age at the last appointment, mean (SD)	1080	60.2 (14.6)
Diagnosis delay, mean (SD)	815	5.0 (8.2)
Disease duration, mean (SD)	824	12.4 (10.0)
Death, N (%)	1080	79 (7.3)
Disease subsets		
Limited cutaneous SSc, N (%)	1080	620 (57.4)
Diffuse cutaneous SSc, N (%)	1080	191 (17.7)
SSc sine scleroderma, N (%)	1080	30 (2.8)
Very early diagnosis of SSc, N (%)	1080	133 (12.3)
Overlap syndromes, N (%)	1080	106 (9.8)
Overlap with polymyositis, N (%)	1079	22 (2.0)
Overlap with dermatomyositis, N (%)	1079	12 (1.1)
Overlap with systemic lupus erythematosus, N (%)	1080	22 (2.0)
Overlap with Sjögren's syndrome, N (%)	1077	24 (2.2)
Overlap with rheumatoid arthritis, N (%)	1080	9 (0.8)
Mixed connective tissue disease, N (%)	1080	29 (2.7)
Classification criteria		
2013 ACR/EULAR classification criteria, N (%)	1035	807 (78.0)
1980 ACR classification criteria, N (%)	904	548 (60.6)

continues on the next page

TABLE I. CONTINUATION

Variable	NPAD	Total
Clinical manifestations		
Skin disease		
Skin thickening, N (%)	1050	789 (75.1)
Skin thickening proximal to the MCP in any location, N (%)	909	432 (47.5)
Skin thickening proximal to the MCP in hand, N (%)	1008	411 (40.8)
Sclerodactyly without skin thickening proximal to the MCP, N (%)	1005	309 (30.7)
Puffy fingers, N (%)	890	390 (43.8)
mRSS in the last appointment, mean (SD)	752	4.7 (7.1)
Calcinosis, N (%)	983	139 (14.1)
Musculoskeletal disease		
Joints involvement (arthritis or arthralgia), N (%)	1002	400 (39.9)
Flexion contractures, N (%)	953	77 (8.1)
Tendon friction rubs, N (%)	922	23 (2.5)
Myositis, N (%)	984	62 (6.3)
Vascular disease		
Raynaud phenomenon, N (%)	1047	978 (93.4)
Digital ulcers, N (%)	1040	365 (35.1)
Pitting scars, N (%)	927	287 (31.0)
Telangiectasia, N (%)	1008	493 (48.9)
Pulmonary arterial hypertension, N (%)	1021	19 (1.9)
Capillaroscopy changes, N (%)	697	547 (78.5)
Early scleroderma pattern, N (%)	540	131 (24.3)
Active scleroderma pattern, N (%)	540	147 (27.2)
Late scleroderma pattern, N (%)	540	82 (15.2)
Lung disease		
Interstitial lung disease, N (%)	870	289 (33.2)
Restrictive syndrome, N (%)	696	31 (4.5)
Lung fibrosis on chest radiograph, N (%)	491	133 (27.1)
Fibrosis on chest CT scan, N (%)	694	109 (15.7)
Ground glass opacities on chest CT scan, N (%)	694	75 (10.8)
Honeycombing on chest CT scan, N (%)	693	33 (4.8)
Gastrointestinal disease		
Oesophageal involvement, N (%)	971	354 (36.5)
Gastric involvement, N (%)	951	116 (12.2)
Intestinal involvement, N (%)	946	51 (5.4)
Heart involvement, N (%)	966	73 (7.6)
Renal involvement, N (%)	961	18 (1.9)
Autoantibodies		
Anti-nuclear antibodies, N (%)	1031	963 (93.4)
Anti-centromere antibodies, N (%)	1043	569 (54.6)
Anti-topoisomerase I antibodies, N (%)	1036	226 (21.8)
Anti-Pm/Scl antibodies, N (%)	665	31 (4.7)
Anti-U1-RNP antibodies, N (%)	693	28 (4.0)
Anti-RNA polymerase III antibodies, N (%)	806	24 (3.0)
Anti-U3-RNP antibodies, N (%)	643	16 (2.5)
Anti-Th/To antibodies, N (%)	606	13 (2.1)
Anti-Ku antibodies, N (%)	633	12 (1.9)
Anti-U11/U12-RNP antibodies, N (%)	613	3 (0.5)

Abbreviations: ACR – American College of Rheumatology, CT – computed tomography, EULAR - European Alliance of Associations for Rheumatology, MCP – metacarpophalangeal joints, mRSS – modified Rodnan skin score, N – number of patients, NPAD – number of patients with available information, RNA – ribonucleic acid, SD – standard deviation, SSc – systemic sclerosis.

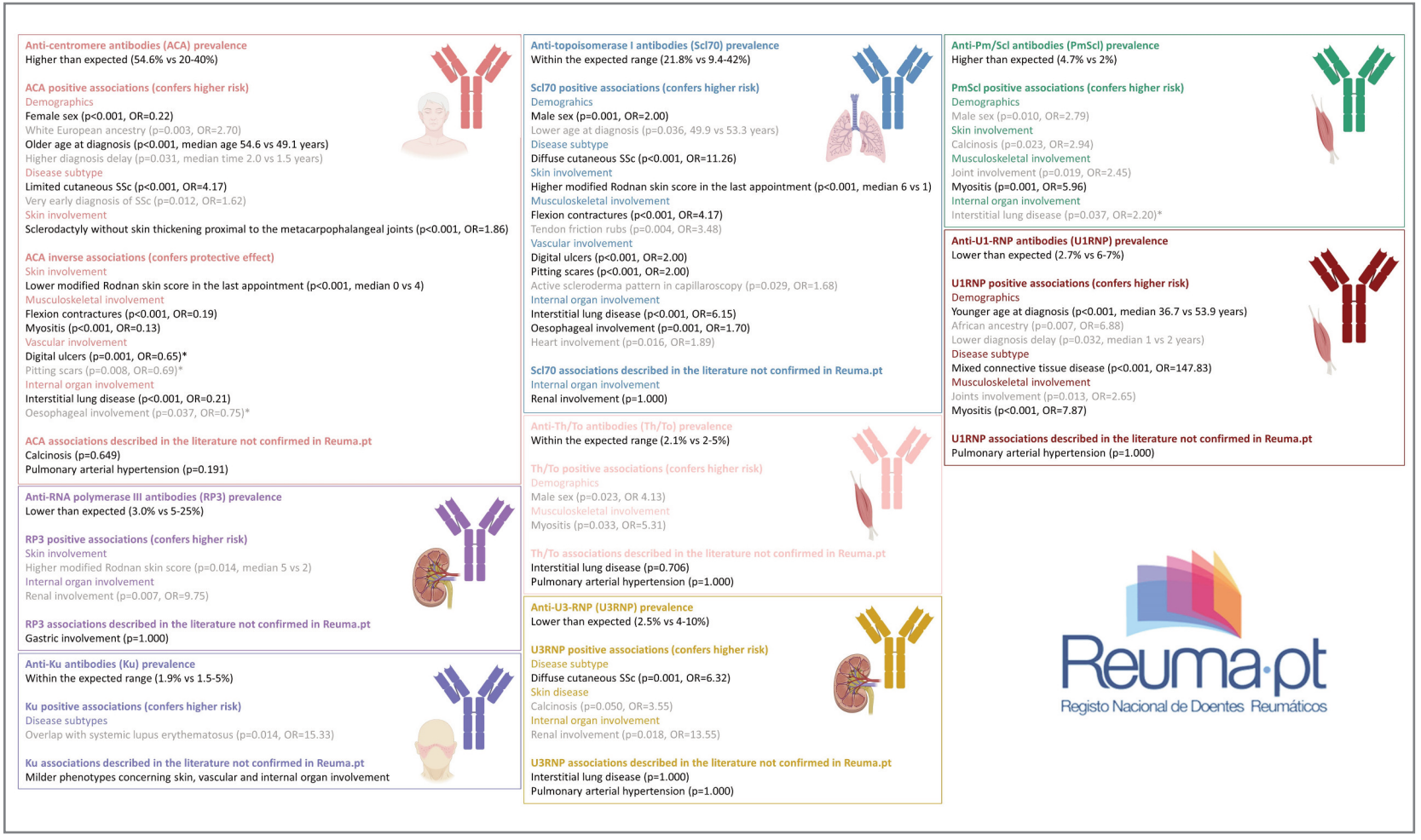


FIGURE.. Clinical associations regarding each of the analysed autoantibodies.

did not have an increased risk of ILD or PAH, contrarily to what was previously reported. These nuances may be specific to the Portuguese SSc population or signal previously reported associations as geographically specific.

086 - DETERMINANTS OF QUALITY OF LIFE IN RHEUMATOID ARTHRITIS AND SPONDYLOARTHRITIS: HOW DO THEY INTERACT FROM A HIERARCHICAL PERSPECTIVE?

Pedro David Carvalho[1, 2, 3], Elsa Vieira-Sousa[2, 4], Marreiros A[3, 5], Pedro MAchado[6, 7, 8]
[1]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal,
[2]Centro Académico de Medicina de Lisboa, Lisboa, Portugal,
[3]Algarve Biomedical Center, Faro, Portugal,
[4]Rheumatology and Bone Metabolic Diseases Department, Centro Hospitalar Lisboa Norte, EPE - Hospital de Santa Maria, Lisbon Academic Medical Centre, Lisboa, Portugal,
[5]Universidade do Algarve, Faro, Portugal,
[6]Rheumatology, Northwick Park Hospital, London North West University Healthcare NHS Trust, London, United Kingdom,
[7]Centre for Rheumatology & MRC Centre for Neuromuscular Diseases, University College London, London, UK., London, United Kingdom,
[8]Centre for Rheumatology, University College of London, London, United Kingdom

TABLE I. MULTIVARIABLE LINEAR REGRESSION ANALYSES TO INVESTIGATE THE ASSOCIATION BETWEEN QOL (ASSESSED BY EQ-5D-3L) AND OTHER DEMOGRAPHIC AND CLINICAL VARIABLES IN THE 3920 PATIENTS WITH RA AND 3984 PATIENTS WITH SPA

Characteristics	Adjusted B* (95% CI) for RA	p-value	Adjusted B* (95% CI) for SpA	p-value
Age			-0.002 (-0.003, -0.001)	0.001
Male gender	0.008 (-0.017, 0.033)	0.533	0.024 (0.002, 0.046)	0.036
Education, university or equivalent	0.001 (-0.013, 0.016)	0.849	-0.017 (-0.033, -0.001)	0.041
BMI	0.002 (0.000, 0.004)	0.045	-0.001 (-0.003, 0.001)	0.593
HLA-B27 positive			0.023 (0.000, 0.046)	0.047
Seropositive for RF or ACPA				
Unequivocal radiological erosion	-0.036 (-0.058, -0.014)	0.001		
Current smoking				
Current alcohol equal or more than 3 units	0.015 (0.004, 0.026)	0.007	0.009 (-0.001, 0.018)	0.067
ASDAS-CRP			-0.064 (-0.077, -0.052)	<0.001
DAS28-CRP-3v	-0.028 (-0.037, -0.019)	<0.001		
MHAQ	-0.216 (-0.247, -0.184)	<0.001	-0.330 (-0.361, -0.298)	<0.001
Work productivity loss (overall work impairment / absenteeism plus presenteeism - %)	-0.003 (-0.003, -0.002)	<0.001	-0.002 (-0.003, -0.002)	<0.001
NSAID intake during the last 3 months	0.017 (-0.004, 0.039)	0.115	-0.046 (-0.067, -0.024)	<0.001
Current cDMARD	0.027 (-0.005, 0.059)	0.099	-0.006 (-0.028, 0.015)	0.568
Current bDMARD	-0.005 (-0.028, 0.018)	0.653	-0.038 (-0.059, -0.017)	<0.001

*Unstandardized coefficients. Legend: ACPA, anti-citrullinated protein antibody; ASDAS-CRP, Ankylosing Spondylitis Disease Activity Score; bDMARDs, biological disease-modifying antirheumatic drugs; BMI, body mass index; cDMARDs, conventional disease-modifying antirheumatic drugs; DAS28-CRP-3v, Disease Activity Score 28-CRP-3 variables; EQ-5D-3L, EuroQOL-5 Dimension 3-level; MHAQ, Modified Health Assessment Questionnaire; NSAID, Nonsteroidal anti-inflammatory drug; RA, rheumatoid arthritis; RF, rheumatoid factor.

Introduction/Aim: To assess the hierarchy of outcomes contributing for quality of life (QoL) in rheumatoid arthritis (RA) and spondyloarthritis (SpA).

Materials and Methods: The COMOrbidities in SPondyloArthritis (COMOSPA) and COMOrbidities in Rheumatoid Arthritis (COMORA) initiatives are two multicentre transversal studies resulting from an international collaboration which enrolled 7904 consecutive patients with SpA and RA from 26 countries. In this study we analysed data on QoL assessed by EuroQOL-5 Dimension 3-level (EQ-5D-3L) in both databases. Both RA and then SpA subgroups were analysed separately. In a first exploratory step, linear regression models were used to identify univariable associations between several demographic and clinical variables and EQ-5D. Variables with a p-value <0.10 in the univariable analysis were tested in linear regression multivariable models. Subsequently, a decision tree was constructed according to an unbiased hierarchical multivariable analysis using the Chi-square Automatic Interaction Detector (CHAID) method, with EQ-5D as dependent variable. We stipulated a minimum requirement of 20 cases for parent branches and a minimum of 10 cases for child branches.

Results: Data on 3920 RA patients and 3984 SpA patients was analysed. In RA subgroup QoL was significantly associated with modified Health Assessment Questionnaire (MHAQ) [adjusted (adj) B=-0.216, (95% confidence interval (CI)= (-0.247,-0.184)], Disease Activity Score 28 (DAS28)-CRP-3 variables [adjB=-0.028, 95%CI=(-0.037,-0.019)], work productivity loss [adjB=-0.003, 95%CI=(-0.003,-0.002)], presence of unequivocal radiological erosion [adjB=-0.036, 95%CI=(-0.058,-0.014)], current alcohol consumption ≥3 units per day [adjB=0.015, 95%CI=(0.004,0.026)] and body mass index (BMI)

[adjB=0.002, 95%CI=(0.000,0.004)] (table 1). In SpA subgroup QoL was significantly associated with MHAQ [adjB=-0.330, 95%CI=(-0.361,-0.298)], Ankylosing Spondylitis Disease Activity Score (ASDAS) [adjB=-0.064, 95%CI=(-0.077,-0.052)], work productivity loss [adjB=-0.002, 95%CI=(-0.003,-0.002)], current NSAID treatment [adjB=-0.046, 95%CI=(-0.067, -0.024)], current bDMARD treatment [adjB=-0.038, 95%CI=(-0.059,-0.017)], age [adjB=-0.002, 95% CI=(-0.003,-0.001)], male gender [adjB=0.024, 95%CI=(0.002,0.046)], universitary education [adjB=-0.017, 95%CI=(-0.033,-0.001)] and HLA-B27 positivity [adjB=0.023, 95%CI=(0.000,0.046)] (table 1). The decision tree revealed MHAQ as the first variable with the most discriminative power on EQ-5D, followed by work productivity loss and disease activity both in RA and SpA patients.

Conclusions: Disability is a major contributor for QoL measured by EQ-5D in RA and SpA patients. Disease activity and work productivity loss also play important roles for QoL in these patients.

092 - HEALTH-RELATED QUALITY OF LIFE AND FUNCTION IN ADULTS WITH JUVENILE IDIOPATHIC ARTHRITIS – COMPARISON WITH ADULT-ONSET RHEUMATIC DISEASES

Filipa Oliveira Ramos [1, 2], Rodrigues AM[3], Martins FM[4], Ana Teresa Melo[1], Francisca Aguiar[5], Luisa Brites[6], Soraia Azevedo[7], Ana Catarina Duarte[8], Carolina Furtado[9], Ana Filipa Mourão[3, 10], Graça Sequeira[11], Inês Cunha[12], Ricardo Figueira[13], José Melo-Gomes[14], Maria José Santos[2, 8], JE Fonseca[1,2]

[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal,
[2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal,
[3]EpiDoC Unit, CEDOC, NOVA Medical School, NOVA University, Lisboa, Portugal,
[4]Sociedade Portuguesa de Reumatologia, Lisboa, Portugal,
[5]Serviço de Reumatologia, Centro Hospitalar São João, Porto, Portugal,
[6]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal,
[7]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal,
[8]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal,
[9]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal,
[10]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal,
[11]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal,
[12]Serviço de Reumatologia, Centro Hospitalar do Baixo Vouga, E.P.E., Aveiro, Portugal,
[13]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal,
[14]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal

Introduction: We have previously shown that most polyarticular and enthesis related arthritis (ERA) juvenile idiopathic arthritis (JIA) patients meet classification criteria for rheumatoid arthritis (RA) and spondylarthritis (SpA), maintain active disease and report functional impairment in adulthood.

Objective: To evaluate functional disability, mental health, fatigue and health related quality of life (HRQoL) among different JIA categories in adulthood and identify differences with adult-onset counterpart rheumatic diseases.

Methods: Cross-sectional analysis nested in a cohort study of JIA patients registered in the Rheumatic Diseases Portuguese Register (Reuma.pt). Functional disability (Health Assessment Questionnaire - Disability Index (HAQ-DI)), mental health symptoms (Hospital Anxiety and Depression Scale (HADS)), fatigue (Functional Assessment of Chronic Illness Therapy – Fatigue Scale (FACIT-F) and health-related quality of life (EuroQol-5D (EQ5D) and Medical Outcomes Study 36-item Short Form (SF-36)) were compared among JIA categories. Adult patients with polyarticular JIA course and with enthesis related arthritis (ERA) JIA were compared respectively to patients with rheumatoid arthritis (RA) and adult-onset spondylarthritis (SpA), matched for gender and age with adjustment for disease duration and activity.

Results: A total of 585 JIA patients were included, mean disease duration 22.8 ± 12.7 years, 38% had active disease and 8.7% had severe disability at the time of the last visit. 10.5% and 4.8% presented anxiety and depression symptoms, respectively, with no differences among all JIA categories. Fatigue was less reported in ERA patients when compared to undifferentiated JIA patients (FACIT-F score 45 [39; 49.5] vs 34 [25; 40]; p=0.045). Persistent oligoarthritis and ERA patients scored higher in EQ5D and in physical component of

TABLE. PREDICTORS OF FLARE IN MULTIVARIATE COX REGRESSION ACCORDING TO EACH OF THE FLARE DEFINITIONS (r-SFI, SLE-DAS, SLEDAI-2K)

	r-SFI	SLE-DAS	SLEDAI-2K
Anti-RNP+	2.11 (1.30-3.42)	2.39 (1.44-3.95)	2.22 (1.11-4.42)
Immunosuppressants	1.96 (1.22-3.15)	2.32 (1.38-3.88)	2.26 (1.12-4.54)
Prednisone*	1.93 (1.19-3.14)	1.99 (1.18-3.35)	2.17 (1.07-4.38)
Blood cytopenias§	2.08 (1.03-4.17)	n.s.	n.s.
Arthritis§	n.s.	n.s.	2.23 (1.12-4.44)

* Prednisone ≤7.5 mg/day as required by LLDAS. § Blood cytopenias; arthritis: cumulative SLE features up to baseline. Risk for each predictor reported as Hazard Ratio (95% Confidence Interval); n.s.: non-significant

the SF-36 when compared to other JIA categories. When we compared JIA to adult-onset rheumatic diseases, we found less disability in polyarticular course JIA when compared to RA (median HAQ of 0.25 (0; 1) vs 0.63 (0.13;1.13); p<0.001) and in ERA JIA when compared to SpA (median HAQ 0 (0;0.44) VS 0.75 (0;1,5); p=0.041). SpA patients had more depression and anxiety symptoms than ERA patients (respectively 14.8% vs 0%; p=0.003 and 21.3% vs 9%; p=0.002). JIA patients had less fatigue symptoms when compared to control patients with adult-onset diseases (FACIT-F score 42 [33.5; 47] vs 40 [29; 47.5]; p=0.041 and 45 [39; 49.5] vs 41 [29; 46]; p=0.010). JIA patients with polyarticular course had better scores on EQ5D and all domains of SF36, than control patients with RA.

Conclusion: Persistent oligoarticular JIA was the category with less functional impairment and with better HRQoL in adulthood. Adults with polyarticular JIA and ERA JIA reported less physical disability and fatigue when compared to adult-onset counterparts. The same pattern was observed for HRQoL regarding polyarticular JIA and RA and for anxiety and depression in ERA JIA when compared to adult-onset SpA. Overall, polyarticular and ERA JIA have lower functional impairment and better quality-of-life in adulthood than adult-onset RA and SpA.

106 - PREDICTORS OF FLARE IN SLE PATIENTS ATTAINING LUPUS LOW DISEASE ACTIVITY STATE: A REAL-LIFE COHORT STUDY OF 292 PATIENTS WITH 36-MONTH FOLLOW-UP

Rita Cunha[1], Liliana Saraiva[2], JAP da Silva[2, 3], Ines L[2, 4]
[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, EPE, Aveiro, Aveiro, Portugal,
[2]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal,
[3]Coimbra Institute for Clinical and Biomedical Research (iCBR) - Faculty of Medicine, University of Coimbra, Coimbra, Portugal,
[4]Faculdade de Ciências da Saúde, Universidade da Beira Interior, Covilhã, Portugal

Background: Lupus Low Disease Activity State (LLDAS) is a target for management of patients with SLE, that should be maintained in the long-term by preventing flares. Stratification of flare risk would be useful to optimize management.
Objectives: To identify predictors of flare in SLE patients attaining LLDAS.
Methods: Patients with SLE fulfilling classification criteria [ACR (1997) and/or SLICC and/or EULAR/ACR], followed at an academic lupus clinic from January 2017 to March 2020 were eligible. Baseline for each patient was the first visit with LLDAS within the study period. Patients never fulfilling LLDAS were excluded. Flares were identified as change from baseline by 3 instruments: revised SELENA flare index (r-SFI); SLEDAI-2K; Systemic Lupus Erythematosus Disease Activity Score (SLE-DAS). Time to first flare up to 36 months was identified separately for each instrument. Predictors of flare were sought through survival analysis, with distinct models for each of the three definitions of flare. Univariate analysis was performed using Kaplan-Meir curves and Log-Rank tests. Tested variables at baseline were: gender; age at time of SLE diagnosis; disease duration; cumulative SLE organ involvement (arthritis; mucocutaneous; renal; neurologic; haematological; anti-phospholipid syndrome); cumulative immunological features (anti-dsDNA; anti-Sm; anti-RNP, anti-phospholipid antibodies; hypocomplementemia); ongoing treatment (hydroxychloroquine; prednisone; immunosuppressants). Variables with p<0.1 were further tested in multivariate Cox regression models. Hazard

ratios (HR) were determined with 95% confidence intervals (95%CI).

Results: From 322 patients in this SLE cohort, 292 (90.7%) fulfilled LLDAS and were included in the analyses (female: 87.3%; mean age: 46.2±14.5 years; previous lupus nephritis: 36.0%; receiving ongoing antimalarials, immunosuppressants, glucocorticoids: 92.8%, 34.6% and 29.8%, respectively. Over follow-up, the proportion of patients with flares according to each definition were: 28.4% (r-SFI), 24.7% (SLE-DAS) and 13.4% (SLEDAI-2K). The r-SFI flares were moderate in 28.9% and severe in 9.6% of the cases. From all patients, 54.1% maintained stable glucocorticoid-free control of the disease, without flares during follow-up. In the multivariate models, the following parameters were independent predictors of flare, as defined by any of the definitions (table 1): anti-RNP+; oral glucocorticoids; immunosuppressants.

Conclusion: Patients attaining LLDAS but requiring ongoing treatment with immunosuppressants and/or glucocorticoids present a higher risk of flare and thus might need a tighter clinical monitoring. Anti-RNP+ was newly identified as a potential biomarker for higher risk of flares. Glucocorticoid-free, stable low disease activity is an achievable target.

REFERENCES:

Mathian A, Pha M, Haroche J, Cohen-Aubart F, Hié M, Pineton de Chambrun M, et al. Withdrawal of low-dose prednisone in SLE patients with a clinically quiescent disease for more than 1 year: a randomised clinical trial. Ann Rheum Dis. 2020;79(3):339-46.

Inês L, Duarte C, Silva RS, Teixeira AS, Fonseca FP, da Silva JA. Identification of clinical predictors of flare in systemic lupus erythematosus patients: a 24-month prospective cohort study. Rheumatology (Oxford). 2014;53(1):85-9.

112 - HOW ARE WE TREATING PATIENTS WITH FRAGILITY FRACTURES? - ASSESSMENT OF PATIENTS OBSERVED AT EMERGENCY DEPARTMENT FOR FRACTURES

Catarina Soares[1], Soraia Azevedo[1], Francisca Guimarães[1], Hugo Parente[1], Joana Ramos Rodrigues[1], Daniela Peixoto[1], José Tavares-Costa[1], Filipa Teixeira[1], Carmo Afonso[1], Daniela Santos-Faria[1]

[1]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal

Background: Fragility fractures (FF) are the main clinical consequence of osteoporosis (OP). OP and FF represent a costly human and socioeconomic burden in all regions of the world with an impact on health condition with morbidity, decreased quality of life and mortality. Several predictive factors have been associated with an increased risk of fracture. A first FF is one of those risk factors, particularly in the 2 first years after the fracture. Despite the effectiveness of available therapeutic options to prevent osteoporotic fractures, they are underused and only a small number of patients are successively referred for therapy.

Objective: To assess the treatment rate in patients who had a FF observed at the Emergency Department (ED) and the predictors for subsequent use of anti-osteoporotic agents (AOA).

Methods: Retrospective monocentric study that included patients with a FF (wrist, hip and vertebrae), observed at the ED in a tertiary center between 1st January 2017 and 31st December 2018. The search for fractures was performed through ICD9 codes and clinical data was reviewed until 31th December 2020. We excluded totally dependent patients or in palliative care, peri-prosthetic fractures and patients with osteometabolic diseases other than osteoporosis. We identified 1673 FF and after calculating a representative sample (90% confidence interval) 457 patients were included. The prescription of AOA and dual-energy X-ray absorptiometry (DEXA) were analyzed. To identify predictors of AOA prescription we performed a multivariate analysis including variables with a significant association in univariate analysis. SPSS was used for statistical analysis and significance level was defined as 2-sided $p<0.05$.

Results: 172 patients with hip fracture, 173 with wrist fracture and 112 with vertebral fracture were included. Most patients were women (79.9%) with a mean age of 77.6 (SD=10.3) years-old at the time of the fracture. Among these patients, 30.6% had already had a previous FF, 16% of patients had a previous DEXA and 7.4% were or had been on AOA.

After the FF, 8.1% of patients did a DEXA and 7.4% started AOA or kept what they were already taking. During the follow-up time, 16.6% of patients had another FF within an average of 1.7 (SD=0.7) years later. Of these, 19.7% had more than one subsequent fracture.

Neurological disorders and the need for hospitalization at the time of the FF were associated with lesser AOA implementation ($p=0.007$ and $p<0.001$, respectively). In turn, we found an association between AOA use after FF and a previous treatment with AOA ($p<0.001$), previous diagnosis of osteoporosis ($p<0.001$), getting a DEXA after the fracture ($p<0.001$),

TABLE. SOCIODEMOGRAPHIC AND CLINICAL CHARACTERISTICS OF THE STUDY POPULATION AT BASELINE

Age at the time of the fracture, years (mean (SD))		77.6 (10.3)
Gender %(n/N)		Female: 79.9% (365/457)
Previous fragility fracture % (n/N)		30.6% (140/457)
Dyslipidemia % (n/N)		60.4% (276/457)
Arterial Hypertension % (n/N)		70.7% (323/457)
Diabetes Mellitus % (n/N)		24.9% (114/457)
Others comorbidities % (n/N)	Inflammatory Rheumatologic disease Hematologic disease Pulmonary disease Neurological disease Cardiac disease Gastrointestinal disease Psychiatric disease Chronic kidney disease Neoplasia	1.8% (8/457) 6.6% (30/457) 15.3% (70/457) 33% (151/457) 32.4% (148/457) 5.9% (27/457) 36.8% (168/457) 7.9% (36/457) 9.8% (45/457)
Need for hospitalization % (n/N)		38.7% (177/457)
Previous bone density scanning % (n/N)		16% (73/457)
Diagnosis of osteoporosis % (n/N)		8.1% (37/457)
Anti-osteoporotic treatment % (n/N)		7.4% (34/457)

SD: standard deviation

use of corticosteroid therapy (p=0.032), a coexisting inflammatory rheumatic disease (p=0.001) and dyslipidemia (p=0.018).

After adjustment for age and gender, the main predictors of use of AOA after a FF were the previous use of anti-osteoporotic treatment (p=0.001), getting a DEXA after the fracture (p<0.001), a coexisting inflammatory rheumatic disease (p=0.006) and dyslipidemia (p=0.015).

Conclusions: The current study provides evidence that individuals who experience fragility fractures are not adequately managed for osteoporosis. Similarly, bone density scanning in these patients has not been widely done. It is necessary to raise global awareness of the prevention, diagnosis and treatment of osteoporosis. The early treatment of these high-risk patients might decrease the disease burden of osteoporosis.

122 - MOLECULAR PROFILING OF RADIOGRAPHIC AXIAL SPONDYLOARTHRITIS PATIENTS REVEALS AN ASSOCIATION BETWEEN INNATE AND ADAPTIVE CELL POPULATIONS AND THERAPEUTIC RESPONSE TO ADALIMUMAB

Daniel Sobral[1], F.R. Fernandes [2], Atlas Sardoo[3], Miguel Bernardes[4], Patrícia Pinto[5], Helena Santos[6], João Lagoas Gomes[7, 8], José Tavares-Costa[9], JAP da Silva[10], João Madruga Dias[11], Alexandra Bernardo[5], Jean-Charles Gaillard[12], Jean Armengaud [12], Vladimir Benes[13], Rita Pinheiro Torres [8, 14], Domingues, L[8], Sara Maia[8], Jaime C. Branco[7, 8, 15], Coelho AV[2], Fernando Pimentel-Santos[8, 14, 15]

[1]UCIBIO, DCV, Faculdade Ciências e Tecnologia - Universidade NOVA de Lisboa, Caparica, Portugal, [2]Instituto de Tecnologia Química e Biológica (ITQB), Lisboa, Portugal,

[3]Universidade de Trás-os-Montes e Alto Douro, Vila Real, Portugal,
[4]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal,
[5]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal,
[6]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal,
[7]Rheumatology Department, Centro Hospitalar Lisboa Ocidental, Lisboa, Portugal,
[8]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal,
[9]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal,
[10]Serviço de Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal,
[11]Reumatologia, Centro Hospitalar Médio Tejo, Torres Novas, Portugal,
[12]Laboratoire Innovations technologiques pour la Détection et le Diagnostic (Li2D), Service de Pharmacologie et Immunoanalyse (SPI), Centre d énergie atomique, Avignon, France,
[13]EMBL, Heidelberg, Germany,
[14]Serviço de Reumatologia, Centro Hospitalar Lisboa Ocidental | Hospital Egas Moniz, Lisboa, Portugal,
[15]NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

Background: The response to treatment in spondylarthropaties is heterogeneous, due to factors yet to be better described. For that reason, it is important to find tools that might help clinicians to decide what is the best available therapeutic option for each patient.
Objectives: The goal of this study is to use comprehensive molecular profiling to characterize clinical response to therapy in a real-world setting. Specifically, to identify molecular biomarkers differentiating good responders and non-responders to TNF inhibitors (TNFi) treatment, using adalimumab, in radiographic axial spondyloarthritis | ankylosing spondylitis (r-axSpA|AS) patients context.
Methods: Whole-blood mRNA and plasma proteins were measured in a cohort of biologic naïve r-axSpA|AS patients (n = 35) from the Bioefficacy study (Biomarkers identification of anti-TNF alpha agent efficacy in AS patients using RNA sequencing and mass spectrometry), pre and post (14 weeks) TNFi treatment using adalimumab. Response to treatment was categorized according to ASAS20. Results of differential expression analysis were used to identify the most enriched pathways and in predictive models to distinguish responses to TNFi.
Results: A treatment-related signature, independent of the type of response, suggests a reduction in inflammatory disease activity. We found genes and proteins robustly differentially expressed between baseline and week 14 in responders, including the GWAS AS-associated genes TNFRSF1A, FCGR2A, TYK2, TBKBP1, IL1R1, IL6R, ICOSLG, IL7R, HHAT and LTBR. Moreover, CRP and HP proteins showed strong and early decrease in the plasma of AS patients, while a cluster of apolipoproteins (APO1, APO2, APO3) showed an increased expression at week 14. Good responders to TNFi treatment tend to have higher expression of innate immunity genes at baseline, and lower expression of markers associated with adaptive immunity, particularly B-cells. A logistic regression model incorporating ASDAS-CRP, gender and Gene x, the top differentially expressed gene at baseline between responders and non-responders, enabled an accurate prediction of response to adalimumab in our cohort (AUC=0.97).
Conclusion: Differences in disease activity and/or innate/adaptive immune cell type composition at baseline may be a major contributor to response to adalimumab in r-axSpA|AS. Alternatively, a model including clinical and gene expression variables could be considered, particularly in patients with mild disease activity.

125 - CLINICAL COURSE AND RISK FACTORS FOR SEVERE/CRITICAL COVID-19 IN PATIENTS WITH RHEUMATIC DISEASES – A MULTICENTER, NATIONWIDE STUDY

Sofia C Barreira[1, 2], Ana Rita Cruz-Machado[1, 2], Matilde Bandeira[1, 2], Catarina Duarte[2], Maria Seabra Rato[3], Bruno Miguel Fernandes[3], Salomé Garcia[3], Filipe Oliveira Pinheiro[3], Miguel Bernardes[3], Nathalie Madeira[4], Cláudia Miguel[4], Rita Pinheiro Torres [5], Ana Bento da Silva[5], Jorge Pestana Lopes[6], Diogo Esperança Almeida[7], Carolina Mazeda[8, 9], Filipe Cunha Santos[10], Patrícia Pinto[11], Marlene Sousa[12], Hugo Parente[13], Graça Sequeira[14], Maria José Santos[6], JE Fonseca[1, 2], Vasco C Romão[1, 2]

[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Hospital de Santa Maria, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal,
[2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade

de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal,
[3]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal,
[4]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal,
[5]Serviço de Reumatologia, Centro Hospitalar Lisboa Ocidental | Hospital Egas Moniz, Lisboa, Portugal,
[6]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal,
[7]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal,
[8]Serviço de Reumatologia, Centro Hospitalar do Baixo Vouga, E.P.E., Aveiro, Portugal,
[9]Instituto de Biomedicina – iBiMED, Universidade de Aveiro, Aveiro, Portugal,
[10]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal,
[11]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal,
[12]Serviço de Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal,
[13]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal,
[14]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal

Background/Purpose: Since the beginning of the COVID-19 pandemic, some studies have addressed risk factors for severe forms of the disease in patients with rheumatic diseases. Higher prednisolone doses and rituximab (RTX) have been associated with an increased risk of severe COVID-19, although not consistently. Our main goal was to assess the clinical features and to identify risk factors for severe COVID-19 in patients with rheumatic diseases.

Methods: Multicenter observational nationwide study of adult patients with rheumatic diseases and confirmed or suspected infection by SARS-CoV-2 prospectively-followed in the Rheumatic Diseases Portuguese Register – Reuma.pt – between march and september 2020. Mild COVID-19 was defined as symptomatic disease without pneumonia; moderate disease if clinical signs of pneumonia without hypoxemia; severe disease as hypoxemic pneumonia and/or need for hospitalization; and critical disease as requiring admission to intensive care unit or death. Clinical features and treatment of patients with mild/moderate vs. severe/critical COVID19 were compared using Pearson Chi-Square, exact Fisher and Mann-Whitney U tests, as adequate.

TABLE. COMPARISON OF PATIENTS WITH SEVERE/CRITICAL VS MILD/MODERATE COVID19

	Severe/critical COVID19 (n=45)	Mild/moderate COVID19 (n=134)	Univariate analysis p-value	Multivariate analysis OR (95%CI); p-value)
Female, N (%)	33 (73.3)	104 (77.6)	0.548	0.73 (0.28-1.88); p=0.510
Age, mean±SD, in years	67.8±14.1	52.0±12.6	**<0.001**	**1.09 (1.06-1.13); p<0.001**
Caucasian, N (%)	39 (86.7)	111 (82.8)	0.166	
Rheumatic disease, N (%)			**0.039**	
Non-inflammatory	8 (17.8)	9 (6.7)		2.01 (0.57-7.12); p=0.280
Inflammatory arthropathy	22 (48.9)	89 (66.4)		
CTD and vasculitis	15 (33.3)	36 (26.9)		1.92 (0.76-4.85); p=0.170
Disease duration, median (IQR), in years	9.7 (14.3)	6.5 (10.0)	0.092	
Disease activity before COVID-19, N (%)			0.270	
Remission	18 (40.0)	43 (32.1)		
Low	13 (28.9)	69 (51.5)		
Moderate	5 (11.1)	16 (11.9)		
High	4 (8.9)	6 (4.5)		
Comorbidities, N (%)				
Obesity	11 (24.4)	27 (20.1)	0.534	
Arterial hypertension	21 (46.7)	28 (20.9)	**0.001**	
Diabetes	10 (22.2)	3 (2.2)	**<0.001**	
Cardiovascular disease	8 (17.8)	6 (4.5)	**0.007**	
Chronic kidney disease	4 (8.9)	2 (1.5)	**0.036**	
Cerebrovascular disease	1 (2.2)	5 (3.7)	0.627	
Asthma	2 (4.4)	0 (0.0)	0.062	
Chronic obstructive pulmonary disease	1 (2.2)	5 (3.7)	0.627	
Interstitial lung disease	1 (2.2)	1 (0.7)	0.441	
Hyperuricemia	1 (2.2)	2 (1.5)	0.573	
Ever smoked	9 (20.0)	5 (3.7)	0.741	
Malignancy	4 (8.9)	4 (3.0)	0.111	
Comorbidity index ≥1*	29 (64.4)	48 (35.8)	**0.001**	1.50 (0.65-3.48); p=0.345
Medications prior to COVID				
Corticosteroids	18 (40.0)	53 (39.6)	0.958	
NSAIDs	2 (4.4)	28 (20.9)	**0.010**	
Methotrexate	14 (31.1)	51 (38.1)	0.475	
Sulphasalazine	4 (8.9)	11 (8.2)	0.887	
Leflunomide	2 (4.4)	9 (6.7)	0.733	
Hydroxychloroquine	6 (13.3)	20 (14.9)	0.793	
Azathioprine	1 (2.2)	5 (3.7)	0.627	
Mycophenolate mofetil	0 (0.0)	1 (0.7)	0.561	
JAK inhibitors	0 (0.0)	2 (1.5)	0.410	
TNF inhibitors	1 (2.2)	23 (17.2)	**0.010**	
Tocilizumab	0 (0.0)	5 (3.7)	0.333	
Ustekinumab	0 (0.0)	1 (0.7)	0.561	
Rituximab	5 (11.1)	2 (1.5)	**0.012**	**13.77 (2.23-85.17); p=0.005**
Belimumab	0 (0.0)	1 (0.7)	0.561	

*At least 1 comorbidity among arterial hypertension, obesity, diabetes, cardiovascular disease, asthma, chronic obstructive pulmonary disease, chronic kidney disease.
CTD – connective tissue disease, JAK – janus kinase; NSAIDs – nonsteroidal anti-inflammatory drugs; TNF – tumor necrosis factor

Independent association between demographic, disease-related and treatment-related variables and COVID-19 severity was evaluated through multivariate logistic regression.

Results: We included 179 patients with confirmed (97.2%) or suspected (2.8%) COVID-19 (Table 1). Forty-five (25.1%) patients developed severe/critical disease, requiring hospitalization, and 10 (5.6%) patients died. In the same time frame, the national rate of hospitalization was 8.4% and mortality 2.6%. Most patients reported direct contact with a positive patient (65.9%). Major symptoms reported were cough, fever, malaise, fatigue, myalgia, headache and anosmia. Lymphopenia and elevated C-reactive protein were the

most frequent laboratory abnormalities. Patients with severe/critical disease were older (p<0.001), had a higher prevalence of arterial hypertension (p=0.001), diabetes (p<0.001), cardiovascular disease (p=0.007) and chronic kidney disease (p=0.036) (Image 1). Considering inflammatory arthritis as the reference category, patients with CTD/vasculitis (33.3% vs 26.9%, OR 1.69, 95%CI 0.79-3.61) and non-inflammatory diseases (17.8% vs 6.7%, OR 3.60, 95%CI 1.25-10.39) had a higher probability of severe/critical COVID-19. Regarding therapy, the proportion of patients under RTX was higher in severe/critical COVID-19 patients (11.1% vs 1.5%, OR 8.25, 95%CI 1.54-44.16). Treatment with tumor necrosis factor inhibitors was associated with less probability of severe/critical COVID-19 (2.2% vs 17%, OR 0.11, 95%CI 0.01-0.84). Treatment with other disease modifying anti-rheumatic drugs was not associated with COVID-19 severity. Age (OR 1.09, 95%CI 1.06-1.13, p<0.001) and treatment with RTX (OR 13.77, 95%CI 2.23-85.17, p=0.005) were independently associated with severe/critical COVID-19 (Image 1).

Conclusion: Hospitalization and mortality in rheumatic patients was higher than the reported in the general national population for the same time frame, although a reporting bias must be considered. Older age and treatment with rituximab were independent risk factors for severe/critical COVID-19 in patients with rheumatic diseases.

132 - ANTIBODY RESPONSE AFTER SARS-COV-2 INFECTION IN PATIENTS WITH RHEUMATIC DISEASES: A MULTICENTER, NATIONWIDE STUDY

Ana Rita Cruz-Machado[1, 2], Sofia C Barreira[1, 2], Marc Veldhoen[3], Matilde Bandeira[1, 2], Catarina Duarte[2], Maria Seabra Rato[4], Bruno Miguel Fernandes[4], Salomé Garcia[4], Filipe Oliveira Pinheiro[4], Miguel Bernardes[4], Nathalie Madeira[5], Cláudia Miguel[5], Rita Pinheiro Torres [6], Ana Bento da Silva[6], Carolina Mazeda[7, 8], Filipe Cunha Santos[9], Marlene Sousa[10], Hugo Parente[11], Maria José Santos[12], JE Fonseca[1, 2], Vasco C Romão[1, 2]

[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal,
[2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal,
[3]Instituto de Medicina Molecular João Lobo Antunes, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal,
[4]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal,
[5]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal,
[6]Serviço de Reumatologia, Centro Hospitalar Lisboa Ocidental | Hospital Egas Moniz, Lisboa, Portugal,
[7]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal,
[8]Instituto de Biomedicina – IBiMED, Universidade de Aveiro (Aveiro), Aveiro, Portugal,
[9]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal,
[10]Serviço de Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal,
[11]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal,
[12]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal

Background: The development and duration of humoral immunity after SARS-CoV-2 natural infection remains of interest. For the general population, available data suggest a robust immune response, able to protect against reinfection for at least 6-8 months. As for the subgroup of patients with rheumatic and musculoskeletal diseases (RMDs), information is lacking. Considering that immunosuppression might preclude an adequate anti-viral response, we aimed to assess the rate of seroconversion after natural infection in patients with RMDs and to find factors that may influence antibody response.

Methods: Multicenter observational study of patients with RMDs prospectively-followed in the Rheumatic Diseases Portuguese Register – Reuma.pt –in the first 6 months of the pandemic in Portugal – March to September 2020. We included all patients with inflammatory and non-inflammatory RMDs with confirmed (PCR-positive) or suspected COVID-19. Patients were asked to collect a blood sample for antibody testing against SARS-CoV-2 at least 3 months after the resolution of infection. All samples were processed in a single center. IgG antibodies recognizing the SARS-CoV-2 receptor-binding domain (RBD) were quantified using ELISA. Seroconversion was assumed for any titer ≥1:50.

Results: Out of 179 included patients, 79 (44%) performed antibody testing. Of these, 65 (82%) had inflammatory RMDs and 14 (18%) had non-inflammatory RMDs – described in Table 1. Blood samples were

TABLE I. DEMOGRAPHIC AND CLINICAL DATA OF PATIENTS WITH INFLAMMATORY AND NON-INFLAMMATORY RMDS WHO PERFORMED SEROLOGY

	Inflammatory arthritis[1] N, %	CTD/Vasculitis[2] N, %	Non-inflammatory diseases[3] N, %	Total N, %	p value
N,%	40, 51	25, 32	14, 18	79, 100	
Age (y) Median (IQR)	53 (16)	58 (15)	61 (14)	57 (14)	**0.026**
Female N,%	26, 65	22, 88	13, 93	61, 77	**0.03**
Treatment[4] (N, %)					
Corticosteroids	17, 43	12, 48	1, 7	30, 38	**0.029**
Azathioprine	1, 3	3, 12	0, 0	4, 5	0.150
Hydroxyxhloroquine	1, 3	11, 44	1, 7	13, 17	**<0.001**
Leflunomide	3, 8	1, 4	0, 0	4, 5	0.522
Mycophenolate	0, 0	1, 4	0, 0	1, 1	0.335
Methotrexate	16, 40	10, 40	1, 7	27, 34	0.063
Sulfassalazine	3, 8	0, 0	0, 0	3, 4	0.219
Belimumab	0, 0	1, 4	0, 0	1, 1	0.335
Rituximab	1, 3	1, 4	0, 0	2, 3	0.735
Tocilizumab	2, 5	0, 0	0, 0	2, 3	0.368
TNFi	9, 23	0, 0	0, 0	9, 11	**0.007**
Ustekinumab	1, 3	0, 0	0, 0	1, 1	0.610
COVID-19 severity (N, %)					0.425
Asymptomatic[5]	5, 13	3,12	0, 0	8, 10	
Mild[6]	8, 20	2, 8	2, 14	12, 15	
Moderate[7]	21, 53	15, 60	7, 50	43, 54	
Severe[8]	6, 15	4, 16	5, 36	15, 19	
Critical[9]	0, 0	1,4	0,0	1,1	
Timing of serology[10] (days) Median (IQR)	256 (123)	225 (161)	234 (65)	237 (125)	0.543
IgG+ (N,%)	33, 83	23, 92	14, 100	70, 89	0.168
IgG Titers (GM±GSD) (min.-max)	1/1436±3.96 (1/100-1/25600)	1/1367±4.262 (1/100-1/25600)	1/1903±4.327	1/1508±4.075	0.051

collected between days 89 and 331 (median time 237, IQR 125 days) after symptom onset or positive PCR test (if asymptomatic). Seventy (89%) patients had positive IgG antibodies, with a geometric mean titer of 1/1508±4.075 (min 1/100- max 1/25600).

No differences were seen in the seroconversion rate between patients with and without inflammatory RMDs. Disease activity status at the time of the infection also did not influence seroconversion. Although

1 – Includes rheumatoid arthritis, psoriatic arthritis (PsA), spondyloarthritis other than PsA, RS3PE, undifferentiated arthritis and microcrystalline arthritis, adult onset Still disease. 2 – Includes systemic lupus erythematosus, undifferentiated connective tissue disease, mixed connective tissue disease, systemic sclerosis, Sjögren syndrome, giant cell arteritis, Behçet disease. 3 – Fibromyalgia, osteoarthritis, osteoporosis, Paget bone disease. 4 – treatment before infection; only DMARDs and corticosteroids were considered; 5 – reference for statistical analysis; 6 - symptomatic disease without evidence of pneumonia; 7 - clinical signs of pneumonia but room-air SpO2 ≥ 90%; 8 - hypoxemic pneumonia and/or need for hospitalization; 9 - requiring admission to intensive care unit or death. 10 – relative to symptom onset or first positive PCR test if asymptomatic. CTD – connective tissue diseases; GM: geometric mean; GSD: geometric SD factor; i: inhibitors; y: years.

TABLE II. DIFFERENCES BETWEEN PATIENTS WHO DEVELOPED IgG ANTIBODIES AND THOSE WHO DID NOT AND BINARY LOGISTIC REGRESSION MODEL FOR PREDICTION OF IgG DEVELOPMENT

	IgG+ (N, %)	IgG – (N, %)	Univariate analysis (p value)	Multivariate analysis OR (95% CI); p value
N,%	70, 89	9, 11		
Age (y) median (IQR)	58 (14)	48 (21)	0.649	1.01 (0.94-1.07); 0.885
Female, N,%	55, 79	6, 67	0.418	1.09 (0.17-7.10); 0.930
Rheumatic diseases: N,%			0.168	
Inflammatory arthritis[1]	33, 47	7, 78		
CTD/Vasculitis	23, 33	2, 22		
Non-inflammatory	14, 20	0, 0		
Disease activity before infection: N,%			1.000	2.29 (0.18-28.81); 0.521
Remission/low	54, 84	8, 89		
Moderate/high	10, 16	1, 11		
Treatment: N, %				
Corticosteroids	30, 43	0, 0	**0.011**	
Azathioprine	4, 6	0, 0	1.000	
Hydroxychloroquine	12, 17	1, 11	1.000	
Leflunomide	4, 6	0, 0	1.000	
Mycophenolate	1, 1	0, 0	1.000	
Methotrexate	24, 34	3, 33	1.000	
Sulfassalazine	3, 4	0, 0	1.000	
Belimumab	1, 1	0, 0	1.000	
Rituximab	2, 3	0, 0	1.000	
Tocilizumab	2, 3	0, 0	1.000	
TNF inhibitors	6, 9	3, 33	0.062	0.13 (0.02-0.91); **0.041**
Ustekinumab	1, 1	0, 0	1.000	
Comorbidities:				
Hypertension	22, 31	1, 11	0.271	
Diabetes mellitus	5, 7	1, 11	0.528	
Cardiovascular disease[2]	6, 9	1, 11	0.586	
Obesity	21, 30	2, 22	1.000	
COVID-19 severity: (N, %)			**0.007**	
Asymptomatic[3]	4, 6	4, 44		15.15 (2.33-98.48); **0.004**
Mild	11, 16	1, 11		
Moderate	39, 56	4, 44		
Severe	15, 21	0, 0		
Critical	1, 1	0, 0		
Timing from symptoms to sampling (days) Median (IQR)	236 (126)	239 (105)	0.875	

1 –reference for statistical analysis; 2 – includes personal history of heart failure and/or ischemic cardiopathy. 3 - reference for statistical analysis. CTD: connective tissue diseases.

DMARD therapy didn't influence seropositivity, the proportion of patients under TNF inhibitors (TNFi) was numerically higher in patients who did not develop IgG antibodies (33.3% vs 8.6%, p=0.062). Of note, all patients treated with corticosteroids (N=30) and rituximab (N=2) developed antibodies. There was no correlation between sample timing and RBD IgG titers. On multivariate analysis, treatment with TNFi (OR 0.13, 95%CI: 0.02-0.91, p=0.041), and symptomatic COVID-19 (OR 15.1, 95%CI 2.33-98.48; p=0.004) were the only variables independently associated with serological response (Image 1).

Conclusion: Most patients with rheumatic diseases developed IgG antibodies against SARS-CoV-2, with medium-to-high titers detected between 3 to 11 months after natural infection. In this population, treatment with TNFi decreased the odds of seroconversion while symptomatic COVID-19 was associated with a higher likelihood of developing a humoral immune response.

258 - EFFECTIVENESS OF A BIOLOGIC TAPERING PROTOCOL FOR RHEUMATOID ARTHRITIS, PSORIATIC ARTHRITIS AND AXIAL SPONDYLOARTHRITIS

Pedro Ávila Ribeiro[1,2], Fernando Saraiva[1,2], Patrícia Martins[1,2], Ana Rita Cruz-Machado[1,2], Maria João Saavedra[1,2], JE Fonseca[1,2], Elsa Vieira-Sousa[1,2]

[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Hospital de Santa Maria, CHLN, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Aim: To investigate in rheumatoid arthritis (RA), psoriatic arthritis (PsA) and axial spondyloarthritis (axSpA) patients in remission, submitted to a progressive biologic tapering protocol, how many maintain remission by 1 year. To assess, as secondary aims, the proportion of successful dose reduction, disease flare rate, elapsed time until flare and outcome after flare, in this population.

Materials and Methods: Patients diagnosed with RA, PsA and axSpA, treated with tumor necrosis factor inhibitors (TNFi), in sustained Disease Activity Score (DAS) 28 remission (DAS28<2.6, if RA or PsA with predominantly peripheral disease) or Ankylosing Spondylitis Disease Activity Score (ASDAS) low disease activity (LDA ASDAS<2.1, if axSpA or PsA with predominantly axial disease), for over 12 months, were asked to increase the regular interval (RI) between TNFi administration by 50% (1.5xRI). If remission was maintained after 6 months, dosing interval was increased again to twice the regular interval (2xRI). Disease activity and patient-reported outcomes were assessed every 3 months; and function, health-related quality of life, peripheral joint ultrasound, radiographs, serum drug levels and anti-drug antibodies were assessed at baseline and 1 year. In case of disease flare (defined as DAS28>2.6/ ASDAS>2.1), the previous treatment frequency was reintroduced by protocol.

Results: 16 RA, 15 PsA and 18 axSpA patients underwent baseline assessment. 11 patients were excluded before they started tapering (9 had clinical flares – 7 of whom were RA patients, 1 had subclinical Doppler+ synovitis on ultrasound, 1 was pregnant). The remaining 38 patients initiated tapering and had a mean follow-up (±SD) of 21.1(±7.6) months. 1 year after ta-

TABLE I. BASELINE DEMOGRAPHIC AND DISEASE CHARACTERISTICS OF THE STUDY POPULATION

Characteristics	RA (n=9)	PsA (n=13)	AxSpA(n=16)
Age, years, mean (SD)	54.3 (9.9)	55.1 (8.4)	50.2 (11.7)
Male gender, n (%)	1 (11.1)	13 (100)	13 (81.3)
Predominant axial involvement, n (%)	-	3 (23.1)	16 (100)
Enthesitis, n (%)	-	10 (76.9)	8 (50.0)
Disease activity, mean (SD) Axial (ASDAS) Peripheral (DAS28)	 - 1.99 (0.35)	 0.93 (0.40) 1.20 (0.52)	 1.31 (0.50) -
Physical function, median (IQR) BASFI HAQ	 - 0.06 (0.13)	 0.35 (0.30) 0.00 (0.00)	 1.55 (3.25) -

ASDAS: Ankylosing Spondylitis Disease Activity Score; axSpA: axial spondyloarthritis; BASFI: Bath Ankylosing Spondylitis Functional Index; DAS28: 28-joint Disease Activity Score; HAQ: Health Assessment Questionnaire; IQR: interquartile range; PsA: psoriatic arthritis; RA: rheumatoid arthritis; SD: standard deviation.

pering, 15 patients were taking TNFi at 2xRI (46.9%), 8 at 1.5xRI (25.0%) and 9 at 1xRI (28.1%). 21 patients had a clinical flare (55.3%) after 8.8(±5.9) months, 12 when taking 1.5xRI (57.1%), 8 on 2xRI (38.1%) and 1 on >2xRI (4.8%) due to an intercurrent infection. After flare, 4 patients (19.0%) recovered remission/LDA with no increase in treatment frequency (increase in disease activity score deemed not related with true disease activity), 3 (14.3%) after increasing treatment frequency to 1.5xRI and 13 patients (61.9%) to 1xRI. 1 patient had to switch TNFi (4.8%).

Conclusion: Most patients on sustained remission/ /LDA tolerated tapering TNFi with no need to increase treatment frequency. In the event of flare, resuming treatment frequency allowed to retrieve a remission/LDA status in all but one patient. Disease flares were frequent even before tapering, despite previously sustained remission, especially in RA.

ACKNOWLEDGMENTS:
TaPRA and TaSpA teams: Inês Cordeiro, Joaquim Pereira, Nikita Khmeliskii, Rita Barros, Rita Machado, Maria João Saavedra, Joana Martinho, Eduardo Dourado, Teresa Melo, Susana Capela, Ana Margarida Monteiro, Augusto Silva, Manuel António, Sofia Barreira, Carla Macieira, Vasco Romão, Rui Lourenço Teixeira, Ana Valido, Cristina Ponte.

264 - A FRACTURE LIAISON SERVICE IMPLEMENTATION AFTER TWO YEARS: A RETROSPECTIVE COHORT STUDY.

Susana P. Silva[1], Carolina Mazeda[1], João Paulo Vilas-Boas[1], Bernardo Santos[1], Graça Costa[1], Maria Céu Morais[1], Ana Filipa Rocha Águeda[2], Gisela Eugénio[1], Anabela Barcelos[1]
[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal,
[2]Reumatologia, Centro Hospitalar Universitário Cova da Beira, Covilhã, Portugal

Background: Osteoporosis (OP) is a major public health problem, which as major economic and social consequences1. It is important to improve identification, diagnosis and treatment of OP. A fragility fracture (FF) is a very important event for patients and their family, leading to loss of quality of life and an increase in mortality2. Implementation of a Fracture Liaison Service (FLS) demonstrated effects in fracture incidence and economic burden3, and healthcare systems are now beginning to recognise the benefits of FLS.

Objective: This study aims to evaluate outcomes before and after FLS implementation in our hospital.

Material and Methods: We conducted a retrospective study on patients, aged 50 or over, who suffered a fragility fracture between 2017 and 2020, before (pre-FLS) and after (post-FLS) FLS implementation, in our hospital. Patients with a dependence status before fracture and patients who died in orthopaedic ward after surgery, were excluded. Demographic, comorbidities, and clinical results were collected from the hospital database. Descriptive analysis used medians and interquartile range (IQR) for continuous data as frequencies and percentages for qualitative variables. Nonparametric tests were used for statistical analysis, with p value ≤ 0.05, with SPSS® software.

Results: A total of 611 patients were included, which 424 were evaluated before and 187 after the FLS implementation (Table 1). Gender and status of dependence before fracture were similar between groups. Age at fracture evaluation (p = 0.002), previous fractures and previous OP treatment were different between patients observed pre-FLS and post-FLS implementation (Table 1). There were more smokers in patients observed in our FLS, but the prevalence of diabetes and thyroid diseases were similar. Furthermore, the type of fracture was also statistically different, although most fracture were trochanteric (62.0% vs 55.1%), followed by vertebral (8.3% vs 13.4%), subcapital (11.3% vs 5.3%), malleolar (4.7% vs 8.6%), femur diaphysis (4.2% vs 2.1%), humeral (3.5% vs 5.3%) and wrist (2.6% vs 2.7%) fracture. Anti-osteoporotic drugs were more frequently prescribed post-FLS implementation (64.7% versus 4.7%; p<0.001). A new fracture occurred in 6 (3.2%) patients post-FLS implementation and 92 (21.7%) patients pre-FLS implementation (p<0.001). Most patients were functionally independent after the fracture (52.6% vs 50.4%). Forty-six (10.8%) and eleven (5.9%) patients died after suffered a FF pre-FLS and post-FLS implementation, respectively.

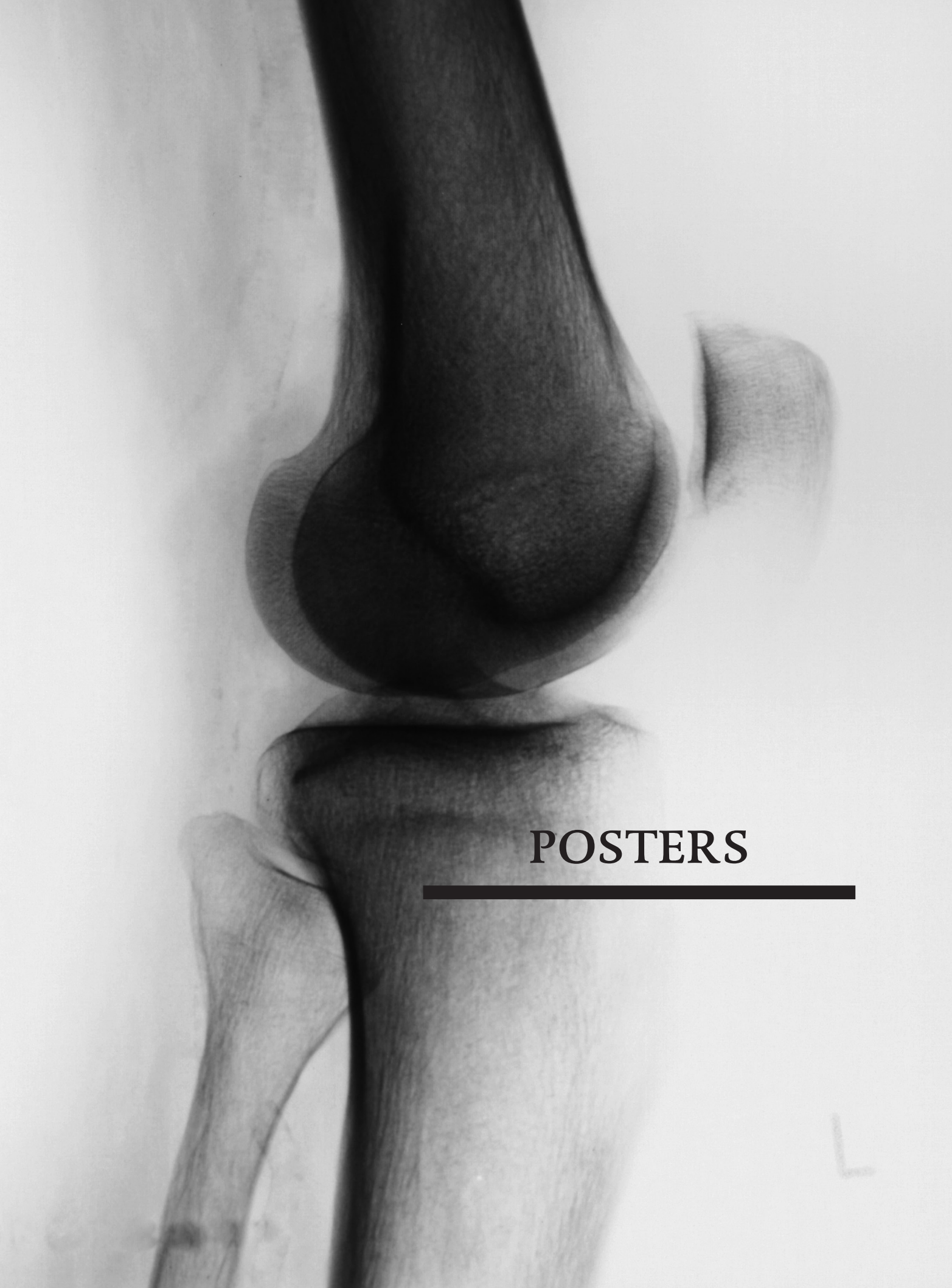
Conclusions: Secondary prevention of osteoporotic fractures has improved since the implementation of the FLS. Anti-osteoporotic drugs prescribing was higher in patients who suffered a FF in post-FLS compared to pre-FLS implementation. However, patient identification and post-fracture assessment timing still need to be improved.

REFERENCES:
1. Bliuc D et al. J Bone Miner Res. 2013;28(11):2317-2324.
2. Kanis J et al. (2018) Broken bones, broken lives: a roadmap to solve the fragility fracture crisis in Europe. International Osteoporosis Foundation.
3. Fuggle NR et al. In: Falaschi P, Marsh D, editors. Orthogeriatrics: The Management of Older Patients with Fragility Fractures. Springer; 2021. Chapter 14.

TABLE I. CLINICAL CHARACTERISTIC OF PATIENTS WITH FRAGILITY FRACTURES OBSERVED BEFORE AND AFTER FRACTURE LIAISON SERVICE IMPLEMENTATION

Clinical characteristics	Pre-FLS 2017-2018	Post-FLS 2019-2020	P value
N 424	187		
Female (%)	355 (83.7%)	158 (84.5%)	0.056
Age at fracture (IQR)	83 (11)	79 (16)	0.002
Status of dependence before fracture			0.094
Partially dependent (Katz 3 and 4)	169 (39.9%)	77 (41.2%)	
Independent (Katz 5 and 6)	255 (60.1%)	110 (58.8%)	
Previous OP treatment	6 (1.4%)	29 (15.5%)	<0.001
Previous fractures	77 (18.2%)	65 (34.2%)	<0.001
Comorbidities			
Current smoker	5 (1.2%)	9 (4.8%)	0.014
Diabetes mellitus	106 (25.3%)	49 (26.2%)	0.056
Thyroid disease	35 (8.3%)	20 (10.7%)	0.331
Alcohol abuse	6 (1.4%)	14 (7.5%)	<0.001
Type of fracture			0.002
Trochanteric fracture	263 (62.0%)	103 (55.1%)	
Subcapital fracture	48 (11.3%)	10 (5.3%)	
Vertebral fracture	35 (8.3%)	25 (13.4%)	
Humeral fracture	15 (3.5%)	10 (5.3%)	
Malleolar fracture	20 (4.7%)	16 (8.6%)	
Femur diaphysis fracture	18 (4.2%)	4 (2.1%)	
Wrist fracture	11 (2.6%)	5 (2.7%)	
Tibia fracture	6 (1.4%)	5 (2.7%)	
Olecranon fracture	5 (1.2%)	2 (1.1%)	
Rib fracture	0 (0.0%)	3 (1.6%)	
Pelvis fracture	1 (0.2%)	0 (0.0%)	
Calcaneus fracture	0 (0.0%)	1 (0.5%)	
Knee-cap fracture	2 (0.5%)	3 (1.6%)	<0.001
Started treatment	20 (4.7%)	121 (64.7%)	<0.001
Status of dependence after fracture			<0.001
Totally dependent (Katz 1 and 2)	28 (6.6%)	30 (16.0%)	
Partially dependent (Katz 3 and 4)	127 (29.9%)	18 (9.7%)	
Independent (Katz 5 and 6)	223 (52.6%)	94 (50.4%)	
New fractures	92 (21.7%)	6 (3.2%)	<0.001
Died	46 (10.8%)	11 (5.9%)	<0.001

Legend: FLS – Fracture Liaison Service; IQR – Interquartile Range.



POSTERS

Trabalho Original

ACTA REUMATOL PORT. 2021:46:??-?? (SUP)

TRABALHO ORIGINAL (COM APRESENTAÇÃO ORAL)

003 - DETERMINANTS OF COVID-19 DISEASE SEVERITY IN PATIENTS WITH UNDERLYING RHEUMATIC DISEASE

Cristiana Sieiro Santos[1], Clara Moriano[1], Carolina Álvarez[1], Elvira Díez[1], Trinidad Sandoval[1]
[1]Reumatología, Complejo Asistencial Universitario de León, León, Spain

Background: Over the month of April, Spain has become the European country with more confirmed cases COVID-19 infection. Studying epidemiological and clinical characteristics of patients with rheumatic diseases infected with SARS-CoV2 is pivotal to clarify determinants of COVID-19 disease severity in patients with underlying rheumatic disease.
Objectives: To describe epidemiological characteristics of patients with rheumatic diseases hospitalized with COVID-19 and determine risk factors associated with mortality in a third level Hospital setting in León, Spain.
Methods: We performed a prospective observational study, from 1st March 2020 until the 1st of December including adults with rheumatic diseases hospitalized with COVID-19 and performed a univariate and multivariate logistic regression model to estimate ORs and 95% CIs of mortality. Age, sex, comorbidities, rheumatic disease diagnosis and treatment and disease activity prior to infection, radiographic and laboratorial results at arrival were analysed.
Results: 4644 patients with COVID-19 were admitted to our hospital, of whom 40 (9%) had a rheumatic or musculoskeletal disease. 53% were women, with a mean age at hospital admission of 75.3 (IQR 68-83) years. The median length of stay was 11 days. A total of 10 patients died (26%) during their hospital admission. Patients who died from COVID-19 were older (median age 78.4 IQR 74.5-83.5) than those who survived COVID-19 (median age 75.1 IQR 69.3-75.8) and more likely to have arterial hypertension (9 [90%] vs 14 [50%] patients; OR 9 (95% CI 1.0-80.8), p 0.049), dyslipidaemia (9 (90%) vs 12 (43%); OR 12 (95% CI 1.33-108), p 0.027), diabetes ((9 (90%) vs 6 (28%) patients; OR 33, p 0.002), interstitial lung disease (6 (60%) vs 6 (21%); OR 5.5 (95% CI 1.16-26), p 0.032), cardiovascular disease (8 (80%) vs 11 (39%); OR 6.18 (95% IC 1.10-34.7) and a moderate/high index of rheumatic disease activity (7 (25%) vs 6(60%); OR 41.4 (4.23-405.23), p 0.04). In univariate analyses, we also found that patients who died from COVID-19 had higher hyperinflammation markers than patients who survived: C-reactive protein (181 (IQR 120-220) vs 107.4 (IQR 30-150; p 0.05); lactate dehydrogenase (641.8 (IQR 465.75 – 853.5) vs 361 (IQR 250-450), p 0.03); serum ferritin (1026 (IQR 228.3 – 1536.3) vs 861.3 (IQR 389 – 1490.5), p 0.04); D-dimer (12019.8 (IQR 843.5 – 25790.5) vs 1544.3 (IQR 619-1622), p 0.04). No differences in sex, radiological abnormalities, rheumatological disease or background therapy, symptoms before admission, duration of stay in hospital between deceased patients and survivors were found. In the multivariate analysis, the following risk factors were associated with mortality: rheumatic disease activity (p=0.003), dyslipidaemia (p=0.01), cardiovascular disease (p=0.02) and interstitial lung disease (p=0.02). Age, hypertension, and diabetes were significant predictors in univariate but not in multivariate analysis.
Conclusions: Our results suggest that comorbidities, rheumatic disease activity and laboratorial abnormalities such as C-reactive protein, D-Dimer, LDH, ferritin elevation significantly associated with mortality whereas previous use of rheumatic medication did not. Inflammation is closely related to severity of COVID-19.

014 - GENDER DIFFERENCES IN CLINICAL FEATURES AND OUTCOMES OF SSC: ANALYSIS OF REUMA.PT/SSC REGISTRY

Raquel Freitas[1], Patrícia Martins[2, 3], Eduardo Dourado[2, 3], Tânia Santiago[4, 5], Francisca Guimarães[6], Bruno Miguel Fernandes[7], Salomé Garcia[7], Beatriz Samões[8], Ana Sofia Pinto[9], Nuno Pina Gonçalves[10, 11], Maria Helena Lourenço[11], Emanuel Costa[12], Margarida Rocha[13], Couto M[14], Ana Catarina Duarte[1], Filipe Araújo[15], Inês Cordeiro[2], Fátima Godinho[1], Catarina Resende[2], Maria João Salvador[5], Ana Cordeiro[1], Maria José Santos[1, 3]
[1]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal
[2]Serviço de Reumatologia e Doenças Ósseas Metabólicas,

Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [3]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [4]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal; [5]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal; [6]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal; [7]Rheumatology Department, Centro Hospitalar de São João, Porto, Portugal; [8]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal; [9]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal; [10]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal; [11]Rheumatology Department, Hospital Egas Moniz (CHLO), Lisboa, Portugal; [12]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal; [13]Rheumatology Department, Centro Hospitalar; Universitário do Algarve, Faro, Portugal; [14]Rheumatology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal; [15]Rheumatology and Osteoporosis Unit, Hospital de Sant´Ana, SCML, Cascais, Portugal

Background: Systemic sclerosis (SSc) is a rare connective tissue disease of unknown aetiology, with a broad spectrum of clinical and laboratory features. Evidence for the role of sex in the clinical manifestations of SSc patients is emerging. Some multicenter cohorts have shown that male SSc patients have more severe disease[1,2].

Objective: To assess the clinical manifestations and survival in the cohort of Portuguese SSc patients according to gender.

Methods: Data from adult SSc patients included in Reuma.pt database up to November 2020 was analysed. Demographic features, SSc subsets, main clinical and immunological features, treatments used and survival data were evaluated and compared between

TABLE 1 – CUMULATIVE CLINICAL AND IMMUNOLOGIC FEATURES AND TREATMENTS USED.

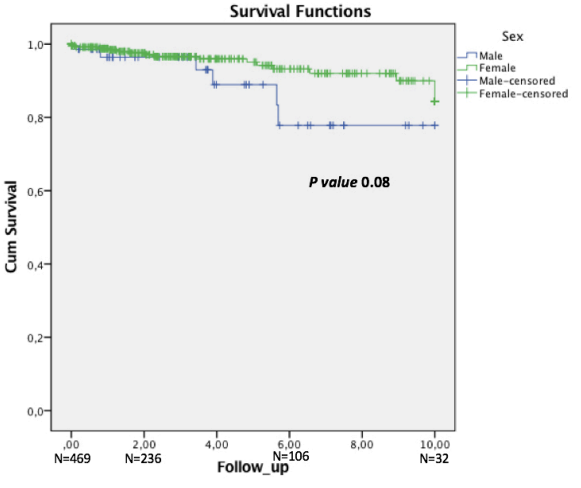
	Whole cohort N=1054	Female N= 922 (87.5%)	Male N=132 (12.5%)	P-value
Limited cutaneous SSc - N (%)	579 (56.3)	525 (58.3)	54 (42.2)	<0.01
Diffuse cutaneous SSc – N(%)	180 (17.5)	136 (15.1)	44 (34.4)	
Skin thickening proximal do MCF/MTF joints– N (%) no= 962	734 (76.3)	634 (75)	147 (83.2)	0.03
Sclerodactyly – N (%) no= 855	547 (64)	473 (63.1)	74 (70.5)	0.14
Raynaud's Phenomenon – N (%) no=1010	943 (93.4)	828 (94)	115 (89.1)	0.04
Musculoskeletal involvement – N(%) no=972	479 (45.4)	410 (45.4)	69 (52.3)	0.09
Myositis – N (%) no=943	54 (5.7)	38 (4.6)	16 (13.3)	<0.01
Cardiac involvement – N (%) no=924	71 (7.7)	65 (8)	6 (5.4)	0.34
Renal involvement – N (%) no= 917	17 (1.9)	13 (1.6)	4 (3.5)	0.17
Gastrointestinal involvement - N(%) no=972	425 (43.7)	362 (42.6)	63 (51.6)	0.06
Gastric involvement – N(%) no=916	114 (12.4)	91 (11.3)	23 (20.4)	<0.01
Pulmonary involvement – N(%) no=915	261 (28.5)	218 (27.2)	43 (37.7)	0.02
Pulmonar arterial hypertension – N(%) no= 871	14 (1.6)	13 (1.7)	1 (0.9)	0.56
Interstitial lung disease – N(%) no=765	218 (28.5)	183 (27.1)	35 (38.9)	0.03
Antinuclear antibodies - N (%) no=1040	934 (89.8)	827 (90.9)	107 (82.3)	0.01
Anti-centromere – N(%) no= 1027	540 (52.6)	509 (56.6)	31 (24.4)	<0.01
Anti-topoisomerase I – N(%) no=1020	214 (21)	174 (19.6)	40 (30.8)	0.01
Anti-RNA polymerase III – N(%) no=710	25 (3.5)	20 (3.3)	5 (5.2)	0.58
Immunomodulators/ Immunossupressants – N(%)	420 (39.8)	363 (39.4)	57 (43.2)	0.40
Calcium channel blockers – N(%)	527 (50)	459 (49.8)	68 (51.5)	0.71
PPIs/Ranitidine/Prokinetics – N(%)	353 (33.5)	309 (33.5)	44 (33.3)	0.97

MCF- metacarpophalageal joints; MTF – metatarsophalangeal joints. Pulmonary arterial hypertension confirmed by right heart catheterisation. Immunomodulators include Methotrexate, Leflunomide, Hydroxychloroquine; Immunosuppressants include Azathioprine, Mycophenolate Mofetil and Cyclophosphamide. P-value comparing female and male.

genders. Survival was calculated for patients included in Reuma.pt within the first two years of diagnosis. Of the 1054 patients included, 716 (68%) fulfilled the ACR/EULAR 2013 classification criteria for SSc.

Results: In total, 1054 adult patients with SSc were included, 132 (12.5%) males. The most common subset was lcSSc (56.3%), followed by dcSSc (17.5%), preclinical SSc (13%), overlap syndrome (9.8%) and 3.3% had SSc sine scleroderma. No differences in demographic features and comorbidities were found between women and men, except for a higher rate of smokers among men (64.5% vs 19.2%; p<0.01). Diffuse cutaneous SSc subtype and antitopoisomerase antibodies were more prevalent in males (Table 1). Raynaud's phenomenon (RP) and skin thickening were the most frequently observed clinical manifestations, RP being more prevalent in females (84.1% vs 94%, p=0.04) and skin thickening proximal to metacarpophalangeal and metatarsophalangeal joints in males (83.2% vs 75%, p=0.03). Additionally, male patients presented significantly more myositis (13.3% vs 4.6%, p<0.01), interstitial lung disease (38.9% vs 27.1%, p=0.03) and gastric involvement (20.4% vs 11.3%, p<0.01). One-third of patients were treated with immunomodulators, with no differences in the pattern of drugs used between genders. During follow-up, more deaths were reported in men (12.1% vs 7.3%, p=0.04). However, the overall 1-, 3- and 5-year survival was not significantly different between men and women (96.4% vs 98.2%, 93% vs 95.9% and 83.4% vs 93.2%, p=0.08, Figure 1).

FIGURE 1 - SURVIVAL FROM DIAGNOSIS OF PATIENTS WITH SSC INCLUDED IN THE COHORT IN THE FIRST TWO YEARS OF DISEASE (N=469)



The X and Y axes represent follow-up time in years and the proportion of patients still on follow-up, respectively.

Conclusion: This study confirms the existence of gender differences in clinical and immunologic features of SSc patients. Although SSc is less common in men, they have a more severe expression of internal organ involvement. Nevertheless, no statistically significant differences were found in survival rates.

REFERENCES

1. Freire M, et al. Clinical and epidemiological differences between men and women with systemic sclerosis: a study in a Spanish systemic sclerosis cohort and literature review. Clin Exp Rheumatol. 2017
2. Peoples C, et al. Gender differences in systemic sclerosis: relationship to clinical features, serologic status and outcomes. J Scleroderma Relat Disord. 2016

039 - CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS HAVE ALTERATIONS IN B AND T FOLLICULAR CELL SUBSETS IN PERIPHERAL BLOOD

Catarina Tomé[1], Filipa Oliveira Ramos[1, 2], Raquel Campanilho-Marques[1, 2], Ana Filipa Mourão[3], Sandra Sousa[4], Ana P. Martins[5], Patrícia Costa Reis[1, 6], Ana Teresa Melo[1, 2], Rui Lourenço Teixeira[1, 2], Miroslava Gonçalves[5], Maria José Santos[1, 4], L Graca[1, 7], JE Fonseca[1, 2], Rita A. Moura[1]

[1]Instituto de Medicina Molecular João Lobo Antunes, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal; [2]Rheumatology Department, Centro Hospitalar Universitário Lisboa Norte, EPE, Hospital de Santa Maria, Lisboa, Portugal; [3]Rheumatology Department, Centro Hospitalar Lisboa Ocidental, EPE, Hospital de São Francisco Xavier, Lisboa, Portugal; [4]Reumatology Department, Hospital Garcia de Orta, EPE, Almada, Portugal; [5]Pediatric Surgery Department, Centro Hospitalar Universitário Lisboa Norte, EPE, Hospital de Santa Maria, Lisboa, Portugal; [6]Pediatric Department, Centro Hospitalar Universitário Lisboa Norte, EPE, Hospital de Santa Maria, Lisboa, Portugal; [7]Instituto Gulbenkian de Ciência, Oeiras, Portugal

Background: Juvenile idiopathic arthritis (JIA) is the most common rheumatic disease in children. Our group has recently demonstrated that extended oligoarticular (eoJIA) and polyarticular JIA (pJIA) mostly evolve to a rheumatoid arthritis (RA) like phenotype in adulthood. Disturbances in B cells, T follicular helper (Tfh) and T follicular regulatory (Tfr) cell immune responses are associated with the pathogenesis of RA, but their exact role in JIA development is not entirely known.

Aims: The main goal of this study was to characterize the frequency and phenotype of B, Tfh and Tfr cells in peripheral blood of children with eoJIA and pJIA when compared to healthy controls and children with persistent oligoarticular JIA (poJIA).
Methods: Blood samples were collected from children with eoJIA (n=5), pJIA (n=11) and poJIA (n=19) treated with disease modifying anti-rheumatic drugs. A group of age-matched healthy children (n=8) was used as control. Peripheral blood mononuclear cells were isolated and the frequency and phenotype of B, Tfh and Tfr cells were evaluated by flow cytometry.
Results: The frequency of B, Tfh and Tfr cells was similar between JIA patients and controls. Children with eoJIA and pJIA, but not poJIA, had higher levels of naïve B cells and lower frequencies of post-switch memory B cells and plasmablasts when compared to controls. Th17-like Tfh cells were significantly increased in all JIA patients when compared to controls. B cell phenotype was similar between JIA patients and controls, but a reduced activated phenotype of Tfh cells was observed in JIA patients in comparison to controls.
Conclusions: Changes in B and Tfh cell subpopulations, but not in Tfr cells, were found in peripheral blood of children with JIA when compared to controls. The increased frequencies of Th17-like Tfh cells detected in JIA when compared to controls suggests a potential role of these cells in JIA pathogenesis. A treatment effect on the activation state of B, Tfh and Tfr cells cannot be excluded.

080 - RISK FACTORS FOR SARS-COV-2 INFECTION IN PATIENTS WITH RHEUMATIC DISEASES – A MULTICENTER, NATIONWIDE STUDY

Matilde Bandeira[1, 2], Ana Rita Cruz-Machado[1, 2], Sofia C Barreira[1, 2], Catarina Duarte[2], Maria Seabra Rato[3], Bruno Miguel Fernandes[3], Salomé Garcia[3], Filipe Oliveira Pinheiro[3], Miguel Bernardes[3], Nathalie Madeira[4], Cláudia Miguel[4], Rita Pinheiro Torres [5], Ana Bento da Silva[5], Jorge Pestana Lopes[6], Diogo Esperança Almeida[7], Carolina Mazeda[8, 9], Filipe Cunha Santos[10], Patrícia Pinto[11], Marlene Sousa[12], Hugo Parente[13], Graça Sequeira[14], Maria José Santos[6], JE Fonseca[1, 2], Vasco C Romão[1, 2]
[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Hospital de Santa Maria, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [3]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal; [4]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal; [5]Serviço de Reumatologia, Centro Hospitalar Lisboa Ocidental | Hospital Egas Moniz, Lisboa, Portugal; [6]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal; [7]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal; [8]Serviço de Reumatologia, Centro Hospitalar do Baixo Vouga, E.P.E., Aveiro, Portugal; [9]Instituto de Biomedicina – iBiMED, Universidade de Aveiro, Aveiro, Portugal; [10]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal; [11]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal; [12]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal; [13]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal; [14]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal

Introduction: Risk factors for SARS-CoV-2 infection have been vastly studied. In the subgroup of patients with rheumatic diseases, consensual results are, however, still lacking. Comorbidities such as diabetes mellitus, hypertension and other cardiovascular risk factors are reported to be associated both with COVID-19 infection and severity. Data indicates that patients with active systemic rheumatic diseases are in general more susceptible to infections, but evidence so far failed to prove that rheumatic diseases per se or certain disease-modifying antirheumatic drugs (DMARDS) are clear risk factors for infection by SARS-CoV-2. Our main goal was to identify risk factors for COVID-19 in patients with rheumatic diseases.
Methods: Multicentre observational nationwide study of adult patients with rheumatic diseases prospectively followed up in the Rheumatic Diseases Portuguese Register – Reuma.pt - in the first 6 months of the pandemic - March to September 2020. We collected demographic, clinical and laboratory data. Patients who developed COVID-19 (COVID-19+) were compared with those who didn't (COVID-19-) using Pearson Chi-Square, Fisher's exact and Mann-Whitney U tests, as adequate. Independent associations between demographic, comorbidities, disease and treatment-related variables and SARS-CoV-2 infection were evaluated through multivariate logistic regression.

Results: 6404 patients had at least one rheumatology appointment registered in Reuma.pt and 179 developed COVID-19 (Table 1). COVID-19+ patients were more frequently females (p=0.005) and Caucasians (p<0.001), had a lower prevalence of inflammatory arthropathies but a higher prevalence of connective tissue diseases (p<0.001), as well as a higher prevalence of cerebrovascular disease (p=0.043) and chronic kidney disease (p=0.014). In this cohort, COVID-19+ patients had a shorter disease duration (p<0.001) and lower disease activity (p<0.001). Regarding treatment, patients under immunosuppressors had lower odds of being infected (p<0.001). Drug sub-analysis showed that methotrexate, tumor necrosis factor inhibitors (TNFi) and tocilizumab were used more frequently in COVID-19- patients. Moderate/high disease activity, compared to remission/low disease activity (OR 0.419, 95%CI 0.269-0.654, p<0.001), use of TNFi (OR 0.160, 95%CI 0.099-0.260, p<0.001) and tocilizumab (OR 0.147, 95%CI 0.053-0.408, p<0.001) were associated with a lower likelihood of having COVID-19. The presence of two or more comorbidities (OR 2.273, 95%CI 1.416 -3.649, p<0.001) was an independent risk factor for SARS-CoV-2 infection.

Conclusion: Having two or more comorbidities was a risk factor for SARS-CoV-2 infection in patients with rheumatic diseases. The low prevalence of inflammatory disease in COVID+ patients might be due to an overreport of COVID+ non-inflammatory patients compared to COVID-, as they are less often registered

TABLE 1 - COMPARISON OF PATIENTS WITH COVID19 AND CONTROLS

	COVID-19+ (n=179)	COVID-19- (n=6225)	Univariate analysis p-value	Multivariate analysis OR (95%CI); p-value)
Female, N (%)	137 (76.5)	4144 (66.6)	**0.005**	NS
Age, median (IQR), in years	55 (20)	56 (22)	0.542	
Caucasian, N (%)	150 (83.8)	4172 (67.0)	**<0.001**	**2.548 (1.419-4.575); p=0.002**
Rheumatic disease by subgroup*, N (%)			**<0.001**	
Inflammatory arthropathy	111 (62.0)	5114 (82.2)		
CTD and vasculites	51 (28.5)	1087 (17.5)		NS
Non-inflammatory	17 (9.5)	9 (0.1)		NS
Rheumatic disease by diagnosis, N (%)			**<0.001**	
Rheumatoid arthritis	48 (26.8)	2430 (39.0)		
Spondyloarthritis	32 (17.9)	1526 (24.5)		
Psoriatic arthritis	20 (11.2)	942 (15.1)		
Systemic lupus erythematosus	12 (6.7)	477 (7.7)		
Osteoarthritis	9 (5)	4 (0.06)		
Vasculitis	8 (4.5)	139 (2.2)		
UCTD	8 (4.5)	46 (0.7)		
Systemic sclerosis	7 (3.9)	213 (3.4)		
Sjogren syndrome	5 (2.8)	133 (2.1)		
Other**	30 (16.8) *	315 (5.1)		
Disease duration, median (IQR), in years	7.1 (11.7)	10.6 (12.5)	**<0.001**	
Disease activity before COVID-19 pandemics***, N (%)			**<0.001**	
Remission	61 (34.1)	1807 (29.0)		
Low	73 (40.8)	1061 (17.0)		
Moderate	21 (11.7)	1200 (19.3)		
High	10 (5.6)	336 (5.4)		**0.419 (0.269-0.654); p<0.001*** **
Not applicable§	17 (9.5)	9 (0.1)		
Comorbidities, N (%)				
≥ 1 comorbidity	77 (43.0)	1554 (25.0)	**<0.001**	
≥ 2 comorbidities	35 (19.6)	423 (6.8)	**<0.001**	**2.273 (1.416-3.649); p=0.001**
Arterial hypertension	49 (27.4)	872 (14.0)	0.204	
Obesity	38 (21.2)	660 (10.6)	0.562	
Ever smoked	34 (19.0)	1271 (20.4)	**0.020**	
Cardiovascular disease	14 (7.8)	207 (3.3)	0.183	
Diabetes	13 (7.3)	193 (3.1)	0.225	
Malignancy	8 (4.5)	116 (1.9)	0.539	

TABLE 1 - CONTINUATION

Chronic obstructive pulmonary disease	6 (3.4)	47 (0.8)	0.060	
Cerebrovascular disease	6 (3.4)	43 (0.7)	**0.043**	
Chronic kidney disease	6 (3.4)	32 (0.5)	**0.014**	
Hyperuricemia	3 (1.7)	28 (0.4)	0.226	
Interstitial lung disease	2 (1.1)	78 (1.3)	0.443	
Asthma	2 (1.1)	55 (0.9)	0.768	
Ongoing treatment class			**<0.001**	
Non-immunosupressors	57 (31.8)	182 (2.9)		
csDMARDs	82 (45.8)	1613 (25.9)		
b/tsDMARDs + csDMARDs	16 (8.9)	1970 (31.6)		
b/tsDMARDs monotherapy	24 (13.4)	1602 (25.7)		
DMARDs subanalysis				
Glucocorticoids	71 (39.7)	2130 (34.2)	1.000	
Methotrexate	65 (36.3)	2422 (38.9)	**0.020**	NS
NSAIDs	30 (16.8)	1077 (17.3)	0.297	
Hydroxychloroquine	26 (14.5)	760 (12.2)	0.913	
TNFi	24 (13.4)	2598 (41.7)	**<0.001**	**0.160 (0.099-0.206); p<0.001**
Sulphasalazine	15 (8.4)	560 (9.0)	0.454	
Leflunomide	11 (6.1)	396 (6.4)	0.662	
Rituximab	7 (3.9)	261 (4.2)	0.722	
Azathioprine	6 (3.4)	128 (2.1)	0.45	
Tocilizumab	5 (2.8)	374 (6.0)	**0.024**	**0.147 (0.053-0.408); p<0.001**
JAKi	2 (1.1)	86 (1.4)	1.000	
Mycophenolate mofetil	1 (0.6)	96 (1.5)	0.376	
Ustekinumab	1 (0.6)	31 (0.5)	1.000	
Belimumab	1 (0.6)	29 (9.5)	1.000	

* For the multivariate analysis, inflammatory arthritis was set as the reference category

**Other diagnoses: adult-onset Still's disease (n=1), antiphospholipid syndrome (n=3); crystal-induced arthropathies (n=4); fibromyalgia (n=3), juvenile idiopathic arthritis (n=2), mixed connective tissue disease (n=3), myositis (n=2), osteoporosis (n=3), overlap syndromes (n=1), Paget bone disease (n=2), polymyalgia rheumatica (n=3), RS3PE (n=1), sarcoidosis (n=2)

*** For the multivariate analysis, remission/low disease activity was set as the reference category

§ In case of non-inflammatory diseases

CTD – connective tissue disease; UCTD – undifferentiated connective tissue disease; NSAIDs – nonsteroidal anti-inflammatory drugs; JAKi – janus kinase inhibitors; TNFi – tumor necrosis factor inhibitors; NS – statistically nonsignificant

in Reuma.pt and non-urgent appointments were delayed during the pandemic. Higher disease activity, use of TNFi and tocilizumab were associated with a lower likelihood of being infected in our cohort. These data support the hypothesis that rheumatic patients do not have an increased risk of SARS-CoV-2 infection and we consider the possibility that patients with higher disease activity had a higher compliance to lockdown measures. Also, a potential protective role of the aforementioned cytokine-inhibiting drugs known to be involved in the pathophysiology of COVID-19 is a current topic of investigation and deserves further acknowledgment.

083 - EFFECTIVENESS OF SECUKINUMAB THERAPY IN REAL-WORLD PSORIATIC ARTHRITIS PATIENTS - DATA FROM THE RHEUMATIC DISEASES PORTUGUESE REGISTRY (REUMA.PT)

José Tavares-Costa[1], Soraia Azevedo[1], Laires PA[2], Salomé Garcia[3], Patrícia Nero[4], Jorge Pestana Lopes[5], Martins-Martinho J [6], Ana Bento da Silva[7], Joana Ramos Rodrigues[1], Graça Sequeira[8], Emanuel Costa[9], Filipe Cunha Santos[10], Carolina Furtado[11], Bernardo Santos[12], Ana Raposo[13], Liliana Saraiva[14], Beatriz Samões[15], Filipe Barcelos[16], Maria José Santos[5], Helena Santos[16, 17]

[1]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal; [2]HE&OR - Novartis Portugal / ENSP – NOVA University of Lisbon, Lisboa, Portugal; [3]Rheumatology

Department, Centro Hospitalar de São João, Porto, Portugal; [4]Rheumatology Department, Hospital CUF Descobertas, Lisboa, Portugal, [5]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal; [6]Rheumatology Department, Centro Hospitalar Universitário de Lisboa Norte, Lisboa, Portugal
[7]Rheumatology Department, Centro Hospitalar Lisboa Ocidental, Lisboa, Portugal; [8]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal [9]Rheumatology Department, Hospital de Braga, Braga, Portugal; [10]Rheumatology Department, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal; [11]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal; [12]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal; [13]Rheumatology Department, Centro Hospitalar Trás-os-Montes e Alto Douro, Vila Real, Portugal; [14]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal; [15]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal; [16]Portuguese Institute of Rheumatology, Lisboa, Portugal; [17]EpiDoc Research Unit, CEDOC, NOVA Medical School, Lisboa, Portugal

Background: Psoriatic arthritis (PsA) affects around 0.31% (95%CI 0.13%-0.48%) of the Portuguese adult population[1]. Secukinumab has demonstrated efficacy across the 6 key PsA manifestations and has been shown to inhibit structural damage and improve physical function[2]. Despite that, real world data on its effectiveness is still missing. The aim was to investigate the effectiveness of secukinumab therapy in multiple domains of PsA patients using real world data.
Methods: PROSAS was a national, multicentric, observational, longitudinal cohort study of patients

TABLE 1: DISEASE ACTIVITY RESPONSE.

	Baseline	3 months	p-value*	6 months	p-value*	12 months	p-value*
PtGA, mean (SD)	62.1 (24)	41.5 (27)	<0.001	44.3 (27)	<0.001	40.2 (27)	<0.001
PhGA, mean (SD)	47.7 (21)	25.9 (23)	<0.001	17.4 (17)	<0.001	21.7 (16)	<0.001
CRP mg/L, median (IQR)	8.1 (13)	3.8 (9.4)	<0.001	3.9 (6.5)	<0.001	4.3 (5.9)	<0.001
ESR, median (IQR)	20.0 (35)	14.0 (18)	<0.001	13.0 (22)	<0.001	14.0 (25)	0.002
BASDAI, mean (SD)	5.7 (2.1)	3.9 (2.7)	0.001	4.0 (2.7)	<0.001	3.2 (2.8)	<0.001
Δ BASDAI, mean (SD)	-	2.1 (2.6)		2.0 (2.0)		1.7 (1.9)	
BASDAI 50, n/N (%)		11/21 (52%)		10/24 (42%)		9/22 (41%)	
ASDAS-PCR, mean (SD)	3.3 (1.1)	2.4 (1.4)	0.008	2.3 (1.0)	0.001	2.1 (1.1)	0.001
Δ ASDAS-PCR, mean (SD)		1.1 (1.1)		1.3 (1.1)		1.5 (1.3)	
ASDAS-PCR CII (Δ > 1.1), n/N (%)	-	8/19 (42%)		7/20 (35%)		7/20 (35%)	
ASDAS-PCR MI (Δ > 2.0), n/N (%)		4/19 (21%)		3/20 (15%)		3/20 (15%)	
DAPSA, mean (SD)	137.0 (50)	57.7 (8.3)	<0.001	87.9 (57)	<0.001	87.9 (55)	<0.001
Δ DAPSA, mean (SD)	-	54.9 (53)		63.0 (60)		56.6 (50)	
DAS28 4V, mean (SD)	4.4 (1.5)	3.3 (1.7)	<0.001	3.1 (1.5)	<0.001	3.1 (1.4)	<0.001
Δ DAS28 4V, mean (SD)	-	1.5 (1.3)		1.4 (1.4)		1.6 (1.5)	
Tender joint count, median (IQR)	5.0 (10)	1.0 (5.0)	<0.001	1.0 (4.0)	<0.001	0.0 (3.0)	<0.001
Swollen joint count, median(IQR)	3.0 (6)	0.0 (3.0)	<0.001	0.0 (2.0)	<0.001	0.0 (2.0)	<0.001
ASAS 20, n/N (%)	-	8/20 (40%)		6/22 (27%)		5/20 (25%)	
ASAS 40, n/N (%)	-	4/20 (20%)		4/22 (18%)		2/20 (10%)	
ASAS 70, n/N (%)	-	3/20 (12%)		1/22 (4.5%)		1/20 (5.0%)	
ACR 20, n/N (%)	-	25/49 (51%)		13/26 (50%)		18/29 (62%)	
ACR 50, n/N (%)	-	10/49 (20%)		6/34 (18%)		8/29 (28%)	
ACR 70, n/N (%)	-	6/49 (12%)		3/43 (7.0%)		1/29 (3.4%)	
EULAR response							
Moderate, n/N (%)	-	20/48 (42%)		15/40 (38%)		16/40 (40%)	
Good, n/N (%)	-	14/48 (29%)		12/40 (30%)		13/40 (33%)	

*p value - Comparisons across different timepoints and baseline;
ASDAS-CRP: Ankylosing Spondylitis Disease Activity Score; ASDAS CII: ASDAS Clinically important improvement; ASDAS MI: ASDAS Major Improvement; BASDAI: Bath Ankylosing Spondylitis Disease Activity Index; CRP: C-reactive protein; DAPSA: Disease Activity in PSoriatic Arthritis; DAS 28 4V: Disease Activity Score 4 variables; ESR: erythrocyte sedimentation rate; IQR: interquartile range; NS: non-significant; PtGA: patient global assessment; PhGA: physician global assessment; SD: standard deviation; Δ: variation between evaluation and baseline

with diagnosis of PsA using real-world anonymous patient-level data from the Portuguese national register database - Reuma.pt. We analyzed PsA patients that started secukinumab treatment between 1st January 2017 and 10th January 2021. We studied data from the following time points: baseline, 3, 6 and 12 months after secukinumab initiation. We collected data on sociodemographic characteristics and therapy efficacy: ACR and ASAS response, patient global assessment (PtGA), physician global assessment (PhGA), C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), Bath Ankylosing Spondylitis Disease Activity Index (BASDAI), Ankylosing Spondylitis Disease Activity Score-CRP (ASDAS-CRP), Disease Activity in PSoriatic Arthritis (DAPSA) and Disease Activity Score (DAS28 4V). Descriptive analysis of continuous variables was reported as mean and standard deviation (SD), or median and interquartile range (IQR) in case of non-normal distribution; categorical variables were displayed as frequency or proportions. For effectiveness analysis we performed paired sample comparisons: if a variable had a normal distribution paired sample t-test were used; if not, the Wilcoxon test (W) was used.

Results: We included 166 patients with PsA, 54.8% (91) were female, 36.7% (61) of patients were biologic-naïve, 24.7% (41) were on 2nd line, and 38.6% (64) had failure to 2 or more biologics. The mean age at last appointment was 52.4 (±11.7) years-old. Most patients presented with peripheral arthritis (58.4% symmetric polyarthritis and 23.4% asymmetric oligoarthritis) and 53% of patients had extra-articular manifestations. Patient age at secukinumab start was 50.8 (±11.7) years-old. Other sociodemographic and clinical characteristics are shown in Table 1. ACR 20 response was attained in 51.0% and 62% of patients at 3 and 12 months, respectively and ASAS 20 response was reached in 40% and 25%, at the same time points. In what comes to PsA specific composite index, we have encountered a significant improvement in DAPSA between baseline and the end of follow-up ($p<0.001$). Significant improvements in disease activity were also observed at 3 months, and were sustained until 12 months, regarding PtGA, PhGA, CRP, ESR, BASDAI, ASDAS-CRP and DAS28 4V (Table 2).

Conclusions: In this real world cohort study, we have confirmed that secukinumab treatment was effective in PsA patients, with significant and sustained improvements in disease activity.

Acknowledgement: This study was supported by Novartis and presented on Behalf of PROSAS Study group.

REFERENCES

1. Canhão H, Rodrigues A, Dias S, Eusébio M, Gouveia N, Sousa R, et al. The Burden of Spondyloarthritis – Paasport A Population-Based Study. Value Heal. 2017;20(9):A548.
2. McInnes IB, Mease PJ, Kirkham B, Kavanaugh A, Ritchlin CT, et al. Secukinumab, a human anti-interleukin-17A monoclonal antibody, in patients with psoriatic arthritis (FUTURE 2): A randomised, double-blind, placebo-controlled, phase 3 trial. Lancet. 2015;386(9999):1137–46.

088 - MORTALITY RISK ASSOCIATED WITH OSTEOPOROTIC FRAGILITY FRACTURE

Soraia Azevedo[1], Hugo Parente[1], Francisca Guimarães[1], Joana Ramos Rodrigues[1], Catarina Soares[1], Daniela Santos-Faria[1], Daniela Peixoto[1], José Tavares-Costa[1], Carmo Afonso[1], Filipa Teixeira[1]
[1]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal

Background: Many studies have shown that mortality increases after hip osteoporotic fracture. Some studies showed this association after a vertebral fracture, but studies that evaluated the global mortality after a major fragility fracture (FF) in global are scarce.

Objectives: To assess predictive factors of mortality occurring after a FF.

Methods: We performed a retrospective monocentric study that included patients with a FF observed at the emergency department (ED) in a tertiary center, between 1st January 2017 and 31st December 2018. Fractures were researched through the 9th International Classification of Diseases codes and FF were identified after revision of the clinical files. One thousand six hundred seventy-three FF were identified. (Image 1) After calculating a representative sample (90% confidence interval), 172 hip, 173 wrist and 112 vertebral fractures were included. Their clinical files until 31th December 2020 (2 to 4 years follow-up after FF) were reviewed. SPSS was used for statistical analysis and significance level was defined as 2-sided $p<0.05$. In multivariate analysis we included variables with a significant association in univariate analysis and those with clinical relevance (reported in others studies).

Results: There was a total of 457 patients evaluated, out of these 79.9% (365) were woman with a mean age of 77.6 (SD=10.3) years-old at the time of their FF. One hundred and twenty patients (26.3%) died during the follow-up period, 35.9% of the men and 23.8% of women inserted in this study. Of these patients 46.5% had a hip fracture and 20.5% a vertebral fracture. Patients took a median of 5 (IQR=4) daily different medications and had a median of 4 (IQR=2)

comorbidities.

We found an association between mortality and male gender (p=0.024), age (older patients had a bigger mortality, p<0.001), hip fracture (p<0.001), more daily medication (p<0.001) and comorbidities (p=0.001), daily oral corticosteroid treatment (p=0.024), normal or low body index mass (BMI) (p<0.001), previous visits to the ED due to falls (p=0.022), type 2 diabetes (p=0.022), cardiac disease (p<0.001), neurologic disease (p<0.001), chronic kidney disease (p<0.001) and no osteoporosis treatment started after FF (p=0.007).

No associations were found between mortality and previous fractures before the fracture included in this study, anxiolytic or antiepileptic treatment, arterial hypertension, dyslipidemia, endocrinopathies, psychiatric disease, hematologic disease, chronic pulmonary disease, other comorbidities or cancer diagnosis not in palliative care.

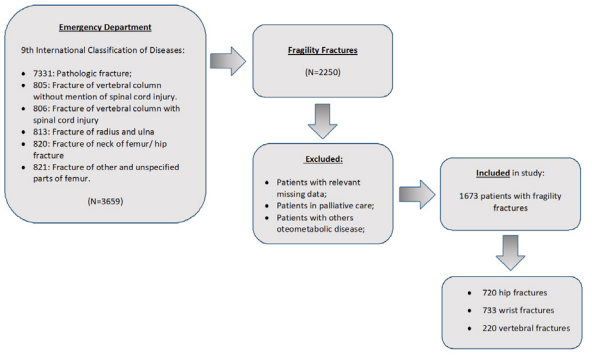
After adjustment for gender, daily medication, comorbidities, anti-osteoporotic treatment, previous visits to the ED due to falls and type 2 diabetes, the main predictors of mortality were age, hip fracture, daily corticosteroid treatment, normal or low BMI and cardiac, neurologic or chronic kidney disease (Table 1).

Conclusions: FF are a very prevalent public health problem that can lead to death. Age, hip fracture and certain comorbidities seem to be associated with higher mortality after a FF. We need to consider these comorbidities and actively search for osteoporosis in these patients to prevent FF. Since most osteoporotic fractures occur during a fall, fall risk reduction may be an important measure to prevent a new fracture.

TABLE 1. MULTIVARIATE ANALYSES: LINEAR MULTIPLE REGRESSION FOR PREDICTIVE FACTORS OF MORTALITY AFTER A FRAGILITY FRACTURE.

Determinants	Unstandardized Coefficients B	Standardized Coefficients Beta	95.0% CI	p-value
Age at fracture time	0.10	1.11	1.07 – 1.15	<0.001
Gender (male)	0.64	1.89	0.98 – 3.66	NS
Number of comorbidities	-0.04	0.97	0.73 – 1.29	NS
Oral corticosteroids	2.30	9.94	1.75 – 56.3	0.009
Previous visits to the emergency service for falls	0.06	1.06	0.56 – 2.03	NS
Hip fracture	0.83	2.29	1.31 – 4.02	0.004
Normal or low body mass index	1.60	4.95	2.22 – 11.0	<0.001
Type 2 diabetes	0.54	1.72	0.89 – 3.320	NS
Cardiac disease	0.61	1.83	1.03 – 3.27	0.041
Neurologic disease	0.57	1.78	1.00 – 3.12	0.049
Chronic kidney disease	1.87	6.49	2.43 – 17.3	<0.001
Number of daily medication	-0.01	0.99	0.83 – 1.13	NS
Anti-osteoporotic treatment after fragility fracture	1.07	2.92	0.58 – 14.7	NS

FIGURE 1. PATIENT FLOWCHART



095 - VISION-RELATED QUALITY OF LIFE IN SPONDYLOARTHRITIS PATIENTS WITH HISTORY OF ACUTE ANTERIOR UVEITIS UNDER TREATMENT WITH GOLIMUMAB: PRELIMINARY RESULTS OF THE GO-VISION OBSERVATIONAL STUDY RHEUMATOLOGY

Inês Leal[1, 2, 3], Patricia Jose[1], Rafael Barão[1], Sofia Sousa Mano[1], Ana Teresa Melo[3, 4], Eduardo Dourado[3, 4], Catarina Tenazinha[3, 4], Cláudia Vaz[5], Joana Fonseca Ferreira[5], Sara Paiva Dinis [5], Filipe Cunha Santos[5], Henrique Fernandes[6], Nikita Khmelinskii[3, 4], Carlos Marques-Neves[1, 2], Elsa Vieira-Sousa[3, 4], JE Fonseca[3, 4]

[1]Serviço de Oftalmologia, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [2]Centro de Estudos das Ciências da Visão, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal; [3]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [4]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [5]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal; [6]Oftalmologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal

Objective: Acute anterior uveitis (AAU) is one of the most common extra-articular manifestations of spondyloarthritis (SpA), causing significant burden in quality of life (QoL). Golimumab (GLM) is a tumor necrosis factor-inhibitor proven to be effective and safe in SpA. The GO-EASY Study provided evidence that GLM decreases the AAU occurrence rate in SpA. Our

aim is to study the impact of GLM in the change of vision-related (VR) QoL in subjects with SpA and past or current AAU.

Methods: Ongoing prospective multicentre observational study (including 8 centres in Portugal) of SpA patients with history of AAU treated with GLM followed-up for 12 months. We intend to recruit in total 30 patients and we report herein the 6 months outcomes for the first 9 patients enrolled. The occurrence of AAU was assessed in the 2 years before GLM treatment was started and the first 6 months of follow-up and calculated for the period at risk for a new AAU. VR QoL was assessed with the self-administered National Eye Institute Visual Functioning Questionnaire-25 (NEI VFQ-25). Adverse events were noted.

Results: Nine patients (66.7% female, 100% TNFi-naive, mean age 46.1±14.4 years (range 22-65)) have completed 6 months of follow-up. Three patients (33%) were also under oral methotrexate. The mean number of AAU flares in the 2 years preceding the start of GLM was 2.2±1.3 (1-4). During the first 6 months of GLM treatment, the AAU incidence rate was reduced from 1.54 to 0.11 per 100 patient-years (incidence ratio-ratio 13.46, 95% CI 2.15–558.00; $p < 0.01$). At baseline and at 24 weeks after GLM onset, the mean overall index NEI VFQ-25 total score was 70.2 and 83.2, respectively; improvement in the NEI VFQ-25 total score was +13±18.2. No significant or new adverse events occurred.

Conclusion: Preliminary data from the GO-VISION study suggest that GLM is safe and effective in patients with SpA and history of AAU, being able to reduce the AAU occurrence rate and potentially increasing VR QoL.

099 - DETERMINANTS OF HEALTH-RELATED QUALITY OF LIFE IN SPONDYLOARTHRITIS AND COMPARISON WITH CHRONIC LOW BACK PAIN – RESULTS FROM A NATION-WIDE STUDY

Helena Santos[1,2], Ana Rita Henriques[2], Jaime C. Branco[2], Rodrigues AM[2], Helena Canhão[2]
[1]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal, [2]EpiDoC Unit, CEDOC, NOVA Medical School, NOVA University, Lisboa, Portugal

Background/Purpose: Spondyloarthritis (SpA) causes pain, fatigue, stiffness, loss of physical function and impaired health-related quality of life (HRQoL).

This study aims to: 1) to compare HRQoL in patients with SpA, chronic low back pain (CLBP) and subjects with no rheumatic disease (noRMD), and 2) to evaluate determinants of HRQoL in SpA patients, in a population-based setting.

Methods: Data from EpiReumaPt, a national health survey with 10 661 adult participants, randomly selected, that were screened for rheumatic and musculoskeletal diseases (RMDs) was used. Subjects were asked about socio-demographic data, lifestyle habits, chronic non-communicable diseases and HRQoL. SpA diagnosis was based on a positive expert opinion (rheumatologist) combined with predefined criteria; SpA disease activity was also collected. CLBP was established by predefined criteria and noRMD by expert opinion. Univariate and multivariate linear regressions were performed to explore factors associated with QoL assessed by EuroQoL- 5 Dimensions 3L (EQ-5D).

Results: We included 92 SpA, 1376 CLBP and 679 with noRMD. The mean age was 48.4 years (SD=13.7) for SpA, 58.8 years (SD=14.6) for CLBP and 45.9 years (SD=15.6) for noRMD. The 3 groups had a female predominance (64.1%, 70.3% and 53.9%, respectively). SpA and CLBP had similar HRQoL reflected by EQ5D-3L index (ß=-0.03, p-value=0.33), much lower when compared to subjects noRMD (ß=-0.14, p-value<0.001) (table I), Patients with SpA had lower scores in all EQ-5D dimensions, similar in patients with CLBP, but much higher than in participants noRMD. Pain, was reported in almost 60% of SpA adults, followed by limitations in mobility. In our cohort, SpA adults showed similar individual perception of health than adults with CLBP by EuroQoL visual analogue scale (ß=0.03, p-value=0.989), that was much lower (higher score=better health) when compared to adults noRMD (ß=-7.49, p-value=<0.001) (table I). Considering the factors associated with HRQoL in SpA, multimorbidity (>=3 chronic non-communicable diseases) (ß=-0.18; [-0.24; -0.03]; p-value<0.001) and an active disease (ß=-0.13; [-0.29; -0.05]; p-value=0.036), were associated with worse HRQoL; on the other hand, regular physical exercise was significantly associated with better HRQoL (ß=0.18; CI 95% [0.1; 0.3]; p-value<0.001) (table II). Our model can explain 35.43% of the variance of HRQoL in SpA subjects.

Conclusion: In this nation-wide study, SpA showed a similar impact in HRQoL than CLBP. An active disease, multimorbidity and regular physical exercise are largely responsible for HRQoL in SpA.

TABLE I. COMPARISON OF QUALITY OF LIFE AND PHYSICAL FUNCTION BETWEEN SUBJECTS WITH SPONDYLOARTHRITIS AND CHRONIC LOW BACK PAIN AND BETWEEN SUBJECTS WITH SPONDYLOARTHRITIS AND WITHOUT RHEUMATIC DISEASES

	SpA n=92	CLBP n=1376	noRMD n=679	Crude OR SpA/CLBP [95% CI]	Crude p-value SpA/CLBP	Adjusted OR SpA/CLBP[a] [95% CI]	Adjusted p-value SpA/CLBP[a]	Crude OR SpA/noRMD [95% CI]	Crude p-value SpA/noRMD	Adjusted OR SpA/noRMD[b] [95% CI]	Adjusted p-value SpA/noRMD
EQ-5D											
Mobility				0.742 [0.465; 1.155]	0.196	1.371 [0.812; 2.275]	0.229	4.34 [2.588; 7.183]	<<0.001	4.54 [2.5; 8.212]	<<0.001
No problems	63 (68.48%)	849 (61.7%)	613 (90.41%)								
Some or extreme problems	29 (31.52%)	527 (38.3%)	65 (9.59%)								
Self-care				0.647 [0.31; 1.208]	0.204	1.374 [0.626; 2.758]	0.397	4.23 [1.833;9.231]	<<0.001	4.856 [1.849; 1.257]	0.001
No problems	82 (89.13%)	1156 (84.13%)	659 (97.2%)								
Some or extreme problems	10 (10.87%)	218 (15.87%)	19 (2.8%)								
Usual activities				0.957 [0.6; 1.492]	0.849	1.592 [0.943; 2.641]	0.075	4.423 [2.635;7.326]	<<0.001	4.65 [2.56;8.44]	<<0.001
No problems	63 (68.48%)	927 (67.52%)	615 (90.56%)								
Some or extreme problems	29 (31.52%)	446 (32.48%)	64 (9.43%)								
Pain/discomfort				1.074 [0.699; 1.672]	0.748	1.351 [0.845;2.187]	0.213	5.15 [3.284; 8.2]	<<0.001	4.726 [2.937; 7.703]	<<0.001
No pain/discomfort	35 (38.04%)	546 (39.74%)	516 (75.99%)								
Moderate or extreme pain / discomfort	57 (61.96%)	828 (60.26%)	163 (24.01%)								
Anxiety/depression				0.973 [0.603; 1.529]	0.908	1.143 [0.681; 1.877]	0.604	1.961 [1.185; 3.173]	0.007	1.499 [0.871; 2.488]	0.135
Not anxious/depressed	65 (70.65%)	958 (70.08%)	557 (82.52%)								
Moderately or extremely anxious/depressed	27 (29.35%)	409 (29.92%)	118 (17.48%)								
	SpA n=92	CLBP n=1376	noRMD n=679	Crude ß SpA/CLBP [95% CI]	Crude p-value SpA/CLBP	Adjusted ß SpA/CLBP[a] [95% CI]	Adjusted p-value SpA/CLBP[a]	Crude ß SpA/noRMD [95% CI]	Crude p-value SpA/noRMD	Adjusted ß SpA/noRMD[b] [95% CI]	Adjusted p-value SpA/noRMD
Quality of life EQ5D score (mean ± sd)	0.69 ± 0.25	0.66 ± 0.27	0.86 ± 0.21	0.03 [-0.03; 0.086]	0.3	-0.03 [-0.08; 0.03]	0.33	-0.167 [-0.213; -0.121]	<<0.001	-0.141 [-0.186; -0.1]	<<0.001
EQ VAS (mean ± sd)	65.28 ± 18.1	60.92 ± 19.86	75.69 ± 17.64	4.36 [0.108; 8.6]	0.04	0.03 [-4.06; 4.12]	0.989	-10.414 [-14.34;6.49]	<<0.001	-7.488 [-11.2; -3.78]	<<0.001

SpA: Spondyloarthritis; CLBP: Chronic low back pain; noRMD: No Rheumatic diseases; EQ-5D: EuroQoL- 5 Dimensions 3L; EQ VAS: EuroQoL visual analogue scale; HAQ: Health Assessment Questionnaire; OR: Odds ratio; CI: Confidance interval

[a]OR adjusted for: gender, age-group, NUTII, education level, employment status, BMI and number of noncommunicable Diseases; [b]OR adjusted for: gender, age-group, NUTII, marital status and number of noncommunicable Diseases

Sample size is not constant due to missing data: SpA - Mobility (n=92); Self-care (n=92); Usual activities (n=92); Pain/discomfort (n=92); Anxiety/depression (n=92); Quality of life EQ5D score (n=91); HAQ score (n=92). CLBP - Mobility (n=1376); Self-care (n=1374); Usual activities (n=1373); Pain/discomfort (n=1374); Anxiety/depression (n=1367); Quality of life EQ5D score (n=1362); HAQ score (n=1376). NoRMD - Mobility (n=678); Self-care (n=678); Usual activities (n=679); Pain/discomfort (n=679); Anxiety/depression (n=675); Quality of life EQ5D score (n=675); HAQ score (n=679)

100 - PREDICTIVE FACTORS OF UVEITIS AND ITS COMPLICATIONS IN A COHORT OF 302 JIA PATIENTS

Ana Teresa Melo[1, 2], Martins-Martinho J [1, 2], Patrícia Martins[1, 2], Rui Ferreira[3], Patricia Jose[3], Sofia Sousa Mano[3], Inês Leal[3], Patrícia Costa Reis[2, 4, 5], Filipa Oliveira Ramos [1, 2, 5], JE Fonseca[1, 2], Raquel Campanilho-Marques[1, 2, 5]

[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [3]Serviço de Oftalmologia, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [4]Serviço de Pediatria, Centro Hospitalar Universitário Lisboa Norte, Hospital de Santa Maria, Lisboa, Portugal; [5]Unidade de Reumatologia Pediátrica, Centro Hospitalar Universitário Lisboa Norte, Hospital de Santa Maria, Lisboa, Portugal

Introduction: Screening and prompt diagnosis are essential to avoid complications, such as cataracts and glaucoma in uveitis.

Objectives: To identify clinical or laboratorial predictive factors of uveitis complications in patients with JIA.

Methods: A retrospective observational study of patients with JIA, registered at the Portuguese Rheumatic Diseases Register (Reuma.pt) was performed. Demographic variables, data on presence of uveitis

and its complications (synechiae, band keratopathy, glaucoma, cataracts, macular edema), clinical features and treatment were collected and complemented with hospital clinical data. Statistical analysis was done using SPSS 26.0, with a significance of $p<0.05$. Univariate analysis was performed using Fisher's exact test, Mann-Whitney U test or Chi-Square. Multivariate analysis was also performed.

Results: We included 302 JIA patients, 59.6% females, with a mean age at JIA onset of 8.4±4.8 years and a mean disease duration of 12.8±11.1 years. Uveitis was identified in 52 of the 302 patients (17.2%), with a mean age at uveitis onset of 11.3± 9.4 years. The mean time since JIA diagnosis until uveitis onset was 5.4± 8.3 years. Only in 2 cases uveitis started before JIA. Uveitis developed only in adulthood in 11 of those patients. Younger age at JIA diagnosis was associated with uveitis (6.2 [4.5] vs 8.9 [4.7], p=0.01). Oligoarticular persistent (opJIA) (23.7% vs 14.1%, p=0.040) and extended form (oeJIA) (39.3% vs 15.0%, p=0.003) were also positively correlated with the presence of uveitis. On the other hand, polyarticular JIA had a lower frequency of uveitis (7.9% vs 20.4%, p=0.013). First uveitis episode at adulthood was more common in JIA ERA patients (15.4% vs 62.5%, p=0.011), whereas uveitis onset <18years old was associated with an opJIA form (36.0% vs 9.0%, p=0.03). Uveitis was more frequent in patients with positive antinuclear antibodies (ANA) (25.8% vs 13.6%, p=0.012), however, there were no significant differences between the ANA title (≤1/160 vs > 1/160) or pattern. We found no association between the development of uveitis and positivity to rheumatoid factor, anticyclic citrullinated peptides, human leukocyte antigen B27 or with inflammatory markers. Multivariate analyses showed that opJIA and oeJIA were independent predictors of uveitis (OR 2.9 95%CI: 1.3-6.6; OR 6.1 95% CI: 2.3-16.2; respectively). Ocular complications occurred in 15 patients out of the 52 patients (28.3%): synechiae occurred in 28.3%, band keratopathy in 22.7%, cataracts in 17%, glaucoma in 15.1%, macular edema in 7.6%. We found an association between oeJIA form and the development of glaucoma (80% vs 22%, p=0.033) and with the need for ocular surgical procedures (75.0% vs 14.8%, p=0.028). Multivariate analyses showed that oeJIA form was an independent predictor of ocular surgical procedures (OR 14.9 95% CI: 1.2-193.4).

Conclusion: Uveitis was more frequent in opJIA and oeJIA patients and was related to ANA positivity, which is consistent with the literature. The need of ocular surgeries and prevalence of glaucoma seems to correlate with oeJIA.

124 - UNDERTREATMENT OF HIP FRAGILITY FRACTURES IN THE PRIMARY CARE SETTING: A 3-YEAR NATIONWIDE PERSPECTIVE FROM PORTUGAL

Beatriz Oliveira Pinto[1], Vítor Trindade Pedrosa[1], Raquel Baptista Leite[2], Mariana Castro Guimarães[1], Filipe Araújo[3]

[1]ARSLVT - ACES Cascais - USF KosmUS, Cascais, Portugal;

[2]ARSLVT - ACES Cascais - USF São Martinho de Alcabideche, Cascais, Portugal, [3]Rheumatology and Osteoporosis Unit, Hospital de Sant´Ana, SCML, Cascais, Portugal

Objectives: The purpose of this study was to determine the treatment rate following hip fractures in the Primary Health Care (PHC) setting in Portugal from 2018 to 2020, and to characterize prescription patterns of anti-osteoporotic drugs in this context.

Material and methods: We conducted an observational, cross-sectional, retrospective study using data from the PHC database Bilhete de Identidade - Cuidados de Saúde Primários (BI-CSP). BI-CSP is a national primary care database (except Madeira and the Azores) presenting information on demographics, health indicators, drug prescription and primary care units' management and organization. Grouped and anonymized data on any pre-specified variable over a period of time (for instance, diagnosis or treatment) can be obtained through the application of filters. The classification tool used is the International Classification of Primary Care, 2nd Edition. We included in our study all individuals registered in the PHC in Portugal who sustained a hip fracture after the age of 50 years. Treatment rates were determined after considering any prescription of an anti-osteoporotic drug to these patients in the PHC, from 2018 to 2020. The frequency of prescription for each type of drug was also assessed. All outcomes of interest were extracted to a data extraction sheet created for this purpose. Descriptive statistics were generated using Microsoft Office Excel- 2016. This study was approved by the local ethics committee.

Results: We identified a total of 44725 hip fractures in Portugal between 2018 and 2020, 33668 (75.3%) in female patients. Most hip fractures were recorded in the North- (17387) and Lisbon- (14353) Regional Health Administrations (RHA). Only 6282 (14.0%) patients

TABLE 1. PRESCRIPTION PATTERNS FOLLOWING HIP FRACTURE IN PRIMARY CARE IN PORTUGAL.

RHA	RIS (%)	AL (%)	IA (%)	ZA (%)	Cal (%)	T (%)	D (%)	R (%)	SR (%)	Total (n)
N	70 3,0%	1500 65,1%	359 15,6%	230 10,0%	0 0%	19 0,8%	99 4,3%	27 1,2%	0 0%	2304
C	20 1,6%	865 67,7%	172 13,5%	86 6,7%	0 0%	11 0,9%	110 8,6%	14 1,1%	0 0%	1278
LVT	66 3,1%	1427 66,0%	323 14,9%	197 9,1%	0 0%	28 1,3%	98 4,5%	22 1,0%	0 0%	2161
ALT	2 0,8%	177 72,8%	45 18,5%	12 4,9%	0 0%	3 1,2%	2 0,8%	2 0,8%	0 0%	243
ALG	3 1,0%	224 75,7%	30 10,1%	24 8,1%	0 0%	1 0,3%	12 4,1%	2 0,7%	0 0%	296
Total (n)	161	4193	929	549	0	62	321	67	0	6282

RHA: regional health administration; N: north region; C: center region; LVT: Lisbon region; ALT: Alentejo Region; ALG: Algarve region; RIS: sodium risedronate; AL: alendronic acid; IA: Ibandroni acid; ZA: zoledronic acid; Cal: salmon calcitonin; T: teriparatide; D: denosumab; R: raloxifene; SR: strontium ranelate

were treated with anti-osteoporotic therapies following hip fracture. The RHA with the highest treatment rate was the Algarve (15.2%) and the lowest was Alentejo (10.9%). National and regional prescription patterns are shown in Table 1. Bisphosphonates were the most frequently used therapies, particularly alendronic acid (66.7%), ibandronic acid (18.8%) and zoledronic acid (8.7%). Denosumab, raloxifene and teriparatide were prescribed by family physicians in 5.1%, 1.1% and 1.0% of patients after hip fracture.

Conclusions: Only a minority of patients received anti-osteoporotic treatment following fragility hip fractures in the primary care setting in Portugal. The most frequently used therapies were, by far, bisphosphonates namely alendronic acid.

137 - SHEAR-WAVE ELASTOGRAPHY AND B-MODE EVALUATION OF MAJOR SALIVARY GLANDS IN PATIENTS WITH PRIMARY SJÖGREN'S SYNDROME

Ana Rita Prata[1], João Pedro Freitas[1], Mary Lucy Marques[2], Flávio Campos Costa[3], Mariana Santiago[1,4], Sara Serra[1], Margarida Coutinho[1,4], Tânia Santiago[1,4], João Rovisco[1,4], JAP da Silva[1,4]

[1]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal; [2]Department of Rheumatology, Leiden University Medical Center, Leiden, Netherlands; [3]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal; [4]Coimbra Institute for Clinical and Biomedical Research (iCBR) - Faculty of Medicine, University of Coimbra, Coimbra, Portugal

Background: Salivary-gland ultrasound has recently shown to help in the diagnosis and monitoring of primary Sjögren's syndrome (pSS)1. Shear-wave elastography (SWE) is a promising tool for the quantitative assessment of tissue stiffness2. However, studies evaluating its role in ultrasound evaluation of pSS are still limited.

Objective: This study aimed to investigate the diagnostic performance of SWE in pSS and its correlation with B-mode findings.

Methods: Cross-sectional study including consecutive patients fulfilling the ACR/EULAR 2016 classification criteria for pSS, followed at our Rheumatology clinic, and age and gender-matched healthy controls. A protocolized clinical and ultrasound evaluation was performed for each patient. The four major salivary glands (parotid and submandibular glands, bilaterally) were assessed by SWE and B-mode modalities by 2 and 3 independent operators, respectively, blinded to the diagnosis. B-mode scans were rated using the Hocevar score3, and mean shear-wave velocity (SWV) values were obtained from 6 different measures for each gland. Student's t-test, chi-square test and Pearson's correlation were performed to compare data, as appropriate. Inter and intra-rater reliability were assessed using intraclass-correlation coefficient (ICC). Cut-off values for differentiating pSS patients from controls were calculated using Receiver-Operating Characteristics (ROC) curves.

Results: We included 50 pSS patients (mean (SD) age: 56.2 (13.7); 98.0% females) and 25 healthy controls (mean (SD) age 53.5 (9.2); 96.0% females) (Table 1). For SWE and B-mode modalities, inter-rater reliability was moderate to good (ICC: 0.64 and ICC:0.78-0.95, respectively), while intra-rater reliability was good to excellent (ICC:0.74-0.83 and ICC:0.95-0.98, respectively). Hocevar scores were significantly higher in pSS patients than in controls ($p<0.001$) in all four glands examined (Table 2). The mean total SWV (2.09 m/s (0.32); $p<0.001$), mean parotid SWV (2.25 m/s (0.53); $p<0.001$) and mean submandibular SWV values (1.86 m/s (0.36); $p<0.001$) were significantly higher in patients than in controls. No significant correlation between Hocevar scores and SWV values was observed. The cut-offs of 1.95 m/s, 1.96 m/s and 1.60 m/s for total, parotid and submandibular SWV

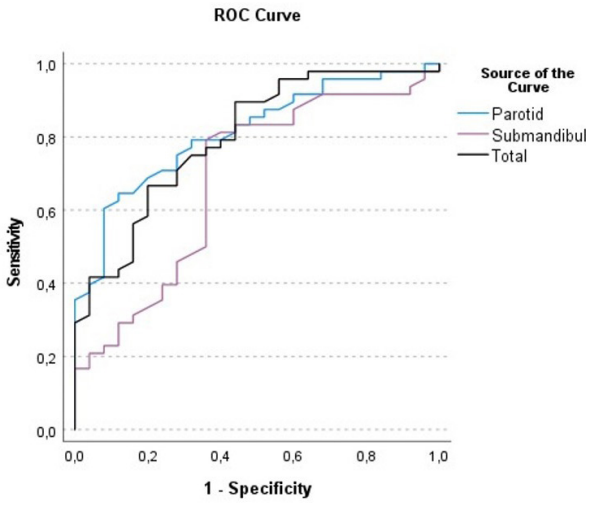
values differentiated pSS patients from controls with a sensitivity of 65.3%, 77.6% and 79.2% and specificity of 80.0%, 68.0% and 64.0%, respectively. Diagnostic performance was similar for total SWV and B-mode Hocevar score (AUROC = 0.85), p= 0.420. For patients with moderate, non-diagnostic, Hocevar scores, total SWV diagnosed pSS with a sensitivity of 75.0% and specificity of 71.0%. The area under the ROC curve (AUROC) was not significantly different between total SWV (AUROC=0.78) and parotid SWV (AUROC=0.81), p=0.696. Both were significantly greater than AUROC for submandibular SWV (AUROC=0.68), p<0.05 (Figure 1).

TABLE 1. SHEAR-WAVE ELASTOGRAPHY AND B-MODE FINDINGS FOR THE FOUR MAJOR SALIVARY GLANDS AT PROTOCOLIZED EVALUATION.

	pSS (N=50)	Controls (N=25)	p value
Shear-wave velocity (m/s)			
Total, mean (SD)	2.09 (0.32)	1.74 (0.24)	<0.001
Left parotid gland, mean (SD)	2.25 (0.40)	1.83 (0.30)	<0.001
Right parotid gland, mean (SD)	2.25 (0.53)	1.83 (0.31)	<0.001
Left submandibular gland, mean (SD)	1.94 (0.45)	1.70 (0.32)	0.020
Right submandibular gland, mean (SD)	1.90 (0.45)	1.61 (0.32)	0.005
Hocevar et al. (range 0-48)			
Total, mean (SD)	22.2 (8.76)	3.72 (3.46)	<0.001
Left parotid gland, mean (SD)	5.14 (2.45)	0.92 (1.26	<0.001
Right parotid gland, mean (SD)	5.68 (2.43)	0.68 (0.85)	<0.001
Left submandibular gland, mean (SD)	5.68 (2.27)	1.28 (1.31)	<0.001
Right submandibular gland, mean (SD)	5.76 (2.29)	0.84 (1.11)	<0.001

pSS- Primary Sjögren's syndrome

FIGURE 1. RECEIVER OPERATING CHARACTERISTICS ANALYSIS FOR PAROTID, SUBMANDIBULAR AND TOTAL MEAN SHEAR WAVE VELOCITY.



Conclusion: SWE may represent a relevant additional tool for the diagnosis of pSS. SWV values did not correlate with B-mode, probably due to the difficulty in scoring severely fibrotic glands using Hocevar score. Larger studies including patients with pSS and patients with sicca syndrome, as well as standardization of SWE protocols, are warranted to assess the role of SWE in the diagnostic algorithm of pSS.

REFERENCES

1. Ramsubeik K et al. Ther Adv Musculoskelet Dis. 2020; 12:1--21
2. Santiago T et al. Clin Exp Rheumatol. 2016; Sep-Oct;34 Suppl 100(5):137-141.
3. Hocevar A et al. Eur J Radiol. 2007;Sep;63(3):379-83.

141 - CLINICAL AND IMMUNOLOGICAL FEATURES OF A PORTUGUESE COHORT OF MIXED CONNECTIVE TISSUE DISEASE

Francisca Guimarães[1], Manuel Silvério-António[2, 3], Martins-Martinho J [2, 3], Ana Teresa Melo[2, 3], Daniela Santos Oliveira[4, 5], Jorge Pestana Lopes[6], André Saraiva[7], Laura Gago[8], Ana Margarida Correia[9], Ana Lúcia Fernandes[10], Sara Paiva Dinis [11], Rafaela Nicolau[12], Susana P. Silva[13], Carlos Costa[14], Tiago Beirão[15], Carolina Furtado[16], Pedro Abreu[17, 18], Carmo Afonso[1], Daniela Peixoto[1]

[1]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal; [2]Rheumatology and Bone Metabolic Diseases Department, Centro Hospitalar Lisboa Norte, EPE - Hospital de Santa Maria, Lisbon Academic Medical Centre, Lisboa, Portugal; [3]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [4]Rheumatology Department, Centro Hospitalar de São João, Porto, Portugal; [5]Faculdade de Medicina da Universidade do Porto, Departamento de Medicina e CINTESIS, Porto, Portugal; [6]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal; [7]Rheumatology Department, Centro Hospitalar Universitário de Coimbra, Coimbra, Portugal; [8]Rheumatology Department, Centro Hospitalar Lisboa Ocidental, Lisboa, Portugal; [9]Rheumatology Department, Hospital de Braga, Braga, Portugal, [10]Rheumatology Department, Hospital de Faro, Faro, Portugal; [11]Rheumatology Department, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal; [12]Rheumatology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal; [13]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal; [14]Rheumatology Department, Centro Hospitalar Trás-os-Montes e Alto Douro, Vila Real, Portugal;

[15]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal; [16]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal; [17]Rheumatology Department, Unidade Local de Saúde de Castelo Branco, Castelo Branco, Portugal; [18]Faculdade de Ciências da Saúde, Universidade da Beira Interior, Covilhã, Portugal

Background: Mixed connective tissue disease (MCTD) represents a rheumatic disorder with overlapping features of systemic lupus erythematosus (SLE), systemic sclerosis (SSc) and polymyositis1. High titers of anti-U1-RNP autoantibodies are mandatory for the diagnosis of this condition.

There has been an effort to better understand this entity and other nationwide studies have been already published. However, national data are not available.

Objectives: To characterize clinical and immunological features of a Portuguese cohort of patients with MCTD.

Methods: Retrospective, multicenter study including adult-onset patients with clinical diagnosis of MCTD and fulfilling at least one of the following classification criteria: Sharp's criteria, Kasukawa diagnostic criteria, Alarcón-Segovia criteria or the Kahn's criteria. SPSS was used for statistical analysis and significance level was defined as 2-sided p<.05.

Results: A total of 98 patients from 16 Portuguese rheumatology centers were included, with a mean age at diagnosis and disease duration of 40.5±13.7 and 7.0±6.5 years, respectively. Most patients were female (87.8%) and Caucasian (70.4%). Raynaud's phenomenon (96.9%), arthralgia/arthritis (94.9%/74.5%) and puffy fingers (60.2%) were the most common and early manifestations. Gastrointestinal (GI), respiratory and muscular involvement were also prevalent, especially during the follow up, affecting 30.6%, 32.7% and 43.9% of the patients, respectively. ANA and anti-U1-RNP were positive in all patients. Clinical and immunological characteristics are described in Table 1. Males were older at symptom's onset (65.0 VS 46.7, p=.035), presenting with more pleural effusion (OR=17.0, 95% CI 1.41-204.69), and positivity to anti-ACPA (OR=20.0, 95% CI: 3.09-129.35). GI involvement occurred more often in Caucasian patients (OR=3.79; 95% CI: 1.024-14.08), while anemia of chronic diseases and weight lost were more frequent in Afro-American patients. These patients were also younger at disease onset than Caucasians (34.1 VS 50.6, p=.01). Most patients fulfilled the Kasukawa classification criteria (91.8%), followed by Alarcon-

TABLE 1. SOCIODEMOGRAPHIC, CLINICAL AND IMMUNOLOGICAL CHARACTERISTICS

Sociodemographic characteristics, n=98	
Female gender, n (%)	86 (87.8)
Female-to-male ratio	7:1
Ethnicity, n (%)	
Caucasian	69 (70.4)
Afro-American	23 (23.5)
Disease characteristics	
Age at first symptom, M±SD, years	36.5±13.1
Age at diagnosis, M±SD, years	40.5±13.7
Disease duration, M±SD, years	7.0±6.5
Comorbidities	
Smoking status, n (%)	
Non-smoker	69 (70.4)
Smoker	7 (7.1)
Past smoker	7 (7.1)
Hypertension, n (%)	25 (25.5)
Obesity, n (%)	3 (3.1)
Dyslipidemia, n (%)	14 (14.3)
Diabetes, n (%)	4 (4.1)
Peripheral arterial disease, n (%)	2 (2)
Stroke, n (%)	2 (2)
Myocardial infarction, n (%)	1 (1)
Pulmonary embolism, n (%)	3 (3.1)
Coronary disease, n (%)	2 (2)
Cancer, n (%)	5 (5.1)
Hematological cancer, n (%)	0 (0)
Solid cancer, n (%)	5 (5.1)
Clinical Manifestations	At presentation
Mucocutaneous system	
Raynaud's phenomenon, n (%)	85 (86.7)
Digital ulcers, n (%)	15 (15.3)
Puffy hands, n (%)	48 (49.0)
Sclerodactyly, n (%)	35 (35.7)
Skin thickening, n (%)	20 (20.4)
Malar rash, n (%)	19 (19.4)
Subacute lupus, n (%)	3 (3.1)
Alopecia, n (%)	9 (9.2)
Nasal/oral ulcers, n (%)	10 (10.2)
Sicca symptoms, n (%)	16 (16.3)
Musculoskeletal system	
Arthralgia/Arthritis, n (%)/n (%)	81 (82.7)/56 (57.1)
Erosive arthritis, n (%)	2 (3.8)
Myositis, n (%)	26 (25.6)
Hematological system	
AIHA, n (%)	2 (2.0)
Anemia of Chronic Disease, n (%)	25 (25.5)

continues on the next page

TABLE 1. CONTINUATION

Leucopenia, n (%)	25 (25.5)
Thrombocytopenia, n (%)	7 (7.1)
Lymphadenopathy, n (%)	8 (8.2)
Respiratory system	
Pleural involvement	1 (1.0)
Pulmonary involvement	13 (13.3)
ILD	-
NSIP	-
UIP	-
LIP	-
other	-
Shrinking lung	-
Muscular	-
Other	-
Cardiovascular system	
Pericarditis	2 (2.0)
Myocarditis	1 (2.0)
Pulmonary hypertension	2 (2.0)
Gastrointestinal system	
Gastroesophageal involvement, n (%)	11 (11.2)
Dysphagia, n (%)	-
Gastroesophageal reflux, n (%)	-
Esophageal dysmotility, n (%)	-
AIH, n (%)	1 (1.0)
Renal System	2 (2.0)
Urinary sediment alterations, n (%)	2 (2.0)
Membranous nephropathy, n (%)	0 (0)
Lupus Nephritis, n (%)	0 (0)
Nervous system	
Headache, n (%)	4 (4.1)
Seizure, n (%)	1 (1.0)
Meningitis, n (%)	1 (1.0)
Neuropathy, n (%)	3 (3.1)
Constitutional symptoms	
Fever, n (%)	14 (14.3)
Night sweats, n (%)	5 (5.1)
Weight loss, n (%)	20 (20.4)
Immunological characteristics	
ANA, n (%)	98 (100)
Anti-U1-RNP, n (%)	98 (100)
Anti-dsDNA, n (%)	21 (21.4)
Anti-SM, n (%)	21 (21.4)
Anti-Ro/SSA, n (%)	31 (31.6)
Anti-La/SSB, n (%)	7 (7.1)
Anti-Scl-70, n (%)	0 (0)
Anti-centromere, n (%)	3 (4.1)
Anti-Jo1, n (%)	0 (0)
Rheumatoid Factor, n (%)	39 (39.8)
Anti-ACPA, n (%)	6 (6.1)
Lupus anticoagulant, n (%)	7 (7.1)
Anti-cardiolipin, n (%)	6 (6.1)
Anti- Beta-2 glycoprotein, n (%)	6 (6.1)
Myositis antibodies, n (%)	9 (9.2)
Direct Combs test, n (%)	5 (5.1)
Complement activation, n (%)	27 (27.6)
ESR elevation, n (%)	71 (72.4)
Hypergammaglobulinemia, n (%)	51 (52.)
Cryoglobulins, n (%)	1 (1.0)
Treatment	
Hydroxychloroquine, n (%)	63 (64.3)
Glucocorticoids, n (%)	71 (72.4)
Methotrexate, n (%)	33 (33.7)
Leflunomide, n (%)	2 (2.0)
Azathioprine, n (%)	15 (15.3)
Mycophenolate Mofetil, n (%)	9 (9.2)
Cyclosporine, n (%)	0 (0.0)
Cyclophosphamide, n (%)	1 (1.0)
Tacrolimus, n (%)	1 (1.0)
Rituximab, n (%)	5 (5.1)
Belimumab, n (%)	0 (0.0)
TNFi, n (%)	0 (0.0)
IVIG, n (%)	2 (2.0)
Iloprost, n (%)	6 (6.1)
Nifedipine, n (%)	35 (35.7)
Number of previous immunomodulator drugs *, M±SD (min-max)	0.54±0.9 (0-5)

continues on the next column

Legend: AIH: Autoimmune hepatitis; AIHA: Autoimmune hemolytic anemia; ANA: antinuclear antibodies; anti-ACPA: anti-citrullinated protein antibodies; Anti-dsDNA: anti-double stranded deoxyribonucleic acid antibody; Anti-SM: anti-smith antibody; anti-U1-RNP: anti-U1 ribonucleoprotein antibody; ESR: erythrocyte sedimentation rate; ILD: Interstitial lung disease; IVIG: Intravenous immunoglobulinLIP: Lymphocytic interstitial pneumonitis; M: mean; NSIP: Nonspecific interstitial pneumonia; SD: standard deviation; TNFi: Tumour necrosis factor inhibitors; UIP: Usual interstitial pneumonia. *except for hydroxychloroquine and glucocorticoids.

Segovia (29.6%), Sharp's (28.6%), and Kahn's criteria (22.4%). After a median follow-up time of 4 (IQR 8) years, 4 deaths were verified (4.1%), mostly (75%) due to infectious complications.

Conclusions: Raynaud's phenomenon, puffy fingers and arthritis were the most common manifestations in Portuguese patients, with similar proportions found in literature. However, our study reported lower prevalence of esophageal dysmotility, renal involvement and serositis and higher proportions of pulmonary hypertension and sclerodactyly than other studies. These differences may be explained by the heterogeneity in the inclusion criteria seen in different studies. This study showed no differences regarding gender and ethnicity regarding most clinical manifestations and immunological features with some exceptions, as reported before.

It still remains controversial if MCTD is a distinct disease. Yet, comparing to SLE and SSc published Portuguese cohorts, we reported different prevalence especially regarding renal and mucocutaneous manifestations, pulmonary hypertension and myositis, showing that there are some distinct clinical features of MCTD.

Here, we characterize the largest cohort of MCTD in Portugal, however more studies are needed to better understand this clinical entity.

147 - THE IMPACT OF ANTINUCLEAR ANTIBODIES INDUCED BY ANTI-TUMOUR NECROSIS FACTOR ALPHA (ANTI-TNF-α) AGENTS ON THE LONG-TERM TREATMENT OUTCOMES IN RHEUMATOID ARTHRITIS PATIENTS

Daniela Santos Oliveira[1, 2], Ana Martins[1], Frederico Rajão Martins[3], Maria Seabra Rato[1], Filipe Oliveira Pinheiro[1], Diogo Guimarães da Fonseca[4], Salomé Garcia[1], Bruno Miguel Fernandes[1], Carlos Vaz[1, 5], Miguel Bernardes[1, 5], Lúcia Costa[1]

[1]Rheumatology Department, Centro Hospitalar São João, Porto, Portugal; [2]Center for Health Technology and Services Research (CINTESIS), Faculty of Medicine, University of Porto, Porto, Portugal, Porto, Portugal; [3]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal; [4]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal; [5]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal

Background: The seroconversion of antinuclear antibodies (ANA) induced by anti-TNF-α therapy remains a matter of concern in rheumatoid arthritis (RA). However, evidence is still scarce regarding the impact of these autoantibodies on the clinical response to treatment.

Aims: This study aimed to explore the impact of ANA induced by anti-TNF-α therapy on the outcomes of treatment in patients with RA over two years of follow-up.

Methods: An observational retrospective cohort study was conducted with two years of follow-up. Patients diagnosed with RA, according to the American College of Rheumatology (ACR) criteria, and registered on the Rheumatic Diseases Portuguese Register (Reuma.pt) who started their first anti-TNFα agent as first biologic between 2003 and 2018 were included. Patients with positive ANA (titer ≥100) and/or positive anti-dsDNA antibodies and/or with a diagnosis of SLE at their first visit were excluded. Demographic, clinical and laboratory data were obtained by consulting Reuma.pt. Disease Activity Score for 28 joints (DAS28), DAS28 delta, Health Assessment Questionnaire (HAQ), HAQ delta were assessed at baseline, 6, 12, 18 and 24 months(M). Clinical response was evaluated by EULAR criteria and three response categories were defined: good, mild and no response. The rate of switch of biological treatment was assessed over 24M. To examine the differences between groups with and without ANA seroconversion independent samples t test, Mann-Whitney U-tests and Chi-square tests were used. Logistic regression models were used to assess the effects of ANA seroconversion on clinical response to treatment over time.

Results: A total of 185 patients (mean age of 49.3±10.9 years old; 85.4% female) with a median follow-up of 7 [4-14] years were included. We found an ANA seroconversion rate (titer≥100) of 77.3% (n=143) with median time of 36 [15-72.3] months. There were no differences among groups regarding age, gender, disease duration, be seropositivity or not and have an erosive disease or not. DAS28 delta was significantly different (p=0.035) between group with positive ANA (2.01±1.29) and negative ANA (1.15±1.51) at 6M. DAS28 was significantly different (p=0.014) between group with positive ANA (5.06±3.39) and negative ANA (3.99±1.43) at 12M. No statistically significant differences were found in other variables. Switch rate was significantly different between patients with ANA seroconversion (median 1[0-1]) versus absence of seroconversion (median 0[0-1]), p=0.025. In the

regression model ANA seroconversion did not predict switch rate and EULAR response over time.
Conclusions: This study showed that the majority of patients with RA treated with an anti-TNF-α agent developed ANA and that their presence may be associated with worse clinical results over time. In fact, previous research suggested that a decrease in anti-TNF-α drug concentration due to the production of autoantibodies may lead to worse outcomes of treatment. Moreover, our data demonstrated that patients with ANA seroconversion had a higher switch rate. Despite these results, there are no differences in the EULAR response between the two groups and ANA seroconversion did not predict this response. Therefore, ANA induced by anti-TNF-α agents should be monitored in patients with RA and its impact on treatment must be considered. Further research is needed to explore these results.

150 - THE IMPACT OF A FRACTURE LIAISON SERVICE ON FIRST-YEAR MORTALITY IN HIP FRACTURE PATIENTS

Sara Paiva Dinis [1], Filipe Cunha Santos[1], Ana Sofia Pinto[1], Gustavo Vieira Factori[2], Carlos Pontinha[3], Paula Rocha[3], Gina Monteiro[3], Isabel Neves[3], Ana Rita Batista[3], Sérgio Caseiro Anunciação[3], Cláudia Vaz[1, 2], Luís Matos[4], Joana Fonseca Ferreira[1, 2]
[1]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal; [2]Faculdade de Ciências da Saúde, Universidade da Beira Interior, Covilhã, Portugal; [3]Serviço de Ortopedia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal; [4]Serviço de Nutrição, Unidade Local de Saúde da Guarda- Hospital Sousa Martins, Guarda, Portugal

Introduction: A Fracture Liaison Service (FLS) is a secondary fracture prevention model by identifying patients at risk for fragility fractures and initiating the appropriate treatment. The introduction of a FLS model was previously described to improve outcomes of osteoporosis-related fractures, namely reducing mortality rates. Our objective is to analyze the effect of the FLS model over the first-year mortality rates following a hip fracture.
Methods: We conducted a case-control study in two populations who suffered a hip fracture during two non-consecutive years, before and after FLS implementation in our center. The first group of patients was diagnosed between January and December of 2017, before FLS implementation. Patients in the second group were evaluated between January and December of 2019 after FLS protocol implementation. Patients' information was withdrawn from our electronic health records database. First-year mortality rates were compared between the two groups using a logistic regression model adjusted for age. Data were analyzed with SPSS 27.0 software.
Results: A total of 366 individuals were included in this study (316 before FLS implementation and 50 after FLS implementation). There were statistically significant age differences between the groups ($p < 0.05$). In the group managed before the FLS implementation, 76.6% were female with a mean age of 84.2 years (SD: 8.6). In the group evaluated in our FLS, 86.0% were female with a mean age of 79.6 years (SD: 8.1). One (2.0%) patient of the FLS group and a total of 77 patients (24.4%) before FLS implementation died during the follow-up period. Patients who were evaluated at our FLS had a lower 1-year mortality compared with patients managed before the implementation of the FLS program [adjusted odds ratio (OR) 0.090; 95% confidence interval (CI) 0.012– 0.669; $p < 0.05$].
Conclusions: We observed significant 1-year-mortality differences following a hip fracture between patients treated before and after implementation of the FLS protocol. A FLS appears to be a successful approach to reduce 1-year mortality in this group of patients.

157 - WHAT DO WE KNOW ABOUT TERIPARATIDE ON DELAYED AND NON-UNION FRACTURE TREATMENT – A SYSTEMATIC REVIEW

Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Tiago Beirão[1], Flávio Campos Costa[1], Romana Vieira[1], Taciana Videira[1], Joana Aleixo[1], P Pinto[1]
[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Background: It is estimated that 5-10% of fractures do not heal properly leading to delayed and non-union fractures, which remains challenging to treat. Teriparatide is a bone anabolic drug approved for the treatment of osteoporosis. Recently, it has been suggested that it can also promote bone fracture healing.
Objective: To systematically identify and review literature evaluating the efficacy and safety of teriparatide on delayed and non-union fractures.
Material and Methods: A systematic literature search was performed in MEDLINE using the query «"fracture"

AND "nonunion" AND "teriparatide"». All retrieved articles were screened by title and abstract and the eligible ones were kept for full-text review. Reference lists were additionally searched. Original research papers encompassing treatment of delayed and non-union fractures with teriparatide in patients aged ≥18 years old were considered for inclusion.

Results: The systematic review identified 63 search results, of which 36 were excluded after title and abstract screening because they did not fit the purpose of the work. After full-text review and addition of 5 articles from reference lists, 28 original research papers were included: 20 case reports, 6 case series and 2 longitudinal uncontrolled studies, in a total of 81 patients, 48 females (59,3%), with a median age [min, max] of 64 [19, 91] years-old. Several fracture sites have been described, with the femur (n = 35) and the tibia (n = 26) being the most frequently affected. As for fracture mechanisms, 30 occurred after trauma and 17 were fragility fractures and, 39 patients were submitted to surgery before teriparatide. The study with the largest number of patients, however, did not describe the mechanisms nor the number of patients with previous surgery. The median time [min, max] between initial fracture and start of teriparatide was 7 [3, 36] months. Most studies used daily injection of 20 µg teriparatide, except 4 studies that used the weekly formulation of 56.5 µg. The median [min, max] duration of treatment was 6 [0.8, 24] months. Overall, 77 (95,1%) had fracture union following a median [min, max] time of 7 [3, 31] months. Only 1 patient (1,2%) reported minor adverse effects with teriparatide, although this information is missing in 22 patients. It should be noted that in 9 patients teriparatide was combined with revision surgery and in 1 patient with low-intensity pulsed ultrasound.

Conclusion: Despite being an off-label use, the existing evidence suggests that teriparatide may be effective and safe for the treatment of delayed and non-union fractures. However, there may be publication bias. Prospective randomized clinical trials are lacking and are needed to confirm these results.

177 - ESCLEROSE SISTÉMICA E TROMBOEMBOLISMO PULMONAR – CASUÍSTICA DE UM CENTRO

Maria Margarida Cunha[1], Ana Catarina Duarte[1], Ana Cordeiro[1], Filipe Vinagre[1]

[1]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal

A esclerose sistémica (ES) é uma doença do tecido conjuntivo com atingimento multissistémico, nomeadamente vascular. Vários trabalhos têm demonstrado o aumento da incidência de tromboembolismo venoso, em particular pulmonar, em várias doenças reumáticas sistémicas.

Descrevemos as características dos doentes com esclerose sistémica e tromboembolismo pulmonar (TEP) num Serviço de Reumatologia.

Foram identificados em Reuma.pt os doentes com seguimento entre 2011 e 2021 em protocolo de Esclerose Sistémica. Foram incluídos doentes que cumpriam os critérios de classificação ACR/EULAR 2013 para ES e formas precoces e/ou overlap de doença. Desses, foram seleccionados os que apresentaram TEP ao longo do curso da doença.

Dos 148 doentes identificados com o diagnóstico de ES, 23 (15.5%) tiveram pelo menos um episódio de TEP. Vinte e dois (95.7%) eram do sexo feminino; à data do diagnóstico de TEP, os doentes tinham idades compreendidas entre os 26 e os 79 anos, com uma mediana de idades de 68; a mediana do tempo decorrido entre o diagnóstico de ES e de TEP foi de 13 meses, sendo que em 7 (30.4%) doentes o TEP ocorreu em simultâneo com o diagnóstico inaugural da doença. Quinze doentes estavam classificados como forma cutânea limitada, 4 como forma cutânea difusa, 2 como forma pré-clínica, 1 como overlap polimiosite/ES e 1 como ES sine escleroderma. Todos apresentavam ANA positivo (100%), 11 (47%) com anticorpos anti-centrómero, 7 (30.4%) com anti-topoisomerase I, 4 (17.4%) anti-SSA, 3 (13%) anti-Ro52, 1 (4.35%) anti-fibrilarina, 1 (4.35%) anti-RNA polimerase III, 1 (4.35%) anti-PM/Scl . Em 12 dos 23 doentes foram pedidos os anticorpos anti-fosfolípidos que foram negativos. À data do TEP, 6 (26%) doentes estavam medicados com corticosteróides orais, 4 dos quais com prednisolona em dose igual ou inferior a 7.5mg/dia ou equivalente, e dois com prednisolona em dose superior a 7.5mg/dia ou equivalente. Apenas 6 (26%) doentes apresentavam um factor provocador clássico para tromboembolismo venoso (3 eram fumadores activos, 2 eram ex-fumadores, um tinha estado recentemente hospitalizado e um apresentava uma trombofilia de alto risco). O TEP foi sintomático em 18 (78.3%) dos 23 doentes, motivando internamento em apenas 2 (8.7%) dos casos, por insuficiência respiratória parcial; nos restantes a manifestação que motivou a investigação diagnóstica foi cansaço ligeiro e arrastado. Nos doentes assintomáticos o motivo de pesquisa de TEP foi

diminuição desproporcional da capacidade de difusão pulmonar para o monóxido de carbono (DLCO) em 3 (13%) dos casos e aumento da pressão sistólica na artéria pulmonar estimada pelo ecocardiograma em 2 (8.7%). Não se verificaram óbitos na sequência do evento tromboembólico. O fármaco anticoagulante mais utilizado foi o rivaroxabano. Apenas 1 doente teve recorrência de TEP. Verificou-se a evolução para hipertensão pulmonar tromboembólica crónica em 3 casos, em doentes com 6, 25 e 54 meses de seguimento pós evento.

O TEP é uma comorbilidade prevalente entre doentes com ES, frequentemente sintomático e com consequente impacto na qualidade de vida. Em alguns doentes pode mesmo haver evolução para hipertensão pulmonar tromboembólica crónica, sendo um factor contributivo para a hipertensão pulmonar. Embora frequentemente sintomático, a clínica pode ser frustre, sendo importante manter uma elevada suspeição para o diagnóstico perante doentes com cansaço e diminuição não explicada e desporpocional da DLCO, com vista ao início atempado da anticoagulação.

180 - MEDICAL CARE IS FREQUENT AND ASSOCIATED WITH OVERUSE OF LOW-VALUE CARE FOR LOW BACK PAIN IN PORTUGAL: RESULTS FROM A NATIONWIDE POPULATION-BASED STUDY

Gomes, LA[1, 2], Cruz, EB[1, 3], Ana Rita Henriques[1, 2], Jaime C. Branco[1, 2, 4], Helena Canhão[1, 2], Rodrigues AM[1, 2]

[1]Comprehensive Health Research Centre, Lisboa, Portugal [2]EpiDoc Research Unit, CEDOC, NOVA Medical School, Lisboa, Portugal; [3]Fisioterapia, Escola Superior de Saúde - Instituto Politécnico de Setúbal, Setúbal, Portugal; [4]Serviço de Reumatologia do Hospital Egas Moniz - Centro Hospitalar Lisboa Ocidental (CHLO- E.P.E.), Lisboa, Portugal

Background: Low back pain (LBP) constitutes an important cause of disability and morbidity in general population. Appropriate care for LBP includes non-pharmacological interventions based on a biopsychosocial framework of patient education and advice. World Health Organization recommends that excessively medical solutions should be avoided. This study aimed to estimate prevalence of medical care seeking and characterize diagnostic workup and management procedures for LBP in the adult Portuguese population. We also aim to compare the medical framework for LBP diagnostic and management between primary and secondary care.

Methods: The present study was conducted under the scope of EpiReumaPt (2011-2013), a population-based study including a representative sample of non-institutionalized Portuguese adults (n=10,661 habitants). A sample of individuals who self-reported history of medical care seeking for LBP within the previous 12 months (n=2,618) were considered. Patients' self-reported data collected through a structured questionnaire was explored to characterize medical care seeking, and diagnostic and management procedures for LBP. Prevalence was computed as weighted proportions, and inference statistics used to compare medical procedures between different levels of care.

Results: A prevalence of medical care seeking for LBP of 38.0% (95%IC, 35.9 to 40.1%) was found. Primary care in isolation (45.3%), multiple care (primary plus secondary care) (28.8%) and secondary care in isolation (25.9%) were the sought levels of care for LBP. Emergency departments (25.9%) and orthopedics (19.4%) were the secondary medical specialties most used. Several distinct structural-based diagnosis and specific/ serious underlying diseases were diagnosed by physicians, mainly supported by laboratory and imaging tests performed to 91.1% of individuals. Disc herniation (20.4%) and osteoarthritis (19.7%) were the most frequent diagnosis, while x-rays (63.7%), clinical history/ observation (44.4%), blood tests (38.2%), urinalysis (34.4%), and CT scans (32.4%) were the most frequent diagnostic procedures. Only 8.5% of individuals were evaluated based on a clinical history/ observation procedure in isolation. Lastly, 75.1% of individuals self-reported being treated for LBP by their physician, 80.4% with oral medication/ pills and 15.3% with injectables. The mean duration of pharmacological treatment was 104.24 (266.80) days. The use of structural-based diagnosis, laboratory and imaging tests, and pharmacological treatment were generally aggravated when secondary care, in isolation or complementarily with primary care, was considered compared with primary care in isolation ($p<0.05$).

Conclusion: Our results show that medical seeking for LBP is frequent and is associated to overdiagnosis through a diagnostic label despite there being no reliable way of determining the pathoanatomical source of pain for the great majority of patients. Medical seeking is also associated with overuse of pharmacological treatment. Funding and delivery actions should be prioritized to assure appropriate care of LBP and reduce this low-value care overuse within local health systems.

181 - THE SPLIT STRATIFIED MODEL OF CARE FOR LOW BACK PAIN – RESULTS FROM AN IMPLEMENTATION STUDY IN THE PORTUGUESE CONTEXT OF PRIMARY HEALTHCARE

Gomes, LA[1, 2, 3], Fernandes, R.[1, 3], Caeiro, C[3], Fernando Pimentel-Santos[4, 5], Jaime C. Branco[1, 2, 5], Rute Sousa[1, 2], Pinto, I.[6], Moniz, R.[6], Helena Canhão[1, 2], Rodrigues AM[1, 2], Cruz, EB[1, 3]
[1]Comprehensive Health Research Centre, Lisboa, Portugal; [2]EpiDoc Research Unit, CEDOC, NOVA Medical School, Lisboa, Portugal; [3]Fisioterapia, Escola Superior de Saúde - Instituto Politécnico de Setúbal, Setúbal, Portugal; [4]NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal; [5]Serviço de Reumatologia do Hospital Egas Moniz - Centro Hospitalar Lisboa Ocidental (CHLO- E.P.E.), Lisboa, Portugal; [6]ACES Arrábida, Setúbal, Portugal

Background: The expected increase of Low Back Pain (LBP) related prevalence and disability represents an urgent challenge for Portuguese health systems. Most episodes of LBP have little or no consequences however, the great disability reported by a substantial minority of patients, those with persistent disabling LBP, explains the disturbing social and economic impact of LBP. A stratified model of care based on the early identification of patients' risk for developing persistent disabling LBP has been shown to be effective for reducing disability of patients who seek primary healthcare. Since this model of care was never tested for the Portuguese context, the present study aimed to compare the effectiveness of a stratified model of care (SPLIT) with usual care for reducing persistent disabling pain of patients with LBP in primary healthcare.
Methods: A controlled before-after study was conducted. Two sequential, independent cohorts of patients with LBP (L03, L84, and L86) were recruited during an 8-month period each in 7 primary healthcare units of ACES Arrábida of ARSLVT, Portugal. The first cohort evaluated the clinical results of usual care, while in the second cohort SPLIT was implemented. Between both cohorts, General Practitioners (GPs) and Physiotherapists were trained for the implementation of SPLIT, which used the STarT Back Screening Tool to stratify patients for matched physiotherapy treatment according to their risk of persistent disabling LBP (low, medium, or high risk). Changes at 2 and 6-month follow-up of disability (RMDQ, 0-24), pain (NPRS, 0-10), and global perception of change (GBRS, -5 to 5) were evaluated as patient-level outcomes. GPs' diagnostic and managing procedures were used as organizational-level outcomes.
Results: In total, 289 patients with LBP were enrolled: 115 in the usual care cohort (mostly treated with medication) and 174 in the SPLIT cohort. Overall, 58.2 and 53.8% of patients in the usual care, and 19.6 and 17.3% in the SPLIT cohort had persistent disabling LBP (RMDQ≥7) at 2 and 6-month follow-up, respectively. After adjustments, SPLIT was associated with a significantly lower probability of patients reporting RMDQ≥7 at 2 (OR: 0.23 [95%IC, 0.13-0.41]) and 6-month (0.21 [0.12-0.40]). The probability of improvement above the minimal clinically important difference was significantly higher for SPLIT compared with usual care for RMDQ (2-month: 4.24 [2.40-7.51]; 6-month: 3.51 [1.95-6.34]), NPRS (2-month: 4.32 [2.48-7.54]), and GBRS (2-month: 3.30 [1.89-5.77]; 6-month: 2.43 [1.36-4.34]). At 6-month, no differences for NPRS were found between SPLIT and usual care (1.46 [0.83-2.57]). After the implementation of SPLIT, imaging tests prescription decreased 30.0% (usual care: 47.8%; SPLIT: 17.8%). Referrals for physiotherapy increased 91.7% (usual care: 8.3%; SPLIT: 100.0%) whereas prescription of pharmacological therapy reduced 32.9% (usual care: 84.3%; SPLIT: 51.4%).
Conclusions: The implementation of SPLIT was associated with a reduction of almost 80% in the probability of patients to develop persistent disabling LBP at 6-month, compared with usual care. A future economic analysis will allow us to explore if this effectiveness was also reflected in lower costs.

184 - COST OF OSTEOPOROSIS-RELATED FRACTURES IN POSTMENOPAUSAL WOMEN IN PORTUGAL

Judite Gonçalves[1], Rodrigues AM[2], Anabela Barcelos[2], Helena Canhão[2], Céu Mateus[3]
[1]NOVA School of Business and Economics, Universidade NOVA de Lisboa, Lisboa, Portugal; [2]CEDOC, Nova Medical School, Lisboa, Portugal; [3]Lancaster University, Lancaster, United Kingdom

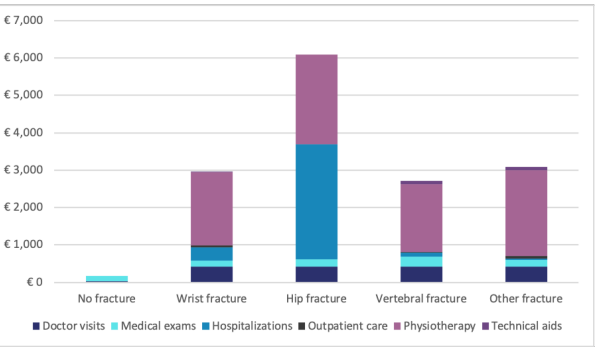
Background: With an aging population, osteoporosis has become a major public health concern. Low impact fractures, the main consequence of osteoporosis, result in increased morbidity and mortality and represent a major and growing economic burden on healthcare systems worldwide (1, 2). Postmenopausal women are at particularly high risk for osteoporosis and low impact fractures (2).

Methods: This retrospective observational study estimated the costs of osteoporotic fractures in postmenopausal (roughly defined as aged 50+) women in Portugal; specifically, direct costs from healthcare consumption in the first year following vertebral, hip, wrist, and/or "other" fracture types. An expert panel (two rheumatologists, one physiatrist, one orthopedist) defined typical healthcare consumption in the year following each type of fracture with a high level of detail (doctor visits by specialty, medical exams, procedures during hospitalization or outpatient care, and physiotherapy). We also considered medical equipment typically prescribed, which goes beyond the payer perspective because costs are borne out-of-pocket by the patients. Yearly incidence rates of non-hip fractures were estimated from the 2013-15 wave of the Epidemiology of Chronic Diseases cohort (EpiDoC), an observational, population-based cohort of non-institutionalized adults in Portugal (3). Incidence of hip fractures was estimated from the 2014 national Diagnosis Related Groups (DRG) database that covers all hospitalizations in the Portuguese National Health Service (NHS). Resources were valued according to the national tariffs practiced in the NHS in 2019, which were multiplied by typical utilization of each resource as defined by the expert panel (specific to each type of fracture), and finally multiplied by the numbers of yearly incident fractures.

Results: We estimated that, in Portugal, about 5,000 wrist, 9,500 hip, 3,500 vertebral, and 39,000 other-site fragility fractures occur each year among women aged 50+. Healthcare costs per patient range from €1,600 for wrist and "other" fractures (e.g., shoulder, lower leg), to € 4,500 for hip fractures. Physiotherapy accounts for the majority of costs, with the exception of hip fractures, where hospitalization costs represent approximately half the total healthcare costs. In Portugal, incident fragility fractures among women aged 50+ years cost an estimated € 116.5 million in direct healthcare costs per year, which compares with € 152 million yearly direct healthcare costs associated with heart failure, also among 50+ women (4). Although hip fractures cost about twice as much as non-hip fragility fractures, they only account for 1/5 of fragility fractures among 50+ women, and about 1/3 of the total estimated direct cost of osteoporosis-related fractures per year in Portugal.

Conclusions: Our results indicate that osteoporosis-related fractures in 50+ women in Portugal represent a significant economic burden, in particular for the NHS. Furthermore, while the focus has traditionally been on hip fractures, the prevention of non-hip fragility fractures, which account for 2/3 of the estimated costs, also deserves attention. Increased awareness amongst physicians, payers, decisionmakers, and patients may help to address the clinical, humanistic and economic burden of osteoporosis.

FIGURE 1. HEALTHCARE COSTS PER PATIENT IN THE YEAR FOLLOWING FRACTURE, BY TYPE OF FRACTURE.



189 - PREDICTORS OF RESPONSE: BASELINE CHARACTERISTICS AND EARLY TREATMENT RESPONSES ASSOCIATED WITH ACHIEVEMENT OF REMISSION AND LOW DISEASE ACTIVITY AMONG UPADACITINIB-TREATED PATIENTS WITH RHEUMATOID ARTHRITIS

Arthur Kavanaugh[1], Zoltán Szekanecz[2], Edward C. Keystone[3], Andrea Rubbert-Roth[4], Stephen Hall[5], Ricardo Xavier[6], Joaquim Polido Pereira[7], In-Ho Song[8], Naomi Martin[9], Yanna Song[8], Samuel I. Anyanwu[8], Peter Nash[10]

[1]Division of Rheumatology Allergy and Immunology, UCSD, San Diego, United States; [2]Division of Rheumatology , Faculty of Medicine, University Of Debrecen, Debrecen, Hungary; [3]MaRS Center, Toronto, Canada; [4]Kantonsspital St Gallen, St Gallen, Switzerland; [5]Emeritus Research and Monash University, Melbourne, Victoria, Australia; [6]Serviço de Reumatologia, Hospital das Clinicas de Porto Alegre, Porto Alegre, Brazil; [7]Rheumatology Department, Hospital de Santa Maria (CHLN), Lisbon Medical and Academic Centre, Lisboa, Portugal; [8]AbbVie Inc. North Chicago, Chicago, United States; [9]AbbVie, Inc, Quebéc, Canada; [10]University of Queensland, Brisbane, Queensland, Australia

Background: Upadacitinib (UPA) 15 mg once daily (QD) has demonstrated efficacy in phase 3 studies of patients with rheumatoid arthritis (RA).1–4 Early

prediction of response to treatment with UPA could help to optimize therapy.

Objectives: To identify baseline (BL) characteristics or Week (Wk) 12 disease activity measures that may predict the achievement of remission (REM) or low disease activity (LDA) at 6 months in patients with RA receiving UPA 15 mg.

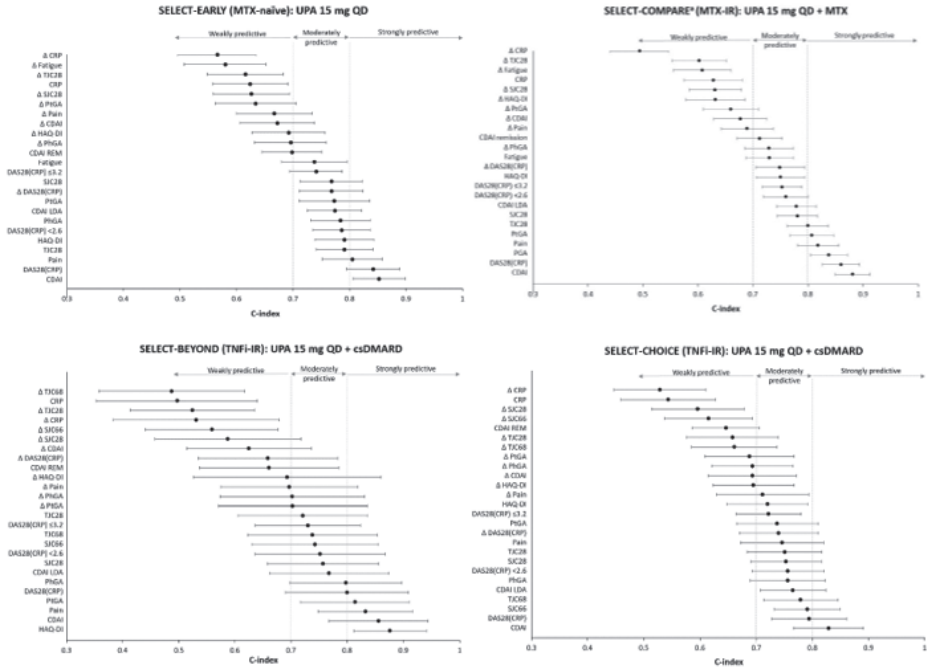
Methods: This ad hoc analysis included patients who were randomized to UPA 15 mg QD, as monotherapy in methotrexate (MTX)-naïve patients (SELECT-EARLY) or in combination with conventional synthetic disease-modifying antirheumatic drugs (csDMARDs), in patients with an inadequate response (IR) to MTX (SELECT-COMPARE) or ≥1 tumor necrosis factor inhibitors (TNFis) (SELECT-BEYOND and SELECT-CHOICE). The association of BL characteristics (including age, disease duration, prior/concomitant treatments, C-reactive protein [CRP], seropositivity, and disease activity) and Wk 12 disease activity parameters with the achievement of Clinical Disease Activity Index (CDAI) REM (≤2.8) or LDA (≤10) at Wk 24 (or Wk 26 in SELECT-COMPARE) was assessed by concordance statistics (C-statistics), or area under the receiver operator characteristic curve. C-index values and 95% confidence intervals were calculated by fitting a univariate logistic regression model for: demographic and BL characteristics, Wk 12 disease activity measures, and change from BL at Wk 12 in disease activity measures. A multivariate

TABLE. ACHIEVEMENT OF CDAI LDA AND REM AT WK 24/26[a]

	SELECT-EARLY	SELECT-COMPARE	SELECT-BEYOND	SELECT-CHOICE
Patient population	MTX-naïve	MTX-IR	TNFi-IR	TNFi-IR
Treatment	UPA 15 mg monotherapy (n=317)	UPA 15 mg + MTX (n=651)	UPA 15 mg + csDMARD (n=146)	UPA 15 mg + csDMARD (n=263)
Efficacy at Wk 24/26[a], n (%)				
CDAI REM (≤2.8)	90 (28.4)	150 (23.0)	16 (11.0)	60 (22.8)
CDAI LDA (≤10)	178 (56.2)	343 (52.7)	73 (50.0)	154 (58.6)

[a] Wk 26 for SELECT-COMPARE only

FIGURE. WK 12 PREDICTORS[a] FOR CDAI REMISSION AT WK 24/26[b]



[a]Determined by a univariate logistic regression model. C-statistics provide a C-index value from 0.5 (chance prediction) to 1 (perfect prediction). Error bars indicate 95% CI. [b] Wk 26 for SELECT-COMPARE only. CRP, C-reactive protein; DAS28(CRP), Disease Activity Score in 28 joints using C-reactive protein; HAQ-DI, Health Assessment Questionnaire-Disability Index; PhGA Physician Global Assessment; PtGA Patients Global Assessment; SJC28/66, swollen joint count in 28/66 joints; TJC28/68, tender joint count in 28/68 joints

logistic regression with stepwise model selection was also performed. The proportion of patients achieving Wk 24/26 CDAI REM/LDA was stratified by ≥50% improvement from BL in swollen and/or tender joint count in 66/68 joints (SJC66/TJC68).

Results: A total of 1377 patients were included in the analysis. Across the 4 studies, CDAI REM and LDA were achieved in 11.0–28.4% and 50.0–58.6% of patients, respectively (Table). BL demographics and disease characteristics were weakly predictive (C-index <0.70) of Wk 24/26 CDAI REM (C-index 0.49–0.69) or LDA (C-index 0.47–0.65), with the exception of BL Health Assessment Questionnaire-Disability Index in SELECT-BEYOND, which was moderately predictive of CDAI REM (C-index 0.73). Changes from BL in disease activity measures at Wk 12 were weakly or moderately predictive of Wk 24/26 CDAI REM (Figure) or LDA. CDAI value at Wk 12 was strongly predictive (C-index >0.80) of Wk 24/26 CDAI REM or LDA. Disease Activity Score in 28 joints using CRP and pain at Wk 12 were strongly predictive of Wk 24/26 CDAI REM (except in SELECT-CHOICE). Physician's global assessment at Wk 12 was the only common predictor in the multivariate regression models for CDAI REM/LDA at Wk 24/26 across the 4 studies. A greater proportion of patients achieving ≥50% improvement in SJC66 and TJC68 at Wk 12 achieved CDAI REM (16.5–37.8% vs 0–9.4%) or LDA (66.0–72.8% vs 20.9–35.7%) at Wk 24/26 than those who did not.

Conclusions: BL characteristics did not strongly predict response to UPA, but composite disease activity scores at Wk 12 predicted Wk 24/26 REM/LDA with UPA 15 mg QD across MTX-naïve, MTX-IR, and TNFi-IR patients. ≥50% improvement in SJC/TJC at Wk 12 was also associated with Wk 24/26 REM/LDA.

References: 1. van Vollenhoven R, et al. Arthritis Rheumatol 2020;72:1607–20; 2. Genovese MC, et al. Lancet 2018;391:2513–24; 3. Fleischmann R, et al. Arthritis Rheumatol 2019;71:1788–800; 4. Rubbert-Roth A, et al. N Engl J Med 2020;383:1511–21.

218 - PREVALENCE AND IMPACT OF UNDIAGNOSED RHEUMATIC AND MUSCULOSKELETAL DISEASES IN PORTUGAL: A SUBGROUP ANALYSIS OF THE EPIREUMAPT STUDY

Nuno Pina Gonçalves[1, 2, 3], Sofia Azeredo[3, 4, 5], Alexandre Sepriano[2, 3], Ana Rita Henriques[4], Tomás Pires[6], Jaime C. Branco[2, 3], Helena Canhão[4, 5, 7], Rodrigues AM[4, 5, 8]

[1]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal; [2]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal; [3]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal; [4]EpiDoC Unit, CEDOC, NOVA Medical School, NOVA University, Lisboa, Portugal; [5]Comprehensive Health Research Center (CHRC), Universidade NOVA de Lisboa, Portugal, Lisboa, Portugal; [6]Faculdade de Ciências Sociais e Humanas - Universidade Nova de Lisboa, Lisboa, Portugal; [7]Unidade de Reumatologia, Hospital de Santo António dos Capuchos - Centro Hospitalar Universitário de Lisboa Central, Lisboa, Portugal; [8]Unidade de Reumatologia, Hospital dos Lusíadas, Lisboa, Portugal

Introduction and objectives: Early diagnosis of rheumatic and musculoskeletal diseases (RMD) is of key importance in achieving effective treatment, improved prognosis and quality of life. The Portuguese national survey of RMD in 2011-2013 (EpiReumaPt) showed that RMD were frequently undiagnosed in Portugal. We aimed to characterize adults with undiagnosed RMD according to gender, age strata and region. Also, we compared health related quality of life (HRQoL), physical function and mental health between undiagnosed RMD, previously diagnosed RMD and no RMD population. Finally, we explored if undiagnosed RMD participants have higher health resources consumption.

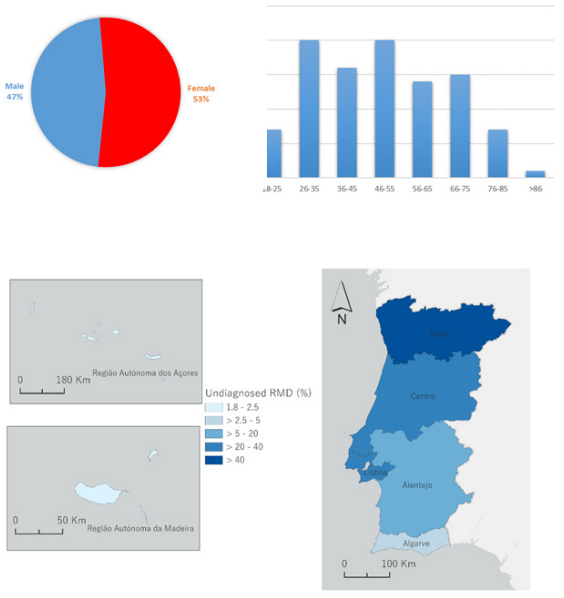
Methods: This study is a subgroup analysis of EpiReumaPt, a nationwide population based survey that determined the prevalence of RMD in Portugal. All participants aged 18 years or older evaluated by a rheumatologist were included. RMD were established by a trained rheumatologist using expert opinion and when possible international classification criteria. Participants were stratified into 3 groups: undiagnosed RMD: individuals who reported having no diagnosis RMD in a self-report questionnaire but had RMD diagnosed by the rheumatologist; previously diagnosed RMD: participants that reported a RMD that was confirmed by the rheumatologist; and non-RMD: participants who were confirmed by the rheumatologist to not have a RMD. Descriptive analysis of the undiagnosed population was weighted according to the study design. The three groups were compared regarding sociodemographic and health-related characteristics, HRQoL (EQ5D), physical function (HAQ), mental health (HADS) and health

TABLE 1: COMPARISON OF HEALTH STATUS BETWEEN PARTICIPANTS WITH AND WITHOUT UNDIAGNOSED RMDS: MULTIVARIABLE ANALYSIS

HRQoL and physical function	Undiagnosed RMD mean (SD)	Previously Established RMD mean (SD)	Non-RMD mean (SD)	B estimates; 95% CI (Undiagnosed RMD *vs* Non-RMD)	Adjusted p value	B estimates; 95% CI (Previously established RMD *vs* Non- RMD)	Adjusted p value
EQ5D (0-1)	0.80 (0.23)	0.61 (0.26)	0.89 (0.18)	-0.07 (-0.10, -0.04)	p<0.001	-0.23 (-0.27, -0.20)	p<0.001
HAQ (0-3)	0.28 (0.53)	0.72 (0.69)	0.09 (0.29)	0.10 (0.05, 0.15)	p<0.001	0.40 (0.33, 0.47)	p<0.001
Mental health	**Undiagnosed RMD n (%)**	**Previously Established RMD n (%)**	**Non-RMD**	**OR; 95%CI (Undiagnosed RMD *vs* Non-RMD)**	**Adjusted p value**	**OR; 95%CI (Previously established RMD *vs* Non- RMD)**	**Adjusted p value**
Anxiety							
Yes	235 (13.1)	325 (23.6)	103 (6.2)	2.3 (1.4, 3.7)	p<0.001	4.8 (2.8, 8.0)	p<0.001
No	1279 (86.9)	1134 (76.4)	801 (93.8)				
Depression							
Yes	111 (5.1)	221 (14.1)	46 (1.9)	1.4 (0.8, 2.4)	p=0.260	2.3 (1.4, 4.0)	p=0.002
No	1403 (94.9)	1238 (85.9)	858 (98.1)				

Sample size is not constant due to missing data: Undiagnosed RMD: EQ5D (n=1497), HAQ (n=1514); Established RMD: EQ5D (n=1450), HAQ (n=1459); Non-RMD: EQ5D (n=899)
p-values were adjusted for age, gender, nomenclature of Territorial Units of Statistics, years of education
NUTSII: Nomenclature of Territorial Units of Statistics (Norte, Centro, Lisboa, Alentejo, Algarve, Lisboa, Madeira and Azores;
EQ5D: European Quality of Life questionnaire five dimensions three levels; HAQ: Health Assessment Questionnaire; HRQoL: health-related quality of life; RMD: rheumatic and musculoskeletal disease.

FIGURE. PREVALENCE OF UNDIAGNOSED RMD IN PORTUGAL, STRATIFIED BY GENDER, AGE STRATUM AND NUTS II



resources consumption (number of clinical visits in the last year). The effect of being undiagnosed or previously diagnosed with an RMD on each outcome was assessed in multivariable models adjusting for age, gender, Portuguese nomenclature of territorial units of statistics and years of education (reference: non-RMD).

Results: In total, 3877 participants were evaluated. Undiagnosed RMD were identified in 29% (n=1514) of Portuguese adults. These were more often women (53%, n=23), among the 46-55 age stratum (20%, n=359) and lived predominantly in the Norte region (41%, n=440). The most prevalent undiagnosed RMD was periarticular disease (34%), followed by knee (18%) and hand (11%) osteoarthritis and osteoporosis (11%). When compared to the non-RMD group, participants with undiagnosed RMD had worse HRQoL (EQ5D: β=-0.07; p<0.001) and higher disability (HAQ: β=0.10; p<0.001). Patients with undiagnosed RMDs were more likely to report anxiety (OR=2.3; p<0.001) and depression (OR=1.4; p=0.260) symptoms as compared to participants with no RMD. Undiagnosed patients were as likely to consult a rheumatologist as those with non RMD (OR=1.7; p=0.159) but were more likely to visit an orthopedist (OR=2.0; p=0.02) and had a higher number of orthopedic appointments (IRR=2.5; p=0.007). Patients with previously diagnosed RMD had even worse EQ5D scores (β=-0.23; p<0.001), HAQ score (β=0.40; p<0.001) and anxiety (OR=4.8; p<0.001) and depression (OR=2.3; p=0.002) symptoms as compared to participants with no RMD.

Conclusions: Patients with undiagnosed RMD are frequent in Portugal and have worse HRQoL, physical function and mental health than people without an RMD. These undiagnosed patients are nonetheless consumers of health resources and tend to seek help from specialties other than rheumatology. Increasing the awareness of RMD might promote their early identification and treatment leading to both personal and societal benefits.

219 - TAXA DE EXECUÇÃO EM 2021 DO PROGRAMA NACIONAL CONTRA AS DOENÇAS REUMÁTICAS (PNCDR): 2004-2014: PORQUE PRECISAMOS DE UM NOVO PNCDR?

Luis Cunha-Miranda[1, 2, 3], Augusto Faustino[1, 2], Helena Canhão[4, 5, 6], Viviana Tavares[3, 7], Jaime C. Branco[3, 4, 8]

[1]Instituto Português de Reumatologia, Lisboa, Portugal; [2]Colégio da Especialidade de Reumatologia da Ordem do Médicos, Lisboa, Portugal; [3]Comissão Científica do Programa Nacional Contra as Doenças Reumáticas 2004-2014, Lisboa, Portugal; [4]CEDOC, NOVA Medical School. Faculdade de Ciências Médicas da Universidade NOVA de Lisboa., Lisboa, Portugal; [5]Rheumatology Department, centro Hospitalar de Lisboa Central, Lisboa, Portugal; [6]Sociedade Portuguesa de Reumatologia, Lisboa, Portugal; [7]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal; [8]Rheumatology Department, Hospital de Egas Moniz, Centro Hospitalar de Lisboa Ocidental, Lisboa, Portugal

Introdução: O Programa Nacional Contra as Doenças Reumáticas foi um programa elaborado entre 2002 a 2004 para integrar o Plano Nacional de Saúde, sendo publicado em 2004.

O PNCDR foi um documento com um horizonte temporal de 10 anos (cessou, por isso, em 2014) cujo desenvolvimento foi dividido em duas fases: a) implementação, durante os primeiros 5 anos e b) consolidação nos 5 anos restantes

Propunha 21 estratégias, sendo 11 de intervenção, 8 de formação e 2 de colheita e análise de informação e tinha como objectivo diminuir o impacto das doenças reumáticas em termos de funcionalidade, trabalho e incapacidade.

Objetivo: O objetivo deste trabalho foi, tendo em conta as 21 estratégias definidas, determinar as taxas de execução parciais e globais do PNCDR.

Material e métodos: Mediante a colaboração de Reumatologistas envolvidos no desenho, implementação e consolidação do PNCDR através de entrevista telefónica

TABELA 1 OBJECTIVOS DO PNCDR E A SUA EXECUÇÃO

ESTRATÉGIAS DE INTERVENÇÃO	Avaliação em 2021
E1 Criação e desenvolvimento de serviços e/ou unidades hospitalares de reumatologia.	Apesar de haver a abertura de 13 serviços de reumatologia a rede em 2021 ainda é deficitária com 42% dos Hospitais do SNS a não terem reumatologia apedar de previsto na RNEHRR
E2 Produção e divulgação de orientações técnicas sobre diagnóstico, acompanhamento e referenciação de doentes reumáticos, nomeadamente no que se refere a: a) Osteoartrose. b) Raquialgias. c) Doenças reumáticas periarticulares. d) Lesões músculo-esqueléticas ligadas ao trabalho. e) Osteoporose. f) Fibromialgia. g) Artropatias microcristalinas. h) Artrite reumatóide. i) Espondilartropatias. j) Doenças reumáticas sistémicas. k) Artrites idiopáticas juvenis.	http://www.acss.min-saude.pt/wp-content/uploads/2016/09/Doencas_Reumaticas.pdf proposta efetuada em 2012 - referenciais de competências e de formação contínua
E3 Produção e divulgação, pelas equipas de saúde escolar, de orientações técnicas sobre identificação de crianças com fatores de risco modificáveis para doenças músculo-esqueléticas, sua referenciação precoce para unidades especializadas em reumatologia e sua integração no ambiente escolar.	Não realizada: no Programa Nacional de Saúde Escolar não existe referência a DRM
E4 Produção e divulgação, pelas equipas de saúde escolar, de orientações técnicas sobre ergonomia do ambiente escolar.	Não realizada
E5 Produção e divulgação de orientações técnicas sobre rastreio transversal oportunístico das alterações da estática e dinâmica músculo-esqueléticas das crianças com 6 anos.	Não realizada
E6 Divulgação periódica, junto dos profissionais de saúde, da localização de consultas de reumatologia em geral e de reumatologia pediátrica, em particular.	Realizada
E7 Produção e divulgação, pelos serviços de saúde ocupacional, de orientações técnicas sobre ergonomia do ambiente laboral.	Lesões Músculo-esqueléticas Relacionadas com o Trabalho Guia de Orientação para a Prevenção PROGRAMA NACIONAL CONTRA AS DOENÇAS REUMÁTICAS Direcção-Geral da Saúde 2008
E8 Produção e divulgação, pelos Centros de Saúde e Ministério da Segurança Social e do Trabalho, de orientações técnicas sobre prevenção de quedas em pessoas idosas.	https://www.dgs.pt/directrizes-da-dgs/normas-e-circulares-normativas/norma-n-0082019-de-09122019-pdf.aspx Apenas para pessoas internadas. Norma na população geral realizada mas não publicada ou publicitada

E9 Elaboração de proposta de norma técnica para a suplementação dietética com vitamina D e cálcio na população idosa.	2008 e atualizada em 2019 https://normas.dgs.min-saude.pt/wp-content/uploads/2019/08/Prevencao-e-tratamento-da-carencia-de-Vit-D_2019.pdf
E10 Validação de critérios de avaliação da funcionalidade do doente reumático.	Realizada embora não especificamente para DR
E11 Elaboração de proposta de modelo de estratificação do acesso de doentes reumáticos a benefícios concedidos em regime especial.	Realizada embora não especificamente para DR com colaboração do PNCDR https://www.dgs.pt/qualidade-e-seguranca/monitorizacao/tabela-nacional-de-funcionalidade1.aspx. Tabela Nacional de Funcionalidades
ESTRATÉGIAS DE FORMAÇÃO	
E12 Promoção, junto das Faculdades de Medicina, do aumento do número de horas de formação de pré e pós-graduação em reumatologia.	Realizada
E13 Promoção, junto da Comissão Nacional do Internato Médico e das administrações hospitalares, do aumento do número de vagas do Internato Complementar de Reumatologia.	Não realizada apesar de proposta As vagas aumentaram pelo aumento de centros com idoniedade e por acção do Colégio da Especialidade
E14 Promoção da formação obrigatória em reumatologia no Internato Complementar de Medicina Geral e Familiar.	Não existe, é opcional apesar de proposta
E15 Promoção da formação específica, na área do sistema músculo-esquelético e das doenças reumáticas, dos profissionais de saúde não médicos, dos técnicos superiores de desporto e dos professores dos diversos níveis de ensino.	Revistas as unidades curriculares de diversos cursos; não existe menção relativamente às doenças reumáticas e músculo-esqueléticas. Proposta feita a diversas faculdades, algumas com eventual resposta positiva
E16 Elaboração de instrumentos pedagógicos, destinados aos profissionais de saúde, sobre identificação precoce da artropatia inflamatória e das doenças reumáticas sistémicas.	Regras de ouro em reumatologia DGS, 2005.
E17 Sensibilização dos empresários e de outros empregadores, bem como dos sindicatos e outras associações laborais para a necessidade de prevenção das doenças reumáticas periarticulares e das lesões músculo-esqueléticas ligadas ao trabalho e para a adoção de medidas que aumentem a adequação da atividade laboral aos condicionalismos de cada doente.	Não realizada
E18 Sensibilização dos profissionais de saúde para as vantagens de a atribuição de benefícios concedidos em regime especial ser baseada nas necessidades específicas de cada doente reumático.	Proposta e realizada mas sem divulgação e avaliação
E19 Desenvolvimento de parcerias multissectoriais para a divulgação, junto da população geral, de informação genérica sobre as doenças reumáticas e sua prevenção e, particularmente, sobre: a) Osteoartrose e raquialgias. b) Fibromialgia c) Hiperuricemia e gota úrica. d) Artropatia inflamatória e doenças reumáticas periarticulares. e) Hábitos para a saúde óssea.	Campanha "Saber que Faz Mover" da SPR, não ligada ao diretamente ao PNCDR Doenças reumáticas: Manual de Auto-ajuda para adultos / Mário Viana de Queiroz. – Lisboa: DGS, 2006. Outras atividades
ESTRATÉGIAS DE COLHEITA E ANÁLISE DE INFORMAÇÃO	
Desenvolvimento de parcerias multissectoriais, com vista à criação de um observatório para as doenças reumáticas, que:	Epireuma.pt, Co. Reuma.pt /EpiDoc ONDOR (até 2014) http://pns.dgs.pt/files/2010/05/ONDOR_Estado_Reumatologia_Portugal-1.pdf
E20 Englobe sistemas de colheita de informação que permitam a obtenção e a análise de dados sobre a prevalência e incidência das doenças reumáticas, assim como sobre a incapacidade temporária e definitiva e absentismo laboral causados por estas doenças ou pelas suas complicações.	EpiDoc/CEDOC/ Nova Medical School
E21 Monitorize os ganhos de saúde resultantes da acção do presente Programa	EpiDoc/CEDOC/ Nova Medical School

Legenda:PNCDR Programa Nacional Contra as Doenças Reumáticas; SPR Sociedade Portuguesa de Reumatologia, Ondor Observatório Nacional das Doenças Reumáticas; CEDOC Chronic Disease Research Centre; A verde objetivos totalmente executados, a laranja parcialmente executados e e a vermelho não executados. NOC e outras publicações. Tratamento Farmacológico da Osteoporose Pós-menopáusica 29/09/2011 027/2011. Prescrição da Osteodensitometria na Osteoporose do Adulto 30/09/2010 001/2010. Agentes Biológicos/Requisitos da "Consulta Especializada" No: 23/DQSDATA: 20/10/08. Prescrição de Agentes Biológicos nas Doenças Reumáticas 30/12/2011 067/2011. Abordagem Diagnóstica da Fibromialgia 27/12/2016 017/2016

ou por email pretendeu-se perceber quais as estratégias que foram efetivamente concluídas até 2021. Foi efetuada uma procura adicional de recomendações, normas técnicas ou outros documentos oficiais do Ministério da Saúde Saúde (MS) e/ou outros organismos e organizações disponíveis online que pudessem confirmar ou excluir a realização das estratégias

Resultados: A taxa de execução foi obtida considerando os objetivos realizados como 1 os não realizados como 0 e os parcialmente realizados como 0.5. Obteve-se uma taxa de execução de 61.9%

Tabela 1 Objectivos do PNCDR e a sua execução

Conclusão: Existem neste momento 12 programas nacionais prioritários e 11 não prioritários e o PNCDR foi excluído em 2014 passando de prioritário para inexistente.

O PNDR teve uma taxa de execução inferior a 2/3 com 6 objectivos não concretizados e 4 apenas parcialmente realizados.

O maior impacto do PNCDR foi talvez o apoio à realização do estudo Epireuma.pt da SPR que conseguiu determinar a prevalência das doenças reumáticas e músculo-esqueléticas (DRM) em Portugal. Contudo a taxa de execução por nós determinada é baixa e suporta a intenção da SPR e de outras entidades em reformular e instituir um novo PNDR.

O PNCDR teve grande parte do seu desenvolvimento fora do âmbito da DGS ou do MS dado que as atividades clínicas e científicas desenvolvidas pelas estruturas da reumatologia incluindo clínicas e académicas da Reumatologia nacional puderam adicionar-lhe um grande contributo. O programa serviu mais como guião para que aquelas estruturas e entidades pudessem contribuir para atingir o nível de execução que obteve, que seria de outra forma muito inferior. É fundamental que o PNCDR seja reativado e complementado cuja urgência tendo em conta a prevalência das DRM, mas igualmente o seu impacto social e previsível agravamento com o envelhecimento populacional.

225 - PEDIATRIC INFLAMMATORY MULTISYSTEM SYNDROME TEMPORALLY-ASSOCIATED WITH SARS-COV-2 – A PORTUGUESE SINGLE CENTRE CASE SERIES

Bernardo Santos[1], Sofia Miranda[2], Ivana Cardoso[3], Francisca Aguiar[4, 5], Mariana Rodrigues[4, 5], Iva Brito[4, 5]
[1]Serviço de Reumatologia, Centro Hospitalar do Baixo Vouga, E.P.E., Aveiro, Portugal; [2]Pediatria, Hospital de Braga, Braga, Portugal; [3]Pediatria, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal; [4]Unidade de Reumatologia Pediátrica e do Jovem Adulto, Centro Hospitalar e Universitário de São João, Porto, Portugal; [5]Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Background: Across the globe, coronavirus disease 2019 (COVID-19) caused by SARS-CoV-2 appears to affect paediatric population in a milder and nonthreatening way, when compared to adults. However, since April 2020 case reports of previously healthy children presenting with unremitting fever, biologic inflammatory syndrome and cardiac dysfunction have been emerging. This syndrome, which has been termed Inflammatory Multisystem Syndrome Temporally Associated with SARS-CoV-2 infection (PIMS-TS), represents a rare complication of COVID-19 in children.

The aim of this study is to describe the clinical, laboratory and imaging characteristics, course, management and outcomes of hospitalized children diagnosed with PIMS-TS in a Portuguese tertiary care hospital.

Methods: A retrospective study including children (< 18 years) that attended our hospital from April 2020 to April 2021 was performed. All the children included fulfilled the case definition of PIMS-TS published by the Centers for Disease Control and Prevention. Sociodemographic and clinical data, laboratory markers and imaging findings were collected. Statistical analysis was performed using SPSS 26.0.

Results: A total of 19 children met the criteria for PIMS-TS, 68% male with a mean age at diagnosis of 8 years old (IQR 5.8-15). They were all caucasian, except for a mixed-race patient, and all previously healthy, except one patient who was obese. Twelve had recent infection by SARS-CoV-2 detected by reverse transcriptase (RT) PCR and 18 had positive IgG serology.

Clinical characteristics are shown in table 1. All had fever at diagnosis, with a median duration of 6 days (IQR 5-6) and 89.5% had mucocutaneous, gastrointestinal and hematological attainment, respectively.

Laboratory findings can be found in table 2. Chest x-ray and pulmonary CT were performed in 84.2% and 31.6% of patients, respectively, with abnormalities in 63.2% and 83.3%; electrocardiogram and echocardiogram were performed in all patients with alterations in 31.6% and 73.7%, respectively. Of note, coronary artery aneurysms (z score >2) were found in 5 cases.

TABLE 1 – SOCIODEMOGRAPHIC AND CLINICAL CHARACTERIZATION OF THE CHILDREN DIAGNOSED WITH PIMS-TS

	PIMS-TS (n=19)
Age, years [a]	8 (5.8 - 15)
Sex, n (%) [b]	
Male	13 (68)
Female	6 (32)
Race/ethnicity, n (%) [b]	
Caucasian	18 (94.7)
Others	1 (5.3)
Medical history of a chronic disease, n (%) [b]	1 (5.2)
Overweight	1 (100)
Clinical features at presentation	
Fever duration, days [a]	6 (5-6)
Muco-cutaneous manifestations, n (%) [b]	17 (89.5)
Maculopapular rash	14 (82)
Conjuntivitis	11 (65)
Pharyngitis	7 (41)
Extremities edema	6 (35)
Cheilitis	6 (35)
Gastrointestinal system, n (%) [b]	17 (89.5)
Abdominal pain	13 (76.5)
Vomiting	13 (76.5)
Diarrhea	12 (70.6)
Hematological alterations, n (%) [b]	17 (89.5)
Aplastic anemia	12 (70.6)
Trombocytopenia	12 (70.6)
Lymphopenia	11 (64.7)
Hypercoagulability	3 (17.6)
Macrophage activation syndrome	1 (5.9)
Respiratory manifestations, n (%) [b]	14 (73.7)
Cough	6 (42.9)
Sore throat	5 (35.7)
Dyspnoea	3 (21)
Pleurisy	3 (21)
Cardiovascular system, n (%) b	12 (63)
Myocarditis	8 (66.7)
Myocardial dysfunction	6 (50)
Coronary aneurysm	5 (33.6)
Chest pain	3 (25)
Lymphoid organs, n (%) b	10 (52.6)
General lymphadenopathies	7 (70)
Hepatomegaly	5 (50)
Splenomegaly	4 (40)
Musculoskeletal system, n (%) b	9 (47)
Myalgia	7 (77.8)
Arthralgia	4 (44.4)
Genito-urinary manifestations, n (%) b	6 (31.6)
Acute kidney injury	2 (33.3)
Gonadal pain	1 (16.7)
Cystitis/urethritis	2 (33.3)
Neurological manifestations, n (%) b	5 (26.3)
Headache	4 (80)

Abbreviations: PIMS-TS, pediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2.
[a] Data showed as median and inter Quartile Range (P25, P75)
[b] Data showed as frequency and percentage

TABLE 2 – LABORATORY RESULTS AT PRESENTATION

Hematology [a]		
	Reference range	
Total white blood cells count, x10^9/L	4.0-11.0	10.19 (7.37-14.85)
Neutrophil count, x10^9/L	1.5-7	10.38 (5.71-11.79)
Lymphocyte count, x10^9/L	1.5-4	0.94 (0.67-1.45)
Hemoglobin, g/dL	12-16	11 (9.4-12.1)
Platelet count, x10^9/L	150-400	156 (104-248)
Inflammatory markers [a]		
	Reference range	
C-reactive protein, mg/L	0-3	218.5 (132-262)
Procalcitonin, ng/mL	0-0.05	2.72 (0.00-8.18)
Ferritin, ug/L	6-140	598 (226-769)
Biochemistry [a]		
	Reference range	
AST, U/L	10-37	52 (38-65)
ALT, U/L	10-37	47 (21-69)
LDH, U/L	135-225	269 (229-306)
Albumin, g/L	38-51	27.1 (24.1-32.5)
Blood total protein, g/L	64.0-83.0	59.7 (53.567.4)
Triglycerides, mg/dL	0-150	169 (119-251)
Total bilirubin, mg/dL	0-1.2	0.65 (0.36-1.41)
Creatinine, umol/L	0.26-0.77	0.52 (0.36-0.9)
Urea, mg/dL	10-34	28 (21-45)
Plasma sodium, mmol/L	135-147	135 (133-138)
Plasma potassium, mmol/L	3.5-5.1	4.3 (3.4-4.8)
Cardiac markers [a]		
Troponin, ng/L	0-34	118 (34-1021)
CK-MB, ng/mL	0.00-6.4	0.7 (0.3-8.1)
Pro-BNP, pg/mL	0-100	238.4 (67.5-525)

Abbreviations: PIMS-TS, pediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2.
[a] Data showed as median and inter Quartile Range (P25, P75)
[b] Data showed as frequency and percentage

Thirty-six percent were admitted in intensive care unit for a median duration of 8 days (IQR 4-9). 42.1% needed respiratory support, 87.5% with supplemental oxygen therapy, 62.5% with mechanical ventilation and 12.5% with non-invasive ventilation.

All patients received intravenous (IV) immunoglobulin, 52.6% IV corticosteroid (CS) pulses and 78.9% IV and oral CS. Other treatments included acetylsalicylic acid (n=18), heparin (n=8) and antibiotic therapy (n=19) - table 3. Seventeen fully recovered and 2 had sequalae: one of them with coronary artery aneurysms and other exertional dyspnea.

Conclusion: In this case series, there was a broad spectrum of clinical symptoms and disease severity, ranging from fever and systemic inflammation to critical care admission with myocardial injury, shock, and development of coronary artery aneurysms.

TABLE 3 – CLINICAL OUTCOMES AND MANAGEMENT.

Clinical course	
ICU admission [b]	7 (36.8)
ICU duration [a]	8 (4-9)
Shock [b,c]	10 (52.6)
Outcome	
Fully recovered [b]	17 (89.5)
Recovered with sequaele [b]	2 (10.5)
Death [b]	0 (0)
Management	
Respiratory support	8 (42.1)
Oxygen supplementation	7 (87.5)
Non-invasive ventilation	1 (12.5)
Mechanical ventilation	5 (62.5)
Pharmacotherapy	
Intravenous immunoglobulin	19 (100)
CCT pulses	10 (52.6)
Parenteral CCT	15 (78.9)
Oral CCT	15 (78.9)
Aspirin	18 (94.7)
Heparin prophylactic	7 (36.8)
Heparin therapeutic	1 (5.3)
Antibiotic therapy	19 (100)
Ceftriaxone	18 (94.7)
Clindamycin	15 (78.9)
Vancomycin	1 (5.3)
Piperacillin/tazobactam	2 (10.5)

Abbreviations: CCT, glucocorticoid; CCT pulses, ≥10mg/kg/day in a single administration; immunoglobulin, anti-inflammatory dose 2g/kg.
[a] Data showed as median and inter Quartile Range (P25, P75)
[b] Data showed as frequency and percentage
[c] Shock was defined as needing inotrope support or fluid resuscitation >20mL/kg

Despite short-term morbidity, there were no mortality cases, with most of them recovering without sequelae. All physicians providing clinical care to children should consider this rare but severe delayed syndrome in paediatric population.

234 - EULAR RESPONSE TO ORIGINAL VS. BIOSIMILAR ETANERCEPT IN PATIENTS WITH RHEUMATOID ARTHRITIS: A NATIONAL BASED (REUMA.PT) STUDY

Cláudia Vaz[1, 2], Ines L[2], Helena Canhão[3]
[1]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, [2]Faculdade de Ciências da Saúde, Universidade da Beira Interior, Covilhã, Portugal; [3]CEDOC, NOVA Medical School, Nova University, Lisboa, Portugal

Background: The use of biosimilars in clinical practice can reduce the financial burden of Rheumatoid arthritis (RA) treatment. Clinical trials support an equivalent response to biosimilar and original etanercept (ETN) treatment. However, data from more heterogeneous patient population in clinical practice is fundamental to assess meaningful differences in effectiveness.
Objectives: To compare EULAR Response to original and biosimilar ETA in patients with RA, from a national (clinical practice based) database (Reuma.pt).
Methods: Observational cohort study including all adult RA patients from the Rheumatic Diseases Portuguese Register (Reuma.pt) who were under treatment with ETN (original or biosimilar) from January 2014 to December 2019. The effectiveness endpoint was a meaningful change in the Disease Activity Score 28 (DAS28-3v) score (EULAR response: ≥1.2) from baseline (when ETN treatment was started) to 6 months. This endpoint was analyzed in the subgroup of patients with DAS 28-3v ≥3.2 at baseline. Association between EULAR response (dependent variable: ≥1.2 vs. <1.2) and clinical/demographic variables at baseline were tested using univariate models (logistic regression LR). Multivariate models (Backward-Wald) were built with variables found to be significant in the univariate analysis. Significant results were considered when $p<0.05$.
Results: A total of 812 patients with RA and receiving ETN were included (33.7% treated with biosimilar). Of these, 489 patients had DAS28-3v available at baseline, with DAS28-3v ≥3.2 in 454 patients. At baseline, the majority were female (82.2%), with mean (±SD) age of 54.0±12.6 years and median disease duration (min-max) of 10.17 (0.31-48.50) years. Odds ratio (OR) for biosimilar ETN was 0.803 (95%CI 0.495-1.030). Demographic and clinical characteristics like age, sex, marital status, schooling years, BMI, erosive index, RF, C reactive protein, comorbidities (HBP, Dyslipidemia, Diabetes, Cardiovascular disease), smoking habits, and use of leflunomide were comparable in both EULAR response groups. ACPA [OR=0.441 (95%CI 0.199-0.973)], sedimentation rate [OR=1.024 (95%CI 1.010-1.038)], use of methotrexate [OR=2.389 (95%CI 1.009-5.661)] and treatment switch [OR=0.456 (95%CI 0.228-0.912)] contributed to different EULAR response. In the multivariate model, only sedimentation rate OR=1.015 (95%CI 1.002-1.027) and use of methotrexate OR=2.425 (95%CI 1.286-

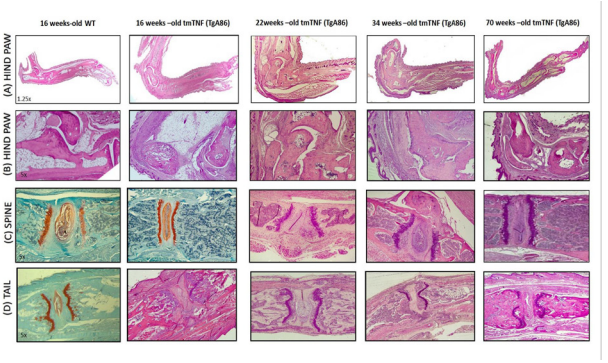
4.569) predicted EULAR response.
Conclusion: These results from a national clinical practice database support a comparable effectiveness of original vs biosimilar ETN in the treatment of RA from baseline to 6 months, but further studies are needed.

262 - PHENOTYPIC, HISTOLOGIC AND IMAGING CHARACTERIZATION OF TRANSMEMBRANE TUMOR NECROSIS FACTOR TRANSGENIC MICE

Elsa Vieira-Sousa[1, 2], Vidal B[2], Soraia Silva[2], Sofio AR[2], Lopes IP[2], Helena Canhão[3], JE Fonseca[1, 2]
[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Hospital de Santa Maria, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [3]CHRC, Comprehensive Health Research Center, CEDOC, NOVA Medical School, Universidade Nova de Lisboa, Lisboa, Portugal

AIM: To characterize tmTNF(TgA86) mice that overexpress mutant transmembrane murine tumor necrosis factor (TNF) and mimic peripheral and axial spondyloarthritis (SpA) features.
Methods: 24 tmTNF(TgA86) mice and 10 littermate controls (WT) were weekly monitored from 4 weeks-of-age (woa) for body weight and macroscopic peripheral arthritis. Mice were euthanized at 16/22/34/70 woa for histology. Hind and forepaws, spine and tail were harvested, and inflammation and bone damage semi-quantitative scores were applied. X-Rays were performed, after euthanasia, to 16 and 70 weeks-old tmTNF(TgA86) mice (VistaScan Combi View). To assess areas of increased tmTNF expression, a TNF inhibitor (certolizumab pegol-like product mice equivalent (AB501)) flurochrome-labelled with alexa680 was administrated to 3 tmTNFmice and 3 WT at 26 woa. Mice were anaesthetized and bioimmunoflourescence (Xenogen-IVIS200) read at 1/3/6/12/24/30hours.
Results: Peripheral arthritis was characterized by progressive swelling and deformity of hind and paws from 4 woa, involving initially the ankles and wrists, and later the tarsus/carpus and digits. The maximum macroscopic peripheral arthritis score was reached at 12-16 woa. Progressive spine deformation, displaying dorsal hunchback and cricked tail was observed. Hampered global growth was translated into lower

IMAGES: PERIPHERAL AND AXIAL HISTOLOGIC CHARACTERIZATION OF TMTNF (TGA86) AT 16, 22, 36 AND 70 WEEKS-OLD.



weight and shortening of mice length (including tail), comparing to littermates. Histologic features, throughout disease progression, at peripheral and axial skeleton are shown in Figure 1. Radiographic assessment showed diminished intervertebral space and vertebral plate erosions at the proximal aspect of the tail, and segmental tortuosity, already noticed at 16 woa. In tmTNF(TgA86) at 70 woa incomplete bridging syndesmophytes could be depicted. Biofluorescence images showed higher uptake of AB501 in the paws and proximal/distal aspects of the tail when comparing with WT non-inflamed areas.
Conclusions: tmTNF(TgA86) mice show phenotypic, histologic, and radiographic features that resemble human SpA. These are associated with a long tmTNF(TgA86) lifespan which can be attractive for assessing new SpA therapeutic interventions.
Acknowledgments: To Leonie M van Duivenvoorde and Dominique Baeten (Amsterdam Academic Medical Center) for providing (tm)TNF (TgA86) mice for breeding and colony establishment. George Kollias from Flemming Institute for authorization for the use of this mice line. This investigator-initiated study was supported (financial & product) by UCB.

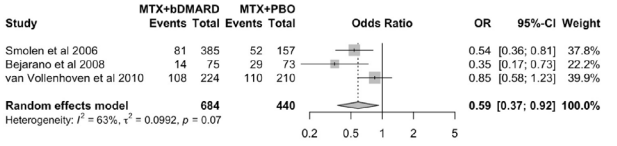
266 - ARE BIOLOGIC AND TARGETED SYNTHETIC DISEASE MODIFYING ANTI-RHEUMATIC DRUGS ASSOCIATED WITH WORK PARTICIPATION IMPROVEMENT IN EARLY RHEUMATOID ARTHRITIS? A SYSTEMATIC REVIEW AND META-ANALYSIS

Mary Lucy Marques[1, 2], Alessia Alunno[3], Louise Falzon[4], Boonen A[5, 6], Sofia Ramiro[2, 7]
[1]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal; [2]Rheumatology,

Leiden University Medical Center, Leiden, Netherlands; [3]Rheumatology Unit, Department of Medicine, University of Perugia, Perugia, Italy; [4]Center for Personalized Health, Northwell Health, New York, United States; [5]Department Internal Medicine, Division of Rheumatology, Maastricht University Medical Center, Maastricht, Netherlands; [6]Care and Public Health Research Institute (CAPHRI), Maastricht University, Maastricht, Netherlands; [7]Rheumatology, Zuyderland Medical Center, Heerlen, Netherlands

Background/Purpose: Treatment of early RA with DMARDs is associated with particularly effective responses, resulting in sustained beneficial outcomes, including clinical and radiographic remission and optimal health related quality of life. The benefit of treatment with biological (b) and targeted synthetic (ts) DMARDs on work participation (WP), a top social role for RA patients, has been less frequently studied. Our aim was to review the evidence of treatment with b-/tsDMARDs in patients with early RA on WP outcomes. Methods: Following PRISMA, a systematic literature review (SLR) was conducted in key electronic databases up to December 2020. The PICOT framework was: (P) RA with disease duration ≤3y; (I) b-/tsDMARDs; (C) any treatment/placebo (PBO); (O) employment status (ES), at-work productivity loss (presenteeism) and/or absence from paid work (sick leave - SL); (T) longitudinal studies (except cost-effectiveness) with any follow-up duration. Two reviewers independently identified eligible studies and extracted data using a self-composed extraction sheet. The Cochrane risk-of-bias tool for RCTs was used for the assessment of risk of bias. Odds ratios (OR) were computed for individual studies, and a pooled OR was obtained by

FIGURE 1. RANDOM-EFFECTS META-ANALYSIS OF THE EMPLOYMENT LOSS IN PATIENTS WITH EARLY RHEUMATOID ARTHRITIS TREATED WITH MTX+BMARD VERSUS MTX IN MONOTHERAPY



bDMARD: biologic disease-modifying antirheumatic drugs; MTX: methotrexate; PBO: placebo.

STUDY CHARACTERISTICS OF THE INCLUDED STUDIES AND OVERVIEW OF THE BETWEEN-GROUP RESULTS

Author (year)	*Selected population*	*Intervention (I)* *Comparator (C)*	*N total (t)* *N employed (e)*	*Assessed Outcome*	*Instrument*	*Follow-up*	*Change (I vs C)$*
Smolen, 2006# ***ASPIRE***	*RA ≤3 y* *csDMARDs* naïve	**I: IFX+MTX** C: PBO+MTX	Nt= 621; Ne= 385 Nt=235; Ne=157	ES SL	Self-reported‡	54 w	+ (nt) + (nt)
Bejarano, 2008#	*RA <2 y* *csDMARD* naïve	**I: ADA+MTX** C: PBO+MTX	Nt=Ne=75 Nt=Ne=73	ES Pres	Self-reported‡	56 w	**+** * **+** *
Anis, 2009 ***COMET***	*RA for 3m-2y* *csDMARDs* naïve	**I: ETN+MTX** C: MTX	Nt=Ne=105 Nt=Ne=100	ES Pres SL	Self-reported‡	52 w	+ (nt) **+** * **+** *
van Vollenhoven, 2010# ***PREMIER***	*RA <3 y* *csDMARDs* naïve	**I_1: ADA+MTX** **I_2: ADA+PBO** C: PBO+MTX	Nt=219; Ne=130 Nt=231; Ne=125 Nt=214; Ne=110	ES Pres SL	Self-reported‡	104 w	**+ (I_1 vs C)*** / + (I_2 vs C)** **+ (I_1 vs C)*** / + (I_2 vs C)** **+ (I_1 vs C)**** / + (I_2 vs C)****
Eriksson, 2013 ***Swefot***	RA <1 y csDMARDs-IR	**I: INF + MTX** C: csDMARD + MTX	Nt=109; Ne=97 Nt=101; Ne=83	SL	Social insurance registry	104 w	+
Emery, 2016 ***OPTIMA*** ***PROWD***	RA <1 y *csDMARDs* naïve	**I: ADA+MTX** C: PBO+MTX	OPTIMA/PROWD: Nt=146/64; Ne=146/64 Nt=174/60; Ne=174/60	ES Pres SL	WPAI-RA	OPTIMA: 24 w PROWD: 26 w	+/- (nt) +/- +/-
Fleischmann, 2016, ***AMPLE***	*RA ≤3 y* csDMARDs-IR	**I: ABA+MTX** C: ADA+MTX	Nt=328 Ne=NR Nt=318 Ne= NR	Pres SL	WPAI-RA	104 w	+§ (nt)
Wiland, 2016 ***PRIZE***	*RA ≤1 y* *csDMARDs* naïve	**I. ETN25/MTX** C_1. PBO+MTX C_2. PBO+PBO	Nt=63 Ne=NR Nt= 65 Ne=NR Nt= 65 Ne=NR	Pres SL	WPAI-RA	117 w	**+ (I vs C_1/C_2)*** **+ (I vs C_1/C_2)***
Schiff, 2017 ***RA-BEGIN***	RA < 2y *csDMARDs* naïve	**I_1: BARI+MTX** **I_2: BARI+PBO** C: PBO+MTX	Nt=159; Ne=67 Nt=215; Ne=117 Nt=210; Ne=94	ES Pres SL	WPAI-RA	52 w	+ (I_1/I_2 vs C) (nt) **+ (24w: I_1/I_2 vs C)**** **(52w: I_2 vs C**)** **+ (24w: I_1/I_2 vs C)*** **(52w: I_2 vs C)***

ABA – abatacept; **ADA** – adalimumab; **BARI** – baricitinib; **csDMARDs** – conventional synthetic disease-modifying antirheumatic drugs; **ETN** – etanercept; **IFX** – infliximab; **IR** – irresponsive; **nt** – significance not tested; **PBO** – placebo; **RA** – rheumatoid arthritis; **SD** – standard deviation; **WPAI** – Work Productivity and Activity Impairment questionnaire.
Studies included in the meta-analysis; *$p \leq 0.05$ vs. Comparator; **$p \leq 0.001$ vs. Comparator; ‡ non-validated instrument; § includes both presenteeism and sick leave in a combined score; **"+":** higher benefit for the intervention group; **"+/-":** no meaningful differences between intervention and comparator groups; **$** when more than one comparator and/or intervention were tested, the respective between-group comparison is presented.

random-effects meta-analysis. Statistical heterogeneity was assessed by I2.

Results: From 5979 records (65 full-text articles screened), 9 studies were included in the SLR (6 RCTs in conventional synthetic (cs)DMARDs-naïve patients; 2 RCTs in bDMARD-naïve patients with inadequate response to csDMARDs; 1 RCT of bDMARD tapering after initial combination with methotrexate (MTX)). Studies assessed WP outcomes with validated and non-validated instruments (Table 1). Interventions comprised 4 bDMARDs (abatacept, adalimumab, etanercept and infliximab) and 1 tsDMARD (baricitinib). Most studies had MTX monotherapy(±PBO) (n=7;78%) as active comparator; in 2 studies bDMARD(±PBO) was added to background MTX. For presenteeism and SL, most between-group comparisons showed improvement in favour of b-/tsDMARDs, but not all comparisons were statistically significant – Table 1. A meta-analysis for ES (weeks 56 to 104) included 3 studies (total of 1124 patients) treated either with adalimumab or infliximab (Figure 1). The pooled OR of the 3 studies showed a lower likelihood of employment loss in patients treated with MTX+bDMARDs (OR: 0.59; 95% CI:0.37 to 0.92) compared to MTX+PBO. High statistical heterogeneity was observed 63% [0.0%; 89.3%].

Conclusion: A protective effect against employment loss was observed in patients with early RA treated with MTX+bDMARDs compared to MTX monotherapy. Although trends towards beneficial effects of b-/tsDMARDs were seen on presenteeism and SL, the methodological heterogeneity hampers clear conclusions. The additional role of b-/tsDMARD in a real world T2T approach should be further studied in long term (preferably population controlled) studies. Efforts to homogenize the design, analysis and reporting of future studies with WP as an outcome by following recently developed points to consider are crucial[1].

REFERENCES:

1. Boonen A, et al. Ann Rheum Dis. 2021 doi: 10.1136/annrheumdis-2020-219523

278 - MEASUREMENT OF VERTEBRAL HOUNSFIELD UNITS IN LOW DOSE COMPUTED TOMOGRAPHY – A RELIABLE METHODOLOGY FOR ASSESSING BONE MINERAL DENSITY AT THE VERTEBRAL LEVEL IN PATIENTS WITH RADIOGRAPHIC AXIAL SPONDYLOARTHRITIS

Mary Lucy Marques[1, 2], Nuno Pereira da Silva[3], Désirée van der Heijde[2], F vanGaalen[2], Sofia Ramiro[2,4]

[1]Serviço de Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal; [2]Rheumatology, Leiden University Medical Center, Leiden, Netherlands; [3]Serviço de Radiologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal; [4]Rheumatology, Zuyderland Medical Center, Heerlen, Netherlands

Background/Purpose: Bone loss paradoxically coexists with bone formation in radiographic axial spondyloarthritis (r-axSpA). Assessing bone mineral density (BMD) in r-axSpA poses challenges, namely because of possible overestimation due to ectopic bone formation. The measurement of Computed Tomography (CT) Hounsfield Units (HU) at the individual vertebral level has been shown to correlate with BMD as measured by Dual-energy X-ray absorptiometry in trauma patients[1]. However, its value in r-axSpA has never been studied. Whole spine low dose CT (ldCT) has been suggested as the ideal method to assess both bone formation and bone loss in r-axSpA. By using ldCT scans, we aimed to cross-sectionally describe HU measurements and their inter-reader reliability at the vertebral level from C3 to L5 in patients with r-axSpA.

Methods: We used ldCT scans of r-axSpA patients included in the Sensitive Imaging in Ankylosing Spondylitis (SIAS) study, a multicenter (Leiden, the Netherlands and Herne, Germany) 2-year prospective cohort. A standardized protocol was applied in both centers and automatic exposure control calibration in ldCT imaging acquisition was used. For the present study, baseline ldCT scans were independently assessed by two trained readers. The HU measurement was performed as described in Figure 1 – using OsiriX software (v6.5.1). Inter-reader reliability was assessed using intraclass correlation coefficients (ICC) absolute agreement, applying two-way mixed effect models. Agreement was assessed using the smallest detectable difference (SDD = 1.96 x SDdifference /(√k), in which SDdifference is the standard deviation of the differences in status scores between two readers, and k=2 (number of readers)) and Bland-Altman plots.

Results: Whole spine ldCT scans from 50 r-axSpA patients (mean (SD) age of 49 (10) years; 43 (86%) male and 42 (84%) HLA-B27 positive) were included. In total, 220 cervical, 588 thoracic and 245 lumbar vertebrae were assessed by both readers. The HU values decreased from the cervical to the lumbar spine vertebrae – Table 1. For both readers, the highest mean

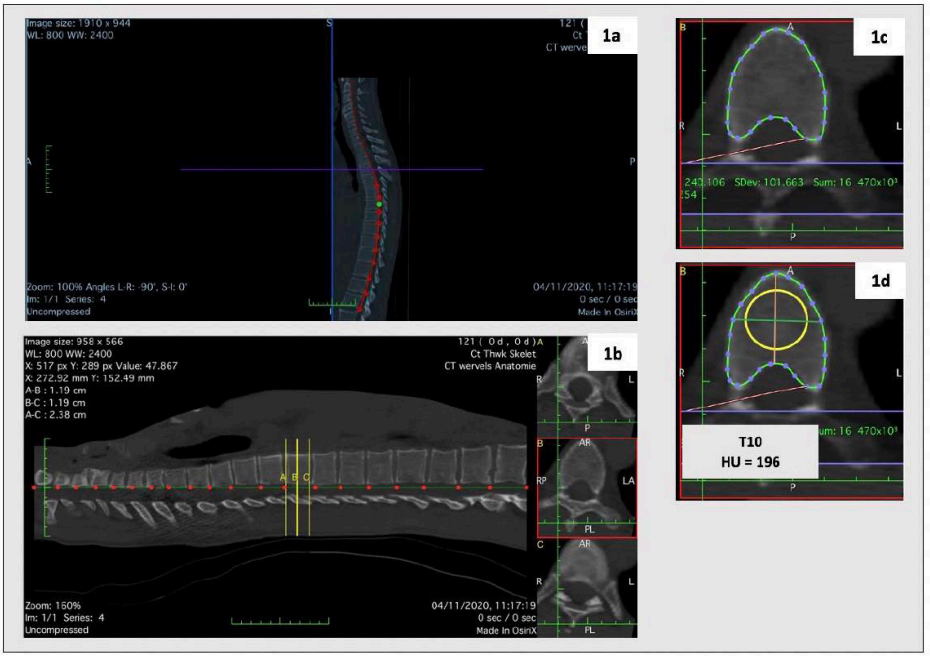


FIGURE 1. METHODOLOGY OF HOUNSFIELD UNITS (HU) MEASUREMENT.

Using a three-dimensional curved-multiplanar reconstruction (3D Curved-MPR) we identify the curve of the spine from the spinal canal (1a). On the obtained sagittal image, the individual vertebral levels are identified. At each vertebral level, two lines of reference are positioned at the superior (yellow line A) and inferior (yellow line C) limits of the vertebra. Equidistant to A and C, the yellow line B will be positioned at the center of the vertebral body (1b). Vertebral HU measurements are taken from a reconstructed cross-sectional slice positioned at the center of the vertebra as defined by the selection process described above (1c). A circular sample region of this slice is used, centered at the body origin and having a diameter equal to 75% of the average of distances posteroinferior-to-anteroinferior, and posterosuperior-to-anterosuperior. The density of the vertebra is calculated as the average image intensity within the sample region, reported in HU (Id).

TABLE 1. DESCRIPTIVE STATISTICS OF HOUNSFIELD UNITS (HU) PER VERTEBRAL LEVEL, ACCORDING TO READER 1 AND 2 MEASUREMENTS

Vertebra§	Mean (SD) HU Reader 1	Range HU Reader 1	Mean (SD) HU Reader 2	Range HU Reader 2
C3	354 (106)	162 to 643	355 (108)	158 to 647
C4	350 (113)	119 to 661	350 (113)	122 to 668
C5	333 (109)	83 to 601	333 (109)	88 to 605
C6	290 (88)	82 to 481	289 (88)	83 to 479
C7	271 (83)	79 to 447	270 (82)	81 to 440
T1	229 (86)	69 to 431	224 (87)	67 to 426
T2	246 (59)	55 to 382	240 (58)	57 to 384
T3	230 (71)	88 to 402	223 (69)	90 to 407
T4	222 (69)	41 to 333	210 (66)	43 to 335
T5	208 (56)	59 to 369	204 (57)	53 to 362
T6	205 (63)	31 to 319	194 (63)	32 to 312
T7	198 (78)	-9 to 353	187 (74)	-10 to 359
T8	185 (68)	7 to 299	177 (65)	3 to 306
T9	194 (72)	-15 to 307	181 (69)	-17 to 304
T10	194 (73)	18 to 375	181 (71)	23 to 372
T11	176 (60)	29 to 305	168 (60)	30 to 302
T12	177 (62)	28 to 367	166 (58)	23 to 372
L1	165 (55)	11 to 325	162 (54)	6 to 317
L2	158 (51)	-13 to 258	153 (51)	-9 to 253
L3	154 (65)	-37 to 345	150 (65)	-34 to 346
L4	163 (84)	-35 to 459	156 (82)	-37 to 461
L5	159 (65)	-12 to 292	158 (63)	-8 to 286

§ Cervical spine values are based on a total of 44 vertebrae scored at each level (C3 to C7) by both readers. The values for thoracic and lumbar spine are based on a total of 50 and 49 vertebrae scored at each level (T1 to L5) by reader 1 and 2 respectively.

(SD) value for HU was obtained at C3 (354 (106) and 355 (108), for reader 1 and 2 respectively), and the lowest at L3 (153 (65) and 150 (65), for reader 1 and 2 respectively). Inter-reader reliability was shown to be excellent (cervical spine ICC(2,1): 0.90 to 1.00; thoracic spine ICC(2,1): 0.97 to 1.00; lumbar spine ICC(2,1): 0.89 to 0.91). SDD varied from 4 to 8. A small degree of systematic error was observed between the two readers, i.e., for the majority of vertebrae, reader 1 scored somewhat higher than reader 2 (mean difference of scores ranging from -0.6 HU to 2.9 HU). The scatter of differences in Bland-Altman plots was homoscedastic at all the vertebral levels.

Conclusion: Low dose CT measurement of HU is a reliable method to assess BMD at each vertebra from C3 to L5. This methodology can aid the future study of bone loss in r-axSpA, a disease affecting the whole spine. Further validation and longitudinal assessment of reliability are warranted in future studies.

REFERENCES

1. Shaun P. Patel, et al. J Clin Exp Orthop. 2016;2(1:14):1–7.

285 - MULTIDISCIPLINARY RHEUMATOLOGY AND DERMATOLOGY CLINIC FOR PSORIATIC ARTHRITIS: FIRST REPORT OF THE EXPERIENCE IN A TERTIARY CENTER

Carolina Ochôa Matos[1, 2], Filipe Santos*[2,3], Augusto Silva*[1, 2], Miguel Alpalhão[4,5], Paulo Filipe[4,5], João Eurico Fonseca[1, 2], Carlos Miranda Rosa[1], Joana Antunes[4,5], Elsa Vieira-Sousa[1, 2]

[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular João Lobo Antunes, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [3]Serviço de Reumatologia da Unidade de Saúde Local da Guarda; [4]Serviço de Dermatologia, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [5]Clínica Universitária de Dermatologia, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina deLisboa Lisboa, Portugal

Introduction: Psoriatic arthritis (PsA) is a heterogeneous chronic inflammatory disease, thus making early and definite diagnosis, as well as remission targets, difficult to establish. Joint Rheumatology and Dermatology PsA Clinics have been associated with numerous benefits for the optimization of disease management and of resources, while improving the collaboration between specialties, and both patients' and physicians' satisfaction.

Methodology: Multidisciplinary PsA Rheumatology and Dermatology Clinic from Centro Hospitalar Universitário Lisboa Norte (CHULN), started in 2010, and up to 2021, a total of 859 visits were performed, corresponding to 525 patients. Patients can be referred due to diagnostic or therapeutic indications, such as suspected PsA or psoriasis (PsO), inadequate control of PsA or PsO disease activity, among other. Both PsA and PsO demographic, disease characteristics and activity measures, as well as diagnostic and therapeutic decisions, are regularly registered by rheumatologists and dermatologists. A cross-sectional analysis of a subset of patients evaluated at their first visit is presented, as a first report assessing the performance of this clinic.

Results: From a preliminary analysis of 93 observed patients, the majority (78%) had a previous PsA diagnosis. Overall, diagnosis was changed in 15% and treatment in 93% of the cases. 41% of the treatment modifications occurred due to poor control of psoriasis activity, 27% due to musculoskeletal symptoms and 12% due to both reasons. In 61% of these cases, the change was because of lack of effect of ongoing treatments. In those with a confirmed PsA diagnosis, the most prevalent PsA subtype was symmetric polyarthritis (47%), followed by olygoarthritis (28%), predominant spondylitis (13%) and predominant distal interphalangeal subtype (11%). There was only one case of arthritis mutilans. In terms of the extra-articular manifestations, 92% of the PsA patients had history of cutaneous psoriasis, while the remaining had isolated nail involvement, present, in total, in 76% of the patients. Until their first visit, 52% had showed enthesitis, 37% dactylitis and 4% anterior uveitis. In the totality of the patients seen, most (65%) manifested with cutaneous/nail symptoms first. Prior to the first appointment, 84% had already been treated with disease-modifying antirheumatic drugs (DMARD), mainly methotrexate. A third (33%) had received treatment with, at least, one biological DMARD.

Conclusion: The first visit at the CHULN Multidisciplinary Rheumatology and Dermatology PsA Clinic, had a relevant impact in the diagnosis and treatment regimens, in a preliminary subgroup of analyzed patients. The full analysis of this cohort in ongoing.

Acknowledgments: To all rheumatology and dermatology trainees that collaborated through the years at the CHULN Multidisciplinary Rheumatology and Dermatology PsA Clinic.

TRABALHO ORIGINAL (COM EXPOSIÇÃO)

002 - SECUKINUMAB THERAPY IN BIOLOGIC--NAÏVE VS. BIOLOGIC-EXPERIENCED PATIENTS: REAL-WORLD EFFECTIVENESS, PERSISTENCE AND SAFETY RESULTS FROM THE RHEUMATIC DISEASES PORTUGUESE REGISTRY (REUMA.PT)

Soraia Azevedo[1], José Tavares-Costa[1], Laires PA[2], Salomé Garcia[3], Patrícia Nero[4], Jorge Pestana Lopes[5], Martins-Martinho J[6], Ana Bento da Silva[7], Joana Ramos Rodrigues[1], Graça Sequeira[8], Emanuel Costa[9], Filipe Cunha Santos[10], Carolina Furtado[11], Bernardo Santos[12], Ana Raposo[13], Liliana Saraiva[14], Beatriz Samões[15], Filipe Barcelos[16], Maria José Santos[5, 17], Helena Santos[16, 18]

[1]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal; [2]HE&OR - Novartis Portugal / ENSP – NOVA University of Lisbon,

Lisboa, Portugal; [3]Rheumatology Department, Centro Hospitalar de São João, Porto, Portugal; [4]Rheumatology Department, Hospital CUF Descobertas, Lisboa, Portugal; [5]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal; [6]Rheumatology and Bone Metabolic Diseases Department, Centro Hospitalar Lisboa Norte, EPE - Hospital de Santa Maria, Lisbon Academic Medical Centre, Lisboa, Portugal, [7]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [8]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, Faro, Portugal, [9]Rheumatology Department, Hospital de Braga, Braga, Portugal; [10]Rheumatology Department, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, ; [11]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal; [12]Rheumatology Department, Centro Hospitalar do Baixo Vouga, EPE, Aveiro, Aveiro, Portugal; [13]Rheumatology Department, Centro Hospitalar ; Trás-os-Montes e Alto Douro, Vila Real, Portugal; [14]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, ; [15]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/ Espinho, Vila Nova de Gaia, Portugal, ; [16]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal; [17]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [18]EpiDoc Research Unit, CEDOC, NOVA Medical School, Lisboa, Portugal

Background: Secukinumab has demonstrated to be efficacious for both psoriatic arthritis (PsA) and ankylosing spondylitis (AS): PASI, ACR and ASAS magnitudes of response have generally been higher in the anti-TNF-naïve population. However, real world data on effectiveness in quality of life (QoL) and function are missing.

Objective: To analyze the differences of secukinumab therapy (effectiveness in disease activity and QoL, persistence and safety profile) in biologic-naïve versus biologic-experienced patients.

Methods: Multicenter (national), observational study of PsA and AS patients using real world anonymous patient-level data from the Portuguese national register database - Reuma.pt. We analyzed data at baseline and after 3, 6 and 12 months after secukinumab initiation, between 1st January 2017 and 10th January 2021. We collected data on sociodemographic characteristics and safety (discontinuation and adverse events); effectiveness [ACR and ASAS response, patient global assessment (PtGA), physician global assessment (PhGA), C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), Bath Ankylosing Spondylitis Disease Activity Index (BASDAI), Ankylosing Spondylitis Disease Activity Score (ASDAS-CRP), Disease Activity in PSoriatic Arthritis (DAPSA) and Disease Activity Score (DAS28 4 V)]; function and QoL [ASQoL (Ankylosing Spondylitis QoL questionnaire), EQ-5D (Euro-QoL-5 dimensions), FACIT-F (Functional Assessment of Chronic Illness Therapy-Fatigue); HADS (Hospital Anxiety and Depression Scale); HAQ (Health Assessment Questionnaire); BASFI and SF-36 (Health Questionnaire Short-Form 36)].

Results: We included 334 patients (166 with PsA and 168 with AS), 68% were biologic-experienced. When we analyzed disease activity, we found that PhGA (p=0.004) and ESR (p=0.002) were significantly lower in naïve patients and more naïve patients reached ACR50 response (p=0.045) at 3 months. At 6 months evaluation, in addition to PhGA (p=0.008) and ESR (p=0.008) that remained significantly lower in naïve patients, PtGA (p=0.009), ASDAS-PCR (p=0.008) and BASDAI (p=0.006) were also lower in naïve patients. In the last evaluation, at 12 months, besides the variables described, also DAS 28 4V was lower in naïve patients (p=0.048) (Table 1).

Regarding QoL and function, at 6 months, BASFI (p<0.001), EQ5D-VAS (p=0.034), SF-36 scores, physical (p=0.013) and mental (p=0.023) domains and ASQoL (p=0.008) were significantly better in naïve patients. After a year of follow-up (12 months evaluation) naïve patients had a better health status (HAQ and ASQoL) (p=0.006 and p=0.003 respectively) and biologic-experienced patients had more anxiety (HADS, p=0.029) (Table 1).

Regarding, drug persistence, although not statistically significant, biologic-experienced patients were more prone to discontinue the drug (18.5% and 11.2%, respectively, p=0.091)

Regarding safety, there were no differences in the number or type of severe (naïve patients: 3/107 vs biologic-experienced: 9/227) or mild (naïve patients: 4/107 vs biologic-experienced: 7/227) adverse effects.

Conclusions: Secukinumab treatment showed a larger magnitude of response (disease activity, function and QoL) in the biologic-naïve population, both in PsA and AS patients. No differences on safety profile were found.

Acknowledgement: This study was supported by Novartis and presented on Behalf of PROSAS Study group.

	Baseline			3 months			6 months			12 months		
	Naïve	Biologic-experienced	p-value	Naïve	Biologic-experienced	p-value	Naïve	Biologic-experienced	p-value	Naïve	-biologic-experienced	p-value
PtGA, mean (SD)	63.1 (25)	66.2 (23)	NS	40.0 (30)	44.6 (25)	NS	37.6 (28)	50.5 (27)	0.009	30.0 (27)	42.0 (27)	0.016
PhGA, mean (SD)	46.5 (19)	50.6 (23)	NS	18.5 (18)	27.8 (19)	0.004	15.8 (16)	24.1 (2.5)	0.008	13.6 (14)	23.0 (17)	0.004
CRP mg/L, median (IQR)	8.0 (14.3)	8.1 (16.6)	NS	3.4 (6.5)	5.2 (11.0)	NS	3.9 (6.5)	2.2 (8.9)	NS	3.5 (5.0)	5.0 (7.6)	0.041
ESR, median (IQR)	18.5 (28)	23 (31)	NS	9 (12)	17 (20)	0.002	11 (18)	19 (30)	0.008	10 (19)	15 (23)	NS
BASDAI, mean (SD)	5.7 (2.1)	5.8 (2.1)	NS	3.8 (2.9)	4.2 (2.4)	NS	3.5 (2.5)	4.9 (2.2)	0.006	3.2 (2.8)	4.3 (2.2)	NS
Δ BASDAI, mean (SD) BASDAI 50, n (%)	-	-	-	2.0 (1.9) 13 (54%)	1.7 (2.0) 38 (45%)	NS NS	2.1 (2.1) 13 (56%)	1.2 (1.9) 31 (35%)	NS NS	2.7 (1.6) 13 (65%)	1.8 (2.1) 45 (49%)	NS NS
ASDAS-CRP, mean (SD)	3.4 (1.1)	3.5 (0.9)	NS	2.3 (1.1)	2.7 (1.1)	NS	2.3 (1.0)	2.9 (1.0)	0.008	2.1 (1.1)	2.6 (1.0)	0.038
Δ ASDAS-CRP, mean (SD) ASDAS-CRP CII (Δ > 1.1), n (%) ASDAS-CRP MI (Δ > 2.0), n (%)	-	-	-	1.1 (1.1) 11 (50%) 3 (14%)	0.9 (0.9) 30 (41%) 9 (12%)	NS NS NS	1.3 (1.1) 8 (47%) 4 (24%)	0.6 (0.8) 19 (25%) 5 (6.5%)	0.003 NS NS	1.5 (1.3) 12 (63%) 6 (32%)	1.0 (1.0) 26 (43%) 12 (20%)	0.046 NS NS
DAPSA, mean (SD)	119.8 (56)	146.0 (45)	0.018	66.9 (55)	93.4 (53)	NS	86.3 (64)	88.9 (54)	NS	69.9 (62)	86.9 (56)	NS
Δ DAPSA, mean (SD)	-	-	-	62.4 (66)	51.8 (62)	NS	56.0 (54)	64.7 (62)	NS	77.7 (56)	46.4 (45)	NS
DAS28 4V, mean (SD)	4.0 (1.6)	4.7 (1.4)	NS	2.8 (1.9)	3.4 (1.3)	NS	2.7 (1.3)	3.2 (1.5)	NS	2.5 (1.2)	3.2 (1.4)	0.048
Δ DAS28 4V, mean (SD)	-	-	-	1.4 (1.7)	1.5 (1.2)	NS	1.1 (0.8)	1.5 (1.5)	NS	2.1 (1.5)	1.3 (1.4)	NS
Tender joint count, median (IQR)	3.0 (5)	4.0 (9)	NS	0.0 (1.0)	1.0 (1.0)	NS	0.0 (2)	1.0 (4)	NS	0.0 (1)	0.0 (3)	NS
Swollen joint count, median(IQR)	2.0 (4)	1.0 (5)	NS	0.0 (1.0)	0.0 (1.0)	NS	0.0 (0)	0.0 (1)	NS	0.0 (0)	0.0 (1)	0.030
ASAS 20, n (%)	-	-	-	9 (38%)	31 (47%)	NS	11 (50%)	19 (26%)	0.041	11 (69%)	22 (40%)	0.049
ASAS 40, n (%)	-	-	-	7 (28%)	16 (23%)	NS	8 (36%)	13 (18%)	NS	8 (47%)	11 (19%)	0.030
ASAS 70, n (%)	-	-	-	5 (19%)	5 (6.8%)	NS	7 (32%)	1 (1.3%)	<0.001	5 (28%)	6 (10%)	NS
ACR 20, n (%)	-	-	-	7 (47%)	18 (42%)	NS	3 (38%)	10 (39%)	NS	9 (82%)	9 (41%)	NS
ACR 50, n (%)	-	-	-	5 (31%)	5 (9.4%)	0.045	1 (11%)	5 (11%)	NS	4 (36%)	4 (15%)	NS
ACR 70, n (%)	-	-	-	2 (9.1%)	4 (6.7%)	NS	0 (0.0%)	3 (4.8%)	NS	1 (9.1%)	0 (0.0%)	NS
MASES, median (IQR)	0 (3)	1 (5)	NS	0.0 (2.0)	0.0 (1.0)	NS	0.0 (2.0)	0.0 (4.0)	NS	0.0 (1.0)	0.0 (3.0)	NS
BASFI, mean (SD)	5.1 (2.6)	6.2 (2.1)	0.011	3.9 (3.1)	4.6 (2.6)	NS	3.0 (2.8)	5.5 (2.4)	< 0.001	3.5 (3.0)	4.7 (2.7)	NS
HAQ, mean (SD)	0.98 (0.8)	1.61 (0.9)	0.001	0.65 (0.6)	1.01 (0.8)	NS	0.86 (0.8)	1.2 (1.1)	NS	0.60 (0.8)	1.3 (0.7)	0.006
EQ-5D index score, mean (SD) EQ-5D VAS, mean (SD)	0.42 (0.2) 44.9 (19)	0.41 (1.0) 49.0 (25)	NS NS	0.45 (0.3) 55.6 (29)	0.42 (0.2) 48.2 (26)	NS NS	0.57 (0.4) 67.8 (18)	0.51 (0.8) 50.0 (23)	NS 0.034	0.53 (0.4) 61.6 (26)	0.43 (0.3) 53.2 (18)	NS NS
SF-36 PCS, mean (SD) MCS, mean (SD)	40.1 (20) 56.1 (21)	29.7 (16) 47.2 (19)	0.011 NS	49.6 (24) 66.4 (19)	38.9 (19) 56 (20)	NS NS	61.6 (27) 71.3 (19)	37.9 (20) 54.8 (22)	0.013 0.023	55.7 (24) 63.7 (21)	43.2 (21) 58 (26)	NS NS
ASQoL, mean (SD)	13.7 (17)	13.3 (11)	NS	8.1 (5.7)	10.0 (6.1)	NS	4.1 (4.4)	9.9 (5.6)	0.008	3.0 (2.3)	8.3 (5.9)	0.003
FACIT-F, mean (SD)	31.2 (26)	25.5 (12)	NS	32.3 (10)	30.4 (11)	NS	33.9 (13)	29.0 (11)	NS	35.8 (10)	32.7 (10)	NS
HADS Anxiety, mean (SD) Depression, mean (SD)	7.0 (3.4) 6.2 (3.7)	8.8 (4.0) 7.6 (4.2)	NS NS	5.6 (3.9) 5.5 (4.1)	7.2 (4.3) 6.4 (4.2)	NS NS	5.4 (3.7) 5.6 (4.1)	8.4 (4.9) 7.3 (4.7)	NS NS	4.2 (2.9) 5.2 (4.1)	7.7 (4.8) 6.7 (5.0)	0.029 NS

ASDAS-CRP: Ankylosing Spondylitis Disease Activity Score; ASDAS CII: ASDAS Clinically important improvement; ASDAS MI: ASDAS Major Improvement; BASDAI: Bath Ankylosing Spondylitis Disease Activity Index; BASFI: Bath Ankylosing Spondylitis Functional Activity Index; CRP: C-reactive protein; DAPSA: Disease Activity in PSoriatic Arthritis; DAS 28: Disease Activity Score; ESR: erythrocyte sedimentation rate; EQ-5D: Euro Quality-of-Life 5 dimensions; FACIT-F: Functional Assessment of Chronic Illness Therapy Fatigue Scale; HADS: Hospital Anxiety and Depression Scale; HAQ: Health Assessment Questionnaire; IQR: interquartile range; MASES: Maastricht Ankylosing Spondylitis Enthesis Score; NS: non-significant; PtGA: patient global assessement; PhGA: physician global assessment; SF-36: Short-Form Health Survey 36; SD: standard deviation; Δ: variation between evaluation and baseline;

005 - HLA-B27 AND UVEITIS ASSOCIATED WITH SPONDYLARTHROPATHIES IN A MULTIDISCIPLINARY AUTOIMMUNE DISEASE UNIT: ARE THERE CLINICAL DIFFERENCES BASED ON THE HAPLOTYPE?

Cristiana Sieiro Santos[1], Isabel Sendino Tenorio[1], Miguel Cordero[1], Carolina Álvarez[1]
[1]Reumatología, Complejo Asistencial Universitario de León, León, Spain

Background: Uveitis is one of the most frequent extraarticular manifestations of spondylarthritis (SpA), characterized by sudden onset, often unilateral, anterior and recurrent and may be the first manifestation of some systemic diseases.
Objectives: To compare the clinical characteristics of human leucocyte antigen (HLA)-B27+ uveitis patients with SpA diagnosis (ASAS criteria) receiving systemic therapies with HLA-B27 negative uveitis patients.
Methods: We performed a retrospective and descriptive study by including patients diagnosed with uveitis associated with SpA attending our Multidisciplinary Autoimmune Disease Unit from January 2000 to January 2020 and compared the clinical profile of patients with uveitis related and not related to the antigen HLA-B27.
Results: We included 73 patients, 78% were HLA-B27 positive and 21% were HLA-B27 negative. When comparing the two groups, patients with uveitis HLA-B27 positive and uveitis HLA-B27 negative, no differences regarding sex were found. HLA-B27 negative patients were older at diagnosis (43.5±8.9 years) than HLA-B27 positive patients (39.1±11.6 years, p=0.04). Regarding articular involvement, peripherical involvement was more frequent in HLA-B27 negative patients (44%) than in HLA-B27 positive patients (13%, p=0.01) however, there was no difference regarding axial involvement or axial and peripherical involvement. Uveitis preceded the articular involvement in 75% HLA-B27 negative patients and in 44% HLA-B27 positive (p=0.047). The time since first uveitis flare was longer in the HLA B27 positive group: 9.12 years (2.75-12.8) versus 3.56 years (1.34-3.5) in the HLA B27 negative group (p 0.02). HLA-B27 positive patients showed a higher percentage of bilateral/alternating involvement (61.4%) as compared with the HLA-B27 negative patients (31.2%, p=0.046). Mean recurrence of episodes of uveitis was 3.1 ± 1.78 years in HLA-B27 positive patients and 2.1 ±1.74 years in HLA-B27 negative, with no significant difference between the two groups (p=0.90). Initial treatment with immunosuppressors was implemented in 82% of the HLA-B27 positive patients vs 87.5% of HLA-B27 negative patients (p=1) and with biologic disease-modifying agents (bDMARDs) in 28% of HLA-B27 positive patients vs 12.5% of HLA-B27 negative patients, with no significant differences in the number of patients that required systemic treatment (p=0.33).
Conclusion: Uveitis related with positive HLA-B27 debuted at a younger age and showed a higher percentage of bilateral/alternating uveitis involvement and a longer period since first uveitis flare. Uveitis onset preceded articular manifestations more frequently in HLA-B27 negative patients. There were no differences between both groups in sex, systemic treatment and mean recurrence of episodes.

TABLE 1: CHARACTERISTICS OF THE 73 PATIENTS

	HLA-B27 Positive (57)	HLA-B27 Negative (16)	p value
Age (years)	39.1±11.6 years	43.5±8.9 years	0.04
Male	36 (64%)	9 (56.3%)	0.772
Axial involvement (Ax)	30 (52%)	5 (31%)	0.163
Peripheral involvement (Pp)	7 (13%)	7 (44%)	0.01
Axial+Pp	20 (35%)	6 (38%)	0.78
Uveitis as disease onset	25 (44%)	12 (75%)	0.047
Time since onset of first uveitis flare (years)	9.12 years (2.75-12.8)	3.56 years (1.34-3.5)	0.02
Bilateral/alternating involvement	35 (61.4%)	5 (31.2)	0.046
Mean recurrence of uveitis episodes	3.1 ± 1.78 years	2.1 ±1.74 years	0.90
csDMARDs	47 (82%)	14 (87.5%)	1
bDMARDS	16 (28%)	2 (12.5%)	0.33

006 - CORRELATION BETWEEN SKIN AND JOINT INVOLVEMENT IN PATIENTS WITH PSORIATIC ARTHRITIS: EXPERIENCE FROM A THIRD LEVEL HOSPITAL

Cristiana Sieiro Santos[1], Clara Moriano[1], Carolina Álvarez[1], Elvira Díez[1], Trinidad Sandoval[1]
[1]Reumatología, Complejo Asistencial Universitario de León, León, Spain

Background: Determining the relationship between severity of skin and joint disease is essential for better phenotyping of patients and determine whether clinical features of arthritis can associate with psoriasis.
Objectives: To characterize the relationship between skin and joint activity in patients with psoriatic arthritis (PsA) and psoriasis (PsO) at enrolment.
Methods: We performed a retrospective and descriptive study including patients diagnosed with

TABLE 1: DEMOGRAPHIC AND CLINICAL CHARACTERISTICS OF 50 PATIENTS

	PASI <10% N = 35	PASI> 10% N=15	OR 95%IC	P value
Age	54.4 ±10.4	53.9 ±12.9	-	0.86
Female	63%	73%	0.62 (0.16-2.34)	0.53
Duration of psoriasis	14.15 ±8.2	16.6±7.9	-	0.03
Duration of arthritis	10.6 ±7.4	12.2 ±7.6	-	0.05
Psoriasis onset	42%	73%	3.67 (1.97-13.8)	0.03
Arthritis onset	17%	20%	1.21 (0.26-5.64)	1
Nail psoriasis	34%	73%	4.27 (1.38-20.14)	0.015
Hair line	29%	67%	5 (1.36-18.35)	0.03
Enthesitis	14%	13%	1.08 (0.19-6.32)	0.96
Dactylitis	20%	33%	0.5 (0.13-0.96)	0.47
HLA B27	34%	13%	3.39 (0.66-17.56)	0.18
Type of arthritis				
Axial	26%	13%	0.44 (0.08-2.36)	0.47
Peripheral	42.8%	73%	6.88 (1.77-26.76)	0.05
Both	31%	13%	0.34 (0.06-1.75)	0.29
Joint involvement				
Back	14%	20%	0.67 (0.14-3.24)	0.68
Shoulders	17%	53%	5.52 (1.44-21.14)	0.02
Elbows	5.7%	33%	8.25 (1.38-49.21)	0.02
Wrists	31%	73%	6 (1.56-23.11)	0.01
Fingers	54%	60%	1.26 (0.37-4.32)	0.77
Knees	29%	47%	2.19 (0.63-7.65)	0.33
Ankles	40%	60%	2.25 (0.65-7.73)	0.22
Toes	46%	53%	1.36 (0.4-4.56)	0.76
Articular pattern				
Polyarthritis	26%	60%	4.33 (1.2-15.61)	0.02
Asymmetric oligoarthritis	29%	27%	0.91 (0.23-3.54)	1
DIP involvement	14.3%	13.3%	0.92 (0.16-5.39)	1
Spondyloarthritis	11.4%	13.3%	1.19 (0.19-7.33)	1
Arthritis mutilans	-	-	-	-
DAS28	2.44 ±0.95	4.1±1.2	-	0.02

PsA with a history of PsO. Age, sex, disease onset and duration, pattern of PsA and PsO, sites of skin and joint involvement were collected. PsA patients were evaluated at enrolment for skin activity by Psoriasis Area and Severity Index (PASI), joint activity by Disease Activity Score 28 (DAS28) for peripheral arthritis and Bath Ankylosing Spondylitis Disease Activity Index (BASDAI) for axial involvement. A PASI>10% was used to define moderate-severe psoriasis (MS-P). We compared clinical characteristics of patients with PsA based on PASI score and evaluated the relationship of skin and joint activity with linear regression.

Results: A total of 50 patients were collected, 64% were women with a mean age of 57.4 ±11.9 years. Patients with MS-P had a long-standing history of PsO (16.6±7.9 vs 14.15±8.2 years, p 0.03) and arthritis (12.2±7.6 vs 10.6±7.6 years, p 0.05). PsO was diagnosed more frequently before arthritis in the group of MS-P (73% vs 42%, p 0.03). Nail (73% vs 34%, OR 4.27 (1.38-20.14), p 0.015) and hairline psoriasis (67% vs 29%, OR 5 (1.36-18.35), p 0.03)

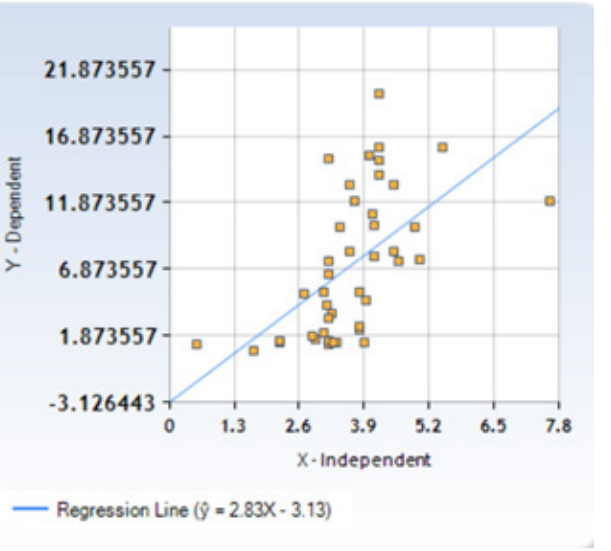


FIGURE. CORRELATION BETWEEN SKIN AND JOINT ACTIVITY

were more frequent in the group of MS-P. Polyarthritis was the most common clinical pattern (60% vs 26%, p 0.02) and peripheral arthritis, in shoulder, elbows and wrists was more associated to patients with MS-P. No significant difference regarding sex, age, arthritis onset, dactylitis, enthesitis or HLA-B27 was found. Patients with MS-P had a higher joint activity for peripheral arthritis (4.1 vs 2.44, p 0.02). The correlation between the skin and joint activity was positive and statistically significant r=0.568 (p 0.02), (graph 1).

Conclusions: Patients with PsA with MS-P associate nail and hairline psoriasis, polyarthritis, and peripheral joint involvement more often. Cutaneous disease activity is correlated with joint activity. Collaboration between dermatologists and rheumatologists is recommended for a proper assessment of psoriatic arthritis with skin involvement.

031 - EFFICACY OF TNF INHIBITORS IN MONOTHERAPY VERSUS IN COMBINATION WITH OTHER DISEASE-MODIFYING ANTI-RHEUMATIC DRUGS IN PORTUGUESE PATIENTS WITH PSORIASIC ARTHRITIS

Filipe Oliveira Pinheiro[1], Bruno Miguel Fernandes[1], Salomé Garcia[1], Maria Seabra Rato[1], Diogo Guimarães da Fonseca[2], Daniela Santos Oliveira[1, 3], Ana Martins[1], Frederico Rajão Martins[4], Pedro Madureira[1], Miguel Bernardes[1, 5], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal; [2]Serviço de Reumatologia,

Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal; [3]Faculdade de Medicina da Universidade do Porto, Departamento de Medicina e CINTESIS, Porto, Portugal; [4]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal; [5]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Background: TNF inhibitors (TNFi) are a key therapeutic weapon in psoriatic arthritis (PsA), and can be used as monotherapy or in combination with other csDMARDs. The optimal use of iTNF in PsA, as monotherapy or in combination therapy with csDMARDs, is still under debate. We aimed to compare the response to treatment with TNFi in monotherapy and combined with csDMARDs, as first biologic, in patients with PsA.

Methods: Retrospective study that included PsA patients followed at our Rheumatology department under TNFi as first biologic, fulfilling CASPAR classification criteria and registered in Reuma.pt. Clinical and laboratory data were collected at the start of the first iTNF and in the last visit of 2019. Disease activity was assessed using CDAI, SDAI, DAS28(4V), BASDAI, ASDAS, and the response measured using the BASDAI50, ASDAS, ASAS, ACR and PsARC responses. Comparison between groups was performed using the chi-square test, Mann-Whitney U/t-test (categorical and continuous variables, respectively). Logistic regression analyses were performed to determine predictors of bDMARD failure, and survival analysis to measure persistence under the first bDMARD regarding csDMARD status at baseline.

Results: We included 99 patients, 47 (47.5% females) with a mean age of 47.9 ± 11.7 years at the start of the first iTNF. Etanercept (41, 41.4%), golimumab (25, 25.3%), adalimumab (22, 22.2%), infliximab (9, 9.1%) and certolizumab (2, 2.0%) were the iTNF started in these patients. Patients who started iTNF as monotherapy had more frequent involvement of axial skeleton compared with combined therapy (54.5% vs 19.5%, p=0.001), were less exposed to corticosteroids (26.3% vs 72.6%, p<0.001) and had higher mean BASMI (3.7±1.8 vs 3.0±0.8, p=0.021) and BASFI (6.7±1.3 vs 4.7±2.5, p=0.036). Patients who were on iTNF monotherapy at the last consultation (43.4%) had lower mean tender (1.0±1.5 vs 3.6±4.3, p=0.002) and swollen (0.2±0.7 vs 0.8±1.0, p=0.012) joint counts, median patient VAS (30±46 vs 50±44, p=0.023), mean CDAI (5.6±4.4 vs 8.7±4.9, p=0.019), SDAI (6.2±4.6 vs 9.1±5.1, p=0.032), and DAS28(4V) (2.2±0.8 vs 2.7±0.9, p=0.047). iTNF failure was not significantly different in both groups. In the regression models, we found that basal DAS28(4V) (OR 1.874, 1.147-3.062 95%CI; p=0,012) was a predictor of first iTNF failure; there were no differences regarding csDMARD status. When evaluating only patients without spondyloarthritis, we found that, at the last visit, iTNF monotherapy patients still had less exposure to corticosteroids (26.9% vs 54.3%, p=0.002), fewer mean tender (0.7±1.0 vs 2.6±4.4, p=0.006) and swollen (0.2±0.7 vs 1.1±2.5, p=0.025) joint counts, with no other differences observed. In the regression models, we found no differences regarding csDMARD status in these patients, while adalimumab (OR 0.009, 0.001-0.139 95% CI; p=0.009) was a negative predictor of bDMARD failure. Survival analysis revealed no differences between mono- and combined therapy.

Conclusion: We can conclude that the differences observed regarding csDMARD status in patients with PsA are mainly due to different patterns of arthritis, namely, predominance of axial involvement. In patients without spondyloarthritis, iTNF monotherapy did not differ significantly in terms of response to treatment and disease activity measures, nor does monotherapy predict bDMARD failure and treatment response. These results suggest that iTNF monotherapy is possible in PsA without compromising treatment response.

032 - BASELINE VITAMIN D LEVELS AND DISEASE ACTIVITY AND RESPONSE IN PORTUGUESE PATIENTS WITH PSORIATIC ARTHRITIS UNDER BMDARD: DOES IT MAKE A DIFFERENCE?

Filipe Oliveira Pinheiro[1], Bruno Miguel Fernandes[1], Salomé Garcia[1], Maria Seabra Rato[1], Diogo Guimarães da Fonseca[2], Daniela Santos Oliveira[1, 3], Ana Martins[1], Frederico Rajão Martins[4], Pedro Madureira[1], Miguel Bernardes[1, 5], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal; [2]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal; [3]Faculdade de Medicina da Universidade do Porto, Departamento de Medicina e CINTESIS, Porto, Portugal; [4]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal; [5]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Background: There is growing evidence that vitamin

D [25(OH)D]) plays an important role in maintaining skeletal health and modulating the immune system. Epidemiological data indicate that vitamin D deficiency is common in immune-mediated rheumatic diseases, especially in rheumatoid arthritis, but there is little data regarding its association with disease activity and response to therapy in patients with psoriatic arthritis (PsA) under bDMARD therapy.

Objectives: We aimed to assess whether 25(OH)D basal levels correlate with disease activity and clinical response to the first bDMARD, at 6 and 12 months of therapy, in a monocentric cohort of patients with PsA.

Methods: This retrospective study was carried out on PsA patients from a Rheumatology department of a tertiary hospital, fulfilling CASPAR criteria and registered in our national database (Reuma.pt), who started the first bDMARD since 2008. Demographic, clinical and laboratory criteria were evaluated at 0, 6 and 12 months of biologic therapy. Disease activity was assessed using CDAI, SDAI, DAS28(4V), BASDAI, ASDAS, DAPSA and the response was measured using the EULAR, BASDAI50, ASDAS, ASAS, ACR and PsARC responses. Correlations were made between absolute serum levels of 25(OH)D and continuous variables, as well as associations between different vitamin D cutoffs and disease activity measures and response criteria. Multiple linear and logistic regression analyses were performed to determine whether vitamin D is a predictor of disease activity and therapeutic response.

Results: We included 81 patients, 41 (50.6%) females; with a mean age of 48.0±11.7 years, a mean disease duration of 9.5±7.4 years and a mean body mass index of 28.4±5.2 kg/m2. Thirteen (16.0%) were smokers. The mean 25(OH)D basal level was 25.5±13.2 ng/ml, 21 (25.9%) had 25(OH)D basal levels ≥30 ng/mL and 31 (38.3%) ≤20 ng/mL. Sixty-two patients (76.5%) were under csDMARD therapy. Golimumab (29, 35,8%), etanercept (28, 34.6%) and adalimumab (10, 12.3%) were the most frequently prescribed bDMARDs. There were only very weak, albeit positive, correlations between 25(OH)D levels and measures of disease activity. The

BASDAI50 response at 6 months was associated with higher basal 25(OH)D levels (29.5±14.5 vs 21.5±10.2 ng/mL, p = 0.013); the ASAS20 (33.9±15.9 vs 24.2±12.8 ng/mL; p = 0.023), ASAS40 (31.9±14.6 vs 25.0±13.8 ng/mL; p = 0.023) and ASAS70 (47.0±4.2 vs 26.6±14.2; p = 0.027) responses at 12 months were associated with higher basal levels of 25(OH)D; basal 25(OH)D levels were ≥ 30ng/mL in a significantly higher proportion of patients who achieved CDAI (38.9% vs 10.5%; p = 0.027) and SDAI (38.9% vs 7.7%; p = 0.008) remission and ASDAS disease inactive (29.4% vs 7.3%; p = 0.040) at 1 year. In the regression models, basal levels of 25(OH)D were found to be predictors of good EULAR responders (OR 1.315, 1.017-1.213 95% CI; p = 0.037) at 6 months. Basal levels of 25(OH)D were not significantly different in patients who discontinued bDMARD and no significant correlations or associations were identified regarding more specific PsA activity measures, such as DAPSA and PsARC, nor were they predictive of these responses.

Conclusion: We can conclude that there is a global trend for an association between higher levels of vitamin

D and lower measures of disease activity and better therapeutic responses to the first biologic. It was possible to find statistically significant associations with some disease activity measures and response criteria that, although primarily designed for other rheumatic diseases, are often used in PsA.

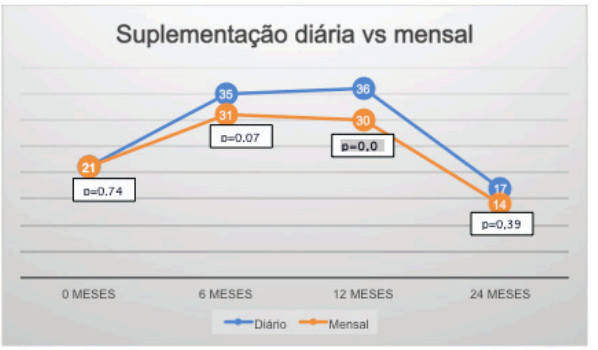
034 - EFICÁCIA DE DIFERENTES SUPLEMENTOS DE VITAMINA D NA OSTEOPOROSE PÓS-MENOPÁUSICA

Filipe Oliveira Pinheiro[1], Maria Seabra Rato[1], Pedro Madureira[1], Iva Brito[2, 3]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal; [2]Unidade de Reumatologia Pediátrica e do Jovem Adulto, Centro Hospitalar e Universitário de São João, Porto, ; Portugal; [3]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Introdução: O papel da vitamina D no metabolismo e saúde ósseos é inequívoco, como demonstrado globalmente na literatura. Sabe-se, no entanto, que níveis baixos de vitamina D são frequentemente reportados na população, pelo que se torna essencial que se determine a forma mais eficaz de suplementação de vitamina D, uma vez que o seu aporte é desprezível através da dieta. Apesar das várias formulações e regimes posológicos existentes, ainda não é claro se existe uma diferença significativa entre a eficácia da suplementação com diferentes fármacos na elevação dos valores séricos de 25-hidroxivitamina D [25(OH) D].

Métodos: Estudo de coorte retrospetivo, realizado em mulheres pós-menopáusicas seguidas em Consulta de Reumatologia de um hospital terciário com osteoporose densitométrica sob suplementação com vitamina



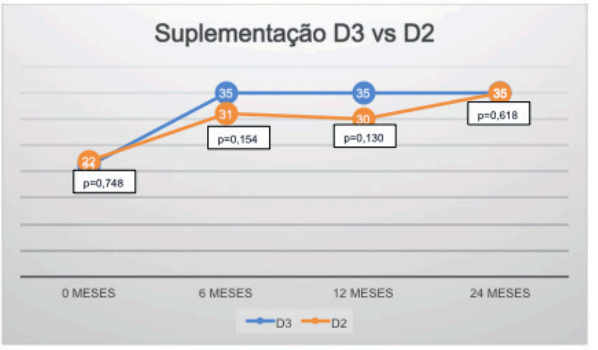


FIGURA.

D. Foram avaliadas caraterísticas demográficas e clínicas, como tipo de suplemento (calcifediol/D2 e colecalciferol/D3), frequência do suplemento (diária e mensal), fármacos antiosteoporóticos, níveis de [25(OH)D], cálcio e albumina, fósforo, magnésio, PTH (medidos aos 0, 6, 12 e 24 meses), existência de episódios de fratura sob suplementação, e valores de densidade mineral óssea e T score na coluna lombar e fémur+colo femoral (medidos aos 0 e 24 meses). A comparação entre grupos (suplementação diária vs mensal, D2 vs D3) foi realizada recorrendo ao teste de Mann-Whitney U; foram feitas análises de regressão para identificar preditores de risco de fratura.

Resultados: Foram incluídos 72 doentes, com idade mediana 65 ± 14 anos, 32 dos quais (44.4%) com osteoporose fraturária. Quarenta e sete (65.3%) iniciaram suplementação com D3 e 25 (34.7%) com D2; trinta e sete doentes (51.4%) faziam suplementação de forma diária e 35 mensal (48.6%). A maioria dos doentes iniciou terapêutica antiosteporótica com alendronato (35, 48.6%), estando os restantes sob zoledronato (14, 19.4%), teriparatide (13, 18.1%), ibandronato (6, 8.3%), pamidronato (2, 2.8%), risedronato (1, 1.4%) e denosumab (1, 1.4%). A [25(OH)D] basal era semelhante nos grupos que iniciaram suplementação (diária 21±12 vs mensal 21±12, p=0,741; D3 21±14, D2 21,7±7, p=0,758). Os níveis de [25(OH)D] foram significativamente mais elevados no grupo sob suplementação diária relativamente à mensal (36±20 vs 30±13, p=0,031), com uma tendência para níveis superiores com D3 vs D2 (35±22 vs 30±13, p=0,130). Ocorreram 9 fraturas em doentes sob terapêutica (12,5%), sem diferenças estatisticamente significativas no que concerne aos grupos diário vs mensal ou D3 vs D2. As análises de regressão permitiram verificar que idade (OR 1.108, IC95% 1.015-1.211; p=0,022) e [25(OH)D] aos 6 meses (OR 1,070, IC95% 1,012-1,131; p=0,017) são preditores do risco de fratura subsequente. Realizando uma análise comparativa dos níveis de vitamina D aos 6 meses, verificou-se que estes eram mais elevados nos doentes que receberam suplementos diários vs mensais (35±14 vs 31±18, p=0,071) e que receberam D3 vs D2 (35±17 D2 31±19, p=0,154).

Conclusão: Nesta coorte constatámos que a suplementação diária foi mais eficaz na elevação dos níveis séricos de vitamina D numa população com osteoporose pós-menopáusica. Os resultados sugerem que a idade e os níveis de vitamina D aos 6 meses podem ser preditores do risco de fratura subsequente. Verificou-se uma diferença estatisticamente significativa com a suplementação diária aos 6 meses relativamente à mensal, e uma tendência global para níveis superiores com a vitamina D3 e com a posologia diária. Estes resultados precisarão, no entanto, de ter confirmação mais robusta.

040 - VITAMIN D LEVEL IN RHEUMATOID ARTHRITIS PATIENTS STARTING A BIOLOGIC DISEASE-MODIFYING DRUG AND ITS CORRELATION WITH DISEASE ACTIVITY AND RESPONSE TO TREATMENT

Salomé Garcia[1], Bruno Miguel Fernandes[1], Filipe Oliveira Pinheiro[1], Maria Seabra Rato[1], Diogo Guimarães da Fonseca[2], Daniela Santos Oliveira[1], Ana Martins[1], Frederico Rajão Martins[3], Georgina Terroso[1], Miguel Bernardes[1], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal; [2]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal; [3]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal

Background: Vitamin D, a fat soluble vitamin that is

mainly involved in the regulation of calcium/phosphate metabolism, has a increasingly understood role in immunomodulatory activity, both in innate and adaptive immune system. In rheumatoid arthritis (RA), vitamin D showed to suppress the proliferation of synoviocytes and to reduce the production of proinfammatory cytokines, in vitro. Recently the hypothesis has been raised that vitamin D has a negative association with RA activity.

Objectives: This study aimed to evaluate the relationship between the 25-hydroxyvitamin D (25(OH) vitD) level, RA activity and response to a first biologic disease-modifying drug (bDMARD).

Methods: This is a longitudinal, retrospective study including consecutive patients with the diagnosis of RA followed at our rheumatology department. Demographic, clinical, and laboratorial data were collected from our national database at baseline, 6 and 12 months after initiation of a first bDMARD. Statistical analysis was performed using SPSS 23.0. Correlations between variables were studied using Spearman correlation analysis and comparison between groups was performed using Wilcoxon and Kruskal-Wallis tests; $p<0.05$ was considered statistically significant.

Results: Mean age of patients (n=236) was 51.5 ± 11.2 years old, 192 (81.4%) were females with a median disease duration of 10.1 [4.7, 16.7] years. Seropositivity for anti-citrullinated protein antibodies was present in 192 (81.4%) patients and for rheumatoid factor in 175 (74.2%). The majority exhibited a very high or high disease activity at baseline (median DAS28 5.75 [4.99 – 6.63]) and 90% (n=212) of them were concomitantly using corticosteroids and/or other disease-modifying anti-rheumatic drugs (117 with methotrexate (MTX), 62 with leflunomide and 32 with sulfasalazine). Regarding bDMARD, 56.8% (n=134) initiated an TNF alpha inhibitor. After 6 and 12 months from a bDMARD initiation there was a significant reduction of ESR, CRP levels, TJCs, SJCs and DAS28 (all p-values< 0.001), as expected. Median baseline serum 25(OH) vitD concentrations was 25.5 [16.5, 30.0] ng/ml; notably, 34.2% of our sample was affected by hypovitaminosis D at baseline (25(OH) vitD< 20 ng/mL). Among our study population 42.5% patients were responders to first bDMARD (23.8% good and 18.7% moderate responders) according to the EULAR response criteria. Disease remission (DAS28<2.6) was achieved by 17.6% of patients. The percentage of good responders was significantly lower in the subgroup of patients with hypovitaminosis D compared to subjects with normal 25(OH) vitamin D levels at baseline (p=0.002), as it was for the percentage of disease remission (p=0.015). The bivariate correlation analyses showed that 25(OH) vit D levels at baseline correlated with CRP levels and good response to RA treatment after 12 months (Table 1). 25(OH) vit D levels at baseline, 6 and 12 months after bDMARD initiation did not correlate with age, BMI, ESV, number of tender or swollen joints, DAS28, HAQ or with SDAI or CDAI at 6 or 12 months of treatment.

TABLE 1. CORRELATIONS BETWEEN 25(OH) VITAMIN D BASELINE LEVEL AND THE DISEASE ACTIVITY

Disease specific measures at 12 months after a bDMARD initiation	25(OH)vitamin D level at baseline
CRP	r=0.201, p=0.028
ESR	NS
HAQ score	NS
DAS28 ESR	r=0.255, p<0.019
CDAI	NS
SDAI	NS

bDMARD: biologic disease-modifying drug ; CDAI: Clinical Disease Activity Index; CRP: C reactive protein; DAS28-ESR: DAS 28- ESR: disease activity score-28 for rheumatoid arthritis with erythrocyte sedimentation rate; ESR: erythrocyte sedimentation rate; HAQ: Health Assessment Questionnaire Disability Index; NS: not statistically significant; SDAI: Simple Disease Activity Index

Conclusion: In patients with RA, basal 25(OH) vit D levels correlated with response to a bDMARD. These results suggest a role of basal vitamin D status in the prediction of disease evolution and support the hypothesis that vitamin D has an immunomodulatory potential.

041 - FRAX AND THE EFFECT OF TERIPARATIDE ON BONE MINERAL DENSITY IN SECONDARY OSTEOPOROSIS

Salomé Garcia[1], Bruno Miguel Fernandes[1], Maria Seabra Rato[1], Filipe Oliveira Pinheiro[1], Diogo Guimarães da Fonseca[2], Daniela Santos Oliveira[1], Ana Martins[1], Frederico Rajão Martins[3], Georgina Terroso[1], Miguel Bernardes[1], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal; [2]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal; [3]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal

Background: Teriparatide has been shown to increase spine and hip bone mineral density (BMD) and to reduce

vertebral and non-vertebral fractures. It is currently not clear whether the effect of teriparatide is dependent on the baseline risk of fracture or osteoporosis (OP) type, a finding that could have an impact on our therapeutic decision.

Objectives: Investigate if there is a relationship between teriparatide effect in BMD and baseline 10-year fracture probability, assessed using FRAX®, in primary and secondary OP patients.

Methods: This is a longitudinal, retrospective study including consecutive patients with the diagnosis of OP treated with teriparatide for 24 months, with a ten-year follow-up period, at our rheumatology department. Demographic, clinical, laboratorial, BMD and occurrence of fracture data were collected. The 10-year risk of osteoporotic fracture was estimated using the fracture risk assessment tool (FRAX) v 4.1 with the Portuguese population reference. Statistical analysis was performed using the software SPSS 23.0. Correlations between continuous variables were evaluated with spearman coefficient. $p<0.05$ was considered statistically significant.

Methods: Eighty patients (88.8% female, median age 65.00 (59; 75)) were included. Forty-nine patients (61.3%) has secondary OP, mainly of cortisonic etiology (61.2%, n=30). Before treatment, median lumbar spine BMD was 0.870 [0.767, 0.964] g/cm2, median T-score of -2.60 (-3.30, -1.90); median total femur BMD was 0.742 [0.667, 0.863] g/cm2, median T-score of -2.10 (-2.80, -1.30); median femoral neck BMD was 0.671 [0.611, 0.787] g/cm2, median T-score of -2.50 [-3.20, -1.85]. Regarding fracture risk, median FRAX-based 10-year major fracture risk (with BMD) at baseline was 16% [10.0; 23], and median hip fracture risk was 7.2% [3.4; 13.8].

The median variation of BMD, after finishing teriparatide treatment, in the spine was 0.107 [0.029; 0.228]; median BMD variation in total femur was 0.013 [-0.013; 0.068] and median BMD femoral neck was 0.046 [-0.002; 0.109]. We observed a numerically superior effect, albeit without any statistical significance, of teriparatide on bone mineral density gain in secondary OP (versus primary OP) at lumbar spine, total femur and femoral neck.

Most patients continued anti-osteoporotic treatment with a bisphosphonate (81.2%, n=65) and, during follow-up, 17 patients had an incident fracture (8 hip fractures and 6 vertebral fractures), median of 5 [1.75, 8.25] years after ending teriparatide. We found a discrete correlation between FRAX-based hip fracture probability and the variation of bone mineral density in total femur (Spearman's coefficient 0.248, $p=0.04$). There was no correlation between FRAX-based major fracture probability and and the variation of bone mineral density in the spine or femur. When we separately analyze the relationship between the variation in total hip BMD and the FRAX-based fracture risk, depending on whether it is a secondary or primary OP, we find that the correlation is stronger and only remains in secondary OP (Spearman's coefficient 0.348, $p=0.03$).

Conclusion: Our data suggest that teriparatide could be an important weapon in the treatment of secondary cause OP, particularly cortisonic, and in patients at high fracture risk, although further larger studies are needed to confirm these findings.

050 - CLINICAL FEATURES AND OUTCOMES OF ENT INVOLVEMENT IN ANCA ASSOCIATED VASCULITIS

Raquel Freitas[1], Henrique Teixeira[2], Fátima Godinho[1], Maria José Santos[1, 3]

[1]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [2]Otorhinolaryngology Department, Hospital Garcia de Orta, Almada, Portugal, [3]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Background: Anti-neutrophil cytoplasmic autoantibody (ANCA) associated vasculitis (AAV) are rare necrotizing vasculitides. Ear, nose and throat (ENT) involvement is common and frequently precede pulmonary or renal involvement. It has been recently reported that ENT involvement is associated with milder forms of AAV and a lower mortality risk [1].

Objetives: To compare clinical features and survival of AAV with and without ENT involvement followed in our center.

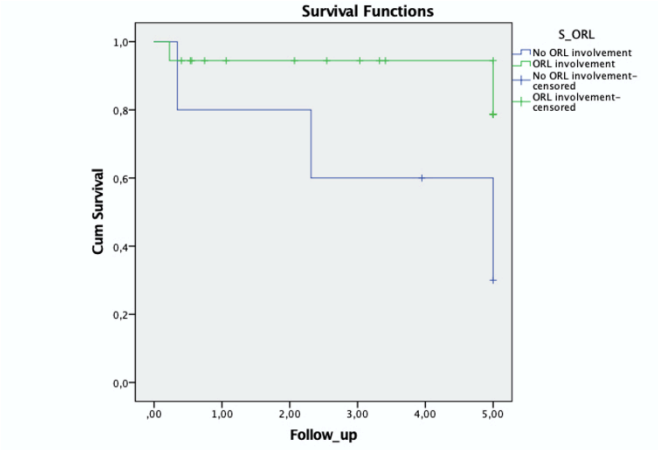
Methods: Demographic features, AAV subtype, main clinical and immunologic characteristics, comorbidities, medication and deaths were analyzed in adult AAV patients registered in Reuma.pt database. Survival was compared between patients with and without ENT.

Results: In total, 25 patients were evaluated, 64% females, mean age at diagnosis 56±16.5 years. The most common AAV subtype was Eosinophilic Granulomatosis with Polyangiitis (40%), followed by Granulomatosis with Polyangiitis (36%) and Microscopic Polyangiitis

TABLE 1 – DEMOGRAPHIC, CUMULATIVE CLINICAL AND IMMUNOLOGIC FEATURES AND TREATMENTS USED. IMMUNOSSUPRESSANTS INCLUDE AZATHIOPRINE, MYCOPHENOLATE MOFETIL AND CYCLOPHOSPHAMIDE. P-VALUE COMPARING PATIENTS WITH AND WITHOUT ENT INVOLVEMENT.

	Whole cohort N=25	ENT involvement N= 18 (72%)	No ENT involvement N=7 (28%)	P-value
Female - N (%)	16 (64)	10 (55.6)	6 (85.7)	0.17
Age at diagnosis – Mean ± SD	56 ± 16.5	54.6 ± 15.7	60.9 ± 20.3	0.47
BMI – Mean ± SD	27.7 ± 6.6	26.6 ± 5.4	32.6 ± 11.7	0.26
Smoker – N (%)	7 (31.8)	7 (43.8)	0	0.07
HTA – N (%)	12 (48)	8 (44.4)	4 (57.1)	0.45
Diabetes – N (%)	6 (24)	4 (22.2)	2 (28.6)	0.56
EGPA – N (%) no=25	10 (40)	7 (38.9)	3 (42.9)	0.25
GPA – N (%) no=25	9 (36)	8 (44.4)	1 (14.3)	
MPA – N (%) no=25	6 (24)	3 (16.7)	3 (42.9)	
PR3-ANCA – N (%)	10 (43.5)	8 (47.1)	1 (16.7)	0.07
MPO-ANCA– N (%)	9 (39.1)	5 (29.4)	5 (83.3)	
General symptoms – N (%)	21 (84)	14 (77.8)	7 (100)	0.29
Muskuloskeletal involvement – N (%)	14 (56)	10 (55.6)	4 (57.1)	0.94
Cutaneous involvemenet – N (%)	14 (56)	10 (55.6)	4 (57.1)	0.94
Ocular involvement – N (%)	5 (20)	4 (22.1)	1 (14.3)	0.65
Pulmonary involvement – N (%)	20 (80)	16 (88.9)	4 (57.1)	0.08
Cardiovascular involvement – N (%)	6 (24)	5 (27.8)	1 (14.3)	0.47
Gastroinestinal involvement – N (%)	7 (28)	6 (33.3)	1 (14.3)	0.34
Genitourinary involvement – N (%)	11 (44)	7 (38.9)	4 (57.1=	0.41
Neurologic involvement – N (%)	16 (64)	11 (61.1)	5 (71.4	0.63
Immunossupressants – N (%)	17 (68)	12 (66.7)	5 (71.4)	0.61
Glucocorticoids – N (%)	23 (92)	17 (94.4)	6 (85.7)	0.49
Rituximab – N (%)	15 (60)	12 (66.7)	3 (42.9)	0.26
Mepolizumab – N (%)	2 (8)	2 (11.1)	0	0.51
Deaths – N (%)	6 (24)	2 (11.1)	4 (57.1)	0.03

FIGURE 1. SURVIVAL



The X and Y axes represent follow-up time in years and the proportion of patients still on respecively

(24%). Anti-PR3 antibodies were present in 43.5% and anti-MPO antibodies in 39.1%. ENT involvement was documented in 76% of the cases and in 64% was one of the inaugural manifestations. The most common signs/symptoms of ENT involvement were sensorioneural deafness (55.6%), nasal ulceration (57.9%), rhinorrhea (47.4%) and epistaxis (33.3%). No differences in demographic and clinical features were found between patients with and without ENT involvement (Table 1). Most patients were treated with glucocorticoids (92%) and two-thirds with immunossupressants and/or Rituximab. Survival rates were better in patients with ENT involvement when compared to patients without ENT involvement (p=0.026, Figure 1)

Conclusion: Our study confirms that ENT involvement

is one of the most frequent clinical feature and is associated with better survival in AAV patients.

REFERENCES

1. Felicetti, M., Cazzador, D., Padoan, R. et al. Ear, nose and throat involvement in granulomatosis with polyangiitis: how it presents and how it determines disease severity and long-term outcomes. Clin Rheumatol 37, 1075–1083 (2018)

052 - GASTROINTESTINAL INVOLVEMENT AND HEALTH RELATED QUALITY OF LIFE IN SYSTEMIC SCLEROSIS PATIENTS

Nádia Marcos[1], Ana Cordeiro[2], Ana Catarina Duarte[2], Sandra Sousa[2], Maria José Santos[2, 3]

[1]Faculdade de Medicina da Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [3]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Background: Systemic Sclerosis (SSc) is characterized by abnormalities in the vascular and immune systems leading to skin thickening and fibrosis in multiple organs. Gastrointestinal (GI) involvement is reported in 90% of SSc patients and can have an enormous impact on the quality of life (QoL). However, evidence is lacking on which patients are more likely to develop GI symptoms and how individual GI symptoms affect QoL.

Objectives: To assess the prevalence of self-reported GI symptoms and to determine the impact of GI involvement on the QoL of SSc patients. Additionally, we aimed to identify demographic, clinical and laboratory characteristics associated with GI involvement.

Methods: Cross sectional study of adult patients with SSc followed at the Rheumatology Department of Hospital Garcia de Orta. Demographic and disease characteristics, current treatment, laboratory tests and self-assessment questionnaires of GI symptoms (UCLA SCTC GITI 2.0 and SHAQ), QoL (SF-36 and EQ-5D) and fatigue (FACIT-F) were collected at last observation using the Reuma.pt registry. Comparisons were made between SSc patients with and without GI involvement, across different GI domains and among limited (lcSSc) and diffuse cutaneous (dcSSc) subtypes.

Results: Of the 92 patients included (mean age 63+-14 years; 93.5% female; median disease duration 8 years, 16.3% with dcSSc), 80 (87%) reported GI symptoms. The most prevalent GI symptom was reflux in 75%. SSc patients with GI symptoms had significantly lower QoL assessed by the questionnaires used and reported more fatigue (Table). Fecal soilage, diarrhea and abdominal distension had the worst impact on patients social function assessed by UCLA SCTC-GITI 2.0. Except for older age at SSc presentation, patients with GI symptoms had similar demographic and clinical characteristics as those without. Also, no differences

TABLE. COMPARISON OF QUALITY OF LIFE AND FATIGUE BETWEEN THE PATIENTS WITH AND WITHOUT GI SYMPTOMS USING SHAQ, SF-36; EQ-5D AND FACIT-F QUESTIONNAIRES.

	Total (n = 92)	GI+, 80 (87%)	GI-, 12 (16.3%)	P value
SHAQ (median±IQR)				
GLOBAL	1.17±1.80	1.17±1.80	0.30±1.05	***0.008***
GI	0.15±0.60	0.23±0.83	0.00±0.00	***0.000***
LUNG	0.00±0.30	0.00±0.30	0.00±0.00	0.190
VASCULAR	0.30±1.50	0.30±1.48	0.30±0.52	0.281
DIGITAL ULCERS	0.00±0.68	0.00±1.13	0.00±0.00	0.179
SF-36 (median±IQR)				
PCS	52.45±47.82	51.09±43.57	93.64±39.72	***0.001***
MCS	55.67±29.30	54.52±30.25	63.85±25.74	0.336
EQ-5D (median±IQR)	0.48±0.41	0.48±0.30	0.77±0.61	***0.049***
FACIT-F (median±IQR)	34.00±17.00	33.00±16.50	48.00±16.30	***0.003***

Legend: IQR, interquartile range; GI+, presence of gastrointestinal symptoms; GI-, absence of gastrointestinal symptoms; SHAQ, Scleroderma Health Assessment Questionnaire; SF-36: Short Form (36) Health Survey; PCS: Physical health component summary score; MCS: Mental health component summary score; EQ-5D: EuroQoL Five-Dimensional; FACIT-F: Functional Assessment of Chronic Illness Therapy – Fatigue

were found regarding SSc subtypes (dcSSc and lcSSc) and autoantibody profile. However, haemoglobin levels were significantly lower in the group reporting GI symptoms.

Conclusion: Our work supports the high prevalence and burden of GI symptoms in SSc patients. Although reflux was the most common, fecal soilage, diarrhea and distension were the most detrimental GI symptoms. The screening of GI symptoms using validated self-reported questionnaires is crucial to identify these patients in a timely manner.

054 - MICOFENOLATO DE MOFETIL NO TRATAMENTO DA DOENÇA PULMONAR INTERSTICIAL ASSOCIADA A CONECTIVITES

Bruno Miguel Fernandes[1], Miguel Bernardes[1], Salomé Garcia[1], Maria Seabra Rato[1], Filipe Oliveira Pinheiro[1], Natália Melo[2], Patrícia Mota[2], Hélder Novais Bastos[2], António Morais[2], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Serviço de Pneumologia, Centro Hospitalar Universitário de São João, Porto, Portugal

Introdução: A doença pulmonar intersticial (DPI) é uma causa importante de morbilidade e mortalidade nas doenças do tecido conjuntivo. A presença de DPI está documentada em até 60% dos doentes com conectivites e varia de acordo com a patologia em questão. O micofenolato de mofetil (MMF) é uma das opções terapêuticas para o tratamento da DPI associada às conectivites.

Métodos: Estudo retrospetivo monocêntrico incluindo todos os doentes com DPI associada a conectivites tratados com MMF. O padrão de DPI foi definido por tomografia computorizada de alta resolução (TC-AR) ou biópsia pulmonar e foram obtidos dados relativos à monitorização da doença pulmonar [TC-AR, capacidade vital forçada prevista (CVFprev) e capacidade de difusão do monóxido de carbono prevista (DLCOprev)] aos 6, 12 e 24 meses de tratamento com MMF.

Resultados: Foram incluídos 25 doentes: 21 do sexo feminino (84%), idade média de 61±14.1 anos, 7 (28%) ex-fumadores, com tempo médio de conectivite de 9.1±7.5 anos e tempo médio de DPI de 29.9±35.6 meses. Catorze doentes tinham esclerose sistémica, cinco tinham síndrome de Sjögren, três artrite reumatóide, dois doença mista do tecido conjuntivo e um lúpus eritematoso sistémico. Relativamente ao padrão de DPI, verificou-se um padrão de pneumonia intersticial não específica em 16 doentes (64%), pneumonia intersticial usual em 7 (28%), pneumonia intersticial linfocítica em 1 doente (4%) e padrão não caracterizável em 1 doente (4%). Descrevemos a evolução da resposta pulmonar ao MMF aos seis (20 doentes), 12 (16 doentes) e 24 (12 doentes) de tratamento com MMF, como é possível observar na tabela 1. Apesar do elevado número de missings, verificamos que a maioria dos doentes apresentou melhoria ou agravamento na CVFprev até 15% aos 6 (50%), 12 (56.3%) e 24 (66.7%) meses de tratamento com MMF. Não se verificou nenhuma diferença de resposta a nível pulmonar entre as diferentes conectivites.

Onze doentes (44%) suspenderam definitivamente o MMF: 5 por intolerância gastrointestinal, 2 por ineficácia terapêutica, 1 por intercorrências infecciosas recorrentes, 1 por diagnóstico de carcinoma do pulmão de pequenas células, 1 por recuperação morfológica e funcional pulmonar e 1 por perda de follow-up.

Conclusão: Dados da literatura sustentam a eficácia do MMF na estabilização da DPI associada a várias conectivites. O nosso trabalho suporta esses dados numa coorte de várias conectivites, apesar do elevado número de missings. Realça-se, ainda, a intolerância gastrointestinal como motivo mais frequente de descontinuação do fármaco.

TABELA 1. EVOLUÇÃO DA RESPOSTA PULMONAR AO MMF AOS 6, 12 E 24 MESES DE TRATAMENTO (MMF: MICOFENOLATO DE MOFETIL).

	6 meses (n=20)	12 meses (n=16)	24 meses (n=12)
DLCOprev			
Melhoria ou agravamento até 10%	6 (30%)	5 (31.3%)	6 (50%)
Agravamento ≥ 10%	5 (25%)	5 (31.3%)	1 (8.3%)
Missing	9 (45%)	6 (37.4%)	5 (41.7%)
CVFprev			
Melhoria ou agravamento até 15%	10 (50%)	9 (56.3%)	8 (66.7%)
Agravamento ≥ 15%	3 (15%)	1 (6.3%)	1 (8.3%)
Missing	7 (35%)	6 (37.4%)	3 (25%)
SC-HR			
Melhoria ou estabilidade	8 (40%)	5 (31.3%)	5 (41.7%)
Agravamento	0 (0%)	1 (6.3%)	3 (25%)
Missing	12 (60%)	10 (62.4%)	4 (33.3%)

055 - TNF INHIBITOR MONOTHERAPY IN RHEUMATOID ARTHRITIS: IS THERE REALLY A DIFFERENCE IN COMPARISON WITH COMBINATION THERAPY WITH CSDMARDS IN REAL-LIFE?

Bruno Miguel Fernandes[1], Salomé Garcia[1], Filipe Oliveira Pinheiro[1], Maria Seabra Rato[1], Diogo Guimarães da Fonseca[2], Daniela Santos Oliveira[1,3], Ana Martins[1], Frederico Rajão Martins[4], Miguel Bernardes[1], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [3]Faculdade de Medicina da Universidade do Porto, Departamento de Medicina e CINTESIS, Porto, Portugal, [4]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal

Background: In Rheumatoid Arthritis (RA), tumor necrosis factor inhibitors (TNFi) in combination with conventional synthetic disease-modifying antirheumatic drugs (csDMARDs) has shown advantages concerning efficacy and immunogenicity in comparison with monotherapy. However, in clinical practice, up to 40% of patients under biological DMARDs (bDMARDs) are on monotherapy.

Objectives: To compare the efficacy outcomes of TNFi in monotherapy and in combination therapy in a RA monocentric cohort.

Methods: Retrospective, cross-sectional study including all the RA patients under TNFi followed at our Rheumatology Department and registered in the national database. Demographic, clinical and laboratorial data and disease activity measures were collected at the last visit of 2019 from each patient. Mann-Whitney U and chi-square tests were used to the comparison analysis between the two groups (continuous and categorical variables, respectively).

Results: A total of 144 patients were included: 84% were females; at the last visit of 2019, the mean age was 56.3±10.9 years and the mean disease duration was 18.3±10.2 years; 73.6% were positive for rheumatoid factor (RF), 81.9% for anti-citrullinated protein autoantibodies (ACPA) and 45.1% had erosive disease. There were no statistically significant differences in these variables between the monotherapy and the combination therapy groups (table 1). Thirty-one patients (21.5%) were under monotherapy with TNFi and etanercept was the most frequent TNFi in both groups (54.8% vs 50.0%; monotherapy and combination therapy groups, respectively). At the start of the first bDMARD, the monotherapy group had a higher disease activity score 28 - 4 variables (DAS 28 4V; 6.083±0.930 vs 5.605±1.043, p=0.039) and a higher simple disease activity score (SDAI; 36.12±11.77 vs 28.76±9.98, p=0.035); also, in the monotherapy group more patients had already started the bDMARD in monotherapy (22.6% vs 2.7%, p<0.001), less patients were under (38.7% vs 73.2%, p=0.001) or had already been treated with (77.4% vs 93.8%, p=0.007) methotrexate, in comparison with the combination group therapy. At the last visit of 2019, the monotherapy group had a higher mean years of duration of iTNF treatment (5.5±5.8 vs 3.4±4.5, p=0.048), a higher mean patient global assessment - visual analogue scale (PGA-VAS; 49±18 vs 39±25, p=0.023), a higher mean prednisolone equivalent dose in mg/day (7.6±6.3 vs 5.6±3.2, p=0.045) and a lower proportion of American College of Rheumatology 50 and 70 responses (ACR 50: 12.9% vs 17.0%, p=0.023; ACR 70: 3.2% vs 10.7%, p=0.045) in comparison with the combination therapy group.

Conclusion: In line with the literature, our real-life results demonstrate some direct (higher PGA-VAS and lower ACR 50 and 70 responses) and indirect (higher current prednisolone equivalent dose) data that suggest that patients with TNFi monotherapy may have a worst disease activity control in comparison with combination therapy.

TABLE 1. DEMOGRAPHIC AND DISEASE-RELATED VARIABLES BETWEEN THE MONOTHERAPY AND THE COMBINATION THERAPY GROUP

	Monotherapy (n=31)	Combination therapy (n=113)
Age - mean±SD	59.1±14.0 years	55.5±9.8 years
Disease duration - mean±SD	20.5±11.2 years	17.7±9.7 years
RF positive - n (%)	20 (60.4%)	86 (76.8%)
ACPA positive - n (%)	25 (80.6%)	93 (85.3%)
Erosive disease - n (%)	15 (48.4%)	50 (44.6%)

056 - IS BASELINE VITAMIN D STATUS RELATED WITH THE RESPONSE TO BDMARDS IN SPONDYLOARTHRITIS PATIENTS?

Bruno Miguel Fernandes[1], Salomé Garcia[1], Filipe Oliveira Pinheiro[1], Maria Seabra Rato[1], Diogo Guimarães da Fonseca[2], Daniela Santos Oliveira[1,3], Ana Martins[1], Frederico Rajão Martins[4], Miguel Bernardes[1], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Rheumatology Department,

Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [3]Faculdade de Medicina da Universidade do Porto, Departamento de Medicina e CINTESIS, Porto, Portugal, [4]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal

Background: Vitamin D is thought to have an important role in immune regulation and is being subject of research in several autoimmune diseases. Some data suggest that vitamin D deficiency is common in Spondyloarthritis (SpA) and may be associated with disease activity and structural damage.
Objectives: To evaluate if there is a relation between baseline vitamin D status and the response to biologic disease-modifying antirheumatic drugs (bDMARDs) in a SpA monocentric cohort.
Methods: Retrospective study including all the SpA patients (ASAS classification criteria) followed at our Rheumatology Department, registered in the national database and treated with bDMARD between June 2008 and July 2020. Demographic, clinical and laboratorial data (including 25-hydroxyvitamin D [25-OHvitD]) at baseline and disease activity measures at 6 and 12 months of treatment with the first bDMARD were collected. Correlations between variables were evaluated by Spearman rank test, Mann-Whitney U test was used to the comparison analysis between groups and univariate logistic regression was used in the prediction analysis.
Results: A total of 195 SpA patients were included: 103 (52.8%) females, 47 (24.1%) smokers and 91 (46.7%) HLA-B27 positive; 139 (71.3%) had Ankylosing Spondylitis, 18 (9.2%) had Inflammatory Bowel Disease Associated SpA and 38 (19.5%) had Undifferentiated SpA. At the time of the first bDMARD, the mean age was 43.5 years (±9.6) and the median disease duration was 12.4 years (0.7-52.7). The mean ASDAS-CPR (Ankylosing Spondylitis Disease Activity Score with C-reactive protein) was 3.9 (±0.8) and, in addition, 61 (31.3%) patients had 25-OHvitD levels below 30 ng/mL and 12 (6.2%) had 25-OHvitD levels below 20 ng/mL. Fifty-three patients (27.2%) were taking NSAIDs (nonsteroidal anti-inflammatory drugs), 77 (39.5%) were under csDMARDs (conventional synthetic disease-modifying antirheumatic drugs). Adalimumab (56%) and golimumab (33.3%) were the most frequently initiated bDMARDs in the first line.

There were no statistically significant correlations between baseline 25-OHvitD levels and ASDAS-CRP at 6 (r=0.031; p=0.714) and 12 months (r=0.035; p=0.672) of bDMARD.

In the subgroup analysis: there were no statistically significant differences in the response to bDMARD at 6 and 12 months evaluated by ASDAS response and ASAS 20, 40 and 70 responses according to the baseline 25-OHvitD levels (25-OHvitD <20ng/mL vs ≥20ng/mL; 25-OHvitD <30ng/mL vs ≥30ng/mL); and there were no statistically significant differences in the baseline 25-OHvitD levels at baseline according to the response to bDMARD at 6 and 12 months of bDMARD (ASDAS: no response vs clinically important improvement or major improvement; ASAS 20: no response vs response).

In the line of these previous results, baseline 25-OHvitD levels did not predict the ASDAS response at 6 (OR 0.97 [0.95-1.00], 95% CI) or 12 (OR 0.98 [0.95-1.01], 95% CI) months of bDMARD.
Conclusion: Despite some data that suggest that lower levels of 25-OHvitD may be associated with higher disease activity in SpA, our results failed to demonstrate that the baseline 25-OHvitD levels can be related or predict treatment response after 6 and/or 12 months of therapy with the first bDMARD in real-life SpA patients.

057 - PREDICTORS OF RESPONSE TO THE FIRST BDMARD IN BIOLOGIC-NAÏVE PATIENTS WITH SPONDYLOARTHRITIS: A COHORT STUDY

Bruno Miguel Fernandes[1], Salomé Garcia[1], Filipe Oliveira Pinheiro[1], Maria Seabra Rato[1], Diogo Guimarães da Fonseca[2], Daniela Santos Oliveira[1,3], Ana Martins[1], Frederico Rajão Martins[4], Miguel Bernardes[1], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [3]Faculdade de Medicina da Universidade do Porto, Departamento de Medicina e CINTESIS, Porto, Portugal, [4]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal

Background: Several markers of response to biologic disease-modifying antirheumatic drugs (bDMARDs) have been identified in Rheumatoid Arthritis. However, data on predictors of response in Spondyloarthritis (SpA) are more limited.
Objectives: To identify predictors of response to bDMARDs in a SpA population.
Methods: Monocentric retrospective study including all the SpA patients (ASAS classification criteria)

TABLE 1. BASELINE CHARACTERISTICS THAT SHOWED STATISTICALLY SIGNIFICANT DIFFERENCES AT BASELINE BETWEEN GROUPS OF PATIENTS WITH VS WITHOUT ASDAS RESPONSES AT 6 AND 12 MONTHS OF BDMARD.

	ASDAS response at 6 months			ASDAS response at 12 months		
	yes	no	p-value	yes	no	p-value
Age at start of bDMARD (mean±SD)	39.6±12.2 years	44.2±10.7 years	p=0.012	40.7±12.8 years	44.2±10.9 years	p=0.035
Age at SpA diagnosis (mean±SD)	32.2±11.1 years	35.8±11.9 years	p=0.023	31.3±10.7 years	35.4±11.2 years	p=0.010
BMI (mean±SD)	25.7±4.3 kg/m^2	28.7±6.0 kg/m^2	p=0.045	25.6±4.3 kg/m^2	28.5±5.7 kg/m^2	p=0.005
CRP (mean±SD)	3.2±3.5 mg/dL	1.1±1.2 mg/dL	p<0.001	3.4±3.5 mg/dL	1.4±1.6 mg/dL	p<0.001
ESR (mean±SD)	36±22	25±20	p<0.001	38±24	27±17	p=0.001
ASDAS-CRP (mean±SD)	4.1±0.8	3.5±0.4	p<0.001	4.2±0.8	3.6±0.8	p<0.001
HLA-B27+	61.5%	26%	p=0.006	60.1%	44.4%	p=0.033
Males	62.3%	35.7%	p=0.004	62.3%	37.5%	p=0.001

(bDMARD: biologic disease-modifying antirheumatic drug; BMI: body mass index; CRP: C-reactive protein; ESR: erythrocyte sedimentation rate).

followed at our Rheumatology Department, registered in the national database and treated with bDMARD between July 2001 and August 2020. Demographic, clinical and laboratorial data at baseline and disease activity measures at 6 and 12 months of bDMARD were collected. Mann-Whitney U test and chi-square tests were used to the comparison analysis between groups (continuous and categorical variables, respectively) and univariate logistic regression was used in the prediction analysis.

Results: A total of 325 patients were included, 178 (54.8%) males, 76 (23.4%) smokers and 164 (50.5%) HLA-B27 positive. Concerning SpA subtypes: 236 (72.6%) had Ankylosing Spondylitis, 31 (9.5%) had Inflammatory Bowel Disease Associated SpA and 58 (17.9%) had Undifferentiated SpA. The mean age at the start of the first bDMARD was 41.7 years (±12.2) and the median disease duration was 12.1 years (0.5-52.7). The mean ASDAS-CPR (Ankylosing Spondylitis Disease Activity Score with C-reactive protein) was 4.0 (±0.8) and most patients (57.2%) exhibited very high disease activity at baseline as evaluated by ASDAS-CRP. Ninety-five (29.2%) patients were taking NSAIDs (nonsteroidal anti-inflammatory drugs) and 131 (40.3%) were under csDMARDs (conventional synthetic disease-modifying antirheumatic drugs), being sulfasalazine the most frequent (28.3%). All patients started iTNF (tumor necrosis factor inhibitors): adalimumab (30.2%) and golimumab (24.6%) were the most frequently started bDMARDs.

At 6 and 12 months of bDMARD, 63.5% and 65.7% of the patients had ASDAS response (clinically important improvement or major improvement). Variables that showed statistically significant differences at baseline between those different groups are presented at table 1. Body mass index (BMI) (OR 0.89 [0.80-0.99], 95% CI) and ASDAS-CRP at baseline (OR 2.8 [1.2-6.6], 95% CI) predicted ASDAS response at 6 months; moreover, only BMI (OR 0.91 [0.83-0.99], 95% CI) predicted ASDAS response at 12 months of bDMARD.

Conclusion: Our results demonstrate that a higher baseline disease activity predicts the response to bDMARDs in SpA. Interestingly, BMI at baseline also predicts ASAS response at 6 and 12 monthes of treatment with bDMARD, in line with some data that suggest an association between BMI and disease activity in SpA.

059 - MUSCULOSKELETAL MANIFESTATIONS IN A COHORT OF 234 INFLAMMATORY BOWEL DISEASE PATIENTS

Bruno Miguel Fernandes[1], Miguel Bernardes[1], Diana Rosa-Gonçalves[2], Fernando Magro[3], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar de São João, Porto, Portugal, [2]Rheumatology, Centro Hospitalar Entre o Douro e Vouga, Santa Maria da Feira, Portugal, [3]Serviço

de Gastrenterologia, Centro Hospitalar de São João, Porto, Portugal

Background: Musculoskeletal symptoms represent the most common extraintestinal manifestations of inflammatory bowel disease (IBD) and a major cause of impaired quality of life in these patients. Spondyloarthritis (SpA) is classically associated with IBD, but other rheumatic manifestations may occur.
Objectives: To characterize musculoskeletal symptoms and rheumatic diseases in an IBD cohort.
Methods: Retrospective monocentric descriptive study including all the patients with IBD consecutively reffered from Gastroenterology to the Rheumatology Department between January of 2013 and December 2020 in a tertiary university hospital. Demographic and clinical data and musculoskeletal symptoms were collected at the time of the first visit in the Rheumatology outpatient center and the rheumatic diseases diagnosed during the entire follow-up were registered.
Results: A total of 234 patients were included, 136 (58.1%) females, 20 (8.5%) smokers. At the first Rheumatology consultation the mean age was 43.6 (±13.7) years and the mean IBD duration was 11.7 (±9.7) years. Concerning IBD: 172 (73.5%) had Crohn's disease and 62 (26.5%) had ulcerative colitis; azathioprine (39.7%), infliximab (28.2%) and mesalazine (26.5%) were the most frequently used drugs; eleven patients (4.7%) were taking glucocorticoids and 106 (45.3%) had already been treated with glucocorticoids.

Regarding musculoskeletal symptoms: 76 (32.5%) patients had peripheral symptoms and 98 (41.9%) had axial symptoms (table 1). Twenty-six (11.1%) patients had radiographic sacroiliitis, 14 (6.0%) had sacroiliitis in computed tomography and 9 (3.8%) in magnetic resonance. Forty-four (18.8%) patients fulfilled Assessment of SpondyloArthritis international Society (ASAS) criteria for axial SpA and 5 (2.1%) for peripheral SpA. Also of note, 16 (6.8%) patients had a previous diagnosis of psoriasis and 5 (2.1%) had uveitis in the past.

Concerning other rheumatic diagnosis, we observed: osteoarthritis in 64 (27.3%), osteoporosis in 16 (6.9%), diffuse idiopathic skeletal hyperostosis in 6 (2.6%), systemic lupus erythematosus in 4 (1.7%), rotator cuff tendinopathy in 2 (0.9%), rheumatoid arthritis, gout, calcium pyrophosphate deposition disease, fibromyalgia, drug-induced lupus, osteitis condensans ilii, Dupuytren's contracture and avascular necrosis of the femoral head in 1 (0.4%), each.

TABLE 1. CHARACTERIZATION OF PERIPHERAL AND AXIAL MUSCULOSKELETAL SYMPTOMS IN PATIENTS WITH INFLAMMATORY BOWEL DISEASE.

		N (%)
Peripheral symptoms	No	158 (67.5%)
	Arthritis / "inflammatory" joint pain	24 (10.3%)
	"Mixed" rhythm joint pain	15 (6.4%)
	"Mechanical" joint pain	29 (12.4%)
	Enthesopathy	8 (3.4%)
Axial symptoms	No	136 (58.1%)
	"Inflammatory" back pain	46 (19.7%)
	"Mixed" rhythm back pain	35 (14.9%)
	"Mechanical" back pain	17 (7.3%)
Total		234 (100%)

Conclusion: Our results demonstrate a high prevalence of musculoskeletal symptoms and rheumatic diseases in patients with IBD. These diagnoses are not limited to the group of SpA and osteoporosis, emphasizing the importance of rheumatologists being alert to other rheumatic diagnoses in patients with IBD.

060 - BONE MINERAL DENSITY, VITAMIN D STATUS AND BONE METABOLISM IN A COHORT OF ADULT PATIENTS WITH INFLAMMATORY BOWEL DISEASE

Bruno Miguel Fernandes[1], Miguel Bernardes[1], Diana Rosa-Gonçalves[2], Fernando Magro[3], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar de São João, Porto, Portugal, [2]Rheumatology, Centro Hospitalar Entre o Douro e Vouga, Santa Maria da Feira, Portugal, [3]Serviço de Gastrenterologia, Centro Hospitalar de São João, Porto, Portugal

Background: Patients with inflammatory bowel disease (IBD) have an increased risk of low bone mineral density (BMD) and bone fractures due to several mechanisms. However, the best management of osteoporosis in this population is yet to determine.
Objectives: To evaluate bone mineral density and other clinical, analytical and demographic features related with the risk of bone fractures in an IBD cohort.
Methods: Retrospective monocentric study including all the patients with IBD consecutively reffered from Gastroenterology to the Rheumatology Department in a tertiary university hospital between January of 2013 and October 2020. Demographic, clinical and analytical

TABLE 1. CORRELATIONS BETWEEN CLINICAL/ANALYTICAL VARIABLES AND THE BMD IN AN IBD POPULATION

	Total hip BMD	Femoral neck BMD	Lumbar Spine BMD
Age	r=-0.356;p<0.001	r=-0.469;p<0.001	r=-0.259;p<0.001
Age at IBD diagnosis	r=-0.254;p<0.001	r=-0.327;p<0.001	r=-0.226;p=0.001
IBD duration	r=-0.147;p=0.031	r=-0.218;p=0.001	n.s.
Hemoglobin	r=0.249;p<0.001	r=0.209;p=0.002	n.s.
Albumin	r=0.189;p=0.005	r=0.208;p=0.002	n.s.
Erythrocyte sedimentation rate	r=-0.231;p=0.001	r=-0.206;p=0.003	n.s.

(BMD: Bone Mineral Density; IBD: Inflammatory Bowel Disease; n.s. not significant).

data and BMD by dual-energy X-ray absorptiometry (DXA) (total hip, femoral neck and lumbar spine) were collected at the time of the first visit in the Rheumatology outpatient center. Correlations between variables were evaluated by Spearman rank test and Mann-Whitney U test was used in the comparison analysis between groups (significance level at p<0.05), using SPSS 25.0 software.
Results: A total of 222 patients were included: 128 (57.7%) females, mean age of 43.4 (±13.6) years, mean IBD duration of 11.6 (±9.7) years. Regarding IBD: 163 (73.4%) had Crohn's disease (CD) and 59 (26.6%) had ulcerative colitis (UC); azathioprine (41.9%) and infliximab (29.8%) were the most frequently used drugs; 10 patients (4.5%) were taking glucocorticoids, 104 (46.8%) had been previously treated with glucocorticoids and 65 (29.3%) had already been exposed to high doses of glucocorticoids (prednisolone equivalent dose ≥ 7.5 mg/day). Ten patients (4.5%) had previous fragility fractures and 32 (14.4%) fulfilled diagnostic criteria of osteoporosis by DXA (T score ≤ -2.5). One hundred eighty-one (81.5%) patients exhibited low levels of 25-hydroxy vitamin D (<30ng/mL), 24 (10.8%) had high levels of parathormone and 150 (67.6%) showed elevated serum concentrations of beta-carboxy-terminal type-1 collagen crosslinks (beta-CTX). Three patients (1.4%) were under treatment with bisphosphonates and 18 (8.1%) were taking calcium and/or vitamin D supplements.

Of interest, serum levels of albumin correlated negatively with beta-CTX (r=-0.401; p<0.001) and positively with osteocalcin (r=0.259; p<0.001). Correlations between clinical/analytical variables and BMD are presented in table 1. Patients under glucocorticoids had lower mean total hip BMD (0.874±0.159 vs 1.008±0.176; p=0.022) and femoral neck BMD values (0.797±0.174 vs 0.933±0.179; p=0.014) in comparison with the group of patients that were not taking glucocorticoids.

No statistically significant differences in BMD values were found between the following subgroups: DC vs UC; normal vs low levels of 25-hydroxy vitamin D; patients exposed vs not exposed to high doses of glucocorticoids.
Conclusion: Our results show an important prevalence of undiagnosed and untreated osteoporosis in patients with IBD. Stronger correlations were found between clinical/analytical variables and femoral neck BMD. Of note are the weak correlations of BMD with acute-phase markers (negative correlation with erythrocyte sedimentation rate and positive correlations with hemoglobin and albumin) and of nutritional status (evaluated by albumin) with bone markers (negative correlation with the bone reabsorption marker beta-CTX and positive correlation with the bone formation marker osteocalcin).

066 - IS THERE AN ASSOCIATION BETWEEN AUTOANTIBODIES INDUCTION AND LOSS OF THERAPEUTIC EFFICACY IN PATIENTS WITH AXIAL SPONDYLOARTHRITIS AND PSORIATIC ARTHRITIS TREATED WITH ANTI-TNF-α AGENTS?

Ana Martins[1], Daniela Santos Oliveira[1], Frederico Rajão Martins[2], Filipe Oliveira Pinheiro[1], Maria Seabra Rato[1], Diogo Guimarães da Fonseca[3], Salomé Garcia[1], Bruno Miguel Fernandes[1], Sofia Pimenta[1, 4], Miguel Bernardes[1, 4], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [3]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [4]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Background: Induction of autoantibodies is frequently observed in patients (pts) treated with a TNF-α blocker. According to other authors, the incidence of induction of antinuclear antibodies (ANA) and anti-double stranded DNA antibodies (anti-dsDNA) varies between 23-57% and 9-33%, respectively. However, it is unknown whether the induction of these autoantibodies affects the pharmacokinetics and bioavailability of biotherapy and, consequently, reduces the efficacy and safety of the drug.
Objectives: To analyze if there is an association between autoantibodies induction and therapeutic efficacy in pts with axial spondyloarthritis (axSpA) and psoriatic arthritis (PsA) treated with anti-TNF-α agents.
Methods: We performed a retrospective analysis of pts with axSpA and PsA treated in our University Hospital with a TNF-α blocker as first biologic agent. We analysed the autoantibodies induction rate and investigated the influence of autoantibodies in therapeutic efficacy after 12 (T12) and 24 (T24) months of therapy. Clinical evaluation, laboratory findings and disease activity and functional scores (Bath Ankylosing Spondylitis Disease Activity Index, AS Disease Activity Score with CPR- ASDAS-CRP, Bath AS Functional Index) were collected from reuma.pt and medical records. For PsA pts, Disease Activity Score-28-CRP, Simple Disease Activity Index, Clinical Disease Activity Index and Health Assessment Questionnaire scores were also collected. Pts with positive ANA (titer>1/100) prior to anti-TNF-α therapy were excluded. Continuous and categorical variables were analyzed using a t-test and a chi-square test, respectively. P-value <0.05 was considered statistically significant.
Results: In the axSpA group, 236 pts were included, 44.5% females, mean age at diagnosis of 42.3 ± 12.4 years and median disease duration of 11.5 (IQR 6.0-21.0) years. Positive ANA were found in 16.9% at T12 and 26.3% at T24 and positive anti-dsDNA in 3.4% at T12 and 3.8% at T24, with similar conversion rates between different anti-TNF drugs and no significant gender difference. A significant difference in ASDAS-CPR was found in axSpA pts with and without ANA at T12 (p=0.047). However, no difference was found in the others disease activity and functional scores at T12. At T24, no significant difference, including ASDAS-CPR, was found. Also, no significant difference was found in pts with and without anti-dsDNA.

In the PsA group, 94 pts were included, 46.8% females, mean age at diagnosis of 46.7 ± 11.7 years and median disease duration of 11.5 (IQR 6.5-16.5) years. Positive ANA were found in 14.9% at T12 and 21.3% at T24 and positive anti-dsDNA in 2.1% at T12 and 3.2% at T24. When comparing the groups with and without ANA and with and without anti-dsDNA at T12 and T24, no significant difference in disease activity and functional scores was found.
Conclusion: This study revealed high rates of serology conversion, similar to the rates described before. The authors found that ASDAS-CPR was higher in axSpA pts with ANA after 12 months of therapy. However, this difference was no longer evident after 24 months. No other significant difference was found. We consider that the induction of autoantibodies may interfere with the response to anti-TNF-α therapy in a short and initial period of time. Long-term follow-up data are lacking to say whether that influence will disappear consistently over the long run, as we observed after 12 months of therapy. However, we can state that, a priori, seroconversion should not lead to treatment suspension because of concerns about loss of efficacy.

067 - SAFETY AND PERSISTENCE OF MONTHLY INTRAVENOUS ILOPROST IN SYSTEMIC SCLEROSIS

Patrícia Martins[1, 2], Eduardo Dourado[1, 2], JE Fonseca[1,2], Vasco C Romão[1, 2], Catarina Resende[1]
[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introduction: Vasculopathy is a crucial feature of systemic sclerosis (SSc). Raynaud's phenomenon (RP) and digital ulcers (DU) greatly impact the patients' quality of life. Intravenous (IV) iloprost is broadly used to treat RP and DU secondary to SSc. However, no internationally accepted standardised protocol on iloprost use is currently available.
Methods: We reviewed the clinical records of patients with SSc-related DU and/or moderate-to-severe RP (more than two attacks/day with at least moderate pain) not responsive to calcium channel blockers (CCB), receiving or that have received IV iloprost infusions from January 1st 2011 to March 31st 2021. There were no restrictions concerning combination treatments or comorbidities. Our protocol for IV iloprost consists of an initial ten-hour infusion of iloprost 200ng/mL

(0.05 mg in 250 ml of 0.9% saline solution) on five consecutive days, followed by a single-day infusion every month. Over the ten hours of infusion, there is a progressive increase of the dose, up to the patient's maximum tolerated dose, ranging from 0.5 to 1.5 ng/kg/min. The infusion rate starts at 4 ml/h and is increased according to the following scheme: 4 mL/h (1st hour), 8 mL/h (2nd hour), 12 mL/h (3rd hour), and then 16 mL/h if tolerated by the patient, until the end of the infusion. Adverse events were assessed by consulting clinical records.

TABLE 1. DEMOGRAPHIC AND CLINICAL DATA OF SSC PATIENTS.

Demographic data	
Female, n (%)	47 (95.9)
Mean age, years ± SD	56.7 ± 18.1
Mean age at diagnosis, years ± SD	47.9 ± 18
Median disease duration, years (range)	13.9 (1-55)
Median disease duration at the beginning of the treatment, years (range)	2.4 (0.3-6)
Clinical subtypes	
dcSSc, n (%)	12 (24.5)
lcSSc, n (%)	27 (55.1)
Sine scleroderma, n (%)	1 (2)
VEDOSS, n (%)	1 (2)
Overlap syndrome, n (%)	8 (16.3)
Auto-antibody	
Anti-Scl 70, n (%)	19 (38.8)
ACA, n (%)	24 (49)
Pm/Scl, n (%)	6 (12.2)
Clinical manifestations	
R, n (%)	49 (100)
Digital ulcers (active/history) , n (%)	42 (85.7)
Telangiectasia, n (%)	28 (57.1)
Calcinosis, n (%)	7 (14.3)
Dysphagia, n (%)	8 (16.3)
Reflux, n (%)	16 (32.7)
ILD, n (%)	18 (36.7)
PAH, n (%)	3 (6.1)
Arthritis/arthralgia, n (%)	18 (36.7)
Concomitant treatment for RP/DU	
Nifedipine/amlodipine, n (%)	45 (91.8)
Bosentan, n (%)	14 (28.6)
Sildenafil, n (%)	5 (10.2)

ACA – anticentromere antibodies; dcSSc - diffuse cutaneous systemic sclerosis; DU - digital ulcers; ILD – interstitial lung disease; lcSSc. - limited cutaneous SSc; PAH – pulmonary arterial hypertension; RP - Raynaud's phenomenon; SD - standard deviation; VEDOSS - very early diagnosis of systemic sclerosis.

Results: Forty-nine patients with SSc have been treated with IV iloprost according to our treatment protocol. Patients' characteristics and clinical features are presented in Table 1. Thirty patients initiated iloprost to treat DUs, 14 to treat RP and 5 to treat both. Sixty per cent of patients in the DU group resolved the DUs within the first month of therapy. RP significantly improved in 64% of patients in the RP group within a month. In the RP+DU group, 60% of patients resolved the DUs and significantly improved RP after three months. Currently, 36 patients are actively undergoing treatment. The reasons for discontinuation in the remaining 13 patients included clinical improvement (N=5), switch to treatment with ambulatory elastomeric pump (N=4), death (N=3) or change of follow-up to another hospital (N=1). Iloprost persistence at two and five years after treatment onset was 95.9% and 83.7% (Figure 1), respectively. Nine adverse events were recorded (18.4% of patients): headache was reported in four patients, hypotension in three patients, tachycardia in one patient and generalised erythroderma in one patient.

Conclusions: SSc patients achieved clinical improvement with a good tolerability profile, leading to a high drug persistence rate. Side effects were managed by adapting the infusion rate. Our data support that

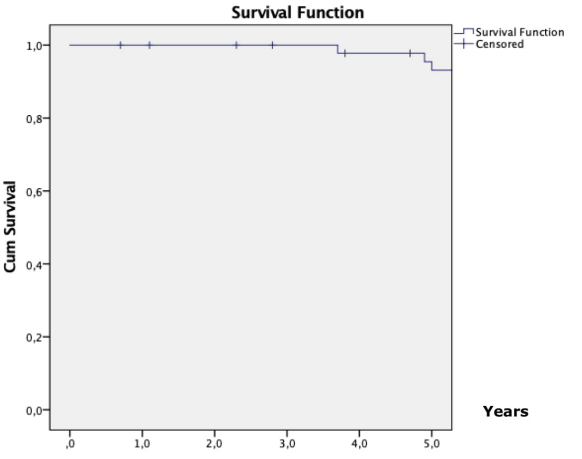


FIGURE 1. KAPLAN-MEIER ANALYSIS OF THE TREATMENT PERSISTENCE OF ILOPROST UP TO FIVE YEARS AFTER TREATMENT ONSET.

monthly single iloprost infusions can be effective, safely administered and adjusted according to the drug tolerance.

068 - CLINICAL CHARACTERIZATION OF PORTUGUESE PATIENTS WITH ANTISYNTHETASE SYNDROME

Patrícia Martins[1, 2], Eduardo Dourado[1, 2], Ana Teresa Melo[1, 2], Beatriz Samões[3], Marlene Sousa[4], Raquel Freitas[5], Maria Helena Lourenço[6], Bruno Miguel Fernandes[7], Emanuel Costa[8], Hugo Parente[9], Frederico Rajão Martins[10], JE Fonseca[1, 2], Vasco C Romão[1, 2], Nikita Khmelinskii[1, 2], Raquel Campanilho-Marques[1, 2], Inês Cordeiro[1, 2]

[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Hospital de Santa Maria, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [4]Rheumatology Department, Centro Hospitalar Universitário de Coimbra, Coimbra, Portugal, [5]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [6]Rheumatology Department, Hospital Egas Moniz (CHLO), Lisboa, Portugal, [7]Serviço de Reumatologia, Centro Hospitalar de São João, Porto, Portugal, [8]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal, [9]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [10]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal

Background: Antisynthetase syndrome (ASyS) may have different clinical phenotypes and outcomes associated with different anti-aminoacyl RNA-synthetase (anti-ARS) antibodies. Its wide clinical spectrum can include inflammatory myopathy, interstitial lung disease (ILD), arthritis, fever, mechanic's hands, and Raynaud phenomenon (RP).

Objective: To describe a nationwide, multicentre cohort of Portuguese patients with ASyS.

Methods: Retrospective analysis of patients with ASyS from nine Portuguese Rheumatology centers. Data on patients' signs and symptoms, laboratory

TABLE 1. PATIENT CHARACTERISTICS ACCORDING TO THE ANTI-ARS.

Variables	Overall, n=70	Jo-1, n=42 (60%)	PL-12, n=11 (15.7%)	PL-7, n=10 (14.3%)	EJ, n=4 (5.7%)	OJ, n=2 (2.9%)
Mean age at onset, yrs	52 ± 15	46.6 ± 14.4	55.2 ± 14.7	56.5±12.5	56.3±11.2	73.5±2.1
Female, n (%)	49 (70)	29 (69)	9 (81.8)	7 (70)	2 (50)	2 (100)
Median age in years at disease onset (IQR)	52 (15-75)	48 (15-70)	59 (20-70)	62 (39-73)	60 (40-65)	73.5 (72-75)
Median follow-up time in yrs (IQR)	3 (0-32)	5 (0-32)	3 (0-13)	1 (1-4)	4 (2-21)	1 (0-2)
Median diagnostic delay in yrs (IQR)	6 (1-33)	7 (1-33)	7 (2-19)	4 (1-23)	1.5 (1-2)	12.5 (2-21)
Myositis, n (%) and Comparison Anti-Jo.1 ARS vs PL-12 and PL-7	36 (51.4)	25 (59.5)	3 (27.3) *p < 0.01	4 (40) p=0.7	3 (75) -	0 -
ILD, n (%) and Comparison Anti-Jo.1 ARS vs PL-12 and PL-7	53 (75.7)	33 (78.6)	8 (72.7) p = 0.98	6 (60) p=0.56	4 (100) -	1 (50) -
ILD pattern - NSIP, n (%)	30 (56.6)	18 (54.5)	6 (75)	3 (50)	1 (25)	0
ILD pattern - UIP, n (%)	6 (11.3)	3 (9.1)	1 (12.5)	1 (16.7)	1 (25)	0
ILD pattern - other specific pattern, n (%)	6 (11.3)	4 (12.1)	0	2 (33.3)	1 (25)	0
ILD pattern - non-specific pattern, n (%)	11 (15.7)	8 (24.2)	1 (12.5)	0	1 (25)	1 (100)
Mechanic's hands (%), n (%)	23 (32.9)	14 (33.3)	3 (27.3)	2 (20)	0	1 (50)
General impairment, n (%)	18 (25.7)	11 (26.2)	3 (27.3)	2 (20)	2 (50)	0
Fever, n (%)	7 (10)	4 (9.5)	2 (20.2)	0	1 (25)	0
Raynaud phenomenon, n (%)	22 (31.4)	11 (26.2)	7 (63.6)	0	2 (50)	0
Arthritis, n (%) and Comparison Anti-Jo.1 ARS vs PL-12 and PL-7	43 (61.4)	29 (69)	5 (45.4) p=0.07	2 (20) *p < 0.01	2 (50) -	1 (50) -
Malignancy, n (%)	4 (5.7)	3 (7.1)	1 (9.1)	0	0	0
Deaths, n (%)	2 (2.9)	1 (2.4)	0	0	0	1 (50)

ILD - interstitial lung disease; IQR- interquartile range; NSIP - Non-specific interstitial pneumonia; UIP - Usual interstitial pneumonia; yrs - years

results, pulmonary radiological findings (computed tomography) and treatment (immunomodulators) were collected.

Results: Among the 70 patients included, 42 patients (60%) were anti-Jo1–positive, 11 (15.7%) were anti-PL12–positive, 10 (14.3%) were anti-PL7–positive, 4 (5.7%) were anti-EJ–positive and 2 (2.9%) were anti-OJ positive. In one patient it was not possible to identify the type of antibody. Antibody overlap was found in 15 patients (21.4%), who were positive for anti-Ro52 antibodies. The general clinical characteristics are shown in Table 1. The diagnostic delay was greater in patients positive for anti-OJ, followed by anti-Jo-1 and anti-PL12. The follow-up was shorter for anti-PL7 and anti-OJ-positive patients. Anti-PL7-positive patients had lower rates of arthritis when compared to anti-Jo1 ($p< 0.01$). When compared with anti-Jo-1 ARS, myositis was less common in anti-PL12 ($p < 0.01$). ILD prevalence was similar in the different ARS subgroups. Glucocorticoids (GCs) were the most frequently used class of drugs. A more conservative treatment plan (e.g. GCs plus methotrexate or azathioprine) was the treatment of choice in ASyS with myositis and/or arthritis involvement. Rituximab or mycophenolate mofetil were preferred when lung involvement occurred. Only two deaths were reported, being one associated with lung neoplasia.

Conclusion. This is the first study investigating the clinical phenotypes of Portuguese patients with ASyS. These results are generally concordant with data retrieved from international cohorts.

069 - SOBREDIAGNOSTICAMOS ARTRITE REUMATÓIDE SERONEGATIVA? O PAPEL DA ECOGRAFIA MÚSCULO-ESQUELÉTICA NA CLARIFICAÇÃO DE ARTROPATIAS SERONEGATIVAS

Diogo Esperança Almeida[1, 2], Emanuel Costa[1, 2], Francisca Guimarães[2], Ana Sofia Pinto[2, 3], Hugo Parente[2], Soraia Azevedo[2], José Tavares-Costa[2], Carmo Afonso[2], Daniela Santos-Faria[2], Marcos Cerqueira[1], Filipa Teixeira[2]

[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal, [2]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [3]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal

Introdução: Vários dados na literatura sugerem que a artrite reumatóide seronegativa (AR-) e a AR com positividade de factor reumatóide (FR) e/ou anticorpos anti-peptídeos citrulinados (antiCCP) (AR+) podem ter mecanismos fisiopatológicos diferentes, estando igualmente estabelecido que a AR+ têm um prognóstico menos favorável que a AR-. A positividade de FR e antiCCP tem também valor diagnóstico na AR. Várias outras artropatias, com mecanismos fisiopatológicos e tratamentos diferentes, podem mimetizar a apresentação da AR. São exemplos as espondilartrites periféricas, a artrite psoriática e as artropatias microcristalinas. Os autores levantam a possibilidade da AR- ser sobrediagnosticada na prática clínica perante uma artropatia inflamatória crónica seronegativa e sugerem que a ecografia músculo-esquelética poderá ajudar a clarificar estes quadros através da identificação e caracterização das lesões mais típicas de cada uma das condições mencionadas.

Objectivo: Comparar a frequência de lesões elementares ecográficas em articulações e enteses encontradas em doentes com AR- e AR+.

Métodos: Estudo transversal com um momento de avaliação ecográfica sistematizada de 10 articulações, 12 pontos de entese e aparelho tendinoso flexor e extensor de 4 dedos das mãos (tabela 1) em doentes consecutivos com AR-. Doentes com AR+ com características demográficas similares foram submetidos ao mesmo processo de avaliação. Os doentes com AR- tinham que cumprir todos os seguintes critérios: história de poliartrite crónica, negatividade de FR e antiCCP, ausência de manifestações extra-articulares e de história familiar de espondilartrite, e ausência de condrocalcinose radiográfica ou de identificação de cristais em artrocenteses prévias.

Resultados: Foram incluídos 23 doentes com AR- e 20 doentes com AR+, sem diferenças significativas quanto a sexo, idade, índice de massa corporal, tempo de evolução de doença e utilização de terapêutica biotecnológica.

Como esperado, considerando a associação de FR e antiCCP a uma evolução erosiva, doentes com AR- tiveram número de articulações com erosões significativamente inferior a doentes com AR+ (mediana 0.0 vs. 3.0, U 391.0, p<.001***), com diferença significativa em 3 das 5 articulações avaliadas (bilateralmente). Foram detectadas erosões em 6.5% das articulações de doentes com AR- e 30.5% das articulações de doentes com AR+.

Doentes com AR- tiveram um número de lesões estruturais ou inflamatórias associadas a entesite significativamente superior a doentes com AR+

(mediana 2.0 vs. 0.5, U 124,0, p=.008**), com a entesite tricipital a ser significativamente mais prevalente em doentes com AR- (p=.016*). Foram detectadas lesões de entesite em 18.1% das enteses de doentes com AR- comparativamente a 8.3% de doentes com AR+. Um doente com diagnóstico de AR- tinha aspectos ecográficos de dactilite não evidente na avaliação clínica.

Adicionalmente, foram detectados focos hiperecogénicos na cartilagem do joelho ou na fibrocartilagem triangular do carpo sugestivos de depósitos de pirofosfato de cálcio em 3 doentes com AR-.

Conclusões: Na presente amostra, foram encontradas lesões ecográficas características de outras artropatias, como entesite/dactilite e deposição de microcristais, numa parte relevante dos doentes diagnosticados com AR-. Os dados sugerem que a AR- poderá ser sobrediagnosticada na prática clínica. Uma avaliação ecográfica sistematizada e regularmente incluída no seguimento poderá contribuir para uma clarificação diagnóstica e consequente melhoria do tratamento em doentes com artropatia inflamatória crónica.

TABELA 1. COMPARAÇÃO DE FREQUÊNCIA DE LESÕES ECOGRÁFICAS ENCONTRADAS EM ARTICULAÇÕES E PONTOS DE ENTESE EM DOENTES COM AR- E AR+.

	AR- (n=23)	AR+ (n=20)	p-value
Número de pontos de entesite por doente – Mdn ± AIQ	2,0 ± 3,0	0,5 ± 2,0	,008**
Tricipital – n (%)	10 (43,5%)	2 (10,0%)	,016*
Quadricipital – n (%)	9 (39,1%)	6 (30,0%)	,381
Patelar superior – n (%)	4 (17,4%)	1 (5,0%)	,219
Patelar inferior – n (%)	-	-	-
Aquiliana – n (%)	10 (43,5%)	4 (20,0%)	,094
Fáscia plantar – n (%)	5 (21,7%)	1 (5,0%)	,127
Dactilite – D2 e 5 bilateral – n (%)	1 (4.3%)	-	-
Número de erosões articulares por doente – Mdn ± AIQ	0,0 ± 1,0	3,0 ± 3,0	<,001***
Apófise estilóide ulnar – n (%)	6 (26,1%)	11 (55,0%)	,052
Metacarpofalângica 2 – n (%)	3 (13,0%)	11 (55,0%)	,004**
Metacarpofalângica 5 – n (%)	1 (4,3%)	10 (50,0%)	,001**
Metatarsofalângica 1 – n (%)	-	3 (15,0%)	-
Metatarsofalângica 5 – n (%)	3 (13,0%)	11 (55,0%)	,004**
Condrocalcinose – fibrocartilagem triangular carpo + joelho – n (%)	3 (9,5%)	-	-
Duplo-controlo – qualquer articulação – n (%)	-	-	-

AR- – artrite reumatóide seronegativa; AR+ – artrite reumatóide com positividade de FR/antiCCP; Mdn – mediana; AIQ – amplitude interquartílica; n (%) – número absoluto (percentagem) de doentes com a lesão indicada.

073 - THE EXPERIENCE OF A MULTIDISCIPLINARY CLINIC FOR SYSTEMIC AUTOINFLAMMATORY DISEASES

Carolina Zinterl[1], Patrícia Costa Reis[2, 3], I Esteves[4], Ana Berta Sousa[5, 6], J Gonçalo[4], JE Fonseca[1, 7, 8, 9], Filipa Oliveira Ramos [1, 7, 8, 9]

[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Hospital de Santa Maria, CHLN, Lisboa, Portugal, [2]Departamento de Pediatria, Hospital de Santa Maria, Centro Hospitalar Universitário Lisboa Norte, Lisboa, Portugal, [3]Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal, [4]Unidade de Infecciologia e Imunodeficiências, Departamento de Pediatria, Hospital Santa Maria - CHULN, Lisboa, Portugal, [5]Serviço de Genética, Departamento de Pediatria, Hospital de Santa Maria, CHULN, Lisboa, Portugal, [6]Laboratório de Imunologia Básica, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal, [7]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal, [8]Instituto de Semiótica Clínica, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal, [9]Centro Académico de Medicina de Lisboa, Portugal, Lisboa, Portugal

Background: Systemic autoinflammatory diseases (SAID) are characterized by inappropriate activation of the innate immune system and include not only monogenic periodic fever syndromes, but also multifactorial conditions. As SAID are rare and represent a diagnostic challenge, a multidisciplinary approach is important to ensure successful diagnosis and adequate follow-up of these patients.

Objective: To describe the functioning of our multidisciplinary SAID clinic and to characterize our clinical experience.

Methods: Our SAID clinic takes place monthly and is managed by pediatric rheumatologists closely collaborating with pediatricians specialized in infectious diseases and immunodeficiencies and one medical geneticist. Patients' data is systematically incorporated into the Rheumatic Diseases Portuguese Register (Reuma.pt). Biological samples are stored in a biobank. We describe our clinical experience based on SAID patients registered into Reuma.pt/SAID between

July 2011 and June 2020.
Results: We have registered 176 patients (58% were male). The median age of disease onset was 3.1 years [1 month; 59 years] and the median time of follow-up was 1.4 years [0; 13 years]. The majority of patients were diagnosed with Periodic Fever, Aphthous Stomatitis, Pharyngitis, Adenitis Syndrome (PFAPA) (133), 20 with undefined SAID (uSAID) and 13 with monogenic SAID, including Familial Mediterranean Fever (FMF) (n=5), Tumor Necrosis Factor Receptor-Associated Periodic syndrome (TRAPS) (n=1), Cryopyrin-Associated Periodic Disease (CAPS) (n=1), and Hyperimmunoglobulin D Syndrome/Mevalonate Kinase Deficiency (HIDS/MKD) (n=2). A genetic test was performed in 31 patients (18%). A mutation responsible for the phenotype was found in 26% of the patients tested. 34 patients (19%) achieved remission.
Conclusions: The most common SAID in our dedicated clinic is PFAPA and these patients have a benign clinical course. FMF is the most common monogenic SAID. The percentage of patients with an identified causal mutation is low.

076 - PULMONARY INVOLVEMENT IN PRIMARY SJÖGREN'S SYNDROME

Ana Martins[1], Sofia Pimenta[1, 2], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Background: Primary Sjögren syndrome (pSS) is a chronic systemic auto-immune disease. Pulmonary manifestations are among the most frequent extraglandular manifestations and include, more frequently, xerotrachea, small airway disease and interstitial lung disease (ILD). ILD include mainly lymphocytic and non-specific interstitial pneumonia (LIP and NSIP, respectively), and, less frequently, usual interstitial pneumonia (UIP) and organizing pneumonia. ILD is an important factor contributing to impaired quality of life and survival rate of pSS patients.
Objectives: To investigate possible associations of pulmonary involvement (PI) and clinical, haematological, serological and histopathological characteristics of patients with pSS. To characterize the symptoms, pulmonary function and histopathological patterns of patients with PI.
Methods: We performed a retrospective analysis of patients with pSS who fulfilled the ACR/EULAR 2016 classification criteria followed in our University Hospital. Demographic data, disease duration, extraglandular involvement and histopathologic data were collected and analyzed. ESSDAI score was calculated. Fisher's exact test/chi-square test and Mann-Whitney U test were used to compare the groups with and without PI. In patients with pulmonary involvement, clinical characteristics, pulmonary function tests with DLCO, 6-minutes' walk test, histopathologic patterns and treatment were collected from medical records. Paired samples t-test was used to compare patients at diagnosis onset and at the last evaluation. P-value < 0.05 was considered statistically significant.
Results: In total, 66 patients were included. PI was documented in 8 patients (12,1%) and disease duration was 3,5 (1,5-7,25) years. Patients with PI were older (p=0,027) and had a higher ESSDAI score (p<0.001) than patients without PI. No other statistically significant difference was found.

In patients with PI (n=8), 7 had chronic cough and 6 had dyspnea. Pulmonary arterial hypertension was found in 1 patient (12,5%). Four patients had UIP, 2 had LIP, 1 had NSIP and 1 had pleuroparenchymal fibroelastosis. Spirometry at diagnosis found a restrictive pattern in 3 pts, an obstructive pattern in 3 pts and was normal in 2 pts. Forced expiratory volume in 1 second (FEV1) was 82,63 ± 25,37 (mean ± SD), forced vital capacity (FVC) was 92,64 ± 27,00, FEV1/FVC ratio was 80,07 ± 21,45 and diffusing capacity

TABLE 1. PULMONARY FUNCTION TEST RESULTS AND 6-MIN WALK TEST AT DIAGNOSIS ONSET AND AT THE LAST EVALUATION IN PATIENTS WITH PULMONARY INVOLVEMENT

Spirometry	At diagnosis onset	At the last evaluation	p-value
FEV1 (%) (mean ± SD)	82,63 ± 25,37	77,61 ± 15,55	0,373
FVC (%) (mean ± SD)	92,64 ± 27,00	84,91 ± 20,75	0,245
FEV1/FVC ratio (%)	80,07 ± 21,45	81,52 ± 25,38	0,734
$DLCO_{SB}$ (%) (mean ± SD)	51,65 ± 21,04	37,00 ± 25,41	0,021
6-min walk test	At diagnosis onset	At the last evaluation	
Distance (mean ± SD)	435,71 ± 66,23	292,67 ± 131,54*	0,025
Oxygen desaturation during the effort (%) (mean ± SD)	7,50 ± 5,32	11,33 ± 6,77	0,018

*2 missings

for carbon monoxide (DLCO) was 51,65 ± 21,04. Comparing the values of spirometry at diagnosis and at the last evaluation (time interval 3,5 years), we found a decline in pulmonary function (FVC decreased by 8.8% and FEV1 decreased by 6.0%) and a quick decrease of DLCO (decreased by 28%), with a significant difference between the 2 evaluations (p=0.021). Functional test also found a quick deterioration, with a significant decrease in the walk distance (p=0.025) and a significant decrease in oxygen saturation during exercise (p=0.018). Regarding the treatment, 4 patients were taking hydroxychloroquine, 7 systemic steroids, 4 rituximab, 2 azathioprine, 1 mycophenolate mofetil and 1 cyclophosphamide. Three patients (37,5%) were receiving supplementary oxygen.

Conclusions: ILD is a frequent manifestation in pSS patients. In this study, patients with PI were older and had more severe disease. As documented in this study, PI in pSS is a serious condition and has a quick progression, with rapid function and functional impairment. An early diagnosis and an adequate treatment are essential to guarantee a better prognosis in these patients.

078 - BEYOND THE JOINT – IMPACT OF SECUKINUMAB TREATMENT IN PSORIATIC ARTHRITIS - DATA FROM THE RHEUMATIC DISEASES PORTUGUESE REGISTRY (REUMA.PT)

José Tavares-Costa[1], Soraia Azevedo[1], Laires PA[2], Salomé Garcia[3], Patrícia Nero[4], Jorge Pestana Lopes[5], Martins-Martinho J [6], Ana Bento da Silva[7], Joana Ramos Rodrigues[1], Graça Sequeira[8], Emanuel Costa[9], Filipe Cunha Santos[10], Carolina Furtado[11], Bernardo Santos[12], Ana Raposo[13], Liliana Saraiva[14], Beatriz Samões[15], Filipe Barcelos[16], Maria José Santos[5], Helena Santos[16, 17]

[1]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [2]HE&OR - Novartis Portugal / ENSP – NOVA University of Lisbon, Lisboa, Portugal, [3]Rheumatology Department, Centro Hospitalar São João, Porto, Portugal, [4]Rheumatology Department, Hospital CUF Descobertas, Lisboa, Portugal, [5]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [6]Rheumatology Department, Centro Hospitalar Universitário de Lisboa Norte, Lisboa, Portugal, [7]Rheumatology Department, Centro Hospitalar Lisboa Ocidental, Lisboa, Portugal, [8]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, [9]Rheumatology Department, Hospital de Braga, Braga, Portugal, [10]Rheumatology Department, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, [11]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal, [12]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [13]Rheumatology Department, Centro Hospitalar Trás-os-Montes e Alto Douro, Vila Real, Portugal, [14]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [15]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [16]Portuguese Institute of Rheumatology, Lisboa, Portugal, [17]EpiDoc Research Unit, CEDOC, NOVA Medical School, Lisboa, Portugal

Background: Secukinumab has proved efficacy in moderate to severe psoriasis and in peripheral arthritis of psoriatic arthritis (PsA) patients.[1,2] Nevertheless, improved functional status, participation in social life, less fatigue and psychological distress, are of high priority for the patients.[3]

Objective: To analyze the effectiveness of secukinumab on quality of life (QoL), functional status and the well-being (physical and psychological) of PsA patients who initiated secukinumab.

Methods: This was a national, observational, multicenter, longitudinal study of patients with diagnosis of PsA using real world anonymous patient-level data from the Portuguese national register database - Reuma.pt.[4] Data from PsA health-related HR-QoL questionnaires as FACIT-F (Functional Assessment of Chronic Illness Therapy-Fatigue); HADS (Hospital Anxiety and Depression Scale); HAQ-DI (Health Assessment Questionnaire-Disability Index) and SF-36 (36-Item Short Form Health Survey), collected at baseline and after 3, 6 and 12 months after secukinumab initiation, between 1st January 2017 and 10th January 2021, were included in the analysis.

Results. In total 166 naïve or biologic exposed PsA patients who initiated treatment with secukinumab, were included in the anal. Overall, 54.8% were women, when secukinumab was started the mean age was 50.8 (SD 11.37) years and patients had a mean disease duration of 11.7 (SD 9.9) years prior to secukinumab initiation. For 36.7% of the patients, secukinumab was the first biologic. An improvement in HAQ-DI was observed at 6 months, and was sustained in 12 months after start secukinumab treatment. On the other hand, anxiety, fatigue and depression levels decreased soon after treatment initiation (at 3 months evaluation). Mean SF-36 scores, of different physical and mental

TABLE I – EFFECTIVENESS ASSESSMENTS WITH SECUKINUMAB THERAPY IN PSORIATIC ARTHRITIS PATIENTS

	Baseline (n=168)	3 months (n=153)	p-value*	6 months (n=138)	p-value*	12 months (n=117)	p-value*
HAQ-DI, mean (SD)	1.3 (0.7)	1.1 (0.7)	NS	1.0 (1.0)	**< 0.001**	1.0 (0.9)	**0.025**
SF-36							
Physical functioning, mean (SD)	43.4 (26.0)	46.0 (27.6)	NS	48.5 (29.6)	NS	49.4 (26.6)	NS
Role limitations due to physical problems, mean (SD)	35.7 (31.5)	40.4 (33.)	NS	43.0 (34.1)	**0.008**	48.2 (39.1)	**0.004**
Bodily pain, mean (SD)	30.4 (14.9)	51.4 (20.0)	**0.002**	45.3 (17.3)	**0.001**	50.3 (22.1)	**<0.001**
General health, mean (SD)	33.3 (12.8)	37.3 (12.6)	NS	35.1 (13.5)	NS	34.8 (18.9)	**0.030**
Vitality/Energy/fatigue, mean (SD)	45.0 (10.7)	50.9 (12.8)	NS	47.7 (13.7)	NS	47.1 (22.9)	NS
Social functioning, mean (SD)	67.6 (24.6)	74.3 (20.0)	**0.034**	72.1 (19.5)	**0.018**	70.6 (29.6)	NS
Role limitations due to emotional problems, mean (SD)	46.1 (38.9)	52.5 (38.8)	NS	58.3 (35.1)	**0.036**	52.9 (45.6)	NS
General mental health, mean (SD)	59.0 (16.6)	66.8 (17.6)	**0.017**	64.9 (21.6)	NS	60.0 (28.3)	NS
SF-36 physical component summary, mean (SD)	35.7 (15.5)	43.8 (18.4)	**0.014**	43.0 (17.6)	**0.002**	45.7 (20.8)	**<0.001**
SF-36 mental component summary, mean (SD)	43.9 (19.0)	61.1 (17.7)	**0.032**	60.7 (17.6)	**0.006**	57.6 (26.5)	NS
HADS							
Anxiety, mean (SD)	7.9 (3.8)	6.0 (3.1)	**0.003**	6.6 (4.5)	**0.011**	7.6 (5.2)	NS
Depression, mean (SD)	7.1 (9.7)	5.1 (3.7)	**0.004**	6.2 (3.7)	NS	7.0 (5.7)	NS
FACIT-F, mean (SD)	28.8 (10.1)	35.2 (6.3)	**0.009**	31.4 (9.7)	NS	29.9 (10.7)	NS

*p value - Comparisons across different timepoints and baseline; NS: non-significant
FACIT-F: Functional Assessment of Chronic Illness Therapy – Fatigue; HADS: Hospital Anxiety and Depression Scale; HAQ: Health Assessment Questionnaire Disability Index; SF-36: 36-Item Short Form Health Survey; SD: Standard Deviation; IQR: InterQuartile Range.

Sample size is not constant - Baseline: SF-36 (n=17); HADS (n=16); HAQ (n=34); FACIT-F (n=14); 3 months: SF-36 (n=17); HADS (n=16); HAQ (n=34); FACIT-F (n=14); 6 months: SF-36 (n=17); HADS (n=15); HAQ (n=40); FACIT-F (n=13);12 months: SF-36 (n=17); HADS (n=13); HAQ (n=34); FACIT-F (n=12);

domains, were better in different study time points, except for vitality and physical functioning, which did not change significantly over time. Bodily pain was consistently better across all study time points. At 12 months of study follow-up, improvements were seen in role limitations due to physical problems, bodily pain and general health. Mean physical SF-36 summary score was better in all study evaluations and mean mental SF-36 summary was better in 3 and 6 months evaluations (Table 1).

Conclusions. Secukinumab treatment in PsA patients led to an early improvement in physical and mental domains of QoL, fatigue, anxiety and depression. Patient perceived health status also had a sustained improvement.

Acknowledgement: This study was supported by Novartis and presented on Behalf of PROSAS Study group.

REFERENCES:

1. McInnes IB, Mease PJ, Kirkham B, Kavanaugh A, Ritchlin CT, Rahman P, et al. Secukinumab, a human anti-interleukin-17A monoclonal antibody, in patients with psoriatic arthritis (FUTURE 2): A randomised, double-blind, placebo-controlled, phase 3 trial. Lancet. 2015;386(9999):1137–46.
2. Langley RG, Elewski BE, Lebwohl M, Reich K, Griffiths CEM, Papp K, et al. Secukinumab in Plaque Psoriasis — Results of Two Phase 3 Trials. N Engl J Med. 2014;371(4):326–38.
3. Gudu T, Gossec L. Quality of life in psoriatic arthritis. Expert Rev Clin Immunol [Internet]. 2018;14(5):405–17. Available from: https://doi.org/10.1080/1744666X.2018.1468252
4. Canhão H, Faustino A, Martins F, Fonseca JE. Reuma. PT - the rheumatic diseases portuguese register. Acta Reumatol Port. 2011;36(1):45–56.

079 - EFFECTIVENESS AND SAFETY OF ORIGINAL AND BIOSIMILAR ETANERCEPT (ENBREL® VS BENEPALI®) IN BDMARD-NAÏVE PATIENTS IN A REAL-WORLD COHORT OF PORTUGAL

Ana Sofia Pinto[1, 2], Maria Margarida Cunha[3], Filipe Oliveira Pinheiro[4], Miguel Bernardes[4], Helena Assunção[5], Martins-Martinho J [6], Catarina Tenazinha[7], Ana Margarida Duarte-Monteiro[7], Susana Silva[8], Frederico Rajão Martins[9], Lígia Silva[10], Couto M[11], Margarida Faria[12], Filipe Araújo[13], Tomás Fontes[14], Daniela Santos-Faria[2], José Tavares-Costa[2]
[1]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, [2]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [3]Reumatology Department, Hospital Garcia de Orta, EPE, Almada, Portugal, [4]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [5]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [6]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [7]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Hospital de Santa Maria, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [8]Rheumatology Department, Hospital Infante D. Pedro (CHBV), Aveiro, Portugal, [9]Rheumatology Department, Hospital

TABLE I – EFFECTIVENESS ASSESSMENTS WITH SECUKINUMAB THERAPY IN PSORIATIC ARTHRITIS PATIENTS

	Rheumatoid Arthritis			Psoriatic Arthritis			Spondyloarhritis		
	Enbrel® (n=645)	Benepali® (n=219)	p-value	Enbrel® (n=267)	Benepali® (n=68)	p-value	Enbrel® (n=368)	Benepali® (n=126)	p-value
Female, n (%)	532 (82.5)	168 (76.7)	0.06	151 (56.6)	34 (50.0)	0.33	176 (47.8)	58 (47.4)	0.73
Age (years), mean (±SD)	52.9 (12.5)	55.0 (12.8)	0.05	48.6 (11.6)	50.0 (11.0)	0.36	43.8 (12.4)	45.0 (13.2)	0.46
Education (years), mean (±SD)	8.2 (4.5)	8.5 (4.1)	0.30	8.7 (4.2)	9.3 (4.6)	0.62	9.9 (3.9)	10.5 (4.6)	0.30
Smoker, n (%)	75 (14.5)	38 (23.0)	0.08	21 (10.4)	6 (13.3)	0.78	65 (24.8)	20 (22.0)	0.21
Alcohol current consumer, n (%)	27 (5.4)	15 (9.1)	0.33	27 (13.6)	5 (11.4)	0.70	20 (7.8)	9 (10)	0.23
BMI (Kg/m^2), mean (±SD)	27.2 (6.8)	25.0 (6.5)	0.15	27.9 (4.9)	27.4 (9.6)	0.89	26,7 (4.5)	26,1 (5.2)	0.84
Comorbidities, n (%)									
Hypertension	113 (25.1)	39 (26.9)	0.67	44 (22.4)	6 (14.3)	0.24	42 (18.7)	13 (18.8))	0.97
Dyslipidaemia	15 (3.3)	7 (4.8)	0.41	8 (4.1)	2 (4.8)	0.69	6 (2.7)	2 (2.9)	1.00
Diabetes	42 (9.3)	14 (9.7)	0.91	16 (8.2)	3 (7.1)	1.00	13 (5.8)	4 (5.8)	1.00
CV disease	31 (6.9)	5 (3.4)	0.13	4 (2.0)	0 (0.0)	1.00	11 (4.9)	3 (4.3)	1.00
HLA B27 positivity, n (%)	-	-	-	22 (20.8)	0 (0.0)	0.01	182 (72.8)	56 (62.9)	0.08
RF positivity, n (%)	386 (72.4)	151 (77.8)	0.14	11 (6.6)	1 (2.4)	0.47	-	-	-
ACPA positivity, n (%)	342 (69.9)	144 (77.4)	0.05	5 (3.8)	0 (0.0)	0.59	-	-	-
Erosive disease, n (%)	295 (57.2)	104 (55.3)	0.25	-	-	-	-	-	-
Disease duration (years), median (IQR)	7.9 (I11.7)	7.2 (10.7)	0.52	7.8 (9.7)	7.4 (8.8)	0.99	9.3 (14.4)	11.1 (15.0)	0.76
Treatment									
cDMARDs, n (%)	505 (78.7)	186 (84.9)	0.04	170 (63.7)	51 (75.0)	0.08	133 (36.3)	40 (31.7)	0.35
NSAIDs, n (%)	214 (33.3)	51 (23.3)	0.01	72 (27.0)	13 (19.1)	0.18	99 (27)	25 (19.8)	0.11
Corticosteroids, n (%)	426 (66.4)	152 (69.4)	0.41	93 (34.8)	27 (39.7)	0.45	73 (19.9)	15 (11.9)	0.04

SD: Standard deviation; BMI: Body Mass Index; CV: Cardiovascular; HLA: Human Leucocyte Antigen; RF: Rheumatoid Factor; ACPA: Anti-Citrullinated Protein Antibodies; cDMARD: conventional Disease-Modifying Anti Rheumatic Drugs; NSAID: Non-Steroid Anti-Inflammatory Drug

de Faro, CHA, Faro, Portugal, [10]Rheumatology, Centro Hospitalar Trás-os-Montes e Alto Douro, Vila Real, Portugal, [11]Rheumatology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [12]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [13]Rheumatology and Osteoporosis Unit, Hospital de Sant´Ana, SCML, Cascais, Portugal, [14]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal

Background: The patent expiration of the original etanercept in Europe has facilitated the development of biosimilar products. Non-significant differences in efficacy and safety were noted in clinical trials which are not expected to influence clinical performance. Nonetheless, daily practice data should be gathered to support the claim for biosimilarity.

Objective: To compare the effectiveness and safety of original and biosimilar etanercept, in bDMARD-naïve patients, measured by persistence rates over 3 years of follow-up

Methods: A retrospective multicenter non-interventional study, using data collected prospectively from Reuma.pt was done, including patients with: age ≥ 18 years old; diagnosis of Rheumatoid Arthritis (RA), Psoriatic Arthritis (PsA) or Spondyloarthritis (SpA) (axial or peripheral); active disease who initiated treatment with etanercept as first line of biological

treatment after 2010. Kaplan-Meyer was used to calculate the persistence rate in treatment. Disease activity at baseline and follow-up data at 6, 12, 18 and 24 months of treatment was compared using the chi-square for categorical variables and t-student or Mann-Whitney tests for continuous variables. Reasons for discontinuing therapy were summarized using descriptive statistics.

Results: We included 1694 patients (413 on Benepali® and 1280 on Enbrel®). The population's baseline characteristics (table 1) were not significantly different among both groups, except concomitant treatment in RA (higher use of conventional DMARDs in Benepali® group and higher use of NSAIDs in Enbrel® group) and in SpA patients (higher use of corticosteroids in Enbrel® group).

At baseline, a higher joint count was found in patients treated with Enbrel® with a statistical difference for swollen (p=0.03) and tender (p=0.01) joints count (SJC and TJC, respectively) in RA and in TJC in SpA patients (p=0.02). In RA patients, CDAI and SDAI were higher in patients who started Enbrel® (p=0.03; p=0.04, respectively). Pain measured by visual analogic scale was higher in SpA patients treated with Benepali® (p=0.03).

The 3-year PR was not significantly different between both treatment groups in RA, PsA and SpA (Figure1). In RA, PR in Benepali® was 72.6%, with a median time-on-drug (TOD) of 28.3 months; for Enbrel® PR was 63.6%, with a median TOD of 27.4 months (p=0.566). In PsA patients, the PR for Benepali® was 70.6%, with a median TOD of 27.6 months, and in Enbrel® 67.0%, with a median TOD of 28.1 months (p=0.743). In SpA patients, the PR were 78.4% for Benepali® (median TOD of 27.4 months) and 71.5% for Enbrel® (median TOD of 28.0 months (p=0.816)).

In RA patients, we did not find differences between both groups for the proportion of patients in remission or low disease activity by CDAI ≤10, SDAI≤11 or DAS28 <3.2 at 6, 12, 18 and 24 months of treatment. For PsA, no differences were found in the same timelines for DAPSA≤14, DAS28<3.2, BASDAI<4, ASDAS<2.1 or PsARC response. Also, in SpA patients, no differences were found in BASDAI<4, BASFI<4, ASDAS<2.1, ASDAS response and BASDAI response in all the timelines with the exception of BASDAI response at 18 months, which was achieved in fewer patients in biosimilar therapy (p=0.02).

Overall, 535 (31.6%) patients stopped etanercept (428 patients on Enbrel® and 107 patients on Benepali®). Discontinuations due to inefficacy were the most frequent, but there were no significant differences between both groups as for adverse events.

Conclusions: Benepali® and Enbrel® showed similar effectiveness and safety in RA, PsA and SpA in our cohort of patients.

081 - DOENÇA PULMONAR INTERSTICIAL NAS DOENÇAS REUMÁTICAS SISTÉMICAS – QUATRO ANOS E MEIO DE CONSULTA MULTIDISCIPLINAR DE PATOLOGIA DO INTERSTÍCIO PULMONAR

Ana Catarina Duarte[1], Ana Cordeiro[1], Miguel Lopes[2], João Cruz[3], Ana Oliveira[4], Ana Soares[5], Maria José Santos[1, 6], Jorge Soares[2]

[1]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal, [2]Serviço de Pneumologia, Hospital Garcia de Orta, Almada, Portugal, [3]Serviço de Radiologia, Hospital Garcia de Orta, Almada, Portugal, [4]Serviço de Anatomia Patológica, Hospital Garcia de Orta, Almada, Portugal, [5]Serviços Farmacêuticos, Hospital Garcia de Orta, Almada, Portugal, [6]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introdução: A doença pulmonar intersticial (DPI) ocorre em várias doenças reumáticas sistémicas (DRS), surgindo sobretudo em doentes com DRS já conhecida.

A DPI associada às DRS é um desafio e a multidisciplinaridade uma mais-valia na gestão destes doentes. Em Dezembro de 2016 foi criada a consulta multidisciplinar de patologia do interstício (CMPI) no Hospital Garcia de Orta, com a colaboração regular da reumatologia.

Objetivo: Descrever os doentes com DRS e suspeita/documentação de DPI, assim como os doentes com DPI cuja investigação etiológica revelou alterações clínicas e/ou analíticas enquadráveis em DRS; ilustrar o contributo da CMPI para o diagnóstico e tratamento destes doentes.

Resultados: Foram avaliados 58 doentes com DRS confirmada/em investigação e suspeita de DIP, tendo 10 sido reavaliados. Destes, 41 (70.7%) eram mulheres, com uma mediana de idades de 72 anos (intervalo interquartil 14.5). Em 45 doentes a DRS era já conhecida (Figura 1) e os restantes 13 tinham manifestações clínicas/alterações analíticas sugestivas de DRS. Contudo, só em 5 foi feito diagnóstico de DRS (2 artrite reumatoide [AR], 1 síndrome Sjögren primária

TABELA 1 – EVOLUÇÃO FUNCIONAL DOS DOENTES SOB TERAPÊUTICA IMUNOSSUPRESSORA E ANTI-FIBRÓTICA

DRS	Fármaco escolhido	Padrão radiográfico	PFR prévios a início de terapêutica		PFR 6-12 meses após alteração terapêutica	
			CVF (%)	DLCO (%)	CVF (%)	DLCO (%)
AR	RTX	UIP	56.5	44.8	55.9	-
	Associação pirfenidona		51.5	30.4	61.8	28.8
AR	RTX	UIP	66.3	62.9	63.6	63.8
AR	RTX	UIP	106	59.5	119.4	53.6
AR	Pirfenidona	UIP	70.1	44	74	57.9
AR	Pirfenidona	UIP	63.4	39.5	65.1	38.7
ES	Associação nintedanib (já sob MMF)	NSIP fibrosante	61.5	30.3	65.1	29
ES	RTX	UIP	56.3	38.7	66.3	42.2
ES	MMF	UIP	40.3	28.4	59	30.2
SSp	MMF	LIP	41.1	-	45.2	-
SSp	MMF	NSIP	81.1	66.1	79.7	55.8
SSp	MMF	LIP	75.7	69.3	90.9	80.1
SSp	Associação pirfenidona (já sob RTX)	NSIP fibrosante	46.8	-	49.2	25.2
LES	MMF	OP	73.6	66.2	79.4	68.1
LES	RTX	NSIP	26.9	20	36.5	-
DMTC	MMF	NSIP	83.4	81.3	88.1	77.8

DRS – doença reumática sistémica; PFR – provas de função respiratória; CVF – capacidade vital forçada; DLCO – capacidade de difusão do monóxido de carbono; AR – artrite reumatoide; ES – esclerose sistémica; SSp- Síndrome de Sjögren primária; LES – lúpus eritematoso sistémico; DMTC – doença mista do tecido conjuntivo; RTX- rituximab; MMF – micofenolato de mofetil; CYC – ciclofosfamida; UIP – pneumonia intersticial usual; NSIP – pneumonia intersticial não específica; LIP – pneumonia intersticial linfocítica; OP – pneumonia organizativa

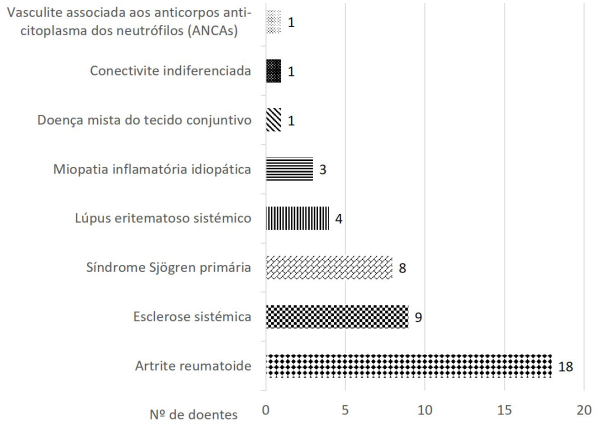
[SSp], 1 síndrome anti-sintetase, 1 granulomatose eosinofílica com poliangeíte - GEPA).

Na maioria dos doentes com DRS conhecida, a tomografia computorizada (TC) torácica permitiu definir o padrão de DPI subjacente - 16 com pneumonia intersticial usual (UIP), 12 com pneumonia intersticial não específica (NSIP; 4 destes com NSIP fibrosante), 3 com pneumonia organizativa e 2 com pneumonia intersticial linfocítica. Duas doentes com SSp tinham TC torácica sugestiva de pneumonite de hipersensibilidade. Um doente apresentava um padrão não classificável por TC tórax, mas pelas comorbilidades e estabilidade clínica/funcional foi decidido não realizar exames invasivos. Uma doente com AR e infiltrados algodonosos migratórios em TCs tórax seriadas foi diagnosticada com bronquiolite folicular (BF), através de biópsia pulmonar cirúrgica. Numa outra doente com esclerose sistémica e NSIP fibrosante, o crescimento de um nódulo pulmonar entre exames levou ao diagnóstico de adenocarcinoma do pulmão.

Em 8 doentes com DRS conhecida a discussão em CMPI excluiu DPI. Um desses doentes, com lúpus eritematoso sistémico, foi diagnosticado com shrinking lung, dado o padrão funcional restritivo desproporcional às alterações imagiológicas.

As decisões terapêuticas em 24 doentes com DPI associada a DRS incluíram o início/ajuste de imunossupressão em 15 doentes. Os fármacos preferencialmente prescritos foram o rituximab e o micofenolato de mofetil, ambos em 7 doentes. Um doente com GEPA iniciou apenas prednisolona (0.5mg/kg/dia). Foi ainda decidido início de terapêutica anti-fibrótica em 10 doentes (7 com padrão UIP e 3 com NSIP fibrosante), mantendo-se em 3 destes imunossupressão concomitante. Na doente com BF decidiu iniciar-se claritromicina, tendo em conta o

FIGURA. DISTRIBUIÇÃO DOS DIAGNÓSTICOS REUMATOLÓGICOS À DATA DA PRIMEIRA APRESENTAÇÃO NA CONSULTA MULTIDISCIPLINAR DE PATOLOGIA DO INTERS



papel imunomodulador. Nos restantes doentes com DPI e DRS, optou-se por manter vigilância funcional. Na tabela apresentamos os valores das provas de função respiratória de 15 doentes antes e após o ajuste terapêutico; os doentes com menos 6 meses de terapêutica foram excluídos.

Dos 58 doentes avaliados, 11 faleceram. A infeção respiratória foi a principal causa do óbito (5 doentes), seguida da progressão da DPI (4 doentes).

Conclusão: A criação da CMPI permitiu agilizar o diagnóstico e o tratamento, muitas vezes com decisões que ultrapassaram as indicações formais dos fármacos. A longo prazo, esperamos que esta abordagem integrada tenha impacto na sobrevida destes doentes.

082 - DISEASE ACTIVITY AND INFLAMMATION FOLLOWING WITHDRAWAL OF CERTOLIZUMAB PEGOL TREATMENT IN AXIAL SPONDYLOARTHRITIS PATIENTS WHO DID NOT EXPERIENCE FLARES DURING THE C-OPTIMISE STUDY

Lianne Gensler[1], Xenofon Baraliakos[2], Ana Lourenço[3], Lars Bauer[4], Bengt Hoepken[4], Thomas Kumke[4], Mindy Kim[5], Robert Landewé[6]

[1]University of California San Francisco, San Francisco, CA, United States, [2]Rheumazentrum Ruhrgebiet, Ruhr-University Bochum, Herne, Germany, [3]UCB Pharma, Paço de Arcos, Portugal, [4]UCB Pharma, Monheim am Rhein, Germany, [5]UCB Pharma, Smyrna, Georgia, United States, [6]Rheumatology, Zuyderland Medical Center, Heerlen, Netherlands

Background: C-OPTIMISE was a phase 3b clinical trial investigating certolizumab pegol (CZP) maintenance dose continuation, reduction or withdrawal following achievement of sustained remission in patients with axial spondyloarthritis (axSpA). During the C OPTIMISE maintenance period, the majority of patients randomised to CZP, either the full or reduced maintenance dose, did not experience disease flares. Conversely, in those who had CZP withdrawn, only a minority of patients remained flare-free.1

Objectives: This post-hoc analysis evaluates disease activity and clinical markers of inflammation in patients who did not experience a disease flare following randomisation to CZP full maintenance dose, CZP reduced maintenance dose or placebo (PBO) during the maintenance period (Weeks 48–96) of C OPTIMISE.

Methods: C-OPTIMISE (NCT02505542) was a multicentre, double-blind, parallel-group, randomised phase 3b study with a 48-week open-label run-in period.1 Adult patients with early (<5 years' symptom duration) active axSpA received open-label CZP 200 mg every 2 weeks (Q2W) for the first 48 weeks; from Week 48, patients who achieved sustained remission (Ankylosing Spondylitis Disease Activity Score [ASDAS] <1.3 at Week 32 or 36 and Week 48) were randomised 1:1:1 to double-blind CZP 200 mg Q2W (full maintenance dose), CZP 200 mg Q4W (reduced maintenance dose) or PBO for a further 48 weeks (maintenance period). A flare was defined as ASDAS ≥2.1 at two consecutive visits or ASDAS >3.5 at any visit. We report ASDAS, Bath Ankylosing Spondylitis Disease Activity Index (BASDAI), and C-reactive protein (CRP) and faecal calprotectin levels during Weeks 48–96 in CZP- and PBO-randomised patients who did not experience a flare (i.e., completed Week 96 on randomised treatment). Missing data were imputed using last observation carried forward.

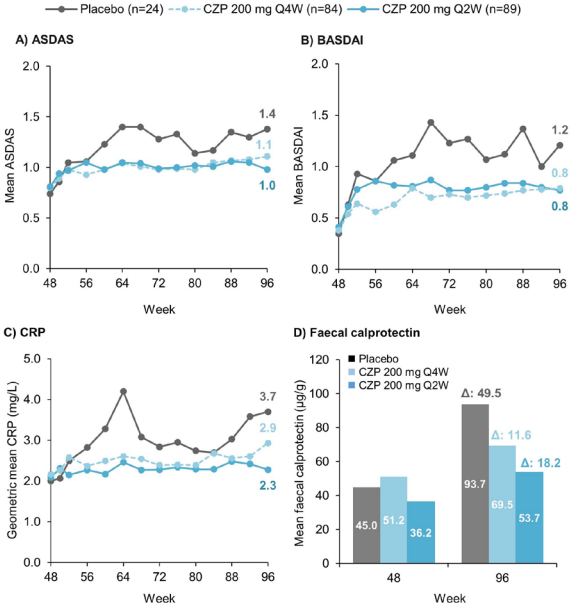
Results: Of 313 patients entering the maintenance period at Week 48, 197 (62.9%) completed Week 96 on randomised treatment without experiencing a flare; of these, 89 (85.6%) and 84 (80.0%) patients were in the CZP 200 mg Q2W and CZP 200 mg Q4W arm, respectively, with only 24 (23.1%) patients randomised to PBO not experiencing a flare. Baseline characteristics of these patients are shown in the Table. During Weeks 48–96, disease activity (ASDAS, BASDAI) and CRP levels were comparable between the CZP full and reduced maintenance dose group, and lower in both CZP arms than in PBO (Figure A–C). From Week 60 up to Week 96, PBO patients who did not flare had

TABLE. BASELINE (WEEK 0) CHARACTERISTICS OF PATIENTS WHO DID NOT EXPERIENCE FLARES DURING THE C-OPTIMISE MAINTENANCE PERIOD

	Placebo (n=24)	CZP 200 mg Q4W (n=84)	CZP 200 mg Q2W (n=89)
Age (years), mean (SD)	29.8 (7.4)	32.9 (6.7)	32.4 (7.2)
Male, n (%)	19 (79.2)	69 (82.1)	69 (77.5)
Time since diagnosis (years)			
Mean (SD)	2.0 (1.8)	2.0 (1.7)	2.5 (1.6)
Median	1.2	1.2	2.7
Symptom duration (years)			
Mean (SD)	2.7 (1.7)	3.4 (1.9)	3.9 (2.9)
Median	2.8	3.5	3.9
ASDAS, mean (SD)	3.4 (0.8)	3.7 (0.8)	3.7 (0.7)
BASDAI, mean (SD)	6.3 (1.1)	6.6 (1.5)	6.4 (1.4)
CRP (mg/L), geometric mean	6.28	7.88	7.35
Faecal calprotectin (µg/g), mean (SD)	71.8 (111.4)	87.1 (110.5)	81.0 (120.0)

SD: standard deviation.

FIGURE. (A) ASDAS, (B) BASDAI, (C) CRP AND (D) FAECAL CALPROTECTIN IN PATIENTS WHO DID NOT EXPERIENCE A FLARE DURING THE C-OPTIMISE MAINTENANCE PERIOD (WEEKS 48-96)



Missing data were imputed using last observation carried forward. Δ values for fecal calprotectin show change from Week 48. ASDAS: Ankylosing Spondylitis Disease Activity Score; BASDAI: Bath Ankylosing Spondylitis Disease Activity Index; CRP: C-reactive protein; CZP: certolizumab pegol; Q2W/Q4W: every 2/4 weeks.

consistently higher mean ASDAS, BASDAI and CRP levels compared with CZP-randomised patients (Figure A–C). Similarly, there was a greater increase in faecal calprotectin levels between Weeks 48 and 96 in the PBO arm compared with both CZP arms (Figure D).

Conclusions: Despite not meeting the threshold for a flare, consistently higher disease activity and increases in serologic and inflammatory biomarkers were observed in PBO-randomised patients who did not experience a flare during the C OPTIMISE study compared to those who remained on CZP. These findings confirm that patients with axSpA who achieve sustained remission benefit from continued CZP treatment, either with the full or reduced maintenance dose, over treatment withdrawal.

Acknowledgements: This study was funded by UCB Pharma. Editorial services were provided by Costello Medical.

REFERENCES

1. Landewé R. Ann Rheum Dis 2020;79:920–8.

085 - SECUKINUMAB 150 MG PROVIDES SUSTAINED IMPROVEMENT IN SIGNS AND SYMPTOMS OF NON-RADIOGRAPHIC AXIAL SPONDYLOARTHRITIS: 2-YEAR RESULTS FROM THE PREVENT STUDY

Denis Poddubnyy[1], Atulya Deodhar[2], Xenofon Baraliakos[3], Ricardo Blanco[4], Eva Dokoupilova[5], Stephen Hall[6], Alan Kivitz[7], MGH van de Sande[8], Anna Stefanska[9], Patricia Pertel[10], Hanno Richards[10], J Braun[11], Maria José Santos[12, 13]

[1]Charité - University Medicine, Berlin, Germany, [2]Oregon Health and Science University, Portland, United States, [3]Rheumatology, Rheumazentrum Ruhrgebiet, Herne, Germany, [4]Division of Rheumatology, Hospital University Marqués de Valdecilla, IDIVAL, Santander, Spain, [5]Department of Pharmaceutical Technology, Faculty of Pharmacy, Brno, Medical Plus, sro, Masaryk University, Uherske Hradiste, Czech Republic, [6]Department of Medicine, Monash University, Melbourne, Australia, [7]Clinical Research Division, Altoona Center for Clinical Research, Pennsylvania, United States, [8]Amsterdam Rheumatology and immunology Center, VU University Medical Center, Amsterdam, Netherlands, [9]Immunology, Hepatology and Dermatology, Novartis Ireland Limited, Dublin, Ireland, [10]Immunology, Hepatology and Dermatology, Novartis Pharma AG, Basel, Switzerland, [11]Department of Rheumatology, Rheumazentrum Ruhrgebiet, Herne, and Ruhr-University Bochum, Bochum, Germany, [12]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [13]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Background: Axial spondyloarthritis (axSpA) is an inflammatory disease characterised by chronic back pain, and it comprises radiographic axSpA and non-radiographic axSpA (nr-axSpA).[1] Secukinumab (SEC) 150mg, with (LD) or without loading (NL), dose significantly improved the signs and symptoms of patients with nr-axSpA in the PREVENT (NCT02696031) study through Week 52.[2]

Objectives: To report the long-term clinical efficacy and safety of secukinumab from the PREVENT study through 2 years.

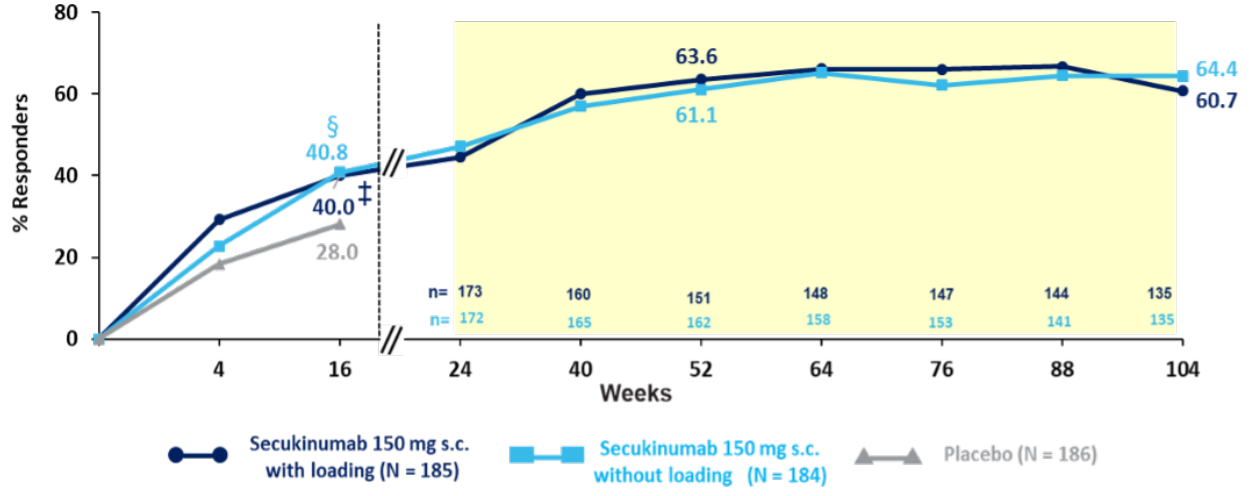
Methods: A detailed study design, key primary and secondary endpoints have been reported previously.[2] In total, 555 patients fulfilling ASAS criteria for axSpA plus abnormal C-reactive protein (CRP) and/or MRI, without evidence of radiographic changes in sacroiliac (SI) joints according to modified New York Criteria for AS were randomised (1:1:1) to receive SEC 150mg with LD, NL, or placebo (PBO) at baseline. LD patients received SEC 150mg at Weeks 1, 2, 3, and 4, and then every 4 weeks (q4wk) starting at Week 4. NL patients received SEC 150mg at baseline and PBO at weeks 1, 2, and 3, and then 150mg q4wk. 90% patients were anti-tumour necrosis factor (anti-TNF) naïve, 57% had elevated CRP and 73% had evidence of SI joint inflammation on MRI. All images

TABLE: SUMMARY OF CLINICAL EFFICACY (OBSERVED DATA)

Endpoints	Week	SEC 150 mg LD (N=185)	SEC 150 mg NL (N=184)	PBO-SEC 150 mg (N=186)
*ASAS40 in anti-TNF-naïve patients, n/M (%)	52[a]	90/137 (65.7)	95/145 (65.5)	85/151 (56.3)
	104	78/123 (63.4)	83/123 (67.5)	83/134 (61.9)
BASDAI change from baseline, mean±SD	52[a]	–3.7±2.8	–3.7±2.6	–3.3±2.4
	104	–4.1±2.6	–3.9±2.6	–3.7±2.5
BASDAI50, n/M (%)	52[a]	90/153 (58.8)	92/163 (56.4)	90/161 (55.9)
	104	88/137 (64.2)	84/136 (61.8)	87/142 (61.3)
ASAS partial remission, n/M (%)	52[a]	46/152 (30.3)	56/163 (34.4)	46/161 (28.6)
	104	51/137 (37.2)	50/135 (37.0)	50/142 (35.2)
ASDAS CRP inactive disease, n/M (%)	52[a]	49/152 (32.2)	58/163 (35.6)	48/160 (30.0)
	104	50/132 (37.9)	53/133 (39.8)	53/142 (37.3)

*For anti-TNF-naïve patients, N=164, LD; 166, NL; 171, PBO-SEC.
[a] total number of evaluable patients including open-label SEC and standard of care (SOC; 2 patients in LD, 1 patient in NL continued on SOC). After Week 52, only patients who continued to receive open-label SEC are presented.
ASAS, Assessment of SpondyloArthritis International Society; ASDAS, Ankylosing Spondylitis Disease Activity Score; BASDAI, Bath Ankylosing Spondylitis Disease Activity Index; M, number of patients with evaluation; N, total randomised patients; n, number of patients who are responders; SD, standard deviation

IMAGES: FIGURE. ASAS40 RESPONSE WAS MAINTAINED THROUGH WEEK 104 IN THE OVERALL POPULATION



were assessed centrally before inclusion. All patients continued to receive open-label SEC 150mg treatment after Week 52. Efficacy assessments through Week 104 included ASAS40 in anti-TNF naïve patients, ASAS40, BASDAI change from baseline, BASDAI50, ASAS partial remission, and ASDAS-CRP inactive disease in the overall population. The safety analyses included all patients who received ≥1 dose of study treatment for the entire treatment period up to Week 104. Data are presented as observed.

Results: Overall, 438 patients completed 104 weeks of study: 78.9% (146/185; LD), 77.7% (143/184; NL) and 80.1% (149/186; PBO). Efficacy results at Week 52 were sustained through Week 104 and are reported in the Table 1. The safety profile was consistent with the previous reports with no deaths reported during the entire treatment period up to Week 104.2

Conclusion: Secukinumab 150mg demonstrated sustained improvement in the signs and symptoms of patients with nr-axSpA through 2 years. Secukinumab was well tolerated with no new or unexpected safety signals.

REFERENCES

1. Strand V, et al. J Clin Rheumatol. 2017; 23(7):383–91.
2. Deodhar A, et al. Arthritis Rheumatol. 2020. Online ahead of print.

086 - DETERMINANTS OF QUALITY OF LIFE IN RHEUMATOID ARTHRITIS AND SPONDYLOARTHRITIS: HOW DO THEY INTERACT FROM A HIERARCHICAL PERSPECTIVE?

Pedro David Carvalho[1, 2, 3], Elsa Vieira-Sousa[2, 4], Marreiros A[3, 5], Pedro MAchado[6, 7, 8]

[1]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal, [2]Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Algarve Biomedical Center, Faro, Portugal, [4]Rheumatology and Bone Metabolic Diseases Department, Centro Hospitalar Lisboa Norte, EPE - Hospital de Santa Maria, Lisbon Academic Medical Centre, Lisboa, Portugal, [5]Universidade do Algarve, Faro, Portugal, [6]Rheumatology, Northwick Park Hospital, London North West University Healthcare NHS Trust, London, United Kingdom, [7]Centre for Rheumatology & MRC Centre for Neuromuscular Diseases, University College London, London, UK., London, United Kingdom, [8]Centre for Rheumatology, University College of London, London, United Kingdom

Introduction/Aim: To assess the hierarchy of outcomes contributing for quality of life (QoL) in rheumatoid arthritis (RA) and spondyloarthritis (SpA).

Materials and Methods: The COMOrbidities in SPondyloArthritis (COMOSPA) and COMOrbidities in Rheumatoid Arthritis (COMORA) initiatives are two multicentre transversal studies resulting from an international collaboration which enrolled 7904 consecutive patients with SpA and RA from 26 countries. In this study we analysed data on QoL assessed by EuroQOL-5 Dimension 3-level (EQ-5D-3L) in both databases. Both RA and then SpA subgroups were analysed separately. In a first exploratory step, linear regression models were used to identify univariable associations between several demographic and clinical variables and EQ-5D. Variables with a p-value <0.10 in the univariable analysis were tested in linear regression multivariable models. Subsequently, a decision tree was constructed according to an unbiased hierarchical multivariable analysis using the Chi-square Automatic Interaction Detector (CHAID) method, with EQ-5D as dependent variable. We stipulated a minimum requirement of 20 cases for parent branches and a minimum of 10 cases for child branches.

Results: Data on 3920 RA patients and 3984 SpA patients was analysed. In RA subgroup QoL was significantly associated with modified Health Assessment Questionnaire (MHAQ) [adjusted (adj) B=-0.216, (95% confidence interval (CI)= (-0.247,-0.184)], Disease Activity Score 28 (DAS28)-CRP-3 variables [adjB=-0.028, 95%CI=(-0.037,-0.019)], work productivity loss [adjB=-0.003, 95%CI=(-0.003,-0.002)], presence of unequivocal radiological erosion [adjB=-0.036, 95%CI=(-0.058,-0.014)], current alcohol consumption ≥3 units per day [adjB=0.015, 95%CI=(0.004,0.026)] and body mass index (BMI) [adjB=0.002, 95%CI=(0.000,0.004)] (table 1). In SpA subgroup QoL was significantly associated with MHAQ [adjB=-0.330, 95%CI=(-0.361,-0.298)], Ankylosing Spondylitis Disease Activity Score (ASDAS) [adjB=-0.064, 95%CI=(-0.077,-0.052)], work productivity loss [adjB=-0.002, 95%CI=(-0.003,-0.002)], current NSAID treatment [adjB=-0.046, 95%CI=(-0.067,-0.024)], current bDMARD treatment [adjB=-0.038, 95%CI=(-0.059,-0.017)], age [adjB=-0.002, 95%CI=(-0.003,-0.001)], male gender [adjB=0.024, 95%CI=(0.002,0.046)], universitary education [adjB=-0.017, 95%CI=(-0.033,-0.001)] and HLA-B27 positivity [adjB=0.023, 95%CI=(0.000,0.046)] (table 1). The decision tree revealed MHAQ as the first variable with the most discriminative power on EQ-5D, followed by work productivity loss and disease

TABLE 1 - MULTIVARIABLE LINEAR REGRESSION ANALYSES TO INVESTIGATE THE ASSOCIATION BETWEEN QOL (ASSESSED BY EQ-5D-3L) AND OTHER DEMOGRAPHIC AND CLINICAL VARIABLES IN THE 3920 PATIENTS WITH RA AND 3984 PATIENTS WITH SPA

Characteristics	Adjusted B* (95% CI) for RA	p-value	Adjusted B* (95% CI) for SpA	p-value
Age			-0.002 (-0.003, -0.001)	0.001
Male gender	0.008 (-0.017, 0.033)	0.533	0.024 (0.002, 0.046)	0.036
Education, university or equivalent	0.001 (-0.013, 0.016)	0.849	-0.017 (-0.033, -0.001)	0.041
BMI	0.002 (0.000, 0.004)	0.045	-0.001 (-0.003, 0.001)	0.593
HLA-B27 positive			0.023 (0.000, 0.046)	0.047
Seropositive for RF or ACPA				
Unequivocal radiological erosion	-0.036 (-0.058, -0.014)	0.001		
Current smoking				
Current alcohol equal or more than 3 units	0.015 (0.004, 0.026)	0.007	0.009 (-0.001, 0.018)	0.067
ASDAS-CRP			-0.064 (-0.077, -0.052)	<0.001
DAS28-CRP-3v	-0.028 (-0.037, -0.019)	<0.001		
MHAQ	-0.216 (-0.247, -0.184)	<0.001	-0.330 (-0.361, -0.298)	<0.001
Work productivity loss (overall work impairment / absenteeism plus presenteeism - %)	-0.003 (-0.003, -0.002)	<0.001	-0.002 (-0.003, -0.002)	<0.001
NSAID intake during the last 3 months	0.017 (-0.004, 0.039)	0.115	-0.046 (-0.067, -0.024)	<0.001
Current cDMARD	0.027 (-0.005, 0.059)	0.099	-0.006 (-0.028, 0.015)	0.568
Current bDMARD	-0.005 (-0.028, 0.018)	0.653	-0.038 (-0.059, -0.017)	<0.001

*Unstandardized coefficients. Legend: ACPA, anti-citrullinated protein antibody; ASDAS-CRP, Ankylosing Spondylitis Disease Activity Score; bDMARDs, biological disease-modifying antirheumatic drugs; BMI, body mass index; cDMARDs, conventional disease-modifying antirheumatic drugs; DAS28-CRP-3v, Disease Activity Score 28-CRP-3 variables; EQ-5D-3L, EuroQOL-5 Dimension 3-level; MHAQ, Modified Health Assessment Questionnaire; NSAID, Nonsteroidal anti-inflammatory drug; RA, rheumatoid arthritis; RF, rheumatoid factor.

activity both in RA and SpA patients.

Conclusions: Disability is a major contributor for QoL measured by EQ-5D in RA and SpA patients. Disease activity and work productivity loss also play important roles for QoL in these patients.

089 - CERTOLIZUMAB PEGOL EFFICACY IN PATIENTS WITH NON-RADIOGRAPHIC AXIAL SPONDYLOARTHRITIS STRATIFIED BY BASELINE MRI AND C-REACTIVE PROTEIN STATUS

Atulya Deodhar[1], Lianne Gensler[2], Stephen Hall[3], Philip C. Robinson[4], Ana Lourenço[5], Bengt Hoepken[6], Lars Bauer[6], Thomas Kumke[6], Walter P. Maksymowych[7]

[1]Oregon Health & Science University, Portland, OR, United States, [2]University of California San Francisco, San Francisco, CA, United States, [3]Cabrini Medical Centre, Monash University and Emeritus Research, Melbourne, Victoria, Australia, [4]University of Queensland School of Clinical Medicine, Brisbane, Queensland, Australia, [5]UCB Pharma, Paço de Arcos, Portugal, [6]UCB Pharma, Monheim am Rhein, Germany, [7]University of Alberta, Edmonton, Canada

Background: This post-hoc analysis from the phase 3 C-axSpAnd study aimed to evaluate whether the response to certolizumab pegol (CZP) in non-radiographic axial spondyloarthritis (nr-axSpA) is impacted by patients' baseline MRI and C-reactive protein (CRP) status.

Methods: C-axSpAnd (NCT02552212) is a 3-year, phase 3, multicenter study including a completed 52-week double-blind, placebo-controlled period.1 Patients were adults with a diagnosis of axSpA, meeting Assessment of SpondyloArthritis international Society (ASAS), but not modified New York, classification criteria, active disease (Bath Ankylosing Spondylitis Disease Activity Index [BASDAI] >4, spinal pain >4), objective signs of inflammation (C reactive protein [CRP] ≥10 mg/L [CRP+] and/or evidence of sacroiliitis on MRI [MRI+]), who had failed ≥2 non-steroidal anti-inflammatory drugs (NSAIDs). Patients were randomized 1:1 to placebo or CZP (400 mg at Weeks 0, 2 and 4, then 200 mg every 2 weeks), which they received in addition to non-biologic background medication for 52 weeks. Adjustments to background medication or switching to open-label CZP (or other biologics) at any point was permitted. We report Ankylosing Spondylitis Disease Activity Score – major improvement (ASDAS-MI) and ASAS 40% response (ASAS40) for CZP-randomized patients according to prespecified subgroups based on MRI/CRP status (MRI+/CRP+, MRI–/CRP+, MRI+/CRP–). Comparisons between MRI/CRP subgroups are descriptive only; Week 12 (ASAS40) and Week 52 (ASDAS-MI) comparisons between CZP and placebo were pre-specified. Missing values, or values collected after switching to open-label treatment, were imputed using non-responder imputation.

TABLE 1: DEMOGRAPHIC AND BASELINE CLINICAL CHARACTERISTICS.

	STS UGPL (n=8)	Saline solution UGPL (n=9)	p-value
Age (years), mean (SD)	49.0	48.4	NS
Sex, female % (n/N)	87.5% (7/8)	66.7% (6/9)	NS
Dominant side, right % (n/N)	100% (8/8)	100% (9/9)	NS
Affected side, right % (n/N)	62.5% (5/8)	66.7% (6/9)	NS
Education levels, % (n/N) Primary education Secondary education Higher education	 37.5% (3/8) 37.5% (3/8) 25.0% (2/8)	 33.3% (3/9) 44.4% (4/9) 22.3% (2/9)	NS
Employment Status, employed % (n/N)	50.0% (4/8)	66.9% (6/9)	NS
Number of comorbidities, median (IQR)	0 (2)	0 (2)	NS
Number of daily medications, median (IQR)	0 (2)	0 (2)	NS
Nocturnal pain, yes %(n/N)	100% (8/8)	100% (9/9)	NS
VAS at rest (0–10), mean (SD)	6.6 (1.3)	5.3 (2.4)	NS
VAS during activities (0–10), mean (SD)	7.8 (1.6)	5.2 (2.7)	0.033
DASH Score, mean (SD)	65 (11)	50 (15)	0.037
DASH-Work Score, mean (SD	77 (10)	59 (27)	NS
EQ5D, mean (SD)	0.2772 (0.4)	0.4765 (0.2)	NS
VAS EQ5D (0–100), mean (SD)	53 (19)	57 (25)	NS
UCLA score, mean (SD)	45 (13)	39 (17)	NS

DASH: Disabilities of the Arm, Shoulder and Hand; EQ5D: EuroQol five-dimensional; VAS: Visual Analogue Scale; UCLA: University of California at Los Angeles; UGPL: ultrasound guided percutaneous lavage; SD: Standard deviation

Results: At baseline, a total of 317 patients were randomized, 159 to CZP (45 MRI+/CRP+, 40 MRI–/CRP+ and 74 MRI+/CRP–) and 158 to placebo (42 MRI+/CRP+, 40 MRI–/CRP+ and 76 MRI+/CRP–). At Week 52, ASDAS-MI was achieved in 47.2% and 7.0% of CZP and placebo-treated patients, respectively, and ASAS40 in 56.6% and 15.8%. When stratified by MRI/CRP status, response rates in all three subgroups for CZP-treated patients were also higher compared to placebo for both ASDAS-MI and ASAS40 at Week 12 and Week 52 (Figure 1). For ASDAS-MI, there was a greater difference in response rates between subgroups compared with ASAS40, with numerically higher response rates in the MRI+/CRP+ and MRI–/CRP+ subgroups vs the MRI+/CRP– subgroup (Figure 1a). Since the ASDAS value is largely dependent on the CRP value, this is to be expected. For ASAS40, the main secondary outcome, a numerically higher response rate was also observed for the MRI+/CRP+ group, while response rates were comparable for the other groups (Figure 1b).

Conclusion: Clinically relevant responses were observed in nr-axSpA patients with either MRI and/or CRP positivity, with the highest response seen in the MRI+/CRP+ subgroup.

REFERENCES

1. Deodhar A. Arthritis Rheumatol 2019;71:1101–11.

Acknowledgements: This study was funded by UCB Pharma. Editorial services were provided by Costello Medical and funded by UCB Pharma.

089 - SAFETY AND TOLERANCE OF SODIUM THIOSULFATE IN CALCIFIC TENDINITIS OF THE ROTATOR CUFF

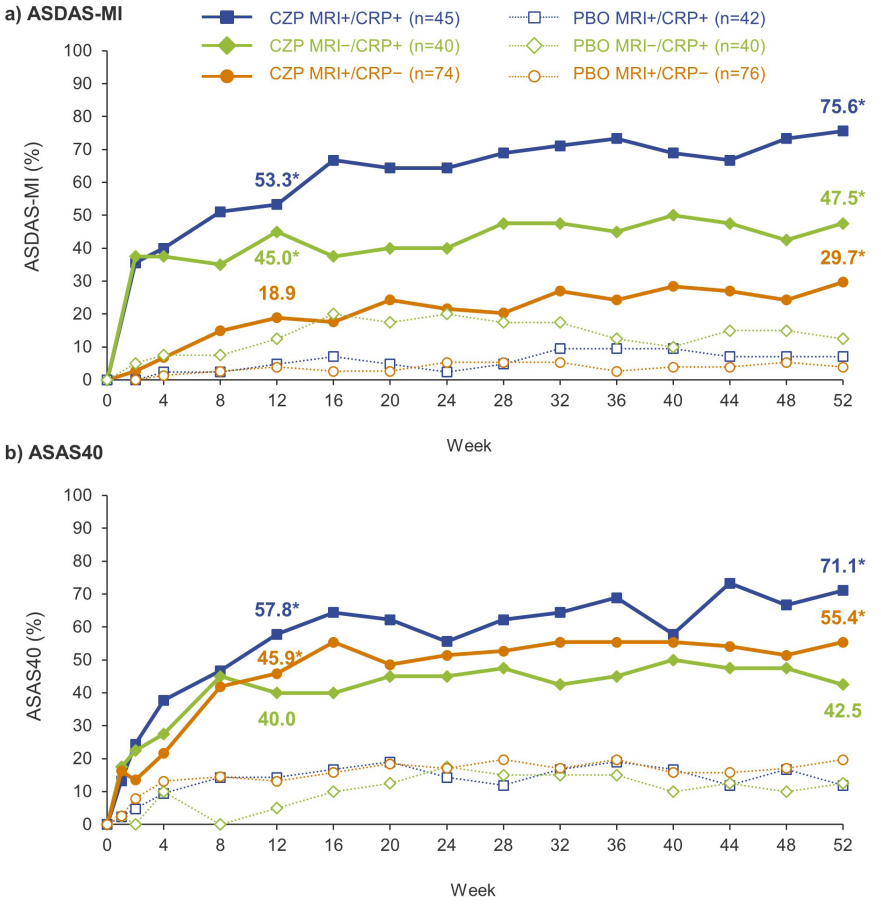
Soraia Azevedo[1], Hugo Parente[1], Emanuel Costa[2], Francisca Guimarães[1], Catarina Soares[1], Joana Ramos Rodrigues[1], Daniela Santos-Faria[1], Daniela Peixoto[1], José Tavares-Costa[1], Carmo Afonso[1], Filipa Teixeira[1]
[1]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [2]Rheumatology Department, Hospital de Braga, Braga, Portugal

Background: Calcific tendinitis of the rotator cuff is one of the most common causes of shoulder pain.[1] Ultrasound guided percutaneous lavage (UGPL) or barbotage or irrigation of calcific tendinopathy is indicated when conservative treatments (physiotherapy, nonsteroidal anti-inflammatory drugs) have failed.[2] Outcomes after UGPL are worse in case of dense calcifications.[3] Recent reports have shown the interest of topical sodium thiosulfate (STS) in the treatment of calciphylaxis, tumoral calcinosis associated with connective tissue diseases, pseudohypoparathyroidism or hyperphosphatemic familial tumoral calcinosis.[4,5,6]

Objectives: To assess the safety and tolerance of UGPL with STS versus with saline solution (standard of care), in calcific tendinitis.

Methods: We performed a double blinded randomized clinical trial, phase II study that included patients with calcific tendinitis and shoulder pain for more than 3 months. Only dense calcifications (type A calcification according to the Molé Classification) with more than 5 mm in diameter, on the standard anteroposterior

FIGURE 1. ASDAS-MI AND ASAS40 RESPONSE IN PATIENTS STRATIFIED BY BASELINE MRI/CRP STATUS



Missing values, or values collected after switching to open-label treatment, were imputed using non-responder imputation. *p<0.001 for CZP vs PBO. ASAS40: Assessment of SpondyloArthritis international Society 40% response; ASDAS-MI: Ankylosing Spondylitis Disease Activity Score – major improvement; CRP: C-reactive protein; CZP: certolizumab pegol; PBO: placebo.

radiographs, were included. Patients were randomized in two groups: STS lavage and saline solution lavage.

SPSS was used for statistical analysis and significance level was defined as 2-sided $p<0.05$.

Results: Seventeen patients were included, 76.5% (13) were women with a mean age of 48.7 (SD=8.8) years-old.

The median duration of pain before the procedure was 12 months (IQR=16), with a minimum of 3 months and a maximum of 48 months. The Yocum test for subacromial impingement were positive in 17 (100%) of patients. The patients reported that they had lost an average of 19 (SD=25) workdays due to the shoulder pain.

Nine patients (52.9%) were randomized to the control group and performed a saline solution UGPL. The other 8 patients (47.1%) were randomized for the treatment group (STS) and performed an UGPL with STS.

Demographic and baseline clinical characteristics and its comparison between STS and saline solution groups are shown in Table 1.

During the procedure, no adverse effects (allergy, pain, fainting or others) were recorded.

At the one-week evaluation, none of the patients reported pain aggravation and there were no new rotator cuff tear or bursitis recorded by ultrasound. There were also no cases of frozen shoulder.

Conclusions: In this preliminary analysis, STS UGPL was well tolerated with no side effects occurring during the procedure and at one-week follow-up. New studies using STS will be needed to conclude on the interest of this molecule in the treatment of calcific tendinitis.

REFERENCES

1. Louwerens JK, et al. Prevalence of calcific deposits within the rotator cuff tendons in adults with and without subacromial pain syndrome: clinical and radiologic analysis of 1219 patients. J Shoulder Elbow Surg.2015
2. de Witte PB, et al. Calcific tendinitis of the rotator cuff: a randomized controlled trial of ultrasound-guided needling and lavage versus subacromial corticosteroids. Am J Sports Med. 2013
3. Krasny C, et al. Ultrasound-guided needling combined with shock-wave therapy for the treatment of calcifying tendonitis of the shoulder. J Bone Joint Surg Br. 2005
4. Ossorio-García L, et al. Intralesional Sodium Thiosulfate to Treat Calciphylaxis. Actas Dermosifiliogr. 2016
5. Jost J, et al. Topical Sodium Thiosulfate: A Treatment for Calcifications in Hyperphosphatemic Familial Tumoral Calcinosis? J Clin Endocrinol Metab. 2016
6. Guigonis V, et al. Treatment of heterotopic ossifications secondary to pseudohypoparathyroid. Ann Endocrinol (Paris).2015

090 - ARE CONVENTIONAL OR BIOLOGIC DISEASE-MODIFYING ANTI-RHEUMATIC DRUGS AN EFFECTIVE TREATMENT FOR HAND OSTEOARTHRITIS?

Ana Martins[1], Sofia Pimenta[1, 2], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Background: Hand osteoarthritis (HOA) is a very common and complex disorder that cause pain and stiffness. This joint disease is very heterogenous and can result in functional limitation and impaired quality of life. There are no disease-modifying drugs approved but previous publications and anecdotal reports suggest some disease-modifying anti-rheumatic drugs (DMARD) as a possible treatment. These treatments are sometimes used off-label in some patients.

Objectives: To systematically identify and review available literature evaluating conventional or biologic DMARD treatment in hand osteoarthritis.

Methods: A systematic literature search was performed in MEDLINE. English language-only key word searches were used with combinations of terms including osteoarthritis, hand, anti-rheumatic drugs, biological therapeutic and disease-modifying anti-rheumatic drug.

All retrieved articles were screened by title and abstract and the eligible ones were kept for full-text review. Reference lists were additionally searched to identify additional potential trials. Only randomized controlled trials (RCTs) evaluating the efficacy of conventional or biologic DMARD intervention in patients aged ≥ 18 years old with HOA and without inflammatory arthritis are included in this review. RCTs evaluating other therapies, presented in duplicate and unpublished RCTs were excluded.

Results: The systematic search identified 39 RCTs since 2010. After title and abstract screening, 29 were excluded. One duplicated RCT was excluded. Nine RCTs were included in the systematic review for full analysis. All studies included are randomized, double-blind and placebo-controlled trials. Eight trials were parallel arm design trials, while one was a cross-over design trial and combined both treatment periods. The drugs studied were hydroxychloroquine (3 studies), methotrexate (1 study), etanercept (1 study), lutikizumab (1 study) and adalimumab (3 studies). In these studies, more than three-quarters were women

and the mean age of participants ranged from 50.2 to 67.5 years. Some authors chose to include only patients with erosive hand osteoarthritis while others included patients with and without erosive hand osteoarthritis. The primary endpoint in the majority of these studies was pain improvement (using VAS or AUSCAN for pain). Secondary endpoints included functional evaluation and structural damage (using x-rays, ultrasound or MRI). The majority of studies failed to reach the primary endpoint, showing that DMARD wasn't more effective than placebo in reducing pain. Three studies (one with methotrexate, one with etanercept and the other with adalimumab) showed a DMARD effect in subchondral bone, with a faster remodeling and less erosive progression in patients treated with DMARD. This effect was more pronounced in joints with inflammation at the baseline.

Conclusions: Neither conventional nor biological DMARDs showed efficacy in reducing pain or improving the functional scores in patients with hand osteoarthritis. Some studies showed that DMARD may have an effect on subchondral bone. They seem to promote a faster remodeling and a decrease in erosive progression in joints with soft tissue swelling or synovitis on the MRI or ultrasound. Specific patients might eventually benefit from DMARD use, especially those with aggressive erosive HOA. However, DMARD shouldn't be routinely used in all patients. A better understanding of pain mechanisms and the role of inflammation in HOA is required in order to identify and develop more effective treatments.

091 - DRUG-INDUCED LUPUS ERYTHEMATOSUS SECONDARY TO ANTI-TNF-α AGENTS IN PATIENTS WITH SPONDYLOARTHRITIS AND PSORIATIC ARTHRITIS

Ana Martins[1], Daniela Santos Oliveira[1], Frederico Rajão Martins[2], Filipe Oliveira Pinheiro[1], Maria Seabra Rato[1], Diogo Guimarães da Fonseca[3], Salomé Garcia[1], Bruno Miguel Fernandes[1], Sofia Pimenta[1, 4], Miguel Bernardes[1, 4], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [3]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/ Espinho, Vila Nova de Gaia, Portugal, [4]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Background: Induction of autoantibodies is frequently observed in patients (pts) treated with TNF-α antagonist and the possible development of drug-induced lupus erythematosus (DILE) remains a matter of concern. The prevalence of DILE secondary to anti-TNF-α therapy is 0.5-1% and clinical features include arthritis/ arthralgia, rash, serositis, fever, myalgias, cytopenias, among others. According to the literature, DILE due to anti-TNF-α agents differs in several ways from the clinical and laboratory findings typically associated with classic DILE.

Objectives: To estimate the incidence of induction of antinuclear antibodies (ANA) and DILE in a monocentric cohort of pts with spondyloarthritis (SpA) and psoriatic arthritis (PsA) treated with anti-TNF-α agents. To describe the clinical and laboratorial features and outcomes of pts with DILE.

Methods: We performed a retrospective analysis of pts with SpA and PsA treated with anti-TNF-α agents, from our University Hospital, between July 2001 and December 2020. Pts with positive ANA (titer>1/100) before the anti-TNF-α therapy were excluded. Because specific criteria for the diagnosis of DILE have not been established, we considered the diagnosis in case of a temporal relationship between clinical manifestations and anti-TNF-α treatment and fulfillment of ACR/EULAR 2019 classification criteria for SLE. In pts with DILE, clinical features, laboratory findings, systemic therapies and outcome after anti-TNF-α discontinuation were collected from reuma. pt and medical records. For clinical and demographic predictors, continuous and categorical variables were analyzed using a two-sided t-test and a fisher's exact test, respectively. P-value <0.05 was considered statistically significant.

Results: In SpA group, 290 pts were included (44.8%females, mean age at diagnosis of 33.3±11.5 years and mean disease duration of 15.1±10.4 years). In PsA group, 116 pts were included (50.0%females, mean age at diagnosis of 40.1±11.0 years and mean disease duration of 13.1±6.8 years). We observed high serology conversion rates (positive ANA in 67.9% and 58.6% of pts with SpA and PsA, respectively), with similar conversion rates between different anti-TNF drugs. Three pts with SpA (1.0%) and 1 patient with PsA (0.9%) developed DILE. Etanercept was the causative agent in 2 cases, infliximab and adalimumab in 1 case, each. Peripheral arthritis (new onset or abrupt worsening) occurred in 2 pts, serositis in 1 patient, constitutional symptoms in 2 pts, subnephrotic

proteinuria in 1 patient, lymphopenia in 2 pts and hypocomplementemia in 1 patient. Specific treatment was prescribed to the 4 pts (oral corticosteroids) and they achieved complete recovery. After anti–TNF-α treatment interruption, no patient had recurrent disease. We observed that pts with DILE had a significantly longer disease duration (p=0.04) and a significantly longer duration of anti-TNF therapy (p=0.04) than pts without DILE.

Conclusion: Despite frequent induction of autoantibodies, the development of DILE secondary to anti–TNF-α agents is rare. Our study demonstrates an incidence rate similar to other studies reported before. The clinical and laboratorial characteristics of our pts with DILE due to anti–TNF-α agents differ from DILE due to more traditional agents, as is described in literature. Overall, pts in this study had mild disease that improved after therapy discontinuation, without recurrence of the disease. It seems that a longer disease duration and a longer period under anti-TNF-α therapy may increase the risk of DILE development.

093 - IN PREVIOUSLY BIOLOGIC-NAÏVE RHEUMATIC PATIENTS WITH DRUG INDUCED LUPUS SECONDARY TO A FIRST ANTI-TNF THERAPY, IS IT SAFE TO SWITCH TO A SECOND ANTI-TNF-A AGENT?

Ana Martins[1], Daniela Santos Oliveira[1], Frederico Rajão Martins[2], Filipe Oliveira Pinheiro[1], Maria Seabra Rato[1], Diogo Guimarães da Fonseca[3], Salomé Garcia[1], Bruno Miguel Fernandes[1], Sofia Pimenta[1, 4], Miguel Bernardes[1, 4], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [3]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/ Espinho, Vila Nova de Gaia, Portugal, [4]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Background: Drug-induced lupus erythematosus (DILE) secondary to anti-TNF-α agents results from an immunogenicity phenomena not yet fully understood and is a rare condition. Withdrawal of anti-TNF- α therapy usually leads to total resolution of symptoms, however sometimes immunosuppression is needed. It is not clear if this condition is drug specific or class related. Therefore, there are doubts about the safety of switching to a second TNF inhibitor: will a further anti-TNF-α agent increase the risk of DILE recurrence?

Objectives: To analyze the outcomes in patients with DILE secondary to an anti-TNF-α agent that switch to a second anti-TNF-α agent.

Methods: We performed a retrospective analysis of patients with spondyloarthritis, psoriatic arthritis and rheumatoid arthritis from our University Hospital, who developed DILE secondary to an anti-TNF-α agent as a first biologic and switch to a second anti-TNF-α agent. Because specific criteria for the diagnosis of DILE have not been established, DILE diagnosis was considered when a temporal relationship between clinical manifestations and anti-TNF alpha treatment was found and ACR/EULAR 2019 classification criteria for SLE were fulfilled. Clinical and laboratorial features and outcomes were collected from the Portuguese Rheumatic Diseases Register (Reuma.pt) and medical records.

Results: Six of 617 patients developed DILE secondary to anti-TNF-α agents (2 secondary to etanercept, 2 to adalimumab and 2 to infliximab). These patients had total resolution of symptoms and autoantibodies (ANA and anti-DNAds), induced by the therapy, disappeared after withdrawal of the anti-TNF-α agent implied.

Afterwards, 4 of these 6 patients switched to a second anti-TNF-α agent: 1 to etanercept, 1 to certolizumab, 1 to adalimumab and another to golimumab. The time interval between the two therapies was 2,0 ± 0,8 months. Regarding the outcomes, in all four patients, no DILE recurrence or autoantibodies induction recurrence was observed. These patients have a good response to the new biotherapy, without side effects reported, and a significant clinical improvement was observed.

Conclusion: Our study results are in agreement with the literature described before. It seems that exist a low rate of DILE recurrence with an alternative anti-TNF-α agent. Thus, this condition seems to be drug specific rather than class related. Therefore, it seems secure to use a second anti-TNF-α agent, even in a short period of time after DILE development. There is no evidence about the best or securest second TNF inhibitor, so any anti-TNF-α agent can be prescribed. A carefully monitoring of symptoms of relapse should be ensured. In conclusion, DILE secondary to a TNF inhibitor should not be an absolute contraindication to the use of a subsequent anti-TNF-α agent.

094 - THE ROLE OF EDUCATION IN AXIAL SPONDYLOARTHRITIS

Luisa Brites[1], Rita Cunha[2], Helena Santos[3], Helena

Donato[4], Fernando Pimentel-Santos[5, 6]
[1]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [2]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [3]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal, [4]Documentation and Scientific Information Service, Centro Hospitalar da Universidade de Coimbra, Coimbra, Portugal, [5]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [6]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

Background: Spondyloarthritis is a heterogeneous group of diseases with impact on patients' daily activities. The current standard of care, along with pharmacological treatment, include regular exercise and patient education.[1] Patient education programs are useful to give patients the skills to manage their rheumatic disease, and could have a potential role in improving patients' related outcomes about disease activity, functional capacity and quality of life.[2]
Objective: This systematic review aimed to synthesize the role of education in medical care of patients with axial spondyloarthritis (axSpA).
Methods: We systematically searched three databases, PubMed, Embase and Web of Science Core Collection, from January 2000 to December 2020, using the following terms: "patient education", "patient counselling", "patient teaching", "patient engaging", "patient empowerment", "health education", "spondyloarthritis", "spondyloarthropaties", "spondylitis" and "ankylosing spondylitis". The "Population (P)", "Intervention (I)", "Comparator (C)", "Outcome (O)", PICO criteria were used. "P", defined as axial spondyloarthritis, "I" as education, "C" as standard of care or physical exercise and "O" as disease activity (the Bath Ankylosing Spondylitis Disease Activity Index (BASDAI), Ankylosing Spondylitis Disease Activity Score (ASDAS), functional status (the Bath Ankylosing Spondylitis Functional Index (BASFI) and Bath Ankylosing Spondylitis Metrological Index (BASMI)), and quality of life (Ankylosing Spondylitis Quality of Life (ASQoL), the EuroQol-5D (EQ-5D), the Short Form 36 Health Survey (SF36)). Only randomized clinical trials were included in the final analysis. Two reviewers independently assessed the identified papers according to the established criteria (Disagreements were solved by the introduction of a third reviewer) and extracted the data.
Results: From the initial 491 studies identified, 6 studies were selected for data extraction and qualitative analysis. The study sample size ranged between 41-65 individuals and all of them had a diagnosis of ankylosing spondylitis. The intervention period ranged between 4-12 weeks and the follow up ranged between 3-12 months. In three studies, the comparator was standard of care, and in the other three was physical exercise. Overall, there is an improvement in BASDAI, BASFI, BASMI, ASQoL and SF-36, after education program application. However, this improvement was statistically significant in only one study relative to BASFI and in another relative to ASQoL.
Conclusion: Education appears to be an important adjuvant as non-pharmacological treatment in patients with axSpA. Further research is still needed for more robust conclusions.

101 - CLINICAL AND IMMUNOLOGY FINDINGS IN PATIENTS WITH PRIMARY SJÖGREN'S SYNDROME – AN EXPERIENCE FROM A TERTIARY PORTUGUESE CENTER

Ana Martins[1], Raquel Miriam Ferreira[1], Daniela Santos Oliveira[1], Sofia Pimenta[1, 2], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Background: Primary Sjögren's Syndrome (pSS) is a chronic autoimmune inflammatory disease, characterized by lymphocyte infiltration in the exocrine gland, and commonly affects women in their 50s. Clinical presentation of pSS can vary considerably, ranging from classic sicca symptoms to systemic symptoms. Patients with pSS present a broad spectrum of serologic features.
Objectives: To describe the clinical features in a cohort of Portuguese patients with pSS and to investigate their association with the serological markers.
Materials and methods: We performed a retrospective analysis of patients with pSS who fulfilled the ACR/EULAR 2016 classification criteria followed in our Rheumatology department until August 2020. Demographic data, disease duration, clinical manifestations (using ESSDAI definitions and domains) and immunological profile were collected and analyzed. Immunological data and extraglandular manifestations were compared using chi-square test. P-value < 0.05

TABLE 1. CLINICAL MANIFESTATIONS IN PATIENTS WITH PSS (USING ESSDAI DEFINITIONS AND DOMAINS).

	No. (%)
Constitutional symptoms	7 (10,6)
Low activity	6 (9,1)
Moderate activity	1 (1,5)
Articular	28 (42,4)
Low activity	18 (27,3)
Moderate activity	9 (13,6)
High activity	1 (1,5)
Lymphoadenopathy	6 (9,1)
Low activity	2 (3,0)
High activity	4 (6,1)
Glandular	8 (12,1)
Low activity	7 (10,6)
Moderate activity	1 (1,5)
Cutaneous	3 (4,5)
Low activity	1 (1,5)
Moderate activity	1 (1,5)
High activity	1 (1,5)
Pulmonary	8 (12,1)
Low activity	2 (3,0)
Moderate activity	3 (4,5)
High activity	3 (4,5)
Renal	1 (1,5)
High activity	1 (1,5)
Muscular	1 (1,5)
Moderate activity	1 (1,5)
Peripheral nervous system	1 (1,5)
Low activity	1 (1,5)
Central nervous system	1 (1,5)
Moderate activity	1 (1,5)
Hematological	9 (13,6)
Low activity	6 (9,1)
Moderate activity	2 (3,0)
High activity	1 (1,5)
Biological	32 (48,5)
Low activity	20 (30,3)
Moderate activity	12 (18,2)

was considered statistically significant.

Results: In total, 66 pSS patients were included, 86,4% were females, with a mean age at onset of 51,47 (±14,9) years and median disease duration of 4 (IQR 2,75-6,25) years. Xerostomia and xerophthalmia were present in 57 (86,4%) and 54 patients (81,8%), respectively. Extraglandular involvement was present in 44 patients (66,7%), with articular manifestation being the most frequent observed, followed by hematological and pulmonary disease (table 1). Regarding the pulmonary histological pattern, 3 patients had usual interstitial pneumonitis, 2 had lymphocytic interstitial pneumonitis, 2 had nonspecific interstitial pneumonitis and 1 had pleuroparenchymal fibroelastosis. Renal involvement was present in 1 patient with focal mesangial proliferative glomerulonephritis. One patient had vasculitis and other had multiple sclerosis-like disease. As complications, lymphoma was documented in 4 patients, 3 non-Hodgkin lymphoma (mantle cell lymphoma, follicular lymphoma and small lymphocytic lymphoma) and 1 Hodgkin lymphoma.

Antinuclear, anti-SSA, anti-SSB antibodies and rheumatoid factor were positive in 58 (87,9%), 46 (69,7%), 24 (36,4%), 19 (28,8%) patients, respectively. Hypergammaglobulinemia was present in 32 patients (48,5%). The presence of anti-SSB antibody was more frequently observed in patients with extraglandular manifestations than in patients with sicca symptoms only (p=0.03). No other significant association between specific clinical features nor serological markers were found.

Conclusion: These results demonstrate once again the female predominance and the beginning of symptoms in the 50s in pSS patients. Articular and hematological were the most common extraglandular manifestations. In this study, anti-SSB antibody was significantly associated with extraglandular manifestations, suggesting that its presence may confer a higher risk of a more severe and systemic disease.

102 - REAL-WORLD EFFECTIVENESS OF SECUKINUMAB THERAPY IN ANKYLOSING SPONDYLITIS - DATA FROM THE RHEUMATIC DISEASES PORTUGUESE REGISTRY (REUMA.PT)

Helena Santos[1, 2], Soraia Azevedo[3], Laires PA[4], Salomé Garcia[5], Patrícia Nero[6], Jorge Pestana Lopes[7], Martins-Martinho J [8], Ana Bento da Silva[9], Joana Ramos Rodrigues[10], Graça Sequeira[11], Emanuel Costa[5], Filipe Cunha Santos[12], Carolina Furtado[13],

Bernardo Santos[14], Ana Raposo[15], Liliana Saraiva[16], Beatriz Samões[17], Filipe Barcelos[1], Maria José Santos[7, 18], José Tavares-Costa[3]

[1]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal, [2]EpiDoC Unit, CEDOC, NOVA Medical School, NOVA University, Lisboa, Portugal, [3]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [4]HE&OR - Novartis Portugal / ENSP – NOVA University of Lisbon, Lisboa, Portugal, [5]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal, [6]Rheumatology Department, Hospital CUF Descobertas, Lisboa, Portugal, [7]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [8]Rheumatology and Bone Metabolic Diseases Department, Centro Hospitalar Lisboa Norte, EPE - Hospital de Santa Maria, Lisbon Academic Medical Centre, Lisboa, Portugal, [9]Serviço de Reumatologia, Centro Hospitalar Lisboa Ocidental | Hospital Egas Moniz, Lisboa, Portugal, [10]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [11]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, [12]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, [13]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal, [14]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [15]Rheumatology Department, Centro Hospitalar Trás-os-Montes e Alto Douro, Vila Real, Portugal, [16]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [17]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [18]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Background/Purpose: Ankylosing Spondylitis (AS) affects 0.5% of the Portuguese adult population. Secukinumab has demonstrated efficacy across axial, peripheral and entheseal manifestations and has been shown to inhibit structural damage and improve physical function. Despite the growing number of patients treated with secukinumab, real-world data about its effectiveness is still scarce.

The purpose of this study is to investigate the effectiveness of secukinumab in different domains of AS patients using real world data.

Methods: PROSAS was a national, multicenter, observational, longitudinal cohort study, using data from the Rheumatic Diseases Portuguese Registry (Reuma.pt). Adult patients with the diagnosis of AS, according to their rheumatologist, registered in the Reuma.pt database, who started secukinumab therapy between 1st January 2017 and 10th January 2021, were included in the study. Sociodemographic characteristics and outcome measures as patient global assessment (PtGA), physician global assessment (PhGA), C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), Bath Ankylosing Spondylitis Disease Activity Index (BASDAI), Ankylosing Spondylitis Disease Activity Score-CRP (ASDAS-CRP), 68 tender joint count and 66 swollen joint count and Maastricht Ankylosing Spondylitis Enthesis Score (MASES), were collected at baseline and at 3, 6 and 12 months after secukinumab initiation.

Descriptive analysis of continuous variables was reported as mean and standard deviation (SD), or median and interquartile range (IQR) in case of non-normal distribution; categorical variables were displayed as frequency or proportions. For effectiveness analysis we performed paired sample comparisons using paired sample t-test were used or Wilcoxon signed-rank test (W), as appropriate.

Results: We included 168 AS patients, with a mean age of 46.9 (±11.3) years, 52.4% (88) were male, 77.0% (87) were HLA-B27 positive and 11.3% (19) had a history of uveitis. Approximately 27.4% (46) were biologic-naïve, 33.3% (56) were previously treated with 1 biologic, and 39.3% (66) with 2 or more biologics. Mean disease duration was 19.1 (±11.1) years, with patients' mean age when secukinumab was initiated of 45.1 (±11.3) years-old. Regarding co-medications, 30.4% (51) were on non-steroidal anti-inflammatories, 33.3% (56) were on conventional synthetic disease-modifying drugs and 20.2% (34) on oral steroids.

ASDAS Clinically Important Improvement was attained in 45.7% and 51.5% of the patients at 3 and 12 months, respectively, and ASDAS Major Improvement at the same timepoints in 10.5% and 25.0% of patients, respectively (table I). Proportions of patients achieving ASAS 20 response and BASDAI 50 responses were 45.7% / 54.9%, and 46.0% / 56.5% at 3 and 12 months, respectively. Enthesitis, evaluated by MASES, showed a significant improvement at 3, 6 and 12 months of follow-up. Significant improvements regarding PtGA, PhGA, CRP and BASFI were also observed at 3 months, and sustained until 12 months.

Conclusions: In this real-world cohort/study, we have

TABLE I – EFFECTIVENESS ASSESSMENTS WITH SECUKINUMAB THERAPY IN ANKYLOSING SPONDYLITIS PATIENTS

	Baseline	3 months	p-value*	6 months	p-value*	12 months	p-value*
PtGA, median (IQR) n available	70.0 (33.0) n= 137	50.0 (43.0) n=94	< 0.001	52.0 (45.0) n=90	< 0.001	33.0 (40.0) n=71	< 0.001
PhGA, median (IQR) n available	50.0 (34.0) n= 122	21 (30.0) n=81	< 0.001	20.0 (24.0) n=79	< 0.001	20.0 (22.0) n=61	< 0.001
CRP mg/L, median (IQR) n available	7.37 (17.5) n=132	5.3 (10.4) n=91	0.011	5.8 (9.4) n=89	0.03	4.6 (8.8) n=70	0.003
ESR, median (IQR) n available	21 (26.0)	17 (18.0) n=93	0.005	19.0 (29) n=89	NS	12.0 (20.0) n=69	< 0.001
BASDAI, median (IQR) n available	6.1 (2.7) n=139	4.2 (3.8) n=89	< 0.001	4.8 (3.8) n=89	< 0.001	3.5 (3.6) n=68	< 0.001
ΔBASDAI, median (IQR) n available		-1.7 (2.6) n=87		-1.2 (2.6) n=87		- 2.4 (3.2) n=64	
BASDAI 50, % (n)		46 (40)		39.1 (34)		56.5 (36)	
ASDAS-CRP, mean (SD) n available	3.5 (0.9) n=127	2.5 (1.3) n=82	< 0.001	2.9 (1.0) n=82	< 0.001	2.4 (1.0) n=64	0.003
ΔASDAS-CRP, mean (SD) n available		-0.94 (0.9) n=76		-0.70 (0.9) n=82		-1.17 (1.1) n=60	
ASDAS-CRP CII (Δ > 1.1), % (n)		43.4 (33)		27.0 (20)		51.7 (31)	
ASDAS-CRP MI (Δ > 2.0), % (n)		10.5 (8)		8.1 (6)		25.0 (15)	
ASAS 20, % (n) n available		45.7 (32) n=70		32.9 (24) n=73		54.9 (28) n=51	
ASAS 40, % (n) n available		27.1 (19) n=70		23.3 (17) n=73		33.3(17) n=51	
ASAS 70, % (n) n available		10.0 (7) n=70		9.6 (7) n=73		19.6 (10) n=51	
BASFI, median (IQR) n available	6.6 (2.9) n=110	4.5 (4.8) n=71	< 0.001	5.0 (4.52) n=73	< 0.001	3.3 (4.9) n=52	< 0.001
Tender joint count, median (IQR) n available	2.0 (5.0) n=131	0.0 (4.0) n=93	< 0.001	0.5 (4.0) n=88	< 0.001	1.6 (4.8) n=73	< 0.001
Swollen joint count, median (IQR)	0.0 (1.0) n=131	0.0 (0.0) n=90	0.002	0.0 (1.0) n=85	0.042	0.0 (1.0) n=71	0.003
MASES, median (IQR)	1.0 (4.0) n=89	0.0 (0.0) n=53	< 0.001	0.0 (3.0) n=61	< 0.001	0.0 (0.0) n=50	0.001

*p value - Comparisons across different timepoints and baseline; NS: non-significant

PtGA – patient global assessment; PhGA – physician global assessment; CRP- C-reactive protein; ESR- Erythrocyte sedimentation rate; BASDAI – Bath Ankylosing Spondylitis Disease Activity Index; ASDAS-CRP - Ankylosing Spondylitis Disease Activity Score; ASDAS CII – ASDAS Clinically Important Improvement; ASDAS MI – ASDAS Major Improvement; ASAS - Assessment in Ankylosing Spondylitis response criteria; BASFI – Bath Ankylosing Spondylitis Functional Activity Index; MASES - Maastricht Ankylosing Spondylitis Enthesis Score

confirmed that secukinumab treatment was effective in AS; the significant improvements in disease activity and function at 3 months, were sustained through 12 months.

Acknowledgement: This study was supported by Novartis and presented on Behalf of PROSAS Study group.

103 - IS MINOR SALIVARY GLAND HISTOLOGY A GOOD BIOMARKER SCORE TO PREDICT THE EXTRAGLANDULAR INVOLVEMENT IN PATIENTS WITH PRIMARY SJOGREN'S SYNDROME?

Ana Martins[1], Raquel Miriam Ferreira[1], Sofia Pimenta[1, 2], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Background: The minor salivary gland biopsy (MSGB) is considered the "gold standard" for the diagnosis of primary Sjögren's syndrome (pSS). The histological hallmark is focal lymphocytic infiltration with the presence of 50 or more lymphocytes per 4 mm2 (focus score ≥ 1). Several studies have documented the association between focus score (FS) and extraglandular involvement, suggesting that FS can be used as a histological index of disease severity and a prognostic tool.

Objectives: To investigate the association between the FS and clinical, haematological and serological manifestations in a cohort of patients with pSS.

Materials and methods: We performed a retrospective analysis of patients with pSS who fulfilled the ACR/EULAR 2016 classification criteria that were submitted to minor salivary gland biopsy (MSGB). Demographic data, disease duration, extraglandular involvement and histopathologic data were collected and analyzed. The severity of histologic changes was graded according to the Chisholm and Mason scoring system and the FS was determined. Fisher's exact test/chi-square test and Mann-Whitney U test were used to compare the patients with and without positive biopsy (FS <1 versus FS ≥ 1). Futhermore, to investigate the association between the quantitative value of focus score in groups with and without each extraglandular manifestations the authors used the Mann-Whitney U test. P-value < 0.05 was considered statistically significant.

Results: A total of 59 pSS patients were included. Regarding the histopathologic pattern, 40 of 59 patients had a FS ≥ 1, while 19 patients had a FS <1. Although described as having FS ≥ 1, the pathologists failed to report the absolute value of FS in 11 of 40 (27,5%) patients. Germinal center-like structures were found in 1 (1,7%) patient. The demographic and clinical features in pSS patients with FS <1 and with FS ≥ 1 are presented in table 1. Anti-SSA was strongly associated with FS <1 (p <0,001). No significant difference in demographic and clinical features were found between the two groups.

When comparing the quantitative value of focus score in groups with and without each extraglandular manifestation, the authors found that patients with articular involvement had a significant higher FS value than patients without articular manifestations (p=0.037). No other significant association was found.

Conclusion: Regarding clinical manifestations, we didn't find a significant difference between patients with FS < 1 and those with a FS ≥ 1. The presence of anti-SSA was strongly associated with a FS <1, which corroborates the importance of a positive anti-SSA for pSS diagnosis in patients with a negative biopsy. We found that a higher FS value was associated with articular involvement. However, this study has limitations such as a small sample and a low incidence of extraglandular manifestations. In conclusion, more studies are needed to clarify the associations found in the literature.

104 - PREDICTIVE FACTORS OF SECUKINUMAB PERSISTENCE IN PATIENTS WITH ANKYLOSING SPONDYLITIS - DATA FROM THE RHEUMATIC DISEASES PORTUGUESE REGISTRY (REUMA.PT)

Helena Santos[1, 2], Soraia Azevedo[3], Laires PA[4], Salomé Garcia[5], Patrícia Nero[6], Jorge Pestana Lopes[7], Martins-Martinho J [8], Ana Bento da Silva[9], Joana Ramos Rodrigues[10], Graça Sequeira[11], Emanuel Costa[5], Filipe Cunha Santos[12], Carolina Furtado[13], Bernardo Santos[14], Ana Raposo[15], Liliana Saraiva[16], Beatriz Samões[17], Filipe Barcelos[1], Maria José Santos[7,18], José Tavares-Costa[3]

[1]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal, [2]CEDOC, EpiDoC Unit, NOVA Medical School, Universidade NOVA de Lisboa, Lisboa, Portugal, [3]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [4]HE&OR - Novartis Portugal / ENSP – NOVA University of Lisbon, Lisboa,

Portugal, [5]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal, [6]Rheumatology Department, Hospital CUF Descobertas, Lisboa, Portugal, [7]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [8]Rheumatology and Metabolic Bone Diseases Department, Hospital de Santa Maria, Centro Hospitalar e Universitário Lisboa Norte, Lisboa, Portugal, [9]Serviço de Reumatologia, Centro Hospitalar Lisboa Ocidental | Hospital Egas Moniz, Lisboa, Portugal, [10]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [11]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, [12]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, [13]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal, [14]Rheumatology Department, Centro Hospitalar do Baixo Vouga, EPE, Aveiro, Aveiro, Portugal, [15]Rheumatology Department, Centro Hospitalar Trás-os-Montes e Alto Douro, Vila Real, Portugal, [16]Rheumatology Department, Centro Hospitalar Universitário de Coimbra, Coimbra, Portugal, [17]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [18]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Background/Purpose: Ankylosing Spondylitis (AS) affects 0.5% of the Portuguese adult population. Despite the growing number of patients treated with secukinumab, real-world data about its persistence and variables associated with treatment persistence are still sparse1.

The purpose of this study is to 1) determine secukinumab 1-year persistence, and 2) to identify predictors of secukinumab non-persistence, in AS patients in Portugal.

Methods: This study was a national, multicenter, prospective cohort study, using secondary data from the Rheumatic Diseases Portuguese Registry (Reuma.pt). Adult patients with the diagnosis of AS, according to their rheumatologist, registered in the Reuma.pt database between 1st January 2017 and 10th January 2021, that received secukinumab therapy were included. One-year persistence rate, according to treatment line of biologic therapy was estimated based on the time of secukinumab discontinuation curve (i.e. secukinumab survival curve) using the Kaplan-Meier (KM) method. Time to secukinumab discontinuation was defined as the time interval between first secukinumab dose and the decision of secukinumab discontinuation clearly identified in the registry (due to any reason).

After univariate analysis, Cox regression analyses with backward selection were performed to identify predictors of non-persistence in the first year of follow-up, with variables described in literature as potentially influencing treatment persistence, and with those found to have significant association in univariate analyses.

Results: A total of 168 patients (52.4% men) with a mean age of 45.1 (SD=11.3) years-old at first injection of secukinumab were analyzed. One-year persistence of Secukinumab was 81.5%, with no differences between the naïve and non-naïve (82.6% and 81.1%; p=0.826). In univariate analysis, an association with secukinumab persistence at 1-year follow-up was found with the number of switches (more switches of biologic therapy were associated with less persistence), concomitant use of classic disease modifying anti-rheumatic drugs (csDMARDs) (csDMARDs were associated with more persistence), age of onset of 1st biologic therapy and of secukinumab (older patients had lower rates of persistence). A better physical component score (PCS) on 36-Item Short Form Survey (SF-36 PCS) at baseline was associated with a higher persistence rate. No other associations were found. We adjusted our model to these variables and to disease duration, baseline disease activity (ASDAS-CRP), line of secukinumab treatment and number of comorbidities. In our final model, only age at onset of 1st biologic therapy (p=0.009) and the number of switches (p=0.008) remained with significant association with non-persistence (table 1).

TABLE 1. MULTIVARIATE ANALYSES: COX REGRESSION FOR PREDICTIVE FACTORS OF NON-PERSISTENCE IN ANKYLOSING SPONDYLITIS.

Determinants	Unstandardized Coefficients B	Standardized Coefficients Beta	95.0% CI	p-value
Age at onset of 1st biologic therapy	0.115	1.122	1.029-1.223	0.009
Number of switches	0.926	2.521	1.279-4.971	0.008
Secukinumab treatment line	2.227	9.276	0.971-8.583	NS
Gender	0.727	2.068	0.450-9.496	NS
Baseline ASDAS-CRP	0.705	2.024	0.587-6.862	NS
SF-36 PCS	-0.037	0.964	0.9021-030	NS
Number of comorbidities	-1.056	0.348	0.079-1.540	NS

CI: Confidence Interval; NS: non-significant; SF-36 PCS: 36-Item Short Form Survey physical component – core; DMARDs: disease modifying anti-rheumatic drugs

Conclusion: This real-world cohort confirms that, in secukinumab treatment, an older initiation of biologic and previous switches are important predictors of non-persistence and highlights the importance of early secukinumab initiation in treatment persistence, in AS patients.

Acknowledgement: This study was supported by Novartis and presented on Behalf of PROSAS Study group.

REFERENCES

1. Michelsen B, Lindström U, Codreanu C, et al. Drug retention, inactive disease and response rates in 1860 patients with axial spondyloarthritis initiating secukinumab treatment: routine care data from 13 registries in the EuroSpA collaboration. RMD open 2020;6(3):1–12.

107 - ARE THERE GENDER DIFFERENCES IN AXIAL SPONDYLOARTHRITIS: DATA FROM A PORTUGUESE SPONDYLOARTHRITIS COHORT

Rita Cunha[1], Assistant Professor[2], Elsa Vieira-Sousa[3, 4], Nikita Khmelinskii[3, 4], Pedro Ávila Ribeiro[3, 4], Couto M[5], Maria Inês Seixas[5], Nádia Martins[5], Miguel Bernardes[6, 7], Ana Martins[6, 7], Ana Bento da Silva[8, 9], Maria Helena Lourenço[8, 9], Cláudia Miguel[10], MD[11], Paula Valente[12], José Tavares-Costa[13], João Rovisco[14, 15], Renata Aguiar[1], Anabela Barcelos[1, 16, 17]

[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, EPE, Aveiro, Aveiro, Portugal, [2]Department of Mathematics and CIDMA - Center for Research & Development in Mathematics and Applications, University of Aveiro, Aveiro, Portugal, [3]Rheumatology Department, Centro Hospitalar Universitário de Lisboa Norte, Lisboa, Portugal, [4]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [5]Rheumatology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [6]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [7]Faculdade de Medicina da Universidade do Porto, Departamento de Medicina e CINTESIS, Porto, Portugal, [8]Rheumatology Department, CHLO - Hospital de Egas Moniz, Lisboa, Portugal, [9]Comprehensive Health Research Center – CHRC, Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, Lisboa, Portugal, [10]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal, [11]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [12]Rheumatology Department, Centro Hospitalar Entre Douro e Vouga, Aveiro, Portugal, [13]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [14]Rheumatology Department, Centro Hospitalar Universitário de Coimbra, Coimbra, Portugal, [15]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal, [16]Public Health Research Centre, NOVA National School of Public Health, Public Health Research Centre, Universidade NOVA de Lisboa, Portugal, Lisboa, Portugal, [17]Comprehensive Health Research Center (CHRC), NOVA Medical School, University of Lisbon, Lisboa, Portugal

Background: Historically, axial spondyloarthritis (particularly ankylosing spondylitis) was considered a men's disease and has been under-recognized in women. Emerging evidence reveals gender differences in pathophysiology, disease presentation and therapeutic efficacy.

Objective: To determine if there are differences between genders in a Portuguese cohort of patients with axSpA as regards clinical manifestations, disease activity, functional capacity, patient related outcomes and radiographic findings.

Methods: Patients with ≥18 years fulfilling the ASAS-Assessment of Spondyloarthritis International Society classification criteria for axSpA and registered in the electronic national database- Reuma.pt were included in a multicentric cross-sectional study. Sociodemographic data, clinical features and radiographic findings were collected from the first record in Reuma.pt. These variables were compared between genders using Mann-Whitney test and Chi-Square test. Variables with a significant association with group variable (gender) were considered in the multiple variable analysis to adjust the gender effect on the outcome variables.

Results: A total of 1995 patients were included, 1114 (55.9%) men and 881 (44.1%) women. Men had a lower median age at disease onset (25.1 vs 28.4, p=0.000) and median age at diagnosis (26.9 vs 30.4, p=0.000) and were more frequently smokers (32.1% vs 15.7%, p=0.000). Comparing to women, men had worse BASMI scores (4.0 vs 3.4, p=0.000), higher levels of CPR (10.5 vs 6.9, p<0.000) and were more often HLA-B27 positive (67.8% vs 54%, p=0.000). In univariable analysis, sacroiliitis on radiograph or/and MRI (95.5% vs 91.7%, p=0.04) was more common in men, however that wasn't confirmed in multivariable analysis.

In contrast, women more frequently had inflammatory bowel disease (8.8% vs 4.9%, p=0.004),

higher levels of ESR (25.0 vs 21.0, p=0.003) and worse PROs- BASDAI (5.7 vs 4.5, p=0.000), PGA (60.0 vs 55.0, p=0.000) and fatigue (6.2 vs 5.4, p=0.000).

Conclusion: Physicians must be aware of differences between genders in axial spondyloarthritis because this could result in less underdiagnosis and misdiagnosis, allow optimization of treatment strategies, and decrease overall disease burden in women with axSpA patients.

REFERENCES

Rusman T, van Vollenhoven RF, van der Horst-Bruinsma IE. Gender Differences in Axial Spondyloarthritis: Women Are Not So Lucky. Curr Rheumatol Rep. 2018;20(6):35.

115 - ENVOLVIMENTO RENAL NA SÍNDROME DE SJOGREN PRIMÁRIA: UMA SÉRIE DE CASOS DE UM SERVIÇO DE REUMATOLOGIA

Adriana Carones[1], Beatriz Mendes[1], Ana Rita Prata[1], Liliana Saraiva[1], Luisa Brites[1], Cátia Duarte[1, 2]

[1]Serviço de Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [2]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal

Introdução: A síndrome de Sjogren primária (SSp) é uma doença crónica auto-imune caracterizada por infiltração linfocitária do tecido epitelial das glândulas exócrinas, bem como locais extraglandulares. A prevalência do envolvimento renal na SSp varia entre 1-14%, sendo mais comum a nefrite túbulo-intersticial e mais rara a glomerulonefrite (1).

O objetivo deste trabalho foi caracterizar o envolvimento renal por glomerulonefrite de uma população de doentes com SSp.

Métodos: Estudo transversal incluindo doentes com o diagnóstico de SSp, cumprindo critérios de classificação ACR/EULAR 2016, com seguimento regular no serviço de Reumatologia do Centro Hospitalar e Universitário de Coimbra.

Procedeu-se à recolha de dados demográficos, clínicos e laboratoriais através da consulta dos processos clínicos e foi realizada uma análise descritiva.

Resultados: Foram incluídos 141 doentes com diagnóstico de SSp, dos quais 4 tinham envolvimento renal. Deste subgrupo, 3 apresentavam diagnóstico histológico de glomerulonefrite membranoproliferativa. Aquando do envolvimento renal a idade média dos doentes e o tempo de evolução da doença era respetivamente de 47,7+/-11,7 anos e 19,0+/-11,0 anos. Dois dos casos identificados apresentavam simultaneamente achados histológicos de nefrite túbulo-intersticial.

Clinicamente verificou-se perfil tensional hipertensivo em dois doentes e edemas periféricos em um doente.

Analiticamente todos os doentes apresentavam positividade para anticorpos antinucleares (ANA), anti-SSA60 e fator reumatóide, sendo que dois doentes apresentavam ainda anti-SSB positivo. A lesão renal aguda e proteinúria foram ambas documentadas em 100% dos doentes. O valor médio de creatinina sérica foi de 2,7+/- 1,5 mg/dl (variação de 1,6+/-1,4 mg/dl face ao valor basal) e o da proteinúria em amostra de 24 horas foi de 2632,6+/-1044,2 mg/dia. Todos os doentes apresentaram hipocomplementemia.

Relativamente à terapêutica, um doente realizou pulsos de metilprednisolona (1000mg/dia, durante 3 dias) e dois doentes prednisolona 1mg/kg/dia. Todos os doentes iniciaram imunossupressão: dois doentes com azatioprina (50mg/dia) e um doente com micofenolato mofetil (1500mg/dia).

Houve recuperação completa da função renal em dois doentes e todos os doentes apresentaram melhoria significativa da proteinúria (< 500mg/dia).

Conclusões: Na SSp pode ocorrer envolvimento renal, o qual se manifesta com maior frequência sob a forma de nefrite túbulo-intersticial. A doença glomerular, apesar de menos comum, acarreta maior morbilidade e implicações prognósticas.

Dado ser frequentemente assintomático é fundamental a monitorização regular da função renal e sedimento urinário em doentes com SSp para diagnóstico e tratamento precoce de modo a melhorar o prognóstico destes doentes.

REFERÊNCIAS

1. François H, Mariette X. Renal involvement in primary Sjögren syndrome. Nature Reviews Nephrology. 2016;12(2):82-93.

120 - MATERNAL AND FETAL OUTCOMES IN PREGNANT WOMEN WITH JUVENILE IDIOPATHIC ARTHRITIS: A SYSTEMATIC LITERATURE REVIEW

Rita Pinheiro Torres[1,2*], Maria Helena Lourenço[1,2*], Agna Neto[4,1,3], Fernando Pimentel-Santos[1,2,3], Inês Silva[1,3], Ana Filipa Mourão[1,2,3], Helena Donato[5], Manuela Costa[1], Jaime C. Branco[3,2,1]

1. Rheumatology Department, Hospital de Egas Moniz, CHLO, Lisboa, Portugal

2. CEDOC, Centro Estudo de Doenças Crónicas, NOVA Medical School| Faculdade de Ciências Médicas, Universidade NOVA de Lisboa (NMS/FCM – UNL) Lisboa, Portugal

3. NOVA Medical School, Faculdade Ciência Médicas, Lisboa, Portugal
4. Rheumatology Department, Hospital Central do Funchal, Madeira, Portugal
5. Serviço de Documentação e Informação Científica, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

Purpose: Juvenile idiopathic arthritis (JIA), one of the most common chronic diseases in children, can be classified in seven different categories according to its onset presentation. Recent data showed an increased risk of preterm birth in women with JIA, despite the small patient samples analysed. Our aim is to describe the current available knowledge on JIA adverse, maternal and fetal, outcomes.
Methods: A systematic literature review was conducted since January 2000 until December 2020, by searching the PubMed and Embase bibliographic databases, using "oligo", "polyarticular RF-negative", "enthesitis-related", "systemic", "psoriatic", "undifferentiated" and "idiopathic juvenile arthritis", "pregnancy or obstetrics or fetal or maternal or reproductive outcome or complications" as mesh terms. The search was limited to articles in English language, presenting a comparator group (healthy individuals or patients without known auto-immune rheumatic diseases) and at least one clinical outcome of interest. Two independent reviewers screened the titles and abstracts followed by a full-text review to assess papers regarding their eligibility.
Results: Ten observational studies out of 1560 references, fulfilled the inclusion criteria, of which, 9 were retrospective and 1 prospective (table 1). A total of 6.214 JIA patients (6.811 pregnancies) and 18.659.513 healthy controls (21.339.194 pregnancies) were included.

Concerning maternal outcomes, delivery by caesarian section (CS) was more frequent among JIA women (4/6 studies). Pre-eclampsia was referred in 3 out of 6 studies and a higher risk of vaginal bleeding and placenta previa in one additional study. No study found an increased risk, for gestational diabetes or hypertension, in pregnant women with JIA.

Regarding fetal outcomes, 8 studies revealed significantly increased of preterm birth (only in first births in one study) but one study didn't show any increased risk. Two studies showed a higher risk of small gestational age (SGA) and in another 2, increased risk for low birth weight (LBW). No evidence demonstrated increased risk of major congenital malformations.
Conclusion: This systematic review suggests an increased risk for pre-eclampsia, preterm birth, delivery by caesarian section, SGA and LBW, among patients with JIA. Conclusions should be carefully interpreted, giving the heterogeneity of studied populations regarding demography, disease type, disease activity, and medication.

121 - METHOTREXATE - IMPLICATIONS OF PHARMACOGENETICS IN THE TREATMENT OF PATIENTS WITH RHEUMATOID ARTHRITIS

Rita Pinheiro Torres [1, 2], Fernando Pimentel-Santos[2, 3, 4], Jaime C. Branco[2, 3, 4]

[1]Serviço de Reumatologia, Centro Hospitalar Lisboa Ocidental | Hospital Egas Moniz, Lisboa, Portugal, [2]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [3]Rheumatology Department, Centro Hospitalar Lisboa Ocidental, Lisboa, Portugal, [4]NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

Background: Methotrexate (MTX) is an anti-folate drug with anti-proliferative and anti-inflammatory effects. MTX proved to be the most highly effective, fast-acting disease modifying anti-rheumatic drug (DMARD), being widely used for the treatment of rheumatoid arthritis (RA) (Song et al., 2014).
Objectives: This review aims to describe the main genetic variants identified concerning proteins that play a role in methotrexate's kinetics and efficiency profile.
Methods: A literature review was conducted since January of 2000 until December 2020, by searching the PubMed and Embase bibliographic databases, employing the following MeSH terms: methotrexate, pharmacogenetics, pharmacokinetics and rheumatoid arthritis. The search was limited to articles in English language. Two independent reviewers screened the titles and abstracts followed by a full-text review to assess papers regarding their eligibility. A total of 48 articles matched the research criteria and were analysed.
Results: Genetic variants of four main proteins, with different functions, have been consistently described. Reduced folate carrier 1 (RFC1), a constitutively expressed folate transport protein that has high affinity for MTX is responsible, almost exclusively, for the transport of folate and MTX into the cell. The most commonly studied variant of the gene is the 80G > A variant (rs1051266), mapped within exon 2, on chromosome 21. It seems to improve RA responses

to MTX, clinical efficacy with long disease remission (Hayashi et al., 2013). ABC transporters are involved in the eflux of MTX from cells. An increased expression and function of these transporters should decrease MTX concentrations in target cells, resulting in lack of therapeutic response. ABCB1 3435 C/T (rs1045642) is a high frequency polymorphism, significantly associated with RA good responses, symptom remission and reduced adverse events, due to MTX treatment (Zhu et al., 2014).Thymidylate synthase (TYMS) is involved in thymidine synthesis. MTX decreases TYMS activity by inhibition and decreasing the access to tetrahydrofolate (THF) cofactors (1). The most common genetic variant of the TYMS gene consists of a 28 bp tandem repeat (rs34743033), with double and triple number of repeats (2R and 3R). The 3R allele genotype was associated with decreased efficacy and increased toxicity (4).The 5,10-methylenetetrahydrofolate reductase (MTHFR) enzyme is indirectly inhibited by MTX. The most common SNPs of the MTHFR gene are C677T (rs1801133) and A1298C (rs1801131). Both are associated with a decreased efficacy and an increased toxicity of MTX (Hughes et al. 2006).

Conclusion: MTX response is affected by many gene variants; the effect of each variant separately is likely to be small. Additionally, gene-gene interaction, enhancing the potential role of linkage disequilibrium. This shows the emerging need for a better gene characterization and to improve the knowledge about variants distribution according to ethnicity, to explain different responses to MTX at an individual level.

128 - CEERI: CENTRO DE ENSAIOS E ESTUDOS EM REUMATOLOGIA - INSTITUTO PORTUGUÊS DE REUMATOLOGIA: PATIENT INCLUSION PERFORMANCE IN RANDOMIZED CONTROLLED TRIALS IN A RHEUMATOLOGY TERCIARY CENTER

Luis Cunha-Miranda[1], Filipe Barcelos[1], Manuela Micaelo[1], José Vaz Patto[1], Manuela Parente[1], Ana Sdofia Santos[1], António Gomes[1], Tânia Borralho[1], Helena Santos[1]

[1]Instituto Português de Reumatologia, Lisboa, Portugal

Introduction: Randomized controlled trials (RCTs) are considered the gold standard for evaluating the efficacy of interventions, and can be very important in clinical setting. Early involvement in drug research is an important activity in Rheumatology practice, allowing the acquisition of knowledge about future drugs, their efficacy and safety, the improvement of clinical skills and practice in the use of new assessment tools..

Centro de Ensaios e Estudos em Reumatologia - Instituto Português de Reumatologia (CEERI) is the trial clinical centre of our rheumatology institution that includes rheumatologists, rheumatology residents, pharmacist, nurses and a clinical trial manager. Our goal was to review our performance as a RCT clinical centre, and improve strategies to obtain the maximum recruitment performance possible

Objective: The purpose of this study is: 1) To review the performance of our center in RCTs, regarding proposed objectives versus randomized patients (including extensions and phase IV studies) 2) To analyse the reasons for underperformance

Methods: We reviewed the number of approved RCTs in our centre taking into consideration the proposed number of patients and the number of randomized patients in those trials. In the underperformed RCTs, local principal investigators accessed the reasons for underperformance.

Results: 173 patients were involved in RCTs, distributed according to table I.

Good or excellent performance was achieved in 65.7 % of the studies. In the 12 underperformed studies, in 4 the recruitment target was completed over 50%. And in 3 were completed less than 50%. In 5 studies we were unable to include patients, 3 due to short recruitment period (CEIC late approval or competitive studies), 1 was in a rare disease and the other had a high number of screen failures due to strict inclusion criteria.

TABLE 1: NUMBER OF STUDIES PER DISEASE

Disease	Nº of RCT	N patients proposed vs screened	N/% of screen failure	Over performed	Achieved proposed number of patients	Under performed
Rheumatoid Arthritis	12	44/61 139%	11/ 15.3%	5	4	3
AS, non radiographic	7	22/31 140%	18/ 36.7%	3	2	2
Psoriasic arthrithis	7	17/22 130%	2/ 8.3%	3	2	2
SLE	4	14/11 78.5%	7/ 38.8%	1	1	2
Sjögren S.	3	6/7 116%	1/ 12.5%	1	1	1
Fibromyalgia	1	5/4 80%	1/ 20%	0	0	1
Giant Cell Arthritis	1	2/0 0%	0	0	0	1
Total	35	108/134 124%	39/ 22,5%	13	10	12

RCT – randomize control trials; AS – ankylosing Spondylitis; SLE Systemic Lupus erythematosus - ; Sjogren S – Sjögren Syndrome

Excluding the 3 studies that were underperformed due to external reasons (not imputable to the center), overall we attained a very good performance in 71.8 % of the studies.

Discussion: A clinical trial centre should look for its strengths and weaknesses. Proposed objectives for clinical trials should be realistic, considering disease specificities, and the center must have an active recruitment strategy. In the past decade, we optimise our clinical resources in order to achieve an excellent performance but strategies in order to reduce underperformance are underway.

131 - PREDICTIVE FACTORS OF FRAGILITY FRACTURES: ASSESSMENT OF PATIENTS OBSERVED AT EMERGENCY DEPARTMENT FOR FRACTURES

Catarina Soares[1], Soraia Azevedo[1], Francisca Guimarães[1], Hugo Parente[1], Emanuel Costa[2], Joana Ramos Rodrigues[1], Daniela Peixoto[1], José Tavares-Costa[1], Filipa Teixeira[1], Carmo Afonso[1], Daniela Santos-Faria[1]

[1]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [2]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

Background: Fractures that result of a fall from a standing height or less are known as fragility fractures (FF) and represent the main clinical consequence of osteoporosis. These fractures are one of the main causes of morbidity and impairment in the elderly. There are some well-recognizes risk factors for FF, including lifestyle factors such as smoking and alcohol intake, systemic glucocorticoids, previous FF and comorbidities such as rheumatoid arthritis.

Objectives: To assess the predictive factors for FF.

Methods: Retrospective monocentric study that included patients with FF (wrist, hip and vertebrae), observed at the emergency department (ED) in a tertiary center between 1st January 2017 and 31st December 2018. The search for fractures was performed through ICD9 codes and clinical data was collected. We excluded totally dependent patients or in palliative care, peri-prosthetic fractures and patients with osteometabolic diseases other than osteoporosis. We identified 1673 FF and after calculating a representative sample (90% confidence interval) 457 FF were included. To identify predictors of FF we performed a multivariate analysis including variables with a significant association in univariate analysis and those with clinical relevance.

TABLE 1: MULTIVARIATE ANALYSES: LINEAR MULTIPLE REGRESSION FOR PREDICTIVE FACTORS OF NEW FRAGILITY FRACTURE

	Unstandardized Coefficients B	Standardized Coefficients Beta	95.0% CI	p-value
Age	0.014	1.014	0.978-1.052	NS
Number of comorbidities	0.114	1.121	0.893-1.408	NS
Visit to the emergency department due to falls	1.485	4.415	1.761-11.069	0.002
Pulmonar disease	0.731	2.077	0.937-4.607	NS
Hematologic disease	0.647	1.909	0.649-5.621	NS
Malignancy	1.038	2.823	1.132-7.041	0.026
Previous diagnosis of osteoporosis in a BMD	0.8	2.225	0.940-5.266	NS

SPSS was used for statistical analysis and significance level was defined as 2-sided $p<0.05$.

Results: 172 patients with hip fracture, 173 with wrist fracture and 112 with vertebral fracture were included. Most patients were women (79.9%) with a mean age of 77.6 (SD=10.3) years-old at the time of the fracture. Sixteen percent of patients had a previous bone mineral density (BMD) test and 7.4% were on antiosteoporotic drugs.

We found an association between the occurrence of a FF and previous visits to the ED due to falls ($p<0.001$), number of comorbidities ($p=0.006$), previous diagnosis of chronic pulmonary disease ($p=0.002$), hematologic disease ($p=0.005$) or malignancy ($p=0.024$) and previous diagnosis of osteoporosis based on BMD test ($p=0.036$).

No associations were found between the number or type of medication taken daily, previous fractures and their localization nor the presence of other specific comorbidities other than those mentioned above.

Multivariate analysis showed that previous visits to the ED due to falls ($p=0.002$) and malignancy ($p=0.026$) remained associated with the occurrence of new FF after the adjustment for comorbidities, smoking, alcoholism and corticosteroid therapy.

Conclusions: Our study showed that previous visits to the ED due to falls and diagnosis of malignancy are independent predictors of FF. In the presence of these comorbidities screening for osteoporosis must be considered and the need for treatment should be assessed.

134 - MUSCLE BIOPSY: MASTER ROLE IN DIFFERENTIAL DIAGNOSIS IN PATIENTS WITH SUSPECTED MYOPATHY

Catarina Soares[1], Soraia Azevedo[1], Francisca

Guimarães[1], Hugo Parente[1], Joana Ramos Rodrigues[1], Filipa Teixeira[1], Daniela Peixoto[1], José Tavares-Costa[1], Carmo Afonso[1], Daniela Santos-Faria[1]
[1]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal

Background: Muscle biopsies are important diagnostic procedures in neuromuscular practice and considered the gold standard for the differential diagnosis of myopathies. In association with clinical and laboratory findings, the muscle biopsy has an important role to a more accurate diagnosis.
Objective: To evaluate the usefulness and safety of muscle biopsies performed in a Rheumatology Unit in patients with suspected myopathy.
Methods: Retrospective analysis of the clinical records of patients submitted to muscle biopsy between January 2010 and January 2021 at our Rheumatology Unit. Demographic, clinical, laboratory, electromyographic and histological data were collected. The histological study was performed in a Neuropathology Specialized Unit.
Results: A total of 55 patients, 25 men and 30 women, with a mean age of 49.9 ±16.7 years, were included. The most common symptoms were myalgia and decreased muscle strength. Other clinical manifestations included asthenia and skin lesions. Most patients (86.4%) also had increased muscle enzymes, particularly creatine kinase. Muscle biopsies were performed mainly in the deltoid muscle. There were no relevant immediate or late complications with this technique.

Of the 55 biopsies, 18 (32.7%) did not show alterations, 9 (16.4%) showed nonspecific alterations and only 1 biopsy was not conclusive because the sample was inadequate. In 2 patients, the histological features did not present specific characteristics of a myopathy but revealed a preferential atrophy of type 2 fibers. Twelve (21.8%) were compatible with inflammatory myopathies, namely polymyositis (9), dermatomyositis (1), inclusion body myopathy (1), and localized nodular myositis (1). In the latter case, the patient presented with atypical symptoms, namely with intermittent episodes of pain, oedema and flushing of different muscle groups. In addition, the muscle biopsy allowed the diagnose of 7 (12.7%) metabolic myopathies (2 McArdle´s diseases and 5 non-specific metabolic disorders), 3 (5.5%) muscular dystrophies (1 Becker´s muscular dystrophy and 2 dystrophinopathy), 1 suspected case of myotonic dystrophy and 1 myopathy associated with statins use were diagnosed. In a patient with an overall decrease in muscle enzymes, the biopsy revealed neurogenic atrophy, without inflammatory infiltrates. Among the 12 patients with inflammatory myopathy, in 9 an electromyography (EMG) was performed, 7 of which presented myopathic findings and 2 had a normal exam. In the other hand, of the 5 patients with muscle biopsy consistent with metabolic myopathy, only 1 had an abnormal EMG.
Conclusion: Although muscle biopsy is an invasive technique, it is safe and useful for the differential diagnosis between the different myopathies, which is fundamental to an appropriate treatment. In this retrospective analysis, it does not seem to exist a relation between the EMG results and the histological findings, and the muscle biopsy was crucial in the final diagnosis in patients with a normal EMG, which is particularly true in the metabolic myopathies.

135 - ANTIRESORPTIVE THERAPY AFTER TERIPARATIDE DISCONTINUATION – WHEN IS THE BEST TIME TO STARTING IT?

Maria Seabra Rato[1], Filipe Oliveira Pinheiro[1], Salomé Garcia[1], Bruno Miguel Fernandes[1], Diogo Guimarães da Fonseca[2], Ana Martins[1], Daniela Santos Oliveira[1], Frederico Rajão Martins[3], Alexandra Bernardo[1], Raquel Miriam Ferreira[1], Miguel Bernardes[1, 4], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [3]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal, [4]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Introduction: Treatment with teriparatide (TPTD) is associated with reduction of fracture risk in patients with severe osteoporosis. This drug can only be used for up to 2 years. After that a treatment course with antiresorptives should be considered, in order to prevent the rebound of bone turnover observed after TPTD discontinuation. In this regard, interest in sequential osteoporosis therapy has grown in recent years but the ideal timing for starting another treatment after TPTD is not well established.
Objective: The aim of this study is to assess if the timing of onset of antiresorptive therapy after TPTD discontinuation has implications in total hip bone mineral density (BMD) and in fracture risk.
Material and Methods: We performed a retrospective cohort study that included patients with severe

osteoporosis treated with TPTD 20mcg/day for 24 months and followed for at least 2 more years in the rheumatology department of a tertiary university hospital. For analysis, demographic and clinical data and results of dual-energy X-ray absorptiometry (DXA) after cessation of teriparatide were used. For comparison between groups Mann-Whitney U test was used.

Results: Fifty-five patients with osteoporosis, with a median age of 68 (32-85) years, were included. Forty-nine patients were female (89.1%). Nineteen patients (34.5%) had primary osteoporosis and 36 (65.5%) glucocorticoid-induced osteoporosis. The median time for initiating antiresorptive treatment was 7 (0-35) months after cessation of TPTD. Forty-three patients (78.2%) started a bisphosphonate, 6 denosumab (10.9%) and 6 patients did not receive any other treatment. The most prescribed bisphosphonate was zoledronate (69.8%). All patients received calcium and vitamin D supplementation. After completion of TPTD regimen 8 patients experienced at least one fragility fracture (14.5%). At follow-up, 37 (67.3%) of patients underwent DXA on average 30.0±15.4 months after starting antiresorptive agents. The median total hip BMD in patients who started antiresorptive therapy in the first 12 months (inclusive) after cessation of TPTD regime was 0,738 (0.587-0.993) g/cm² and the median total hip BMD of patients who started therapy after one year of discontinuation of TPTD was 0.683 (0.390-0.813) g/cm². This difference is marginally significant (p=0.067). The median time in starting antiresorptive treatment is higher in patients with new fragility fractures after TPTD than in patients without new fractures however this difference was not statistically significant (10.0 [2-35] vs 6.0 [0-35] months; p=0.393, respectively).

Conclusion: Although this study is unable to show that anti-resorptive treatment should be started in the first year after discontinuation of TPTD, it is promising since the difference between the medians in the total hip BMD values obtained until one year and after one year are marginally significant. These results can be linked to the small sample size and highlight the need for further studies in this area.

136 - VITAMIN D SERUM CONCENTRATION VARIES ACCORDING TO DISEASE ACTIVITY IN SPONDYLOARTHRITIS

Maria Seabra Rato[1], Salomé Garcia[1], Bruno Miguel Fernandes[1], Filipe Oliveira Pinheiro[1], Diogo Guimarães da Fonseca[2], Ana Martins[1], Daniela Santos Oliveira[1], Frederico Rajão Martins[3], Alexandra Bernardo[1], Miguel Bernardes[1, 4], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [3]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal, [4]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Introduction: Several studies have shown dissimilar results for the relationship between serum 25-hydroxyvitamin D concentration (25-OH-D) and disease activity in spondyloarthritis (SpA).

Objective: This study aims to assess whether vitamin D levels vary according to disease activity in patients with SpA before and after starting treatment with biologic disease-modifying anti-rheumatic drugs (bDMARDs).

Patients and Methods: An observational retrospective study was performed in SpA patients followed in the Rheumatology department of a tertiary university hospital. Demographic and clinical data were collected from the Rheumatic Diseases Portuguese Register (Reuma.pt). Patients were assessed for 25-OH-D levels before and after 6 months of treatment with the first bDMARD. Correlation between 25-OH-D levels and disease activity measured by Ankylosing Spondylitis Disease Activity Score (ASDAS) at baseline and after 6 months were assessed using student's t-test for two samples and one-way ANOVA and with post hoc tests for multiple comparisons.

Results: A total of 189 patients were included. Ninety-seven patients were females (51.3%). The mean age at diagnosis was 34.8±11.2 years and the median disease duration at the start of the first bDMARD was 4.9 years (min:0.1; max:46.0). All patients fulfilled the ASAS criteria for SpA. Nonsteroidal anti-inflammatory drugs were used by 102 patients (54.0%) and conventional synthetic DMARDs by 69 patients (36.5%). At 6 months, 188 patients were treated with tumor necrosis factor inhibitors and one with interleukin-17 inhibitor. According to ASDAS criteria, at baseline 36.8% of patients had high disease activity and 59.5% had very high disease activity. After 6 months of treatment with bDMARD 14.7% of patients have inactive disease, 21.6% low disease activity, 36.3% high activity and 12.6% very high disease activity. The mean value of 25-OH-D at baseline was significantly lower in the group of patients with very high disease activity

compared to the patients with high disease activity (21.9±11.1 ng/ml vs 26.1±11.6 ng/ml, p= 0.02). At 6 months of treatment the mean value of 25-OH-D in inactive, low, high and very high disease activity was 31.0±17.1ng/ml, 28.5±11.2ng/ml, 25.8±10.8ng/ml and 19.3 ±9.5ng/ml, respectively. There was a statistically significant difference between the groups, as determined by one-way ANOVA (p=0.001). A post hoc Dunnett T3 test revealed that patients with very high disease activity have significantly lower mean 25-OH-D levels (19.29±9.5) than patients with inactive disease (31.0±17., p=0.025) and low activity (28.5±11.2, p=0.009). Among the groups with high and very high disease activity, the significance is only marginal (p=0.068).

Conclusion: Vitamin D serum concentration varies according to disease activity in SpA. In fact, SpA patients with lower levels of 25-OH-D are associated with higher rates of disease activity, even in patients treated with biologics agents. It is important to be aware of vitamin D level as it can play a role in the management and treatment of the disease, mainly in the most severe patients.

140 - BONE FRAGILITY RISK FACTORS IN A PEDIATRIC RHEUMATOLOGY DEPARTMENT - DATA FROM A SINGLE-CENTER

Ana Teresa Melo[1, 2], Catarina Tenazinha[1, 2], Filipa Oliveira Ramos [1, 2, 3], Patrícia Costa Reis[2, 3, 4], JE Fonseca[1, 2], Raquel Campanilho-Marques[1, 2, 3]

[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Unidade de Reumatologia Pediátrica, Centro Hospitalar Universitário Lisboa Norte, Hospital de Santa Maria, Lisboa, Portugal, [4]Serviço de Pediatria, Centro Hospitalar Universitário Lisboa Norte, Hospital de Santa Maria, Lisboa, Portugal

Introduction: Low bone mineral density (BMD) is an under-recognized complication of chronic illness in childhood. Reducing the frequency of fragility fractures requires increased attention to risk factors, early intervention, and additional research to optimize therapy and potentially prevent their occurrence.

Objectives: To identify and characterize children with risk factors for low BMD followed in a Paediatric Rheumatology unit.

Methods: A retrospective observational study was conducted by consulting all the clinical records of patients followed at a Pediatric Rheumatology Unit, from January 1st 2020 to April 30th 2021. The following criteria were considered risk factors for low BMD: active chronic inflammatory disease (defined as moderate or high disease activity for at least 3 months duration), corticotherapy (>3month duration), reduced mobility, puberty disorders, metabolic diseases and previous fractures.

Results: Five hundred and sixty-six patients were analyzed, 71 (12.5%) of whom had risk factors for low BMD. Forty patients (56.3%) were female, with a median age of 13.7 [2-21] years. Nine patients with osteogenesis imperfecta (OI) were identified (2 of whom had also reduced mobility). Four other patients had reduced mobility (all had Duchenne muscular dystrophy diagnosis and all medicated with corticosteroids); a malabsorption syndrome was identified in 6 patients (half of them had a metabolic genetic disease diagnosis). Active chronic inflammatory diseases were identified in 41 patients. From those, 37 were also exposed to corticosteroids. The most frequent active chronic inflammatory diseases identified were juvenile idiopathic arthritis (n=12) and juvenile systemic lupus erythematosus (n=12), followed by idiopathic uveitis (n=3), juvenile dermatomyositis (n=3) autoimmune hepatitis (n=2), mixed connective tissue disease (n=2), and ANCA vasculitis (n=2).

If possible, patients were stimulated to practice physical exercise. An adequate dietary calcium intake and vitamin D supplementation were advised. Nineteen patients (26.7%) had at least one fragility fracture identified of whom 10 patients had a Z score of -2. Four of these patients were currently treated with alendronate, 5 with zoledronic acid (4 biannually and 1 annually) and 1 patient was treated with denosumab. None of these patients had subsequent fractures, except a girl with OI type VI that had previously been treated with pamidronate, but maintained repetitive fractures and for that reason was started on denosumab.

Discussion/Conclusion: In our cohort, inflammatory chronic diseases were the main risk factors for low BMD, demonstrating the impact of inflammation on bone, potentiated with steroid intake. An adequate treatment of the underlying illness, exercise, a sufficient calcium intake and vitamin D status are essential to prevent osteoporosis. If general measures fail to prevent further bone loss and fracture, pharmacologic therapy may be considered.

145 - ADESÃO À DIETA MEDITERRÂNICA EM DOENTES COM ARTRITE REUMATÓIDE: RELAÇÃO COM A ATIVIDADE DA DOENÇA, CAPACIDADE FUNCIONAL E IMPACTO GLOBAL

Sofia Charneca[1], Margarida Ferro[1], João Vasques[1], Eduardo Dourado[2, 3], Martins-Martinho J [2, 3], Ana Margarida Duarte-Monteiro[2, 3], Catarina Sousa-Guerreiro[1, 4], JE Fonseca[2, 3]

[1]Laboratório de Nutrição, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [4]Instituto de Saúde Ambiental, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal

Introdução: As estratégias alimentares e nutricionais têm sido reportadas como promissoras intervenções na gestão da artrite reumatoide (AR)[1]. A dieta mediterrânica (DM), rica em nutrientes e compostos bioativos com propriedades anti-inflamatórias e antioxidantes, tem efeitos benéficos na redução da dor e na capacidade funcional em indivíduos com AR[2].

Objetivo: Avaliar a relação entre a adesão à DM (AdDM) e a atividade da doença, capacidade funcional, e impacto global da doença em doentes com AR.

Métodos: Estudo observacional prospetivo e transversal, incluindo doentes diagnosticados com AR que cumprissem critérios de classificação ACR/EULAR 2010. A AdDM foi avaliada através da aplicação do questionário 14-item Mediterranean diet assessment tool, desenvolvido no âmbito do estudo Prevención con Dieta Mediterránea (PREDIMED). O score obtido define o grau de AdDM: um score ≥10 indica boa adesão; entre 6 e 9 indica adesão moderada; e ≤5 indica baixa adesão à DM. A atividade da doença foi determinada com o Disease Activity Score-28, DAS28-PCR. A capacidade funcional e o impacto global da doença foram avaliados utilizando os questionários Health Assessment Questionnaire (HAQ) e Rheumatoid Arthritis Impact of Disease (RAID), respetivamente. A análise estatística foi efetuada com recurso aos testes estatísticos T-Student e Mann-Whitney U, para comparações entre grupos, consoante a normalidade da distribuição dos dados.

Resultados: Um total de 98 indivíduos com AR (74% mulheres, n=72) com uma média de idade de 61,8±9,9 anos foram incluídos no estudo. Os indivíduos avaliados estavam sob metotrexato (77%), corticosteroides em dose baixa (60%), anti-inflamatórios não esteroides (34%) e/ou fármacos biotecnológicos (20%). A mediana do score de AdDM foi de 8,0±2,2, tendo sido verificada uma boa AdDM em 31% dos indivíduos e uma baixa AdDM em 12% dos indivíduos. A mediana do score do HAQ foi de 0,9±0,7, a mediana do score de DAS28-PCR (n=92) foi de 2,3±1,0 e a mediana do score do RAID (n=96) foi de 4,7±2,2. Indivíduos com uma baixa AdDM apresentaram valores significativamente mais elevados de DAS28-PCR quando comparado com aqueles que apresentam AdDM moderada a alta (média de 3,8±1,5 vs. 2,4±0,8, p<0,001). Acrescenta-se que não foram verificadas diferenças significativas na terapêutica (dose de corticoides e metotrexato) entre

FIGURA 1. MEDIANAS DA ATIVIDADE DA DOENÇA (DAS28-PCR) CONSOANTE O GRAU DE ADESÃO À DIETA MEDITERRÂNICA

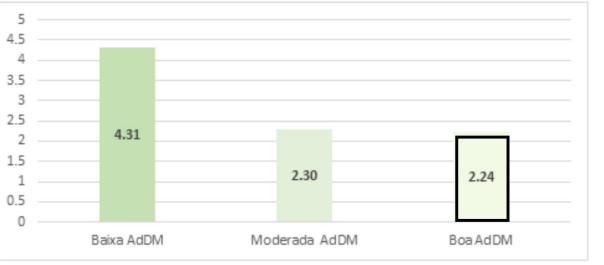


TABELA 1. CARACTERIZAÇÃO DA AMOSTRA CONSOANTE O GRUPO DE AF

	T1 (n=35)	T2 (n=35)	T3 (n=33)
Idade	65,31 ± 7,77	61,09 ± 9,96	56,61 ± 8,33
Ângulo de fase	4,44 ±0,44	5,48 ± 0,23	6,54 ± 0,67
Sexo			
Masculino	4 (11,4%)	5 (14,3%)	18 (54,5%)
Feminino	31 (88,6%)	30 (85,7%)	15 (45,5%)
Terapêutica farmacológica atual			
Metotrexato	22 (62,9%)	30 (85,7%)	29 (90,6%)
Hidroxicloroquina	4 (11,4%)	7 (20,0%)	4 (12,5%)
Sulfassalazina	6 (17,1%)	1 (2,9%)	4 (12,5%)
Corticoides em dose baixa	19 (54,3%)	22 (62,9%)	16 (50,0%)
Anti-inflamatórios não esteroides	11 (31,4%)	16 (45,7%)	10 (31,3%)
Biotecnológicos	7 (20,0%)	7 (20,0%)	7 (21,9%)

Idade e Ângulo de Fase: Média ± DP
Sexo e terapêutica farmacológica atual: Frequência (Percentagem)

grupos consoante a adesão à DM (p=0,667 e p=0,664, respetivamente). Os indivíduos com uma AdDM baixa/moderada apresentaram uma pior capacidade funcional, comparativamente àqueles com uma boa AdDM (p=0,042). Por fim, verificou-se uma tendência não estatisticamente significativa para níveis mais elevados de RAID nos doentes com baixa/moderada AdDM (média de 3,8±2,3 vs. 4,6±2,2, p=0,122).

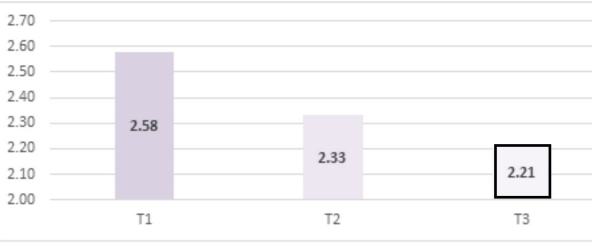
Conclusão: Os nossos resultados apoiam a tese de que uma maior AdDM está associada a menor atividade da doença e a melhor capacidade funcional em doentes com AR. A DM poderá constituir uma medida não farmacológica adjuvante ao tratamento da AR. Estudos de intervenção para comprovar esta hipótese deverão ser desenvolvidos.

TABELA 1. CARACTERIZAÇÃO DA AMOSTRA CONSOANTE O GRUPO DE AF

	T1 (n=35)	T2 (n=35)	T3 (n=33)
Idade	65,31 ± 7,77	61,09 ± 9,96	56,61 ± 8,33
Ângulo de fase	4,44 ±0,44	5,48 ± 0,23	6,54 ± 0,67
Sexo			
Masculino	4 (11,4%)	5 (14,3%)	18 (54,5%)
Feminino	31 (88,6%)	30 (85,7%)	15 (45,5%)
Terapêutica farmacológica atual			
Metotrexato	22 (62,9%)	30 (85,7%)	29 (90,6%)
Hidroxicloroquina	4 (11,4%)	7 (20,0%)	4 (12,5%)
Sulfassalazina	6 (17,1%)	1 (2,9%)	4 (12,5%)
Corticoides em dose baixa	19 (54,3%)	22 (62,9%)	16 (50,0%)
Anti-inflamatórios não esteroides	11 (31,4%)	16 (45,7%)	10 (31,3%)
Biotecnológicos	7 (20,0%)	7 (20,0%)	7 (21,9%)

Idade e Ângulo de Fase: Média ± DP
Sexo e terapêutica farmacológica atual: Frequência (Percentagem)

FIGURA 1. MEDIANA DA ATIVIDADE DA DOENÇA (DAS28-PCR) DE ACORDO COM OS GRUPOS DE ÂNGULO DE FASE



REFERÊNCIAS

1. Philippou E, Petersson SD, Rodomar C, Nikiphorou E. Rheumatoid arthritis and dietary interventions: Systematic review of clinical trials. Nutr Rev. 2021;79(4):410–28.
2. Forsyth C, Kouvari M, D'Cunha NM, Georgousopoulou EN, Panagiotakos DB, Mellor DD, et al. The effects of the Mediterranean diet on rheumatoid arthritis prevention and treatment: a systematic review of human prospective studies. Rheumatol Int. 2018;38(5):737–47.

148 - ÂNGULO DE FASE NA ARTRITE REUMATÓIDE – UMA NOVA VARIÁVEL COM VALOR PROGNÓSTICO?

Margarida Ferro[1], Sofia Charneca[1], João Vasques[1], Eduardo Dourado[2, 3], Ana Margarida Duarte-Monteiro[2, 3], Martins-Martinho J [2, 3], Catarina Sousa-Guerreiro[1, 4], JE Fonseca[2, 3]

[1]Laboratório de Nutrição, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [4]Instituto de Saúde Ambiental, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal

Introdução: O ângulo de fase (AF), que representa a relação entre a resistência e a reatância obtidas na avaliação da composição corporal por bioimpedância elétrica (BIA), é reconhecido como um indicador da integridade celular[1]. As alterações da composição corporal em doentes com AR são muito prevalentes e, uma vez que a evidência sugere que o AF está relacionado com a progressão e prognóstico de doenças inflamatórias crónicas, equaciona-se se um AF mais reduzido poderá ter utilidade clínica enquanto preditor de pior prognóstico na artrite reumatoide (AR)[2].

Objetivo: Estudar a associação entre o AF e a atividade da doença, impacto global da doença e a capacidade funcional em indivíduos com AR.

Métodos: Estudo observacional prospetivo e transversal, incluindo doentes diagnosticados com AR que cumprissem critérios de classificação ACR/EULAR 2010. A composição corporal foi avaliada por BIA com recurso ao equipamento Bodystat QuadScan 4000. A

atividade da doença foi determinada através do Disease Activity Score-28, DAS28-PCR. A capacidade funcional e o impacto global da doença foram avaliados através dos questionários Health Assessment Questionnaire (HAQ) e Rheumatoid Arthritis Impact of Disease (RAID), respetivamente. Através da estratificação do AF em tercis, a amostra foi dividida em três grupos: indivíduos com valores de AF mais baixos (AF≤5º, T1), AF intermédios (5,1º≤AF≤5,8º, T2) e AF mais elevados (AF≥5,9º, T3). A análise estatística foi efetuada com recurso aos testes estatísticos T-Student e Mann-Whitney U, para comparações entre grupos, de acordo com a normalidade da distribuição dos dados.

Resultados: Foram avaliados 103 indivíduos com AR (74% mulheres, n=76) com uma média de idades de 61,1±9,4 anos. Relativamente à terapêutica farmacológica, 80% dos doentes estavam sob metotrexato, 56% sob dose baixa de corticosteroides, 36% sob anti-inflamatórios não esteroides e 21% sob terapêutica com biotecnológicos. O valor médio do AF foi de 5,5±1,0º, a mediana do score de DAS28-PCR (n=85) 2,3±1,0, a mediana do score do RAID (n= 101) 4,6±2,3 e a mediana do score do HAQ 0,8±0,7. Indivíduos com maior AF (T3) estavam medicados com doses superiores de metotrexato quando comparado com o grupo com um menor AF (T1). Verificou-se uma tendência não estatisticamente significativa para níveis mais elevados de atividade da doença para AF mais baixos (T1 vs. T3, p=0,057). Doentes com menor AF apresentavam um maior impacto global da doença (média de 5,1±2,0 em T1 vs. 2,8±2,0 em T3, p<0,001) e também pior capacidade funcional (mediana de 1,1±0,6 em T1 vs. 0,6±0,5 em T3, p<0,001).

Conclusão: Neste estudo, valores de AF mais baixos estão associados a pior capacidade funcional e maior impacto global da AR. No que respeita à atividade da doença, parece existir tendência para uma associação com o AF, podendo a terapêutica farmacológica ter influenciado esta relação. Estes resultados suportam a continuação da exploração do AF como indicador prognóstico na AR.

REFERÊNCIAS

1. Norman K, Stobäus N, Pirlich M, Bosy-Westphal A. Bioelectrical phase angle and impedance vector analysis - Clinical relevance and applicability of impedance parameters. Clin Nutr. 2012;31(6):854–61.
2. Pineda-Juárez JA, Lozada-Mellado M, Ogata-Medel M, Hinojosa-Azaola A, Santillán-Díaz C, Llorente L, et al. Body composition evaluated by body mass index and bioelectrical impedance vector analysis in women with rheumatoid arthritis. Nutrition. 2018;53:49–53.

149 - THE ASSOCIATION BETWEEN AUTOANTIBODY LEVELS AND THE OUTCOMES OF ANTI-TUMOUR NECROSIS FACTOR ALPHA TREATMENT IN RHEUMATOID ARTHRITIS - A RETROSPECTIVE COHORT STUDY WITH TWO YEARS FOLLOW-UP

Daniela Santos Oliveira[1, 2], Ana Martins[1], Frederico Rajão Martins[3], Maria Seabra Rato[1], Filipe Oliveira Pinheiro[1], Diogo Guimarães da Fonseca[4], Salomé Garcia[1], Bruno Miguel Fernandes[1], Carlos Vaz[1, 5], Miguel Bernardes[1, 5], Lúcia Costa[1]

[1]Rheumatology Department, Centro Hospitalar de São João, Porto, Portugal, [2]Center for Health Technology and Services Research (CINTESIS), Faculty of Medicine, University of Porto, Porto, Portugal, Porto, Portugal, [3]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [4]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [5]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal

Background: In rheumatoid arthritis (RA), autoantibodies namely anticitrullinated protein antibodies (Anti-CCP) have prognostic value, independently predicting radiologic progression. However, the evidence is still controversial about how the autoantibody levels change over time and their role in treatments outcomes and in monitoring disease activity in RA.

Aims: This study aimed to characterize the changes of autoantibodies levels (rheumatoid factor (RF) and Anti-CCP) over time and to explore the association between these autoantibodies and the outcomes of the first anti-tumour necrosis factor alfa (anti-TNF-α) therapy as first biologic agent in RA.

Methods: An observational retrospective cohort study was conducted with two years of follow-up. Patients with diagnosis of RA according to American College of Rheumatology (ACR) criteria and registered on Rheumatic Diseases Portuguese Register (Reuma.pt) who started their first anti-TNFα agent (as first biologic) between 2003 and 2018 were included. Patients with positive RA (>30 UI/mL) and/or positive Anti-CCP (>10 U/mL) at their first visit were included. Demographic, clinical and laboratory data were obtained by consulting Reuma.pt. Disease Activity Score for 28 joints [DAS28(3v); DAS28(4v); DAS28(3v; C-Reactive Protein (CRP)), DAS28(4v; CRP), delta DAS28(4v)], Health Assessment Questionnaire (HAQ), delta HAQ, Anti-CCP and RF levels were assessed at baseline, 12 and

24 months. To examine the differences between Anti-CCP and RF levels at baseline, 12 months and 24 months the Wilcoxon test for paired samples was performed. In order to correlate the Anti-CCP and RF levels with DAS28 variables, delta DAS28(4v), HAQ and delta HAQ at baseline, 12 months and 24 months, a correlation coefficient, Spearman's coefficient, was used.

Results: A total of 116 patients (mean age of 50.2±10.4 years old; 85.3% female) with RA were included with a median disease duration of 10.5 [5-18.5] years and a follow-up time of 8 [5-14] years. About 49% of patients were FR and Anti-CCP positivity, 38% only FR positivity and 13% only Anti-CCP positivity. At baseline, 64 (55.2%) patients had an erosive disease and 50 (43.1%) had extra-articular manifestations. Compared to the baseline (160[74.8-496]), FR levels decreased significantly at 12 months (121[49.1-321.8]) and 24 months (107.5[43.3-332]) with a p=0.017 and p=0.029, respectively. There were no differences in Anti-CCP levels over time. No correlation was found between FR/Anti-CCP levels and different DAS28 variables, DAS28(4v) delta, HAQ, and HAQ delta at 12 months and 24 months.

Conclusions: We found that in patients with RA treated with a first anti-TNF-α agent as first biologic, FR levels decreased at 12 months and 24 months follow-up. However, our study failed to demonstrate a correlation between autoantibodies levels and disease activity and HAQ variables. In fact, previous research demonstrated that there is an association between autoantibodies levels and disease activity in RA, nonetheless not being static and increasing with signs of inflammation at baseline. So, further research with large samples is needed to explore this correlation considering the adjustment for confounding inflammatory variables, namely number of swollen/tender joints and morning stiffness.

152 - FOCUSED EXTRACORPOREAL SHOCK WAVES IN RHEUMATOLOGIC PERIARTICULAR PATHOLOGY

Rita Cunha[1], Luís Oliveira[2], José Luis Carvalho[2], Inês Natário[2]

[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [2]Reabilitação Musculo-Esquelética, Centro de Reabilitação do Norte, Vila Nova de Gaia, Portugal

Introduction: In Rheumatology clinical practice, periarticular pathology is very common and affects the patients' quality of life. Many patients are refractory to conventional treatment done in the clinical setting (e.g. corticoanesthesic infiltrative procedures). Therefore, other approaches are needed. Extracorporeal shock wave therapy is a non-invasive treatment by application of shock waves to injured tissue with the intention to reduce pain and to promote regeneration and healing processes.1 Some of The International Society for Medical Shockwave Treatment (ISMST) standard indications are calcific tendinopathy of the shoulder, plantar fasciitis, lateral humeral epicondylitis, greater trochanter pain syndrome (GTPS), patellar tendinopathy and Achilles tendinopathy.

Objectives: To perform a descriptive analysis of the effect of this treatment on pain measured by visual analog scale (VAS) in different periarticular pathology.

Methods: Consecutive patients with periarticular pathology common seen in rheumatologic setting (calcified tendinopathy of the shoulder, plantar fasciitis, lateral humeral epicondylitis, greater trochanter pain syndrome (GTPS)) attending shock wave outpatient clinic of Centro de Reabilitação do Norte, from September 2020 to March 2021 were included. The treatment consists of 3 sessions, using focused shock waves with energy levels ranges from 0.15-0.30 mJ/mm2 and the number of pulses range from 1500-2500 depending on the pathology.

In selected cases, it may be beneficial to complement focal shockwave therapy with radial (pneumatic) shockwaves in order to promote myotendinous elongation.

Medical records were reviewed to retrieve the following data: sociodemographic (age and gender), initial and final VAS and previous treatment. Statistical analyses were conducted using SPSS version 25. Continuous variables were described with mean/median ± standard deviation (SD) and categorical variables with relative frequency.

Results: Fifty-one patients (22 patients with calcified shoulder tendinopathy, 11 with plantar fasciitis, 12 with lateral humeral epicondylitis and 6 with GTPS) were included. Thirty-three (64.7%) were female, mean age (±SD) of 56.6(±8.8) years, median initial pain VAS (±SD) of 7.0(±1.8) and median final pain VAS (±SD) of 4.0(±7.0). Fourteen patients already did other procedures before shock wave therapy (8 with calcified shoulder tendinopathy, 3 with lateral humeral epicondylitis, 2 with plantar fasciitis and 1 with GTPS).

At 3rd session, the mean (±SD) difference between final and initial pain VAS was 2.7±2.2 for lateral humeral epicondylitis, 2.3±1.2 for plantar fasciitis,

2.0±1.8 for GTPS and 1.7±2.5 for calcific tendinopathy of the shoulder.

Conclusion: In this descriptive analysis the biggest decreased in pain VAS was obtained for lateral humeral epicondylitis and plantar fasciitis. In the literature, shock wave therapy has an excellent success rate (ranged from 65 to 91%) and negligible side effects and complications. The therapeutic effect of shock waves can be continuing weeks after finished the procedure (generally 4-6 weeks). For this reason, the pain evaluation did in the 3rd session and the result of the intervention can be underestimated. Rheumatologists should be aware of this procedure, as an alternative non-invasive technique, and its indications.

REFERENCES

1. Crevenna, R., Mickel, M., Schuhfried, O. et al. Focused Extracorporeal Shockwave Therapy in Physical Medicine and Rehabilitation. Curr Phys Med Rehabil Rep 9, 1–10 (2021)

154 - ANXIETY AND DEPRESSION IN SSC – ASSESSING FUNCTION, QUALITY OF LIFE AND GASTROINTESTINAL INVOLVEMENT

Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Tiago Beirão[1], Miguel Guerra[2], Romana Vieira[1], Flávio Campos Costa[1], Taciana Videira[1], Joana Aleixo[1], P Pinto[1]

[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [2]Rheumatology Department, Centro Hospitalar e Universitário Cova da Beira, Covilhã, Portugal

Background: Systemic Sclerosis (SSc) is a chronic disease with multi-organ manifestations that may contribute to disability and low quality of life[1]. Therefore, anxiety and depression are more frequent in SSc patients than in general population[2].

Objectives: To assess the prevalence of anxiety and depression in a SSc cohort and to evaluate its correlation with disability, quality of life and assessment of gastrointestinal (GI) involvement scores.

Methods: A cross-sectional study was conducted evaluating a cohort of SSc patients. All patients answered to the Hospital Anxiety and Depression Scale (HADS) questionnaire. A cut-off score < 8 was considered normal. Questionnaires for assessment of disability (Health Assessment Questionnaire (HAQ) and Scleroderma HAQ (SHAQ)), quality of life (36-Item Short Form Health Survey (SF-36) and EuroQol-5D (EQ-5D)) and gastrointestinal involvement (University Of California, Los Angeles, Scleroderma Clinical Trials Consortium Gastrointestinal Scale (UCLA SCTC GIT) 2.0) were also obtained. Clinical data was analyzed using IBM SPSS Statistics 26®.

Results: We included 20 patients, 17 females [n = 17 (85%)], median (min, max) age was 52.5 (28, 75) years-old. Regarding disease classification, 13 (65%) had limited SSc, 4 (20%) had diffuse SSc and 3 (15%) had early SSc. A score ≥ 8 was found in 14 (70%) patients on HADS-A [median (min, max) = 9 (2, 19)] and in 12 (60%) patients on HADS-D [median (min, max) = 8 (1, 15)]. Depressive patients had significantly worst scores on the measures of function, such as HAQ and lung and gastrointestinal involvements and patient global assessment of SHAQ, of quality of life, such as EQ-5D and physical functioning, role physical, bodily pain, vitality, social functioning and mental health domains of SF-36, and on the UCLA SCTC GIT 2.0 scale. Anxious patients had significantly worst scores on social functioning and mental health domains of SF-36 and on the UCLA SCTC GIT 2.0 scale.

Conclusion: The prevalence of depression and anxiety on SSc patients is high and should not be neglected. Overall disability and multiorgan manifestations, particularly GI involvement, may contribute to a low quality of life and consequently to depression and anxiety.

REFERENCES

1. Firestein & Kelley's Textbook of Rheumatology 2-Volume Set, 11th Edition
2. Brett D. Thombs et al. Depression in Patients With Systemic Sclerosis: A Systematic Review of the Evidence. Arthritis & Rheumatism (Arthritis Care & Research) Vol. 57, 2007, pp 1089–1097

156 - THE IMPACT OF FATIGUE ON SYSTEMIC SCLEROSIS PATIENTS

Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Tiago Beirão[1], Miguel Guerra[2], Romana Vieira[1], Flávio Campos Costa[1], Taciana Videira[1], Joana Aleixo[1], P Pinto[1]

[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário Cova da Beira, Covilhã, Portugal

Background: Fatigue is a frequent and commonly undervalued symptom among rheumatic disease, including Systemic Sclerosis (SSc)[1,2].

Objectives: To determine the prevalence of severe

fatigue in a SSc cohort and to evaluate how it correlates with disability, quality of life and mental illness.

Methods: A cross-sectional study was conducted evaluating a cohort of SSc patients. Fatigue was evaluated using Functional Assessment Chronic Illness Therapy (Fatigue) (FACIT-F) questionnaire. A value < 30 was defined as severe fatigue. Questionnaires for assessment of disability (Health Assessment Questionnaire (HAQ) and Scleroderma HAQ (SHAQ)), quality of life (36-Item Short Form Health Survey (SF-36) and EuroQol-5D (EQ-5D)) and mental illness (Hospital Anxiety and Depression Scale (HADS)) were also filled. Clinical data was obtained and analysed using IBM SPSS Statistics 26®.

Results: We included 20 patients, 17 females [n = 17 (85%)], median (min, max) age was 52.5 (28, 75) years-old. Regarding disease classification, 13 (65%) had limited SSc, 4 (20%) had diffuse SSc and 3 (15%) had early SSc. The median FACIT-F score was 34 (3, 48). The prevalence of severe fatigue was 40% (n = 8). Fatigue had a moderate negative correlation with HAQ (τ = -0.641; p < 0.001) and a weak negative correlation with lung (τ = -0.345; p = 0.039) and gastrointestinal (τ = -0.419; p = 0.011) involvements and with patient global assessment (τ = -0.325; p = 0.047) subtopics of SHAQ. A moderate positive correlation was found between FACIT-F and EQ-5D (τ =0.625; p < 0.001) and physical functioning (τ = 0.560; p = 0.001) and vitality (τ = 0.777; p < 0.001) domains of SF-36. The remaining SF-36 domains had a weak positive correlation with FACIT-F (Table 1). Regarding mental illness, there was a moderate negative correlation between FACIT-F and HADS-D (τ = -0.638; p < 0.001) and HADS-A (τ =-0.535; p = 0.001).

Conclusion: Severe fatigue is frequent among SSc patients. The greater the fatigue, the greater the disability, the lower the quality of life and the worse the score on the scale of depression and anxiety.

REFERENCES

1. F. Basta, A. Afeltra, D.P.E. Margiotta. Fatigue in systemic sclerosis: a systematic review. Clin Exp Rheumatol 2018; 36 (Suppl. 113): S150-S160
2. Sarah Hewlett, Emma Dures, And Celia Almeida. Measures of Fatigue. Arthritis Care & Research Vol. 63, No. S11, November 2011, pp S263–S286

164 - IMPROVEMENT IN FATIGUE, ANXIETY AND DEPRESSION WITH SECUKINUMAB TREATMENT IN ANKYLOSING SPONDYLITIS - DATA FROM THE RHEUMATIC DISEASES PORTUGUESE REGISTRY (REUMA.PT)

Soraia Azevedo[1], José Tavares-Costa[1], Laires PA[2], Salomé Garcia[3], Patrícia Nero[4], Jorge Pestana Lopes[5], Martins-Martinho J [6], Ana Bento da Silva[7], Joana Ramos Rodrigues[1], Graça Sequeira[8], Emanuel Costa[9], Filipe Cunha Santos[10], Carolina Furtado[11], Bernardo Santos[12], Ana Raposo[13], Liliana Saraiva[14], Beatriz Samões[15], Filipe Barcelos[16], Maria José Santos[5, 17], Helena Santos[16, 18]

[1]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [2]HE&OR - Novartis Portugal / ENSP – NOVA University of Lisbon, Lisboa, Portugal, [3]Rheumatology Department, Centro Hospitalar de São João, Porto, Portugal, [4]Rheumatology Department, Hospital CUF Descobertas, Lisboa, Portugal, [5]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [6]Rheumatology and Bone Metabolic Diseases Department, Centro Hospitalar Lisboa Norte, EPE - Hospital de Santa Maria, Lisbon Academic Medical Centre, Lisboa, Portugal, [7]Rheumatology Department, Centro Hospitalar Lisboa Ocidental, Lisboa, Portugal, [8]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, [9]Rheumatology Department, Hospital de Braga, Braga, Portugal, [10]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, [11]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal, [12]Rheumatology Department, Centro Hospitalar do Baixo Vouga, EPE, Aveiro, Aveiro, Portugal, [13]Rheumatology Department, Centro Hospitalar Trás-os-Montes e Alto Douro, Vila Real, Portugal, [14]Rheumatology Department, Centro Hospitalar Universitário de Coimbra, Coimbra, Portugal, [15]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [16]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal, [17]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [18]EpiDoc Research Unit, CEDOC, NOVA Medical School, Lisboa, Portugal

Background: A recent systematic review and meta-analysis shows an increased risk of depression and anxiety among patients with Ankylosing Spondylitis (AS). (1) Furthermore, fatigue is one of the most important factors that predict poor quality of life in AS. (2)

Objective: To analyze the effect of secukinumab treatment on fatigue, anxiety and depression in AS

patients.

Methods: This was a national, observational study of patients with the diagnosis of AS using real world anonymous patient-level data from the Portuguese national register database Reuma.pt. (3) Outcome measures: FACIT-F (Functional Assessment of Chronic Illness Therapy-Fatigue) and HADS (Hospital Anxiety and Depression Scale), were collected at baseline and at 3, 6 and 12 months after secukinumab initiation.

Results. We included 168 patients with AS treated with secukinumab, both naïve and non-naïve to biologic treatment. Overall, 52.4% were men and when secukinumab was started the mean age was 45.1 (SD=11.3) years-old, with a disease duration of 17.3 (SD=11.2) years. Only in 27.4% of patients secukinumab was the first biologic.

When we analyze patient evaluation of fatigue, a significantly improvement in FACIT-F was seen at 3 months, and sustained at 6 and 12 months after secukinumab initiation.

Anxiety and depression decrease significantly early in treatment course (at 3 months evaluation). (Table 1)

Conclusions. Secukinumab treatment in AS patients led to a fast and sustained improvement in fatigue, anxiety and depression.

Acknowledgement: This study was supported by Novartis and presented on Behalf of PROSAS Study group.

REFERENCES

1. Zusman EZ, Howren AM, Park JYE, Dutz J, De Vera MA. Epidemiology of depression and anxiety in patients with psoriatic arthritis: A systematic review and meta-analysis. Semin Arthritis Rheum. 2020;50(6):1481–8.
2. Dean LE, MacFarlane GJ, Jones GT. Five potentially modifiable factors predict poor quality of life in ankylosing spondylitis: Results from the Scotland registry for ankylosing spondylitis. J Rheumatol. 2018;45(1):62–9.
3. Canhão H, Faustino A, Martins F, Fonseca JE. Reuma. PT - the rheumatic diseases portuguese register. Acta Reumatol Port. 2011;36(1):45–56.

175 - DOENÇA DE STILL DO ADULTO: EXPERIÊNCIA DE UM CENTRO

Ana Isabel Maduro[1], André Saraiva[1], Helena Assunção[1], Ana Rita Prata[1], Sara Serra[1], JAP da Silva[1,2]

[1]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [2]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal

Introdução: A doença de Still do Adulto (DSA) é uma doença inflamatória sistémica rara (1:100.000) de etiologia desconhecida, que se caracteriza por grande heterogeneidade quanto à apresentação clínica, evolução e gravidade.

Objetivo: Descrever as manifestações clínicas, parâmetros laboratoriais, tratamento e evolução de pacientes com DSA num centro terciário de Reumatologia.

Métodos: Análise retrospetiva dos casos de DSA em doentes internados no Serviço de Reumatologia do Centro Hospitalar e Universitário de Coimbra (CHUC) entre 2005 e 2020, através da consulta do processo clínico. Todos os doentes cumpriram os critérios diagnósticos de Yamaguchi.

Resultados: Foram incluídos 9 doentes, dos quais 5 (55.6%) eram do sexo feminino. A mediana de idade ao diagnóstico foi de 30 anos, tendo variado entre os 18 e os 58 anos. Todos os doentes (n=9; 100%) apresentavam febre e artralgias no momento do diagnóstico. Outras manifestações comuns à apresentação foram odinofagia (n=8; 88.9%), mialgias (n=7; 77.8%), sintomas constitucionais (n=6; 66.7%), exantema (n=5; 55.6%), artrite (n=5; 55.6%); hepatomegalia (n=5; 55.6%) e adenopatias (n=5; 55.6%). Pleurite e pericardite foram

TABLE 1: ANXIETY, DEPRESSION AND FATIGUE AFTER 3, 6 AND 12 MONTHS OF SECUKINUMAB TREATMENT.

	Baseline (n=168)	3 months (n=157)	p-value*	6 months (n=144)	p-value*	12 months (n=110)	p-value*
HADS							
Anxiety, mean (SD)	8.5 (3.9)	7.4 (4.6)	0.005	8.2 (5.0)	0.046	7.6 (5.2)	0.006
Depression, mean (SD)	7.5 (4.5)	6.6 (4.4)	0.038	7.3 (4.9)	0.091	7.0 (5.7)	0.005
FACIT-F, mean (SD)	25.7 (12.1)	28.2 (11.9)	0.008	29.3 (13.1)	0.008	33.4 (11.2)	<0.001

*p value - Comparisons across different timepoints and baseline; NS: non-significant
FACIT-F: Functional Assessment of Chronic Illness Therapy – Fatigue; HADS: Hospital Anxiety and Depression Scale; SD: standard deviation;
Sample size is not constant - Baseline: HADS (n=39); FACIT-F (n=41); 3 months: HADS (n=25); FACIT-F (n=26); 6 months: HADS (n=24); FACIT-F (n=22); 12 months: HADS (n=21); FACIT-F (n=21);

observadas em 4 (44.4%) e 3 (33.3%) dos doentes, respetivamente. Analiticamente, todos os pacientes apresentavam leucocitose (média 21.01 ± 8.41 x109/L) com neutrofilia e elevação da ferritina (média 3727.78 ± 4634.51 ug/L), da proteína C reativa (média 16.80 ± 7.99 mg/dl) e da velocidade de sedimentação (média 82 ± 25.93 mm/h). Observou-se elevação das enzimas hepáticas em 6 (66.7%) doentes. Quanto ao tratamento, foram administrados anti-inflamatórios não esteróides (AINEs) a 7 doentes (77.8%) e todos (n=9; 100%) receberam glucocorticóides (GCT) (prednisolona entre 0.5 a 1mg/Kg). Em 6 doentes (66,7%) foi associado um DMARD clássico (metotrexato na maioria dos casos), por falha ou dependência de GCT. O tratamento com biológicos foi necessário em 6 casos (66,7%). Dos doentes a fazer biológico, o anakinra foi a primeira escolha, tendo sido realizado switch para tocilizumab em 3 deles (por ausência de resposta ou reação adversa). Dos casos com maior gravidade destaca-se uma doente que desenvolveu pleurite exuberante à apresentação, com insuficiência respiratória e necessidade de suporte ventilatório mecânico. De referir, ainda, o caso de uma doente com DSA refratária à terapêutica que apresentou, 4 anos após o diagnóstico, hipertensão pulmonar e síndrome de ativação macrofágica com disfunção multiorgânica e desfecho fatal.

Conclusão: A DSA é uma patologia com envolvimento sistémico e, embora incomum, deve ser considerada em pacientes com febre de origem desconhecida, impondo-se a exclusão de outras doenças autoimunes, infeciosas e neoplásicas. Segundo a literatura, os GCT são eficazes no controlo das manifestações clínicas em cerca de 60% dos pacientes e os biológicos apenas considerados para casos refratários ao tratamento imunossupressor convencional. Curiosamente, nesta coorte, a maioria dos doentes necessitou de tratamento com agentes biológicos, traduzindo casos mais graves da doença e com pior prognóstico. De facto, embora o prognóstico da DSA seja geralmente favorável, podem ocorrer complicações com risco de vida, cujo reconhecimento precoce e atuação imediata terão um impacto significativo na qualidade de vida e na sobrevida dos doentes.

186 - IMPACT OF AGE ON PATIENT REPORTED OUTCOMES AFTER UPADACITINIB TREATMENT OF MODERATE-TO-SEVERE RHEUMATOID ARTHRITIS

Luís Cunha-Miranda[1], Ana Cordeiro[2], Daniela Santos-Faria[3], Fernando Pimentel-Santos[4, 5], Filipe Vinagre[2], Helena Santos[1], José Tavares-Costa[6], Nikita Khmelinskii[7, 8], P Pinto[9], Rui Lourenço Teixeira[7, 8], Santiago Andres Rodrigues Manica[4, 5], Taciana Videira[9], ana rita cravo[10]

[1]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal, [2]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal, [3]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [4]Serviço de Reumatologia, Hospital de Egas Moniz, Centro Hospitalar de Lisboa Ocidental, Lisboa, Portugal, [5]CEDOC, NOVA Medical School, Lisboa, Portugal, [6]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [7]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Hospital de Santa Maria, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [8]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [9]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [10]Medical, AbbVie, Portugal, Lisboa, Portugal

Background: Several studies suggested age impact on drug response, in several diseases and in rheumatoid arthritis (RA). Older patients (pts) have a worse prognosis and a more difficult disease to treat. Patient-reported outcomes provide additional information on how a treatment impacts pts' life and are pivotal when evaluating the efficacy of RA treatments. Upadacitinib (UPA) is a selective and reversible JAK inhibitor approved for moderate to severe RA in adult patients with inadequate response or who are intolerant to disease-modifying anti-rheumatic drugs (DMARDs).

This study aimed to characterize UPA impact on self-reported pain, disability, and fatigue, among RA pts aged 65 years or older.

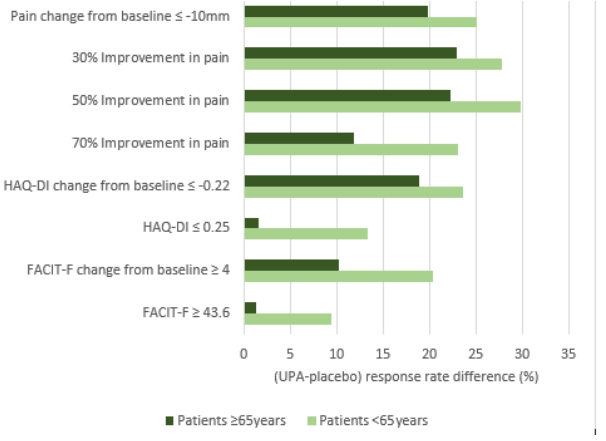
Methods: Pooled analysis from 3 double-blind, multicenter, phase 3 studies evaluating UPA in adult pts with moderately to severely active RA and inadequate response to conventional synthetic DMARDs (SELECT-NEXT study), biologic DMARDs (SELECT-BEYOND study) or methotrexate (SELECT-COMPARE study). Data from baseline to week 12 of pts receiving UPA 15mg once daily (n=1036) and placebo (PCB, n=1041) was pooled from each study into a specific analysis set.

UPA effects after 12 weeks of treatment were estimated within age subgroups (≥65y vs. <65y), regarding pain (visual analog scale, VAS), disability (Health Assessment Questionnaire-Disability Index, HAQ-DI),

TABLE: RESPONSE RATES AND PLACEBO-ADJUSTED TREATMENT EFFECTS AT WEEK 12

	Patients <65y			Patients ≥65y		
	Placebo	UPA	UPA – placebo difference (95%CI)	Placebo	UPA	UPA – placebo difference (95%CI)
Pain change from baseline (mm), LS mean	-13.0	-31.0	-18.0 (-20.4, -15.6)	-17.1	-29.5	-12.4 (-17.2, -7.6)
Pain change from baseline ≤ -10mm	50.1	75.2	25.1 (20.5, 29.7)	52.7	73.2	19.9 (11.1, 28.7)
30% improvement in pain	37.0	64.7	27.8 (23.1, 32.4)	40.7	64.5	23.0 (14.0, 32.1)
50% improvement in pain	21.5	51.2	29.8 (25.3, 34.2)	25.2	47.7	22.2 (13.4, 30.9)
70% improvement in pain	10.9	33.9	23.1 (19.3, 27.0)	13.7	25.7	11.8 (4.5, 19.2)
HAQ-DI change from baseline, LS mean	-0.29	-0.63	-0.34 (-0.39, -0.28)	-0.32	-0.51	-0.19 (-0.29, -0.09)
HAQ-DI change from baseline ≤ -0.22	49.9	73.4	23.6 (19.0, 28.2)	48.2	67.5	18.9 (9.7, 28.1)
HAQ-DI ≤ 0.25	9.8	23.2	13.4 (9.8, 16.9)	10.6	12.6	1.6 (-4.3, 7.6)
FACIT-F change from baseline, LS mean	4.1	8.9	4.8 (3.8, 5.8)	4.5	7.0	2.5 (0.7, 4.3)
FACIT-F change from baseline ≥ 4	44.4	64.8	20.4 (15.3, 25.6)	46.1	56.8	10.3 (0.0, 20.7)
FACIT-F ≥ 43.6	16.4	25.9	9.4 (5.2, 13.7)	20.2	21.6	1.3 (-7.2, 9.8)

FIGURE: UPADACITINIB TREATMENT EFFECTS (PLACEBO-ADJUSTED) AT WEEK 12, BY AGE GROUP



and fatigue (Functional Assessment of Chronic Illness Therapy - Fatigue Scale, FACIT-F). Response rates at week 12, i.e., percentage of pts reporting improvements and improvements ≥ minimum clinically important differences (MCID), were determined. Treatment effects and 95% confidence intervals (CI) between group difference of binary PROs were based on Mantel-Haenszel estimation (adjusting for study). Changes in least squares mean (LSM) from baseline to week 12 were based on a mixed effect for repeated measures model.

Results: The analysis set included 1636 pts <65y (UPA: 822; PCB: 814) and 441 pts ≥65y (UPA: 214; PCB: 227). Among older age group, 16% (n=71) were ≥75 years. The majority of pts were females (UPA:81%; PCB:79%), diagnosed ≥5 years (56% in both groups) and presented clinical disease activity index ≥22 (89% in both groups). Treatment groups were similar regarding demographic, RA and treatment

characteristics at baseline (overall and by age group).

UPA treatment resulted in statistically higher PCB-adjusted effects regarding pain change from baseline ≤ -10mm (≥65y: 19.9% vs. <65y: 25.1%) and other pain-related outcomes. Considering self-reported disability, UPA treatment resulted in a statistically higher proportion of pts than PCB with HAQ-DI change from baseline ≤ -0.22 (≥65y: 18.9% vs. <65y: 23.6%) and, only for pts <65y, a statistically significant proportion with HAQ-DI ≤ 0.25 (13.4%). The PCB-adjusted effect of UPA treatment on fatigue improvement was statistically significant only for pts <65y: compared with placebo, 20.4% UPA pts <65y showed a FACIT-F change from baseline ≥4 and 9.4% UPA pts <65y showed FACIT-F ≥43.6.

Conclusion: Although placebo-adjusted treatment effects resulted less evident in pts aged 65y or older, UPA treatment showed statistically significant improvements of self-reported pain and disability. Age is an important factor when looking at patient-reported outcomes and UPA 15mg has shown significant improvements in both younger and elderly RA pts.

197 - VITAMIN D LEVELS IN PSORIATIC ARTHRITIS PATIENTS UNDER BDMARD: A REAL-LIFE COHORT STUDY

Frederico Rajão Martins[1], Daniela Santos Oliveira[2, 3], Ana Martins[2], Diogo Guimarães da Fonseca[4], Filipe Oliveira Pinheiro[2], Maria Seabra Rato[2], Bruno Miguel Fernandes[2], Salomé Garcia[2], Miguel Bernardes[2], Lúcia Costa[2]

[1]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [3]Center for Health Technology and Services Research (CINTESIS), Faculdade de Medicina da Universidade do Porto, Porto, Portugal, Porto, Portugal, [4]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Introduction: The scope of manifestations of psoriatic arthritis (PsA) may include several skeletal features due to abnormal bone metabolism, in which vitamin D plays a vital role. Previous studies showed mild association between vitamin D and psoriatic arthritis. There hasn't been consensual data regarding the impact of this finding on patient and disease related outcomes, especially on those under biologic therapy (bDMARD).

Objectives: To assess outcomes in PsA patients under first bDMARD according to baseline levels of 25- OHvitD.

Methods: All patients with the definite diagnosis of psoriatic arthritis according to the CASPAR criteria, who started treatment with bDMARD without concomitant vitamin D supplementation, being followed up in a tertiary hospital's rheumatology department, were included in this analysis. Patients without serum 25-OHvitD level evaluation at either baseline, 6 or 12 months after the introduction of bDMARD were excluded. Socio-demographic data, disease and patient-related outcomes were assessed at the same timesets. Correlations between variables were assessed using Spearman's rank test. Fisher's exact test and Mann Whitney U tests were used for subgroup analysis.

Results: A total of 56 patients were included, mostly females (58.9%), with an asymmetric oligoarthritic pattern or with a predominant axial involvement (79.6%), under non-steroidal anti-inflammatory drugs (70.5%) or conventional DMARD (80.4%) at the time of bDMARD introduction. Most patients were treated with golimumab (33.9%), etanercept (30.4%) or adalimumab (17.9%), with an average age at bDMARD initiation of 48.9 +/- 11.7 years, after a median interval after diagnosis of 9.0 (0.6 – 63.8) years. Mean ASDAS CRP was 3.6 +/- 1.1 and, in addition, baseline levels of 25-OHvitD were on average 28.9 +/ 14.9 ng/mL, with 57.1% of patients with values below 30 ng/mL and 35.7% below 20 ng/mL.

There were no statistically significant correlations between vitamin D levels at baseline and ASDAS CRP at 6 (r= - 0.05; p = 0.77) or 12 (r = 0.04; p = 0.82) months, BASDAI at 6 (r = 0.231; p= 0.36) or 12 (r = -0.02; p = 0.95) months, BASMI at 6 (r = 0.07; p = 0.78) or 12 (r = 0.74; p = 0.74) months, BASFI at 6 (r = 0.16; p = 0.52) or 12 (r = 0.26; p = 0.72) months and enthesitis scores at 6 and 12 months of bDMARD.

No significant differences were found in response to bDMARD therapy at 6 and 12 months evaluated by ASDAS response and ASAS response criteria, as well as regarding BASDAI, BASFI, BASMI, acute phase reactants, enthesitis scores and pain assessment measures at the same time sets, according to baseline 25-OHvitD levels.

In subgroup analysis, there were significantly higher ASDAS CRP (U = 15; p = 0.03), patient VAS (U = 18; p = 0.03), physician VAS (U = 11; p = 0.03), CDAI (U 4.5; p < 0.01) and SDAI (U = 4; p < 0.01) scores at 6 months of biologic therapy in the 25-OHvitD ≥ 20 ng/mL group vs 25-OHvitD < 20ng/mL. No difference was statistically significant between groups with 25-OHvitD < 30 ng/mL vs 25-OHvitD ≥ 30 ng/mL.

Conclusion: Our study failed to show significant

correlations between baseline 25-OHvitD levels and therapeutic response measures in psoriatic arthritis patients undergoing a first bDMARD therapy. There were some differences of uncertain clinical relevance regarding ASDAS CRP, SDAI CDAI and pain assessment scores at 6 months of treatment between patients according to the 20 ng/mL cut off value.

198 - THYROID HORMONE REPLACEMENT AND OSTEOPOROSIS IN EUTHYROID POST-MENOPAUSAL WOMEN. EXPERIENCE FROM A SINGLE CENTER

João Paulo Vilas-Boas[1], Carolina Mazeda[1], Susana P. Silva[1], Barcelos A.[1], Maria Céu Portelada[1], Graça Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar do Baixo Vouga, E.P.E., Aveiro, Portugal

Objective: To evaluate the bone mineral density (BMD) and number of fractures in osteoporotic post-menopausal women treated with thyroid hormone replacement therapy (THR) in a cross-sectional study. Long term THR has been suggested to be inversely correlated with BMD and increased risk of fractures. It is still unclear if THR has a negative effect on bone remodeling even in sustained euthyroid patients.

Material and Methods: Data regarding 49 post-menopausal osteoporotic women with ongoing THR treatment was collected from a single Portuguese center orthopedic ward between 2019 and 2021. Patients treated for at least one year with thyroxine, regardless of the dose, due to primary or secondary hypothyroidism and with normal thyroid hormone levels (T4 and TSH) were included. Patients with a potential secondary osteoporosis risk factor were excluded. BMD was measured using a DEXA scanner if considered clinically appropriate. History of fragility fractures was questioned. Patients were compared with a control comprised of post-menopausal osteoporotic woman without thyroid pathology, using the Mann-Whitney U Test. The level of significance was chosen as < 0.05.

Results: Regarding the intervention group at the time of investigation, the mean replacement dosage of levothyroxine was 0.124±0,088 mcg/day. The mean time of treatment with THR was 9,2±5,7 years. No statistically differences were found regarding age and BMI between both groups (table 1).

Discussion: In this cohort no statistically significant differences were found regarding number of fragility fractures of any site and BMD between euthyroid osteoporotic post-menopausal women treated with THR and osteoporotic pos-menopausal woman without thyroid pathology. Due to the transversal nature of this study no data from previous hormone thyroid status was recorded. This study indicates that THR maintaining a euthyroid state in hypothyroid patients results in no increased bone loss or risk of fracture in post-menopausal women. For robust conclusions, we need large prospective cohort studies.

TABLE

Feature	Intervention Group (n=49)	Control Group (n=164)	p-value
Age	x̄ 76,1±SD 8,6	x̄ 79,6±SD 10,3	0,428
BMI	x̄ 28,27±SD 4,75	x̄ 26,83±SD 4,47	0,207
Number fragility fractures	x̄ 1,32±SD 0,6	x̄ 1,29±SD 1,1	0,135
T score femoral neck	x̄ -3,05±SD 2,1	x̄ -2,17±SD 3,3	0,236
T score lumbar spine	x̄ -2,58±SD 0,9	x̄ -2,80±SD 1,65	0,428

211 - CLINICAL FEATURES AND MANAGEMENT IN OSTEOGENESIS IMPERFECTA

Raquel Freitas[1], Sandra Sousa[1], Fátima Godinho[1]

[1]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal

Background: Osteogenesis imperfecta (OI) is a group of rare inherited disorders of connective tissue with the common feature of bone fragility and recurrent fractures. Extraskeletal manifestations can also occur, such as blue or grey discoloration of the sclera, dentinogenesis imperfecta, hearing loss, cardiac and respiratory involvement. A multidisciplinary approach is needed to optimize management of OI, with input from geneticist, pediatrician, rheumatologists, physiatrist, orthopedic surgeons, otorhinolaryngologists, and other allied health professionals.

Objective: To describe demographic, clinical features and treatments used in OI patients of our center.

Results: In total, 31 patients with OI were included, with 20 (64.5%) female, median age at diagnosis of 2 (IQR of 1-6) years. The most common subtype of OI was type I (77.4%). Clinical features differences were found between type I OI and other types (Table 1). Type I OI had less number of bone fractures (6.5 versus 17.5, p<0.01), vertebral fracture (8.7% versus 50%, p=0.0046), bone deformities (34.8% versus 83.3%, p=0.0046) and need for walking aids (4.3%

TABLE 1 – DEMOGRAPHIC, CUMULATIVE CLINICAL FEATURES AND TREATMENTS USED IN OI.

	Whole cohort N=31	Type I OI N= 24 (77.4%)	Other types OI* N= 7 (22.6%)	P-value
Female sex– N (%) no=31	20 (64.5)	15 (62.5)	5 (71.4)	0.51
Family history – N(%) no=28	21 (75)	18 (85.7)	3 (42.9)	0.043
Age at diagnosis – Median (IQR) no=30	2 (1-6)	2 (1-9.8)	2 (1-5.25)	0.67
Current age – Median (IQR) no=31	28 (20-44)	24 (20.5-44.8)	28 (17-29)	0.87
First DEXA (z-score) – Mean ± SD no=23	-4.57 ± 1.9	-4.29 ± 1.6	-5.36 ± 1.2	0.25
Last DEXA (z-score) – Mean ± SD no= 22	-1.50 ± 1.7	-1.2 ± 1.2	-2.95 ± 2.7	0.29
Number of fractures - Median (IQR) no=28	7.5 (5-14.25)	6.5 (5-10.25)	17.5 (9.75-85)	<0.01
Vertebral fracture – N(%) no=29	5 (16.7)	2 (8.7)	3 (50.0)	0.046
Age of last fracture – Median (IQR) no=28	13 (8-22.75)	13 (7-22)	12 (9.5-46)	0.73
Bone deformities - N(%) no=28	13 (44.8)	8 (34.8)	5 (83.3)	0.047
Need for walking aids – N(%) no=29	6 (20.7)	1 (4.3)	5 (83.3)	<0.01
Discoloration of the sclera – N(%) no=29	19 (65.5)	18 (78.3)	1 (16.7)	0.011
Dentinogenesis imperfecta – N(%) no=26	7 (26.9)	5 (25)	2 (33.3)	0.53
Cardiac involvement – N(%) no=22	3 (13.6)	2 (11.1)	1 (25)	0.47
ENT involvement – N(%) no=27	6 (20.7)	4 (17.4)	2 (33.3)	0.56
Respiratory involvement – N(%) no=22	1 (3.8)	0	1 (25)	0.15
Bisphosphonates – N(%) no=30	27 (90)	20 (87)	7 (100)	0.43
Current bisphosphonates – N(%) no=28	9 (32.1)	5 (22.7)	4 (66.7)	0.06
Strontium ranelate – N(%) no=29	4 (13.8)	2 (8.7)	2 (33.3)	0.18
Denosumab – N(%) no=29	2 (6.9)	1 (4.3)	1 (16.7)	0.38
Teriparatide – N(%) no=29	1 (3.4)	0	1 (16.7)	0.21
Orthopedic surgery – N(%) no=31	19 (61.3)	13 (54.2)	6 (85.7)	0.14
Loss of follow up/Discharge patients – N(%)	14 (45.2)	12 (50)	2 (28.6)	0.29

N - number; no - Number of patients with available information; SD - Standard deviation; IQR - Interquartile range; DEXA – Bone densitometry . P-value comparing type I OI to others type of OI. *Other types OI included 2 patients with type III OI, 1 patient with type IV OI, 1 patient with type V OI, 2 patients with type VI OI and 1 patient with type VII OI).

versus 83.3%, p<0.01). Nevertheless, type I OI had higher rate of discoloration of the sclera (78.3% versus 16.7%, p=0.01). ENT involvement was present in 20.7%, mostly with hearing loss, however routine audiologic evaluation in asymptomatic patients was not always performed. Cardiac involvement was found in 13.6% but many patients did not have an routine echocardiogram or electrocardiogram evaluation. Family history of OI was more prevalent in type I OI (85.7% versus 42.9%, p=0.043). Most patients (90%) were treated with bisphosphonates during follow-up and 32.1% are still currently being treated. Approximately half of patients lost follow-up or were discharged, especially when transitioning to adult consultations and/or remained without fractures for a long period of time.

Conclusion: Management of OI patients is complex and challenging due to being a rare disease with a heterogeneity of clinical features. Best practice requires a multidisciplinary approach and a regular follow-up visit. In our cohort we found less than expected routine evaluation for extraskeletal involvement and higher than expected loss of follow up or discharge. To address and improved this issues we are working in a clinical protocol for evaluation and monitorization of OI patients in a rheumatology consultation.

216 - OUTCOMES IN RHEUMATOID ARTHRITIS PATIENTS UNDER TOCILIZUMAB AS FIRST BDMARD: A REAL-LIFE MONOCENTRIC COHORT STUDY

Frederico Rajão Martins[1], Ana Martins[2], Daniela Santos Oliveira[2, 3], Diogo Guimarães da Fonseca[4], Maria Seabra Rato[2], Filipe Oliveira Pinheiro[2], Salomé

Garcia[2], Bruno Miguel Fernandes[2], Lúcia Costa[2], Miguel Bernardes[2]
[1]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [3]Center for Health Technology and Services Research (CINTESIS),, Faculdade de Medicina da Universidade do Porto, Porto, Portugal, Porto, Portugal, [4]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Background: Rheumatoid arthritis (RA) is one of the most frequent systemic inflammatory rheumatic diseases, being constantly assessed regarding new new therapeutic targets and therapies. Tocilizumab (TCZ) is one of the latest biological disease-modifying antirheumatic drugs (bDMARDs) approved for RA's treatment, usually as a second line agent in daily clinical practice.
Objectives: Evaluate the different disease and patient reported outcomes in patients undergoing treatment with tocilizumab as the first biologic therapy.
Methods: All patients with a definite RA diagnosis who had undergone treatment with TCZ as the first biologic therapy were included in this analysis. Diverse socio-demographic, disease and patient-related data were assessed at baseline, 6 and 12 months of treatment with TCZ, and posteriorly extracted from the Portuguese register of rheumatic diseases (Reuma.PT). Statistical analysis included non-parametric tests such as Wilcoxon test and univariate analysis using linear and logistic regression models.
Results: Fifty-one patients were included, 88.2% females, with a median age at introduction of TCZ of 53.5 +/- 10.4 years; mainly seropositive for either rheumatoid factor (66%) or anti citrullinated peptide antibody (ACPA; 68%), with an erosive disease (75.6%) and concomitantly treated with a conventional synthetic disease modifying anti-rheumatic drug (csDMARD) (70.5%). During follow-up there was a statistically significant reduction at 6 and 12 months of TCZ treatment regarding DAS28 (4 variables) (4v) and DAS28(4V)-CRP scores ($p < 0.001$), SDAI ($p < 0.001$), CDAI ($p < 0.001$), 68/66 tender and swollen joint counts (TJC/SJC) ($p < 0.001$), ESR and CRP ($p < 0.001$), patient and physician VAS ($p < 0.001$) and HAQ score ($p = 0.01$ at 6 months and $p < 0.001$ at 12 months). A majority of patients showed good EULAR response at 6 (52.6%) and 12 (56.3%) months, as well as moderate to high mean improvement in ACR core set measures at 6 (53.3±22.7) and 12 (54.3±25.2) months. Seventy-five percent of patients remained under tocilizumab with an average treatment duration of 48.8±37.7 months. There was a significant reduction in DAS28(4V), DAS28(4V)-CRP, CDAI, SDAI, TJC and SJC, ESR, CRP, patient and physician VAS and HAQ scores between 6 and 12 months of therapy ($p < 0,001$). ACR and EULAR responses didn't differ significantly between assessments at 6 and 12 months. In the absence of a representative number of RA patients on TCZ monotherapy, it wasn't possible to draw conclusions about the need to use combined therapy with a csDMARD for better clinically significant response. A higher degree of ACR response at 6 months was associated with higher serum rheumatoid factor levels (OR 1.13, $p < 0.05$) at baseline, while a lower degree of response was seen with higher TJC ($p = 0.05$) and HAQ score ($p < 0.01$). ACR response at 12 months was lower in patients with erosive disease at baseline ($p < 0.05$). Regarding EULAR response criteria at 6 months, there was a negative association with higher TJC ($p < 0.05$), while at 12 months the negative trend was associated with ESR levels ($p < 0.05$) and HAQ scores ($p < 0.05$) at baseline.
Conclusion: There seems to be evidence of good therapeutic response to TCZ in bDMARD naïve RA patients assessed at 6 months from baseline, without evidence of significant improvement of response measures further down the line. Basal serum rheumatoid factor levels, TJC, HAQ scores and the presence of erosive disease may have some predictive value on the therapeutic response.

220 - PERSISTÊNCIA DOS SINTOMAS DE INFEÇÃO POR SARS-COV-2 EM DOENTES REUMÁTICOS – PERSPECTIVA DE UM CENTRO

Ana Lúcia Fernandes[1], Nathalie Madeira[2], Cândida Silva[2], Susana Fernandes[2], Luís Cunha-Miranda[2], Cláudia Miguel[2], Maria Jesus Mediavilla[2], Helena Santos[2], Vera Las[2], Manuela Micaelo[2], Manuela Parente[2], Joana Borges[2], José Vaz Patto[2]
[1]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, [2]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal

Introdução: Face à pandemia por SARS-CoV-2 (COVID-19), nas doenças reumáticas, tendo em conta a disfunção subjacente do sistema imunitário e/ou o uso de imunossupressores (no caso das doenças

inflamatórias sistémicas), existe uma preocupação adicional acerca dos seus outcomes.

Um dos aspetos a ter em consideração, consiste na persistência dos sintomas de COVID-19, após a sua aparente resolução.

Objetivo principal: Descrever a persistência de sintomas de SARS-CoV-2 dos doentes reumáticos.

Métodos: Trabalho descritivo, single-centre (Instituto Português de Reumatologia). Foram selecionados doentes com doenças reumáticas inflamatórias (DRI) e não inflamatórias (DRNI) constantes do Reuma.pt, com infeção confirmada de SARS-CoV-2, ocorrida e resolvida entre Março e Novembro de 2020.

Resultados: Foram incluídos 17 doentes (n=17), 8 do sexo masculino (47%) e 9 do sexo feminino (53%); a média de idades foi 60,9 anos (mín=36; máx=77); 12 doentes tinham DRI (71%) e 5 DRNI (29%); a maioria dos doentes (n=12; 71%) tinha comorbilidades associadas.

Os principais diagnósticos foram espondilartrite axial e periférica (n=4; 24%) e osteoartrose (n=4; 24%); as restantes patologias foram representadas por 1 doente cada (6%): artrite reumatóide (AR) seropositiva, AR seronegativa, lúpus eritematoso sistémico, dermatomiosite, doença indiferenciada do tecido conjuntivo, espondilartrite associada a doença inflamatória do intestino (Chron), sarcoidose e espondilartrose. Dentro das DRI, a maioria dos doentes (n=6; 50%) encontrava-se em remissão, 5 (42%) em baixa actividade e 1 (8%) doente com atividade moderada.

Do total, 4 doentes (24%) estiveram internados, apenas 1 (6%) com DRI. Dentro das DRI, 11 doentes (92%) tiveram sintomas no início da infeção: os mais frequentes (n=6) foram fadiga, mal-estar geral, tosse e disgeusia; no grupo das DRNI, todos os doentes (n=5; 100%) tiveram sintomas, sendo o mais frequente a fadiga (n=4).

Ao fim de 2 meses, apenas 2 doentes (12%), 1 com DRI e 1 com DRNI, mantiveram sintomas da infeção prévia, nomeadamente anósmia, disgeusia e xerostomia.

Todos os 17 doentes (100%) tiveram como desfecho cura sem sequelas, não sendo aqui incluído a persistência de sintomas.

Discussão e conclusão: no presente trabalho foi encontrada uma baixa prevalência de sintomas, após 2 meses de infeção por SARS-CoV-2. Carfì A et al1 publicaram um estudo case-series com 143 doentes onde mostraram que, em média 60.3 dias (DP 13.6) após o início do primeiro sintoma, 87.4% dos doentes apresentava a persistência de pelo menos 1 sintoma, particularmente fadiga e dispneia. Townsend L et al2 também demonstraram a persistência significativa de fadiga, após a fase aguda da doença, em indivíduos com infeção anterior por SARS-CoV-2. Uma hipótese possível para estas situações será uma exacerbação pós infeciosa de comorbilidades pré-existentes, ainda que subclínicas ou latentes. Até à data, desconhece-se ainda se o designado "Long-Covid" consiste numa ou várias entidades distintas, em adição aos danos sequelares já conhecidos desta patologia. Estudos com maior robustez científica poderão tentar esclarecer com maior precisão a prevalência destes sintomas e a sua eventual causa.

REFERÊNCIAS

1. Carfì A, Bernabei R, Landi F. Persistent Symptoms in Patients After Acute COVID-1. JAMA. 2020 Aug 11;324(6):603-605. doi: 10.1001/jama.2020.12603.
2. Towsen L, Dyer, AH, Jones K et al. Persistent fatigue following SARS-CoV-2 infection is common and independent of severity of initial infection. PLoS One. 2020 Nov 9;15(11): e0240784. doi: 10.1371/journal.pone.0240784. eCollection 2020.

227 - PSORIATIC DISEASE – A HETEROGENOUS DISEASE WHO NEEDS A MULTIDISCIPLINARY CARE!

Bernardo Santos[1], Joana Antão[2], Manuela Loureiro[3], Anabela Barcelos[1]

[1]Serviço de Reumatologia, Centro Hospitalar do Baixo Vouga, E.P.E., Aveiro, Portugal, [2]Master´s Student in Medical Statistics, Universidade de Aveiro, Aveiro, Portugal, [3]Dermatologia, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal

Introduction: Rheumatologists and Dermatologists usually manage Psoriatic Arthritis (PsA) and Psoriasis (PsO) separately, but early diagnosis and integrated management could achieve better outcomes with gains in quality of life of the patients. A multidisciplinary care is essential to achieve these goals.

Objectives: The aim of this study is to describe a model of integrated multidisciplinary approach for early diagnosis and management of PsA patients.

Methods: A retrospective study including patients that attended the multidisciplinary clinical from January 2019 to December 2020 was performed. Patients with PsO complaining articular symptoms and patients with articular symptoms with cutaneous lesions suspected to be PsO was referred to this clinical. Demographic, clinical data and disease activity measures were collected. Descriptive, Student's t and Fisher test were estimated.

TABLE 1 - SOCIODEMOGRAPHIC AND CLINICAL CHARACTERIZATION OF THE PATIENTS DIAGNOSED WITH PSORIATIC DISEASE AFTER EXPERT EVALUATION

	Psoriatic Disease N=40
Age (years) [a]	49.5 (37; 63.50)
Sex, n (%)[b]	
Female	18 (45.0)
Male	22 (55.0)
Psoriaisis FH, n (%)[b]	19 (47.5)
Spondyloarthropathy FH, n (%)[b]	7 (17.5)
Tobacco use, n (%)[b]	4 (10.0)
Nourishment, n (%)[b]	
Normal	7 (20.0)
Overweight	13 (37.1)
Obesity Class I	10 (28.6)
Obesity Class II	5 (14.3)
Comorbidities, n (%)[b]	
Diabetes Mellitus	3 (7.5)
Hypertension	10 (25.0)
Dyslipidemia	9 (22.5)
Metabolic Syndrome	6 (15.0)
Depression	9 (22.5)
Current Treatment, n (%)[b]	
Topical	33 (82.5)
csDMARDs	23 (57.5)
bDMARDs	13 (32.5)
adalimumab	4 (10.0)
golimumab	1 (2.5)
sekucinumab	5 (12.5)
ustekinumab	3 (7.5)
Treatment plan, n (%)[b]	
Mantaining	17 (42.5)
Starting	20 (50)
csDMARDs	9 (22.5)
bDMARDs	11 (27.5)

[a] Data showed as median and inter Quartile Range (P25, P75)
[b] Data showed as frequency and percentage

Results: A total of 50 patients were referred to multidisciplinary clinical. In 40 patients were confirmed the diagnosis of PsO. Twenty-two (55%) were male with a median age of 49.5 ± 11.5 years. Family history of psoriasis was present in 19 (47.5%) and 7 (17.5%) had spondyloarthritis family history. Obesity and overweight were the comorbidities most

TABLE 2 – CLINIMETRY OF PATIENTS WITH PSORIATIC DISEASE

	Psoriatic Disease N=40
Phenotype, n (%)[b]	
Skin psoriasis	9 (22.5)
Articular involvement	31 (77.5)
CASPAR criteria	31 (100)
PASI, n (%)[b]	
Mild	27 (67.5)
Moderate	5 (12.5)
Severe	8 (20.0)
PROMs [a]	
DLQI	7 (3.75; 12.25)
APsQol	6 (4; 12.5)
HAQ	0.69 (0; 1.28)
5Q-5D	0.39 (0.28; 0.65)
	Psoriatic Arthritis N=31
Duration arthritis (years), n (%)[b]	
No previous diagnosis	1 (3.2)
<5	6 (19.4)
5-10	5 (16.1)
>10	19 (61.3)
Duration skin (years), n (%)[b]	
No previous diagnosis	1 (3.2)
<5	2 (6.5)
5-10	5 (16.1)
>10	23 (74.2)
Articular involvement, n (%)[b]	
Axial	0 (0.0)
Peripheral	28 (90.3)
Both	3 (9.7)
Extra-articular features, n (%)[b]	
Enthesitis	6 (19.4)
Dactylitis	7 (23.3)
Hands	1 (3.3)
Feet	6 (20.0)
Uveities	1 (2.5%)
BASDAI [a]	3.73 (3.73; 5.19)
BASFI [a]	2.13 (2.13; 4.72)
DAS-28, n (%)[b]	
Active disease	8 (25.8)
Moderate	7 (22.5)
Low	10 (32.2)
Remission	6 (19.3)

[a] Data showed as median and inter Quartile Range (P25, P75)
[b] Data showed as frequency and percentage

TABLE 3 - SOCIODEMOGRAPHIC AND CLINICAL DATA OF PATIENTS WITH AND WITHOUT PSORIATIC ARTHRITIS.

	PsA N=31	PsO N=9	p-value
Age (years)	51.42 ± 11.12	42.89 ± 11.01	0.047[a]
Sex, n (%)			
Female	16 (51.6)	2 (22.2)	0.148[b]
Male	15 (48.4)	7 (77.8)	
Psoriasis in relatives, n (%)			
Yes	14 (45.2)	5 (55.6)	0.711[b]
No	17 (54.8)	4 (44.4)	
Spondyloarthropathy in relatives, n (%)			
Yes	7 (77.4)	0	0.174[b]
No	24 (22.6)	9 (100)	
Tobacco			
Yes	2 (6.5)	2 (22.2)	0.213[b]
No	29 (93.5)	7 (77.8)	
Nourishment, n (%)			
Normal	5 (16.1)	2 (22.2)	0.732[b]
Overweight	11 (35.5)	2 (22.2)	
Obesity Class I	8 (25.8)	2 (22.2)	
Obesity Class II	3 (9.7)	2 (22.2)	
Comorbilities, n (%)			
Diabetes Mellitus			
Yes	2 (6.5)	1 (11.1)	0.545[b]
No	29 (93.5)	8 (88.9)	
Hypertension			
Yes	9 (29.0)	1 (11.1)	0.404[b]
No	22 (71.0)	8 (88.9)	
Dyslipidemia			
Yes	6 (19.4)	3 (33.3)	0.394[b]
No	25 (80.6)	6 (66.7)	
Depression			
Yes	13 (41.9)	0	0.049[b]
No	18 (58.1)	9 (100)	

a T-test student
b Fisher test

found, 42.9% and 37.1%, respectively, followed by hypertension 25%, dyslipidaemia and depression 22.5%. Thirteen patients (32.5%) presented moderate to severe PASI. Thirty-one patients (77.5%) met criteria for PsA according CASPAR criteria. Prolonged cutaneous and articular disease (>10 years duration) was found in 74.2% and 61.3% patients, respectively. Peripheral disease without axial involvement was present in 90.3%. The most frequent extra-articular manifestation was dactylitis (23.3%) and enthesitis (19.4%). Severe or moderate articular disease activity was present in 48.3% of the patients.

Comparing diagnosed PsO and PsA patients there was a statistically significant difference at the mean age and the presence of depression (p= 0.047 and p=0.049, respectively); there was no statistically significant differences in family history of PsO (p=0.711) and spondyloarthritis (p=0.174), nutritional status (p=0.732) and comorbidities such as diabetes mellitus (p=0.545), hypertension (p=0.404), dyslipidaemia (p=0.394) and depression (p=0.089). In our multivariated logistic regression model we found female gender (p=0.016) and prolonged articular disease (>10 years durations) (p= 0.039) as risk factors for the development of severe psoriatic arthritis.

Conclusions: Despite the small number of patients observed in our multidisciplinary clinical, we found that this multidisciplinary care may identify disease in an early stage and offers a more comprehensive treatment approach to an heterogenous and unpredictable disease.

REFERENCES

1. Luchetti MM, Benfaremo D, Campanati A, Molinelli E, Ciferri M, Cataldi S, Capeci W, Di Carlo M, Offidani AM, Salaffi F, Gabrielli A. Clinical outcomes and feasibility of the multidisciplinary management of patients with psoriatic arthritis: two-year clinical experience of a dermo-rheumatologic clinic. Clin Rheumatol. 2018 Oct;37(10):2741-2749
2. Theodorakopoulou E, Dalamaga M, Katsimbri P, Boumpas DT, Papadavid E. How does the joint dermatology-rheumatology clinic benefit both patients and dermatologists? Dermatol Ther. 2020 May;33(3):e13283

228 - DRIVING FACTORS FOR THE UTILISATION OF HEALTHCARE SERVICES BY PEOPLE WITH OSTEOARTHRITIS: RESULTS FROM A NATIONWIDE POPULATION-BASED STUDY

Costa, D[1, 2, 3], Rodrigues AM[2, 3], Cruz, EB[2, 4], Helena Canhão[2, 3], Jaime C. Branco[2, 3, 5], Nunes, C[1, 2]

[1]Public Health Research Centre, NOVA National School of Public Health, Public Health Research Centre, Universidade NOVA de Lisboa, Portugal, Lisboa, Portugal, [2]Comprehensive Health Research Center (CHRC), Universidade NOVA de Lisboa, Portugal, Lisboa, Portugal, [3]CEDOC, EpiDoC Unit, NOVA Medical School, Universidade NOVA de Lisboa, Lisboa, Portugal, [4]Fisioterapia, Escola Superior de Saúde - Instituto Politécnico de Setúbal, Setúbal, Portugal, [5]Rheumatology department,, Hospital de Egas Moniz, Centro Hospitalar de Lisboa Ocidental, Lisboa, Portugal

Background: Worldwide, the current management of knee osteoarthritis (K-OA) seem segmented, heterogeneous, high-cost and often not based on current best evidence. The absence of epidemiological data regarding the utilisation of healthcare services may conceal the need for improvements in the management of K-OA. The aim of this study is to explore the profiles of healthcare services utilisation by people with K-OA and to analyse their determinants, according to Andersen's behavioural model.
Methods: We analysed a sample of 978 participants diagnosed with K-OA from the population-based study EpiReumaPt. Sociodemographic, health-related and utilization of health services data was collected with a structured interview, and the diagnosis of K-OA was validated by a rheumatologist team. With the Two-step Cluster procedure, we primarily identified different profiles of healthcare utilisation according to the services most used by patients with K-OA in different settings: general practitioner, physiotherapy and orthopaedic surgeon appointments, and hospitalization. Secondly, we analysed the determinants of each profile membership, using multinomial logistic regression, according to the predisposing characteristics, enabling factors and need variables ($p<0.005$; 95%CI).
Results: We identified three profiles of healthcare utilisation: "HighUsers"; "GPUsers"; "LowUsers". "HighUsers" represents more than 35% of the sample, and are also the participants with higher utilisation of overall medical appointments (4.43±6.65, $p<0.001$). "GPUsers" represent the participants with higher utilisation of general practitioner appointments. Low users was used as the reference profile in the multinomial regression model. The determinants associated with HighUsers profile membership were: younger age (OR=0.96, 95%CI 0.95, 0.99) and to live in Portugal Mainland (OR=0.43, 95%CI: 0.24, 0.77) as predisposing characteristics; to have additional health coverage (OR=0.65, 95%CI 0.43, 0.98) and to be employed (OR=0.55, 95%CI 0.31-0.97) as enabling factors; have higher number of comorbidities (OR=1.12, 95%CI 0.31, 0.97), worse quality of life (OR=0.33, 95%CI 0.14, 0.79), worse physical function (OR=1.59, 95%CI 1.10-2.23) and have no regular physical exercise (OR=0.57, 95%CI 0.37, 0.88) as need variables.

Moreover, the determinants associated with GPUsers profile membership were: to live in the centre region (OR=2.11, 95%CI 1.21, 3.68) or in Portugal Mainland (OR=0.42, 95%CI 0.21, 0.83), as predisposing factors; have higher number of comorbidities (OR=1.22. 95%CI 1.11, 1.33), presence of anxiety symptoms (OR=1.09, 95%CI 1.03, 1.14) and no regular physical exercise (OR=0.55 95%CI 0.34, 0.89), as need variables. Need variables accounted for the higher variation in AUC and McFadden pseudo-R2, however predisposing characteristics and enabling factors remained significative in the final multinomial models.
Conclusions: Healthcare utilisation by people with knee osteoarthritis is not driven only by clinical needs. The predisposing characteristics and enabling factors associated with healthcare utilisation reveal inequities in the access to healthcare and variability in the management of people with knee osteoarthritis. Research and implementation of whole-system strategies to improve equity in the access and quality of care are paramount in order to diminish the impact of osteoarthritis at individual-, societal- and economic-level.

230 - IMPAIRED BONE HEALTH IN PATIENTS WITH PRIMARY SJOGREN'S SYNDROME IN A SMALL RHEUMATOLOGY CENTER

Pedro Abreu[1, 2], Marina Oliveira[2]
[1]Rheumatology Department, Unidade Local de Saúde de Castelo Branco, Castelo Branco, Portugal, [2]Faculdade de Ciências da Saúde, Universidade da Beira Interior, Covilhã, Portugal

Introduction: Primary Sjögren's syndrome is a chronic and slowly progressive autoimmune disorder. Scientific evidence has shown that patients with primary Sjögren's Syndrome have a higher prevalence of reduced bone mineral density. Therefore, these patients are more likely to develop osteopenia and osteoporosis and, consequently, fragility fractures.
Objectives: To determine the prevalence of reduced bone mineral density in patients diagnosed with primary Sjögren's Syndrome followed at the Rheumatology department of the Local Health Unit in Castelo Branco and investigate whether there is an association between patients' clinical features with known risk factors for osteopenia and osteoporosis.
Methods: This observational and retrospective investigation studied all patients diagnosed with primary Sjögren's Syndrome followed at the Rheumatology department of the Local Health Unit in Castelo Branco. Bone mineral density assessment was obtained by osteodensitometry and the diagnosis of osteopenia and osteoporosis was defined according to the World Health

Organization criteria. Patients' clinical profile is analyzed and it is investigated the influence of age, years of disease duration, body mass index and use of corticosteroids in bone loss, by logistic regression.
Results: A total of 38 patients were enrolled in the study. Women to men ratio is 9:1. The mean age is 60,82 years. The mean duration of primary Sjögren's Syndrome is 4,76 years. Corticosteroids are used with a frequency of 70,3%. 57,9% of individuals are overweight or obese. The prevalence of reduced bone mineral density is 34,3%. No statistically significant association was found (p-value>alpha) between the considered variables and bone mineral density, meaning that it is not influenced by age, years of disease duration, body mass index or use of corticosteroids.
Conclusion: Approximately one third of patients with primary Sjögren's Syndrome have reduced bone mineral density. This is not associated with age, years of duration of the primary Sjögren's syndrome, body mass index or use of corticosteroids.

231 - DETERMINANTS OF CLINICAL AND RADIOGRAPHIC SEVERITY IN PEOPLE WITH HIP OR KNEE OSTEOARTHRITIS: A CROSS-SECTIONAL POPULATION-BASED STUDY

Costa, D[1, 2, 3], Nunes, C[1, 2], Cruz, EB[2, 4], Rute de Sousa[2, 3], Helena Canhão[2, 3], Jaime C. Branco[2, 3, 5], Rodrigues AM[2, 3]
[1]Public Health Research Centre, NOVA National School of Public Health, Universidade NOVA de Lisboa, Portugal, Lisboa, Portugal, [2]Comprehensive Health Research Center (CHRC), Universidade NOVA de Lisboa, Portugal, Lisboa, Portugal, [3]EpiDoc Research Unit, CEDOC, NOVA Medical School, Lisboa, Portugal, [4]Fisioterapia, Escola Superior de Saúde - Instituto Politécnico de Setúbal, Setúbal, Portugal, [5]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal

Background: Osteoarthritis (OA) a leading and multifactorial cause of disability, being clinical and radiographic severity associated with individual and socio-economic burden. The aim of this study is to estimate the prevalence of hip and/or knee osteoarthritis (HKOA) in Portugal, characterise the population according to clinical and radiographic severity profiles and identify their determinants.
Methods: We analysed a representative sample of 1081 participants with validated diagnosis of HKOA, from the population-based study EpireumaPt. Data were collected in a three-stage approach: 1) structured interview, when sociodemographic, lifestyle and health-related data were collected; 2) clinical appointment with a rheumatologist; 3) diagnosis validation by a team of rheumatologists. The potential determinants were defined as the sociodemographic and anthropometric, health-related and lifestyle variables, including anxiety and depression, measured with the Hospital Depression and Anxiety Scale. The outcomes of this study are the clinical and radiographic severity, both collected in the clinical appointment. Clinical severity was measured with the Knee Injury and Osteoarthritis Outcome Scale (KOOS) and with the Hip Disability and Osteoarthritis Outcome Scale (HOOS), and categorized by tertiles in worse, middle and better. Radiographic severity was measured with Kellgren-Lawrence classification as mild, moderate and severe. We estimated weighted prevalence of HKOA by age class and sex. The determinants associated with clinical and radiographic severity were analysed with ordinal logistic regression ($p<0.05$, 95%CI). The analysis was performed with SPSS 26.0 complex samples.
Results: HKOA affects 14.1% (95%CI: 12.6, 15.7) of the Portuguese adults, increasing in people ≥55 years old, and in women. Most of people have mild (48.5%) radiographic severity. People with worse clinical severity are older adults (64.39±0.70), women (75.2%), low educated (38.1%), with overweight (39.0%) or obesity (45.9%), with high comorbidities count (3,65±2.04), and with heterogeneous radiographic severity. The determinants associated with worse clinical severity are: to have 55-64 years old (OR=2.13 95%CI: 1.29, 3.80), higher comorbidities count (OR=1.16 95%CI: 1.06, 1.27;), temporarily disabled (OR=8.28 95%CI: 2.14, 32.04) or retired (OR=2.95 95%CI: 1.34, 6.46), overweight (OR=2.11; 95%CI: 1.18, 3.78), or obesity (OR=2.90; 95%CI: 1.60, 5.25), and depression symptoms (OR=1.07 95%CI: 1.02, 1.12). Drinking alcohol (OR=0.66 95%CI: 0.44, 0.97), and to have primary/basic education (OR=0.52; 95%CI: 0.33, 0.80) or secondary/superior education (OR=0.35 95%CI: 0.20; 0.61) are inversely associated with worse clinical severity. The determinants associated with worse radiographic severity are: to have 65-74 years old (OR=3.67 95%CI: 1.50, 9.03) or ≥75 years old (OR=3.15 95%CI: 1.22, 8.14), and worse tertile of HOOS/KOOS5 (OR=5.19 95%CI: 2.72, 9.90). Female sex (OR=0.65 95%CI: 0.42; 1.00) and to live in Lisbon (OR=0.23 95%CI: 0.11, 0.47) or in the Centre (OR=0.35 95%CI: 0.20, 0.62) are inversely associated

with worse radiographic severity.
Conclusion: HKOA is highly prevalent in Portugal, as well as risk factors associated with the severity of the disease. This study raise concerns on the need for effective preventive and management strategies to avoid HKOA progression and the escalating of individual and socio-economic burden.

232 - OSTEOPOROSE EM DOENTES COM ARTRITE REUMATÓIDE: DEXA E A AVALIAÇÃO DE CUMPRIMENTO DAS GUIDELINES NUMA UNIDADE DE REUMATOLOGIA

Pedro Abreu[1, 2], Sara Vida[2]
[1]Rheumatology Department, Unidade Local de Saúde de Castelo Branco, Castelo Branco, Portugal, [2]Faculdade de Ciências da Saúde, Universidade da Beira Interior, Covilhã, Portugal

Introdução: A osteoporose (OP) caracteriza-se pela redução da densidade mineral óssea, alteração da microarquitetura e resistência ósseas e aumento do risco de fraturas vertebrais e não-vertebrais. A artrite reumatóide (AR) e a terapêutica prolongada com corticóides são fatores de risco para a OP. O exame utilizado, para o diagnóstico da OP é a absorciometria radiológica de dupla energia (DEXA), que quantifica a densidade mineral óssea. Apesar de não existir um consenso universal, segundo as guidelines da NOF, Associação Americana de Endocrinologistas e da Direção Geral de Saúde, em doentes com AR deve-se proceder à realização periódica da DEXA, com um intervalo de 24 meses. Estas indicações têm como objetivo o diagnóstico precoce da OP e o acompanhamento da evolução após instituição da terapêutica.
Objetivos: Identificar os doentes com AR e OP da Unidade de Reumatologia do Hospital Amato Lusitano. Avaliar o cumprimento das guidelines quanto à realização de DEXA: inicial e repetição ao fim de 24 meses.
Métodos: Estudo retrospectivo, englobando os utentes seguidos na Unidade de Reumatologia do Hospital Amato Lusitano, com o diagnóstico de AR codificado entre março de 2011 e dezembro de 2017. Foram incluídos 118 doentes e os dados foram recolhidos através da consulta dos processos clínicos. A análise estatística foi realizada com recurso ao Statistical Package for the Social Sciences (SPSS), versão 25, com um nível de significância α definido a 0.05.
Resultados: Amostra de 118 doentes com AR, dos quais 33,1% com diagnóstico de OP. A suplementação com cálcio e/ou Vitamina D foi o tratamento mais prescrito em 86,4% (n=102) dos doentes, seguido dos bifosfonatos em 29,7% (n=35) e desonumab em 4,2% (n=5) dos doentes. A maioria dos doentes realizou DEXA à data do diagnóstico (78,0%; n=92), mas esta só foi repetida em 28,0% (n=33) dos casos. O tempo médio para a repetição da DEXA foi de 50,9 meses e apenas 5,1% repetiram até 2 anos.
Conclusão: As guidelines para realização de DEXA não estão a ser alcançadas. Os doentes que não são respondedores ao tratamento anti-osteoporótico poderão não estar a ser identificados e, com isso, poderão incorrer num risco aumentado de fractura. Os resultados deste trabalho permitirão aumentar a vigilância daqueles doentes sobre a necessidade de repetição do exame de forma que o tratamento anti-osteoporótico seja eficaz. Será importante entender os motivos por detrás desta situação e tentar arranjar soluções.

236 - O IMPACTO DA OBESIDADE NA AVALIAÇÃO DAS ENTESES NOS DOENTES COM ESPONDILARTRITE - UM ESTUDO DE VIDA REAL

Filipe Oliveira Pinheiro[1], Pedro Madureira[1], Maria Seabra Rato[1], Diogo Guimarães da Fonseca[2], Salomé Garcia[1], Bruno Miguel Fernandes[1], Ana Martins[1], Daniela Santos Oliveira[1, 3], Frederico Rajão Martins[4], Miguel Bernardes[1, 5], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [3]Center for Health Technology and Services Research (CINTESIS),, Faculdade de Medicina da Universidade do Porto, Porto, Portugal, Porto, Portugal, [4]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [5]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Introdução: A entesite é uma caraterística clássica da espondilartrite e a sua avaliação pode ser realizada recorrendo a diversos índices, como o Maastrich Ankylosing Spondylitis Enthesitis Score (MASES), o Leeds Enthesitis Index (LEI) e o Spondyloarthritis Research Consortium of Canada Enthesitis Index (SPARCC). Apesar de haver alguns dados relativos ao papel da obesidade na avaliação de atividade da espondilartrite, sobretudo em doentes sob antagonistas de TNF-alfa, pouco se conhece acerca do impacto

na avaliação das enteses. O objetivo deste estudo é determinar se a obesidade se associa a uma atividade mais elevada ao nível das enteses nos doentes com espondilartrite.

Materiais e Métodos: Estudo transversal que avaliou doentes com diagnóstico de espondilartrite, registados na plataforma Reuma.pt, sob terapêutica biotecnológica, aquando da sua primeira avaliação relativa ao DMARD realizado atualmente. Foram avaliados dados demográficos, clínicos e laboratoriais. Foram efetuadas análises de correlação de Pearson entre índices de entesite e valores de Índice de Massa Corporal (IMC), comparação entre grupos de obesos (IMC >30) e não-obesos e índices de entesite (Mann-Whitney U), e análises de regressão logística para identificar preditores de índices de entesite mais elevados.

Resultados: Foram incluídos neste estudo 273 doentes, 123 dos quais (45,1%) do género feminino. Sessenta e quatro (23,4%) eram fumadores e 45 (16,5%) referiam consumo etílico durante a maior parte/todos os dias da semana. Relativamente aos DMARDs realizados, 57 (23,8%) faziam golimumab, 54 (22,6%) adalimumab, 52 (21,8%) infliximab, 45 (18,8%) etanercept, 18 (7,5%) certolizumab, 11 (4,6%) secucinumab e 2 (0,8%) ustecinumab. Apresentavam uma média de idades de 49,0±10 anos, com IMC 23,9±4,1 kg/m2, LEI mediano de 0±2, SPARCC mediano de 1±4 e MASES mediano de 1±4, com VS e PCR medianas de 30±32 mm/1ªh e 1,4±2,2 mg/L, respetivamente. Verificou-se uma correlação moderada entre IMC e MASES (0.428, p<0,001) e SPARCC (0.328, p=0.005), não tendo sido obtida correlação estatisticamente significativa com LEI. Os doentes com obesidade obtiveram avaliações de entesite mais elevadas em todos os índices utilizados – MASES 2±5 vs 0±1, p<0.001, SPARCC 2±5 vs 0±2, p=0.001, e LEI 0±2 vs 0±1, p=0.049. As análises de regressão logística permitiram determinar que a obesidade se associa a MASES mais elevados (OR 1.638, IC95% 1.225-2.191, p=0.001), de forma independente da idade, sexo, VS e PCR, mantendo-se a significância estatística quando se ajusta para aquelas variáveis (OR 1.665, IC95% 1.315-2.108, p<0.001). Essa relação não foi obtida para os outros índices.

Conclusão: Nesta amostra verificou-se que a obesidade se associa a índices de entesite mais elevados nos doentes com espondilartrite, sobretudo MASES, de forma independente da idade, sexo e parâmetros inflamatórios. Os autores realçam, a este propósito, o papel importante que a ecografia musculoesquelética pode ter na avaliação do envolvimento das enteses nos doentes com espondilartrite, mormente nos doentes obesos.

238 - CRENÇAS ACERCA DA MEDICAÇÃO NUMA POPULAÇÃO DE DOENTES COM ARTROPATIA INFLAMATÓRIA

Ana Margarida Correia[1], Emanuel Costa[1], Diogo Esperança Almeida[1], Joana Leite Silva[1], Joana Sousa-Neves[1], Ana Ribeiro[1], Marcos Cerqueira[1]
[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

Introdução: As crenças que os doentes apresentam relativamente à medicação, nomeadamente acerca da sua necessidade e às preocupações relativas a possíveis efeitos adversos, são um fator determinante para a adesão e o sucesso terapêutico.

Objetivos: Numa população de doentes com Artrite Reumatóide (AR) ou Artrite Psoriática (AP), determinar que fatores sociodemográficos e clínicos se associam a crenças relacionadas com a medicação, quer atendendo à necessidade, quer atendendo às preocupações com a mesma.

Metodologia: Estudo observacional, com uma amostra constituída por 58 doentes (sexo feminino 72,4%, masculino 27,6%; idade média = 51,0 ± 13,2 anos; escolaridade < 9 anos 39,7%, > ou = 9 anos 60,3%). Destes, 33 tinham diagnóstico de AR e 25 AP (média de anos de evolução da doença = 8,2 ± 7,3 anos; média da atividade de doença pelo DAS28-PCR ou DAS28-PCR 3 variáveis = 2,32 ± 0,95). Entre março e junho de 2021, foi aplicado um breve questionário com variáveis sociodemográficas e clínicas, como o uso de DMARD´s clássicos ou biológicos e a sua via de administração. Foi aplicada, de forma anónima, a Escala de Crenças Específicas Sobre a Medicação (BMQ), adaptada do Beliefs About Medicines Questionnaire, composta por 2 secções: secção 1 - Necessidades Específicas, crenças dos doentes acerca da necessidade da medicação; secção 2 – Preocupações Específicas, crenças relacionadas com os perigos de dependência e toxicidade ou efeitos secundários a longo prazo. Pontuações mais altas (escala de 5 a 25 pontos), na secção 1 e 2, refletem a crença de maior necessidade e de maior preocupação relativamente à medicação, respetivamente. A média de pontuações na secção 1 e na secção 2 da BMQ foi de 9 e 15,7 pontos, respetivamente. Estatística: descritiva, Teste t-Student, Correlação de Pearson e ANOVA, p<0,05.

Resultados: Doentes com AP e com idade mais jovem apresentaram uma pontuação média mais elevada na

secção 2 da BMQ (p= 0,049 e p<0,01, respetivamente). As variáveis sexo masculino, escolaridade ³ 9 anos e menor tempo de evolução da doença mostraram também uma tendência à associação com a pontuação na secção 2 da BMQ, com valores p próximos da significância estatística (p=0,091, p=0,074 e p=0,094, respetivamente). Na análise multivariada com as variáveis anteriores, apenas a patologia revelou associação estatisticamente significativa com a pontuação na secção 2 da BMQ (B=4,269; p<0,001; IC 95%=[2,060-6,477]). Verificou-se ainda uma correlação estatisticamente significativa (p=0,007) entre as pontuações na secção 1 e na secção 2 da BMQ. Na análise separada da amostra por patologia, verificou-se, nos indivíduos com AR, uma associação entre a via de administração subcutânea dos fármacos e a maior preocupação com os efeitos adversos da medicação (p=0,007), e entre a baixa atividade de doença e maior crença na necessidade específica de medicação para a doença (p=0,006).

Conclusão: Neste estudo, os doentes com AP mostraram maior preocupação com os possíveis efeitos adversos da medicação do que os doentes com AR; tal associação verificou-se mesmo após análise multivariada com outras variáveis com associação a essa secção da BMQ. Os doentes com AR que fazem fármacos subcutâneos têm mais receio do potencial tóxico dos fármacos, e aqueles com mais baixa atividade de doença têm maior necessidade de cumprir a terapêutica prescrita, o que pode ser explicado pela eficácia prévia da medicação no controlo da doença. Verificou-se ainda que, quanto maior é a crença na necessidade de medicação, maior é a preocupação com os seus possíveis efeitos nocivos a longo prazo.

244 - DOENÇAS REUMÁTICAS NA GRAVIDEZ: CASUÍSTICA DE UM CENTRO

Rafaela Nicolau[1], Nádia Martins[1], Maria Inês Seixas[1], Couto M[1], Paulo Monteiro[1]

[1]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Introdução: A consulta pré-concecional e o seguimento da mulher com doença reumática inflamatória (DRI) durante a gravidez desempenham um papel fundamental no controlo da atividade da doença e na prevenção de complicações materno-fetais.

Métodos: Estudo retrospetivo, longitudinal e descritivo da avaliação pré-concecional e das gestações de mulheres seguidas na consulta de Reumatologia num centro hospitalar entre os anos de 2017-2021.

Resultados: Foram incluídas 76 mulheres correspondendo a um total de 136 gestações, das quais 71 foram acompanhadas nesta consulta. 47 (61,8%) tiveram consulta pré-concecional.

A idade média das grávidas foi de 34 anos e o tempo médio de evolução da doença de 63,5 meses. 41% (n=25) tinham o diagnóstico de espondilartrite (SpA), 22,9% (n=14) de lúpus eritematoso sistémico (LES), 13,1% (n=8) de artrite reumatoide (AR), 6,6% (n=4) de síndrome de sjögren primário e 3,3% (n=2) de doença indiferenciada do tecido conjuntivo. 13,1% (n=8) apresentavam outros diagnósticos: síndrome dos anticorpos antifosfolipídicos (SAF) primário; abortos de repetição; défice proteína C; púrpura trombocitopénica idiopática; doença mista do tecido conjuntivo; doença de Behçet e polimiosite. 19,7% (n=12) apresentavam anticorpos anti-SSA/Ro, 4,9% (n=3) Anti-SSB e 13,1% (n=8) anticorpos antifosfolipídicos (AcAF) com 1,6% a cumprir critérios de SAF secundário. 4 (6,6%) mulheres eram hipertensas. No que diz respeito à terapêutica, a salazopirina foi utilizada em doses até 2000 mg id em 16,9% (n=12) das gestações com AR e SpA. 6 mulheres (24%) com SpA engravidaram sob terapêutica biotecnológica (certolizumab (n=4), ustecinumab (n=1) e golimumab (n=1)). A prednisolona foi utilizada em doses ≤ 7.5 mg id em 38% (n=27) das gestações; a azatioprina em doses até 150 mg id em 15,5% (n=11) e a hidroxicloroquina 400mg id em 35,2% (n=25). O ácido acetilsalicílico (100-150 mg id) foi utilizado em 21 gestações e foi associado à heparina de baixo peso molecular em doses até 200 mg id em 5 delas. 3 gravidezes ocorreram sob terapêutica com metotrexato, suspenso antes das 10 semanas de gestação. Não foi necessária terapêutica em 12 casos (16,9%). Na avaliação do outcome materno, verificou-se que a maioria das mulheres se encontrava em remissão no momento da conceção (valores médios: DAS28 1,49; ASDAS 1,35; SLEDAI-2K 1,7; ESSDAI 0). Durante a gravidez, verificou-se um agravamento em 22,9% dos casos (n=14), que predominou nas doentes com SpA (50%, n=7; ASDAS médio 3,33). 2 gestações complicaram com o desenvolvimento de pré-eclâmpsia (3,7%). 53 (85,4%) resultaram em nados vivos, com partos de termo na sua maioria (96,2%). A idade gestacional média do parto foi de 38,5 semanas, com um peso médio dos recém-nascidos de 3213 gramas. Foram registados 2 casos de prematuridade tardia às 36 semanas (3,7%), 5 (8%) de aborto, 3 (4,8%) de gravidez anembriogénica e 1 gravidez ectópica (1,6%). Não se

verificaram complicações cardíacas fetais nas mulheres seropositivas para o anti-SSA/B. Houve exacerbação da doença no periodo pós-parto (1 mês) em 8 mulheres (17,3%): 5 delas com AR (DAS28 médio 3,27) e 3 com SpA (ASDAS médio 2,79).

Discussão/Conclusão: Os resultados apresentados são comparáveis aos reportados na literatura. A maioria das doentes estava em remissão momento da conceção, o que se relaciona com a probabilidade de sucesso da gestação. Salienta-se desta forma, a importância da avaliação pré-concecional da mulher com DRI em idade fértil e do seguimento multidisciplinar durante a gravidez.

253 - ROLE OF VITAMIN D STATUS IN DISEASE ACTIVITY IN RHEUMATOID ARTHRITIS PATIENTS TREATED WITH BDMARDS – DATA FROM A RHEUMATOLOGY CENTER

Diogo Guimarães da Fonseca[1], Filipe Oliveira Pinheiro[2], Maria Seabra Rato[2], Bruno Miguel Fernandes[2], Salomé Garcia[2], Ana Martins[2], Daniela Santos Oliveira[2, 3], Frederico Rajão Martins[4], Miguel Bernardes[2, 5], Lúcia Costa[2]

[1]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [3]Center for Health Technology and Services Research (CINTESIS), Faculty of Medicine, University of Porto, Porto, Portugal, Porto, Portugal, [4]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [5]Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Background: Vitamin D (vit-D) is a fat-soluble vitamin, mainly involved in the regulation of calcium metabolism, and has gained increasing interest in recent years because of its potential role in immunomodulatory activity. Recent data suggest that it is negatively associated with disease activity in rheumatoid arthritis (RA), however this is not yet fully understood.

Objectives: This study's aim was to investigate if there is any correlation between vit-D serum levels at baseline, before taking the first biological disease-modifying antirheumatic drug (bDMARD), and at 6 months after, with disease activity in a cohort of RA patients.

Methods: This is a cross-sectional study, including all the RA patients taking the first bDMARD with evaluation of the vit-D status at baseline and 6 months after biologic therapy at our Rheumatology Department and registered in the national database (Reuma.pt). Demographic, clinical and laboratorial characteristics and disease activity measures were collected from the baseline visit and after 6 months of treatment with the first biologic. For the statistical analysis, two groups were defined, based on the serum levels of 25(OH) vit-D, considering the most common cut-off of 30 ng/mL. For comparison analyses, chi-square test was used for categorical variables and Mann-Whitney U and T-tests were applied for continuous variables.

Results: Seventy-seven patients were included, 58 (75.3%) were females; the mean age was 54.24 ±11.0 years and seropositivity was founded in 65 (84.4%) for anti-citrullinated protein antibodies and in 58 (75.3%) for rheumatoid factor. The first bDMARD most commonly prescribed were etanercept (28.6%) and rituximab (26%). Regarding the vit-D status at baseline, the mean serum level was 28.35 ± 18.21 ng/mL, with the majority of patients having vit-D insufficiency (63.6%). After 6 months with the first bDMARD remission or low activity were achieved in 29.9% of the patients, using DAS28; in 42.9% and 46.8%, with CDAI and SDAI, respectively. Vit-D serum levels at 6 months were 26.81 ±11.72, with the majority of patients still with vit-D insufficiency (62.3%).

At baseline, patients with vit-D insufficiency had greater patient VAS (79.00 ± 19,14 vs 71.71 ± 21.95), greater erythrocyte sedimentation rate (ESR) (40.67 ± 23.17 vs 32.46 ± 26.09) and greater Health Assessment Questionnaire (HAQ) score (1.75 ± 0.609 VS 1.61 ± 0.659) with neither of them having statistical significance. However, when comparing CRP levels at 6 months, it achieved statistical significance (1.05 ± 1.79 VS 1.41 ± 5.22; p=0.026). The same tendency was confirmed when analyzing vit-D levels at 6 months. Patients with vit-D insufficiency presented greater patient VAS (55.33 ± 28.82 vs 42.86 ± 28.28), greater ESR (26.19 ± 21.57 vs 21.00 ± 20.38) and greater HAQ score (1.35 ± 0.662 VS 1.34 ± 0.705), although without statistical significance. However, it did achieve statistical significance when comparing baseline DAS28 and HAQ (5.60 ± 0.91 VS 5.38 ± 1.31; p=0.013 and 1.76 ± 0.53 VS 1.59 ± 0.75; p=0.007, respectively).

Conclusion: Our data failed to demonstrate a statistically significant association between vit-D serum levels at baseline and at 6 months with disease activity in our RA sample. However, it revealed a positive trend of vit-D insufficiency related to higher activity disease. Interestingly, it showed that vit-D insufficiency after 6 months of bDMARD treatment is related to higher

DAS28 and HAQ at baseline. Nonetheless, we insist it is of paramount importance to conduct larger studies to confirm these findings.

263 - WHICH PATIENTS WITH SPONDYLOARTHRITIS WILL NEED DOSE ESCALATION OF SECUKINUMAB TO 300MG MONTHLY? – RETROSPECTIVE STUDY

Diogo Guimarães da Fonseca[1], Maria Seabra Rato[2], Filipe Oliveira Pinheiro[2], Salomé Garcia[2], Bruno Miguel Fernandes[2], Frederico Rajão Martins[3], Daniela Santos Oliveira[2, 4], Ana Martins[2], Miguel Bernardes[2, 5], Lúcia Costa[2]

[1]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [3]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [4]Center for Health Technology and Services Research (CINTESIS), Faculty of Medicine, University of Porto, Porto, Portugal, Porto, Portugal, [5]Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Background: Secukinumab (SEC) is a fully human monoclonal antibody against interleukin-17A, approved in several countries for the treatment of ankylosing spondylitis (AS) and psoriatic arthritis. It is known that some patients benefit from increasing the monthly dose of SEC from 150mg, the most commonly used dose, to 300mg. However, the baseline clinical characteristics that differentiate these patients are not yet fully understood.

Objectives: This study aimed to investigate whether there are any variables at the beginning of biologic therapy that might predict a greater probability of having to increase the dose of SEC to 300mg in order to obtain a response to treatment.

Methods: This is a retrospective cohort study, including all the spondyloarthritis and psoriatic arthritis patients under SEC at our Rheumatology Department and registered in the national database (Reuma.pt). Demographic, clinical and laboratorial characteristics and disease activity measures were collected from the first visit before the patient began SEC. For comparison analyses, chi-square test was used for categorical variables and Mann-Whitney U and T-tests were applied for continuous variables. Multivariate regression analyses assessed the impact of selected variables on the need to increase the dose of SEC to 300mg.

Results: Thirty-two patients with a mean age of 53±11.96 years were included, 19 (58%) were females and 16 (48.5%) had psoriasis. Twenty-seven (81.8%) patients were under a nonsteroidal anti-inflammatory drug, 11 (33.3%) were under corticosteroid and 11 (33.3%) were under conventional synthetic disease-modifying antirheumatic drug (DMARD); 25 (75,8%) had previously been treated with a biological DMARD. The mean patient and physician baseline VAS were 74,39±19,77 and 47,55±23,38, respectively; the mean erythrocyte sedimentation rate and C-Reactive Protein were 26,33±22,62 mm/hr and 10,81±16,88 mg/dL, respectively; the mean swollen and tender joint count were 1,30±1,63 and 3,67±3,14, respectively; the mean Bath Ankylosing Spondylitis Disease Activity Index (BASDAI) and Ankylosing Spondylitis Disease Activity Score (ASDAS) were 6,18±2,06 and 3,41±0,84, respectively; the mean BASMI and BASFI were 4,22±1,58 and 6,28±2,53, respectively; the mean Maastrich Ankylosing Spondylitis Enthesitis Score (MASES) was 2,85±3,23. Nineteen patients (57.6%) had the dose of SEC increased to 300mg. At the baseline visit, the patients which had their SEC monthly dose increased to 300mg were more frequently men (12 vs 2, p=0.005) and had psoriasis (12 vs 4, p=0.049). On the other hand, these patients also exhibited lower MASES values (2±1.089 VS 4±0.501, p=0.022). A regression analysis was conducted, estimating the relationships between the outcome binary variable of the monthly dose of SEC and the following predictors: gender, psoriasis, MASES value and use of corticosteroid. Female gender (OR 0.070, CI95% 0.005-0.890; p=0.040) and absence of psoriasis (OR 0.104, CI95% 0.011-0.952; p=0.045) were predictors for maintaining SEC at a dose of 150mg monthly.

Conclusion: Our data suggest that the most common characteristics of patients in need of increasing the monthly dose of SEC from 150 to 300 mg in order to achieve a better treatment response are: male gender, coexistence of psoriasis and lower MASES value at baseline. The first two variables remained statistically significant in a multivariate model of regression analysis. Nonetheless, we insist it is of paramount importance to conduct larger studies to confirm these findings.

265 - CHARACTERIZATION OF IDIOPATHIC INFLAMMATORY MYOPATHIES FOLLOWED IN A RHEUMATOLOGY CENTER

Vanessa Fraga[1], Sandra Sousa[1], Maria José Santos[1, 2]

[1]Rheumatology Department, Hospital Garcia de

Orta, Almada, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introduction: Idiopathic inflammatory myopathies (IIM) is a heterogeneous group of disorders characterized by acute or subacute proximal motor weakness, elevated serum muscle enzymes, presence of myositis associated autoantibodies (MAAs) and/or myositis-specific autoantibodies (MSAs), abnormal neurodiagnostic studies, abnormal MRI of shoulder and pelvic girdles. Often there are extra-muscular organ manifestations.

Objective: To characterize patients diagnosed with IIM in our Rheumatology department.

Material and Methods: Clinical and laboratory data from patients with the diagnosis of IIM were retrieved from Reuma.pt/Myositis as well as from electronic health records. We performed a cross-sectional descriptive analysis of all patients followed up to May 2021.

Results: A total of 25 patients were included, mostly Caucasians (n=16) and females (76%, n=19). The mean age at disease onset was 50.2±19 years and 80% were diagnosed within 1 year or less from the beginning of symptoms. Dermatomyositis was the most common subgroup (52%, n=13), followed by anti-synthetase syndrome (32% n=8), polymyositis and scleroderma/polymyositis overlap (16% n=4).

The most common manifestations were muscular (84%), cutaneous (72%), interstitial lung disease (52%), articular (48%), dysphagia (36%) and constitutional symptoms (32%).

TABLE 1 - ANALYSIS OF SERUM MUSCLE ENZYMES AT DIAGNOSIS AND AT LAST EVALUATION

Analysis	Mean value at diagnosis ± standard deviation	Mean value at last evaluation ± standard deviation
Creatinine kinase (CK)	1333 ± 1892	347 ± 658
Myoglobin	551 ± 495	238 ± 544
Aspartate aminotransferase (AST)	71 ± 55	78 ± 209
Alanine aminotransferase (ALT)	55 ± 40	39 ± 64
Lactic dehydrogenase (LDH)	683 ± 495	414 ± 367

At diagnosis, most patients (64%, n=16) had elevated CK, AST, ALT and LDH, the mean values at diagnosis and last evaluation are presented in table 1. The vast majority (92%, n=23) had MAAs (84% ANA, 40% Anti-SSA/Ro52) and 48% MSAs (Anti-Jo1 25%, Anti-EJ 16.7%).

Eighteen (72%) patients performed MRI of shoulder and pelvic girdles, 67% of them had muscular abnormalities compatible with myositis. About 82% patients with EMG had an exam which showed inflammatory myopathy. Twenty percent had skin biopsy which was compatibly with dermatomyositis. Only 12% had muscle biopsy, 100% being compatible with myositis.

Among patients with dermatomyositis, 61.5% were diagnosed with cancer in the same year or in the next 2 years, except one patient who was diagnosed 29 years after the initial diagnosis. Breast cancer and basal cell carcinoma were found to be the most common.

All patients were treated with glucocorticoids and 52% received IVIg. Immunosuppressants/immunomodulators included methotrexate (68%), hydroxychloroquine (36%), azathioprine (36%), cyclophosphamide (16%) and mycophenolate mofetil (16%). Eight patients received biological therapy (rituximab or tocilizumab)

Currently, 56% of patients are in clinical remission, 32% are deceased and 12% have active disease.

Conclusions: Inflammatory myopathies is a group of heterogeneous disorders. In our department, muscle MRI and EMG were the most common diagnostic exams. The presence of MAA and MSAs are helpful to establish the diagnosis and also to identify distinct subtypes of disease. The prevalence of malignancy in patients with Dermatomyositis is high especially in the first 2 years.

277 - SERÁ QUE OS PADRÕES DE PUBLICAÇÃO DOS INTERNOS DE REUMATOLOGIA NA ACTA REUMATOLÓGICA PORTUGUESA (ARP) SE ALTERARAM NAS ÚLTIMAS 3 DÉCADAS?

Luis Cunha-Miranda[1, 2], Alexandra Cardoso[3], Catarina Cortesão[1], Cândida Silva[1], Augusto Faustino[1, 2]

[1]Instituto Português de Reumatologia, Lisboa, Portugal, [2]Colégio da Especialidade de Reumatologia da Ordem do Médicos, Portugal, Portugal, [3]Nutrition Department, Instituto Português de Reumatologia, Lisboa, Portugal

Introdução: ARP é a revista científica da Sociedade Portuguesa de Reumatologia (SPR) sendo publicada desde 1974 e tem sido um dos ex-libris da atividade científica da Reumatologia Portuguesa. Nos últimos anos a ARP conseguiu obter a indexação à MEDLINE® (2006) e a obtenção de factor de impacto (2009) que em muito contribui para a atratividade da revista. Para os internos de Reumatologia a publicação na ARP durante o internato é quase obrigatória, não só como objetivo curricular mas igualmente como forma de melhoria das capacidades científicas e reforço da ligação à especialidade.

Objetivos: Avaliar a evolução dos padrões de publicação dos internos de reumatologia de 1990 a 2020 com foco na ARP.

Métodos: Foram avaliados 40 curriculum finais (CV) de exame de especialidade provenientes de 7 centros da reumatologia portuguesa entre 1990 a 2020 e revistos os números de publicações na ARP assim como em outras revistas nacionais e internacionais, em 1ª autoria e em co-autoria.

Resultados: Dos 40 considerados, 10 eram anteriores ao ano 2000, 8 da década de 2000-2010 e 22 entre 2010 e 2020.

O maior número de publicações sob a forma de artigo ocorreu na década de 2010-2020 (N=43), das quais 16 como primeiro autor em revistas internacionais. A evolução de publicações entre décadas foi significativamente superior nos artigos em co-autorias, em revistas internacionais (p=0.001).

No que diz respeito às publicações exclusivas na ARP (N=28), aumentaram significativamente nos últimos anos (p=0.027), verificando-se uma melhoria tendencialmente positiva nas publicações como primeiro autor, e com maior destaque nos artigos publicados em co-autoria (p=0.040).

Nas últimas décadas, as publicações em outras revistas nacionais (excluindo a ARP) diminuíram progressivamente (p=0.004), com as publicações internacionais a não apresentarem variação significativa. Na década de 2010-2020, as publicações na ARP foram significativamente superiores às restantes publicações nacionais (p <0.0001).

Discussão e Conclusão: Nesta nossa amostra não parece existir diferença entre as décadas avaliadas no que diz respeito ao número total de publicações. Existe uma melhoria no padrão de publicação, com maior número de co-autorias em revistas internacionais e um aumento da publicação de artigos na ARP, em detrimento de outras revistas nacionais. Se a primeira conclusão pode ser parcialmente explicada pelo aumento das cooperações entre serviços no âmbito do Reuma.pt, a segunda conclusão terá de ter em conta o desaparecimento de algumas revistas nacionais na área da Reumatologia. De salientar que o trajeto de indexação e da obtenção de factor de impacto em muito valorizaram a Acta Reumatológica Portuguesa e a tornaram atrativa para a publicação de trabalhos científicos por parte dos internos de Reumatologia tendo um amplo retorno.

TABELA 1. PUBLICAÇÕES SOB A FORMA DE ARTIGO NA ARP, RESTANTES REVISTAS NACIONAIS E OUTRAS PUBLICAÇÕES (NACIONAIS E INTERNACIONAIS) AO LONGO DOS TRÊS GRUPOS DE DÉCADAS CONSIDERADAS.

Publicações sob a forma de artigo	Publicações na ARP	Outras Publicações Nacionais	Total de Outras Publicações	P value*1
Grupo I: <2000 (N=30)				
	2.6±2.0 [0;6]		5.7±5.4 [1;17]	0.190
	2.6±2.0 [0;6]	4.3±3.5 [1;12]		0.315
Grupo II: 2000-2010 (N=8)				
	4.8±2.5 [1;8]		3.0±1.8 [0;5]	0.195
	4.8±2.5 [1;8]	2.3±2.1 [0;15]		0.065
Grupo III: 2010-2020 (N=22)				
	5.7±3.4 [0;14]		5.1±6.2 [0;29]	0.128
	5.7±3.4 [0;14]	1.1±1.5 [0;5]		<0.0001
P value*2	0.027	0.004	0.670	

Legenda: média ± desvio padrão [mínimo; máximo]; *1 teste não paramétrico: Mann-Whitney; *2 teste não paramétrico: Kruskal-Wallis.

279 - MIND THE GAP: EARLY REFERRAL TO RHEUMATOLOGY IN PANDEMIC TIMES

Luis Cunha-Miranda[1], Ana Cordeiro[2], Maria Jesus Mediavilla[1], Maria Margarida Cunha[2], Hugo Parente[3], José Tavares-Costa[3]

[1]Instituto Português de Reumatologia, Lisboa, Portugal, [2]Serviço de Reumatologia, Hospital Garcia de

Orta, Almada, Portugal, [3]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal

Introduction: Early referral to rheumatology and decrement of diagnostic gap time are important strategies to reduce disability and to improve quality of life for almost all of rheumatic patients. Covid 19 had a tremendous impact on all of the national health systems, reducing most of the programmed clinical activity. Most of the patients were unable or unwilling to attend the limited hospital/ health centre appointments that were available, increasing the diagnostic gap.

Patient's referral In Rheumatology comes mostly from general practitioners (GP). In the pandemic setting GPs were overwhelmed with "Trace Covid" as well as attending suspected Covid patients with clear influence on daily routines and consequently on the referral for other specialties.

Objectives: To determine the impact of Covid 19 in the number of referrals sent to three different rheumatology departments in Portugal.

Methods: We reviewed the number of CTH (Consulta a Tempo e Horas – timely appointments system) referral episodes in two time periods: from January 2019 to January 2020 (Covid free time) and from March 2020 to March 2021 (Covid period).

Results: Table 1 Number of referrals to rheumatology services in two selected time periods

Conclusion: This is a very simple study evaluating the impact of Covid-19 in a very important activity of our clinical practice. As expected we found a decrease on the clinical referrals from GPs and other physicians to Rheumatology departments with on average a decreased of 38.5% (26.5% - 50.5%).

The differences between hospitals could be due to the eventual higher impact of the Covid pandemia in different hospitals/ areas or different health centre linkage to rheumatology services.

A clear strategy for increasing new referrals and to decrease the diagnostic gap that has surely increased should be acknowledged and confirmed all of our hospitals.

TABLE 1. NUMBER OF REFERRALS TO RHEUMATOLOGY SERVICES IN TWO SELECTED TIME PERIODS

Hospital/ Rheumatology unit	Covid free period	Covid period	Variation %
Instituto Português de Reumatologia	1226	606	-50.5%
Hospital do Conde de Bertiandos	1107	814	-26.5%
Hospital Garcia de Orta	1851	1151	-37.8%

284 - TOWARDS A HOLISTIC APPROACH TO EARLY ARTHRITIS PATIENTS: DOES MENTAL HEALTH RELATES WITH DISEASE ACTIVITY?

Rui Lourenço Teixeira[1, 2], Vasco C Romão[2], Maria João Saavedra[2], JE Fonseca[1, 2]

[1]Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal, [2]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Early arthritis encompasses the potential to evolve into various rheumatic diseases. Some of them may conditionate early joint damage and a significant impact in quality of life. As highlighted in national and international epidemiologic studies, chronic arthritis patients suffer significantly from anxiety and depression symptoms, which also contribute to impaired quality of life. Over the past years, patient reported outcomes (PRO) have gained importance as instruments allowing to measure the impact of these problem.

We aim to describe the evolution of an early arthritis patient's cohort, characterizing the progression of their disease activity and their quality of life, with a focus on mental health.

Patients were selected from our early arthritis cohort. They were evaluated clinically and analytically, and asked to fill PRO's questionaries regarding pain, functional disability, fatigue, depression and anxiety symptoms and mental health status. Patients were followed up during 12 months. Disease activity was registered and corelated with PRO's measures at baseline and one year afterwards.

60 patients were included, mostly female. The average follow-up was 12 months. As expected, there was a significant decrease in disease activity, with 73% of patients in remission (DAS28-PCR) after the 12-month follow-up. There was an inverse correlation between the final SF-36-Mental Health and the initial DAS28(3V/PCR). The majority (59%) of patients progressed to rheumatoid arthritis.

The improvement of patients' mental health status correlates with baseline disease activity.

An early diagnosis and intervention are essential.

However, an effective treatment does not seem to suffice to actually improve patients' mental health and quality of life. A patient-centered, holistic and multidisciplinary maybe the key to achieve success.

287 - PATIENTS FUNCTIONALLY CAPACITY AFTER OSTEOPOROTIC FRACTURES

Carolina Mazeda[1], Susana P. Silva[1], Joao Paulo Vilas-Boas[1], Bernardo Santos[1], Graça Costa[1], Maria Céu Morais[1], Barcelos A.[1]

[1]Rheumatology Department, Hospital Infante D. Pedro (CHBV), Aveiro, Portugal

Objetives: Osteoporotic fractures imposes a significant health care burden globally with the medium and long-term functional consequences being catastrophic. This study aimed to analyze the relationship between type of fracture, co-morbidities and functional dependence.
Material and Methods: This was a retrospective cohort study that involved patients with a fragility fracture observed in a Fracture Liaison Service. Socio-demographic, clinical data and patients' outcomes, after twelve months, were collected. The data covered the period between 1 January 2019 to 31 December 2020 and was collected from the hospital database. A general descriptive analysis was performed, p-value <0.05 was statistically significant.
Results: A total of 187 patients were included (158 females and 29 males) averaging 76.98±11.99 years old. Most frequent comorbidities included hypertension, diabetes mellitus, dyslipidemia, chronic renal disease, thyroid pathology and depressive syndrome. Osteoporosis risk factors, such as alcohol consumption and corticotherapy, were also evaluated. The majority of fractures were hip fractures (62,6%), followed by vertebral (15%), tibia (13.4%), humerus (6.4%), and wrist (2.7%). In the pre-fracture period, most patients were autonomous (58.8%) and only 12.3% were total functionally dependent. After one year of follow-up, a general decrease in autonomy was observed (43.3% were autonomous, 19.3% were total functionally dependent, 5.9% died and 7.5% were lost for follow-up). No statistically significant differences between the two periods were found (p=0.704). In 22% of patients who were functionally independent before the fracture (81 patients), they became partially dependent for activities of daily living, after one year of follow-up. The type of fracture is associated with alterations in functionally dependence (p=0.015). No statistically relevant differences were found between different co-morbidities (p>0.05). The use of systemic corticotherapy were related to vertebral fracture (p=0.02).
Conclusion: FLS has a fundamental roll to promote an early patients recovery to the pre-fracture functionally capacity and counsel the patient about the potential rehabilitative benefits.

292 - OUR EXPERIENCE WITH A MULTIDISCIPLINARY RHEUMATOLOGY/ PNEUMOLOGY CLINIC

Carolina Mazeda[1], Susana P. Silva[1], Joao Paulo Vilas-Boas[1], Margarida Ferreira[2], PG Ferreira[3], Renata Aguiar[1], Barcelos A.[1]

[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [2]Radiology, Centro Hospitalar Baixo Vouga, Aveiro, Portugal, [3]Pneumology, Centro Hospitalar Baixo Vouga, Aveiro, Portugal

Introduction: Interstitial lung disease (ILD) is a common feature of connective tissue diseases (CTD) that usually are associated with significant morbidity and mortality. The heterogeneity of clinical manifestations, radiological presentations and disease severity makes its diagnosis and management complex. Optimal care of patients with CTD-ILD requires collaboration and close interaction by the rheumatology and pulmonology departments. The assessment in a multi-disciplinary clinic (MDC) aims to improve the early diagnosis and treatment of CTD-ILD with individualized plans in order to improve the prognosis and survival of patients.

The MDC of Rheumatology/Pneumology at Centro Hospitalar Baixo Vouga is held monthly and includes an ILD dedicated pulmonologist, a rheumatologist, a radiologist and a pathologist in a shared space. The main objective of our study was to characterize the population referred to MDC and analyze its impact on the diagnosis and treatment of patients.
Methods: This retrospective study included all patients seen in a MDC between January 2017 and December 2020. Data collected for each patient included demographic data, auto-imunity status, symptoms at diagnosis, lung function at diagnosis, established/ presumptive diagnoses and treatment undertaken. A descriptive analysis was performed.
Results: A total of 99 patients were referred to MDC (70 females and 29 men) with a median age at first referral of 70 years (interquartile range 59–76 years) and had a median number of consultations per patient of 1.2. Seventeen patients were referred by Pulmonology for pulmonary disease, analytical changes (2 with anti-Ro52, 1 with anti-P17, 1 with DFS70, 1 with anti-

TABLE 1. RESULTS OF THE MULTIDISCIPLINARY CLINIC IN THE COHORT OF PATIENTS ANALYZED

Category		Diagnosis	Patients (%)	Laboratory screening			HRCT pattern					Non-specific radiological changes
				Anti-CCP	FR	ANA	UIP	NSIP	OP	LIP	Other	
CTD		Rheumatoid Arthritis	28 (28.3)	78.6%	78.6%	10.7%	53.6%	14.2%			14.3%	17.8%
		Systemic Sclerosis	20 (20.2)			100%	10%	65%		10%	5%	10%
		Sjögren's syndrome	13 (13.1)			100%		23.1%		15.4%	23,10%	38.4%
		Undifferentiated Connective Tissue Disease	6 (6.1)				16.7%	16.7%			16.7%	50%
		Systemic Lupus Erythematosus	3 (3)					33.3%			66.7%	
		Small Vessel Vasculitis	3 (3)								100%	
		Antisynthetase Syndrome	3 (3)						66.7%		33.3%	
		Spondyloarthritis	2 (2)									100%
		Anti-SRP Myopathy	1 (1)					100%				
		Dermatomyositis	1 (1)						100%			
		Mixed Connective Tissue Disease	1 (1)					100%				
		Relapsing Polychondritis	1 (1)						100%			
Other ILD	Idiopathic	IPF	4 (4)			45.5%						
	Myscellanea	PH	7 (7.1)									
	Sarcoidosis	Sarcoidosis	6 (6.1)									

ILD - interstitial lung disease; CTD-ILD -connective tissue disease associated ILD; HRCT - high resolution computed tomography; IPF - idiopathic pulmonary fibrosis; HP - hypersensitivity pneumonitis; UIP - usual interstitial pneumonia; NSIP - non-specific interstitial pneumonia; COP - cryptogenic organizing pneumonia; LIP - lymphoid interstitial pneumonia; RF - rheumatoid factor; anti-CCP - anti-cyclic citrullinated peptide antibodies; ANA - Antinuclear antibodies

CENP-B and 2 with anti-Jo1 positives) and doubts to have an underlying CTD. In all of them, this possibility was excluded. The remaining 82 patients were already diagnosed with CTD (Table 1), but in seventeen of these patients the interstitial involvement was not confirmed. Rheumatoid arthritis was the commonest rheumatologic disease with lung manifestation, followed by systemic sclerosis. The average follow-up time was 22.55±29.23 months. At baseline, the patients had an average mMRC of 1.18 and the mean of FVC was 85.25±20.66% and DLCO was 61.30±18.35%. Regarding the HRCT pattern observed (Table 1), the most prevalent was usual interstitial pneumonia (UIP) pattern and nonspecific interstitial pneumonia (NSIP). As for the immunosuppression regimen, most patients started an induction with prednisolone followed by maintenance therapy with mycophenolate mofetil (25.3%), rituximab (11.1%), azathioprine (9.1%) or cyclophosphamide (6.1%).

Conclusion: The multidisciplinary clinic allows an integrated and synergistic approach, which brings value to patients by simplifying the care of these complex patients. This approach allows for greater sensitivity and faster diagnosis allowing patients to receive appropriate treatment sooner and, consequently, to improve quality of life and outcomes. Further research into its cost-effectiveness in this setting is required as well as assessing patient satisfaction.

293 - CORRELATION BETWEEN ARTERIAL STIFFNESS AND NAILFOLD CAPILLARY MICROSCOPIC ABNORMALITIES IN SYSTEMIC SCLEROSIS: RESULTS FROM A SINGLE CENTRE CROSS-SECTIONAL STUDY

Carolina Mazeda[1], Susana P. Silva[1], Joao Paulo Vilas-Boas[1], Renata Aguiar[1], Barcelos A.[1], José Mesquita Bastos[2]

[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, EPE, Aveiro, Aveiro, Portugal, [2]Cardiologia, Centro Hospitalar do Baixo Vouga, EPE, Aveiro, Portugal

Introduction: Systemic Sclerosis (SSc) is a rare connective tissue disease with several systemic manifestations and organ involvement characterized by immune dysfunction, vascular abnormalities and fibrosis. The microvascular damage represents the earliest morphological and functional process of the disease and is a prominent feature of SSc easily accessible by a non-invasive tool, a nailfold video-capillaroscopy (NVC). While microangiopathy is well-documented in SSc, the macrovascular involvement has not been completely clarified. Several studies suggested that SSc might be linked to an increased risk of developing cardiovascular disease. Arterial stiffness is an independent predictor of cardiovascular events, and the carotid-femoral pulse wave velocity (PWV) are considered the gold standard for your measurement.

TABLE 1. BASELINE CLINICAL CHARACTERISTICS

Age, years (mean±SD)	58.95±8,75
Sex (M/F), n	5/17
Duration disease (mean±SD)	51.59±44.59
Diffuse cutaneous, n (%)	7 (31.8)
Modified Rodnan skin score (mean±SD)	10.82±10.55
Digital ulcers, n (%)	11 (50.0)
Anti-Scl70 antibodies, n (%)	7 (31.8)
Anticentromere antibodies, n (%)	11 (50.0)
Anti-RNA polymerase III antibodies, n (%)	3 (13.6)
Pulmonary involvement, n (%)	8 (36.4)
Gastrointestinal involvement, n (%)	9 (40.9)
Articular involvement, n (%)	2 (9.1)
Nailfold video-capillaroscopy pattern	
Nonspecific	1 (4.5)
Early	8 (36.4)
Active	7 (31.8)
Late	6 (27.3)

The aim of this study is to assess changes in arterial stiffness and its relationship with nailfold video-capillaroscopy abnormalities in patients with SSc.

Methods: All patients included in the study fulfilled the ACR/EULAR 2013 classification criteria for SSc and were followed in the Centro Hospitalar do Baixo Vouga outpatient rheumatology clinic between 2012 and 2020; sex and age-matched healthy controls were also enrolled in the study. Socio-demographic and clinical data were collected. Each participant underwent arterial stiffness studies (PWV and augmentation index (AI)) and NVC (nonspecific, early, active or late). A descriptive analysis was performed one-way ANOVA and Kruskal-Wallis test were used to compare continuous variables and Fisher's exact test for categorical variables, respectively. p-value ≤ 0.05 was statistically significant.

Results: Twenty-two patients were included (17 female and 5 men) averaging 58.95±8.75 years old with a mean disease duration was 51.59±44.59 years. Baseline clinical characteristics of all patients with SSc are listed in table 1. History of digital ulceration, disease subtype (limited vs diffuse), pulmonary involvement and positivity for anti-topoisomerase-I antibodies were found to be statistically correlated with microangiopathic severity identified in NVC ($p<0.05$).

PWV is higher in SSc patients than controls (8.08±1.66 vs 6.85±1.84; p=0,01), however PWV levels did not differ significantly between NVC patterns (p=0.362). Values of AI was also higher, however this did not reach statistical significance (p=0.35).

Conclusion: Despite the small sample size, wall stiffness parameters measurements don't seem to correlate to microangiopathic features. Although we were able to demonstrate significant difference in PWV between two groups, but it was insufficient to detect difference in AI. This sug¬gests that patients with SSc may have an increased prevalence of subclinical atherosclerosis, however more studies with larger sample size are warranted and may be beneficial to assess its evolution over time in order to understand its impact on the clinical outcome.

Casos Clínicos

ACTA REUMATOL PORT. 2021:46:??-?? (SUP)

CASOS CLÍNICOS (COM APRESENTAÇÃO ORAL)

015 - HOARSENESS AS THE PRESENTATION OF IGG4-RELATED DISEASE WITH VOCAL CORD AND MEDIASTINAL INFILTRATION

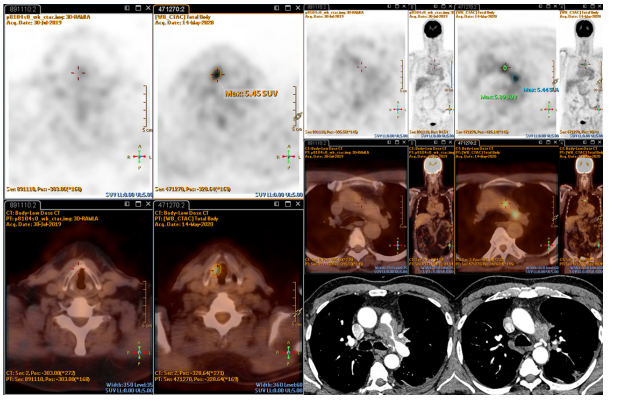
Eduardo Dourado[1, 2], Ana Silva Martins[3], Inês Cordeiro[1, 2], JE Fonseca[1, 2]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Serviço de Hematologia e Transplantação de Medula, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introduction: Immunoglobulin G4-related disease (IgG4-RD) is a fibroinflammatory infiltrative condition characterised by a dense lymphoplasmacytic infiltrate rich in IgG4-positive plasma cells, storiform fibrosis, and elevated serum IgG4 concentrations. IgG4-RD has been described in virtually every organ system.

Clinical vignette: A 64 years-old man presented to the otorhinolaryngology clinic in December 2016 complaining of progressive hoarseness and asthenia in the previous 18 months. He had a previous history of smoking habits, alcoholism and Helicobacter pylori-positive gastritis. A computed tomography (CT) of the chest showed an infiltrative mediastinal mass, and the patient was referenced to the Pulmonology clinic. Between 2017 and 2018, six chest CTs and four biopsies (one endobronchial ultrasound-guided biopsy, one video-assisted thoracoscopic biopsy and two open-surgery biopsies) were performed. The first three biopsies were reported as inconclusive or nonspecific. The fourth biopsy revealed fibro-adipose tissue with local lymphoplasmacytic infiltrate and hyalinised fibrous tissue fragments with inflammatory infiltration composed of lymphocytes, some eosinophils and abundant plasmacytes. A whole-body fluorodeoxyglucose positron emission tomography (FDG-PET)/CT imaging performed in 2018 showed hypermetabolic foci in the right vocal cord, with a maximum SUV of 8.21, and in the aortopulmonary window, with a maximum SUV of 5.9. The patient was then referenced to the Haematology clinic due to a suspected lymphoma. A revision of the biopsied tissues was requested for immunohistochemical evaluation. The sample was negative for Reed-Sternberg cells (CD30+ CD15+), and IgG4 immunostaining revealed more than 50 IgG4-positive plasma cells per high-power field and a ratio of IgG4- to IgG-bearing plasma cells higher than 40%. Serum IgG4 was 521 (normal range 3-201) mg/dL and serum c3 was 84 (normal range > 90) mg/dL. The diagnosis of IgG4-RD was assumed, and the patient was started on 20 mg of prednisolone daily in May 2018. The hoarseness and asthenia resolved and a FDG-PET/CT performed in July 2019 showed marked improvement of the previously described hypermetabolic foci. The prednisolone dose was then slowly tapered from 20 mg to 5 mg daily. In May 2020, the patient reported a recurrence of hoarseness and asthenia and an unintended weight loss of 3 kg in the previous month. A re-evaluation with FDG-PET/CT confirmed relapse of the disease in the right vocal cord and mediastinum. The patient was then referred to the Rheumatology clinic. The prednisolone dose was increased to 0.6

FIGURE 1 - COMPARISON OF THE WHOLE-BODY FDG-PET/CT IMAGING FROM JULY 2019 AND MAY 2020; CT ANGIOGRAPHY OF THE MEDISTINAL MASS.



mg/kg/day (40 mg) for four weeks and then tapered (10 mg every two weeks until 20 mg/day, then 5 mg every two weeks until 10 mg/day, and finally 2.5 mg every two weeks). The symptoms improved initially, but in December 2020, immediately after tapering the prednisolone dose from 7.5 to 5 mg/day, the patient presented with another clinical relapse, characterised again by hoarseness, asthenia and weight loss of 4 kg over one month. Due to this aggravation, rituximab was started with a good clinical response.

Discussion: Laryngeal involvement as a primary feature represents an extremely infrequent expression of IgG4-RD, and vocal cord involvement is even rarer. If left untreated, this patient's prognosis was poor, considering the risks of upper airway obstruction and haemodynamic complications from the mediastinal involvement.

020 - MEPOLIZUMAB NO TRATAMENTO DA GRANULOMATOSE EOSINOFÍLICA COM POLIANGEÍTE – A EXPERIÊNCIA DE UM CENTRO

Ana Catarina Duarte[1], Maria José Santos[1, 2]
[1]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introdução: A granulomatose eosinofílica com poliangeíte (GEPA) é uma vasculite necrotizante, granulomatosa, em que os eosinófilos têm um papel central na fisiopatologia da doença. Os glicocorticoides mantêm-se a base do tratamento, embora muitos doentes necessitem da associação de outros imunossupressores. O mepolizumab (MPZ) é um anticorpo monoclonal que bloqueia a interação da IL-5 com o seu receptor na superfície dos eosinófilos, reduzindo a produção e sobrevivência destas células. Um ensaio clínico mostrou benefício no tratamento de doentes com GEPA recidivante ou refratária à terapêutica convencional, mas os dados de vida real são escassos. Apresentamos a nossa experiência no tratamento da GEPA com MPZ através de 2 casos clínicos.

Caso 1: Homem, 46 anos, com GEPA com envolvimento sistémico (febre, perda ponderal), pulmonar (infiltrados pulmonares migratórios), vias aéreas superiores (obstrução nasal), sistema nervoso periférico (SNP; mononeuropatia múltipla) e cutâneo (púrpura). Analiticamente com eosinofilia periférica > 10%, subida de parâmetros inflamatórios (PIs) e anticorpos anti-citoplasma dos neutrófilos (ANCAs) negativos. Tratado inicialmente com prednisolona (PDN) 1mg/kg/dia e ciclos mensais de ciclofosfamida (500-1000mg/m2) durante 6 meses, com boa resposta. Um mês após fim de ciclofosfamida, estando o doente sob PDN 5mg/dia, verificou-se reaparecimento das queixas de obstrução nasal, com subida de eosinofilia periférica e PIs. Iniciou azatioprina (AZA; titulada até 2mg/kg/dia) e aumentou PDN para 10mg/dia, verificando-se recidiva dos mesmos sintomas com doses de PDN inferiores a 10mg/dia. Por este motivo iniciou MPZ (300mg/mês), estando assintomático ao final de 6 meses de terapêutica, com contagem de eosinófilos e PIs normais, e já sem PDN.

Caso 2: Mulher, 67 anos, com GEPA com envolvimento sistémico (febre, anorexia, perda ponderal), pulmonar (asma), cardíaco (miocardite, derrame pericárdico), gastrointestinal (GI; náuseas, vómitos, diarreia), SNP (mononeuropatia múltipla) e cutâneo (púrpura). Analiticamente com eosinofilia periférica > 10%, subida de PIs e ANCAs negativos. Inicialmente medicada com PDN 1mg/kg/dia e imunoglobulina humana endovenosa por antecedentes de Aspergilose bronco-pulmonar e isolamento recente de Staphylococcus aureus meticilino-resistente no exsudado nasal, com boa resposta ao fim de 6 ciclos mensais. A salientar diagnóstico de Diabetes mellitus induzida pela corticoterapia e fratura de fragilidade de 2 vértebras lombares. Iniciou posteriormente AZA 1mg/kg/dia (intolerância GI a doses superiores), associada a PDN 7.5mg/dia, tendo apresentado ocasional agravamento das queixas de pieira, com necessidade de subida transitória de PDN. Ao final de 18 meses de AZA, a doente apresentou recidiva das queixas GI, febre, púrpura, aumento de eosinofilia periférica e PIs.

Assumiu-se provável vasculite mesentérica, pelo que retomou PDN 1mg/kg/dia e iniciou rituximab (2x1g). Após 5 meses de rituximab, sob PDN 7.5mg/dia, a doente apresentou clínica de insuficiência cardíaca aguda e a investigação complementar revelou envolvimento do endomiocárdio pela doença. Iniciou terapêutica diurética e fez switch para MPZ, encontrando-se assintomática, incluindo do ponto de vista cardíaco, sem eosinofilia periférica e com PIs normais, ao final do primeiro mês de tratamento.

Conclusões: os casos descritos ilustram o papel promissor do MPZ no tratamento da GEPA, incluindo em doentes refratários a várias terapêuticas e com manifestações graves. Além disso, o seu uso permite agilizar a redução/suspensão da corticoterapia.

062 - GRANULOCYTE COLONY-STIMULATING FACTOR RELATED LARGE VESSEL VASCULITIS - A RARE DIAGNOSIS

Ana Margarida Duarte-Monteiro[1, 2], Martins-Martinho J [1, 2], Ryan Costa Silva[3], Lígia Peixoto[3], Joana Lobato[4], Catarina Abreu[5, 6], Nikita Khmelinskii[1, 2], José Carlos Romeu[1, 2], Cristina Ponte[1, 2]

[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Serviço de Medicina I, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [4]Instituto Português de Oncologia de Lisboa Francisco Gentil, Lisboa, Portugal, [5]Serviço de Oncologia Médica, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [6]Faculdade de Medicina da Universidade de Lisboa, Lisboa, Portugal

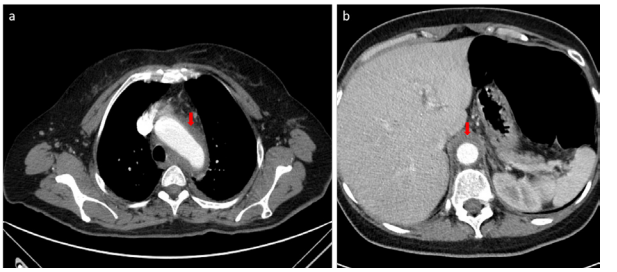
Introduction: Patients under chemotherapy may develop large vessel vasculitis (LVV) as a potential side effect of cytotoxic therapy or as a paraneoplastic manifestation of the underlying disease. Granulocyte colony-stimulating factors (G-CSF) such as pegfilgrastim are frequently used in association with chemotherapy regimens due to risk of neutropenia and can also be associated with LVV. We report two cases of LVV associated with G-CSF.

Case 1: A 56-year-old woman with breast carcinoma (luminal B type), submitted to surgical treatment, was being treated with adjuvant chemotherapy. In her last session it was changed to AC dose dense protocol (doxorubicin, cyclophosphamide and pegfilgrastim). Three days later, she presented with anorexia, asthenia, and fever (38.7ºC), as well as left cervical and dorsal pain with pleuritic characteristics and pain, paresthesia and claudication of the left arm. There was no pulse asymmetry. Inflammation markers were elevated (erythrocyte sedimentation rate [ESR] 103 mm; C reactive protein [CRP] 22.9 mg/dl). Screening for infection was negative. Angio-CT was performed documenting involvement of the aortic arch and initial segment of the left subclavian artery by a hypodense sleeve suggesting a localized vasculitis process (Fig1a). The patient was started on intravenous (IV) methylprednisolone 1g/day for three days, followed by prednisolone 60 mg/day. Five days later she was asymptomatic and her inflammatory markers had decreased (ESR 61 mm; CRP 4.8 mg/dl). The patient awaits control angioCT, however ultrasound of the temporal, facial, axillary, and subclavian arteries demonstrated no active inflammation. The patient was able to taper corticosteroids and resumed the same chemotherapy regimen, excluding pegfilgrastim, showing no signs of relapse two months later.

Case 2: A 48-year-old woman with stage IV non-germinal center diffuse large B cell lymphoma was started on R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) chemotherapy regimen associated with methotrexate and pegfilgrastim. Two weeks after the third cycle the patient reported severe lumbar pain. Physical examination was unremarkable. CRP was elevated (30 mg/dl) and screening for infection was negative. A lumbosacral CT revealed a hypodense sleeve surrounding the abdominal aorta and its major branches, suggestive of vasculitis (Fig1b). She was started on IV methylprednisolone 80mg/day for three days and then oral prednisone 60mg/day. The patient improved clinically and CRP decreased to 14.8mg/dl. After corticosteroid tapering, the same chemotherapy regimen was reinitiated, excluding pegfilgrastim. Three months later she underwent abdominal MRA, PET, and ultrasound of the temporal axillary and subclavian arteries, showing no signs of vasculitis.

Both patients had previous recent imaging studies showing no signs of vasculitis. Considering the temporal relation between the start of chemotherapy protocol including G-CSF, the swift response to corticosteroids, and the absence of relapse even after tapering and resuming chemotherapy, the diagnosis of G-CSF induced vasculitis was established.

FIGURE. A)THORACIC CT: HYPODENSE SLEEVE INVOLVING THE AORTIC ARCH; B)LUMBOSACRAL CT: HYPODENSE SLEEVE SURROUNDING THE ABDOMINAL AORTA



Conclusion: LVV induced by G-CSF may occur in patients under chemotherapy. Differential diagnosis with other causes of raised inflammatory markers and/or fever, such as infection, may be challenging and require a high degree of clinical suspicion. Early identification of the probable causal factor and adjustment of the chemotherapy regimen with G-CSF discontinuation is of paramount importance for the successful management of these patients.

116 - A RARE CASE OF CEREBELLAR DEGENERATION DUE TO PRIMARY SJOGREN'S SYNDROME

Rita Cunha[1], Marcelo Aveiro[2], Tatiana Rodrigues[2], Joana Domingues[3], Ana Oliveira[2]

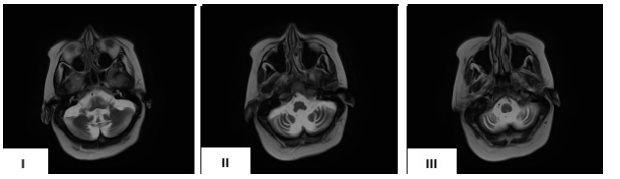
[1]Rheumatology Department, Hospital Infante D. Pedro (CHBV), Aveiro, Portugal, [2]Internal Medicine, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [3]Neurology, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal

Introduction: Sjogren's syndrome (SS) is an autoimmune pathology characterized by salivary and lacrimal glands and possible systemic multi-organ manifestations. The reported prevalence of neurological manifestations in PSS varies widely from 10 to 60% with peripheral neuronopathy being the most common.

Clinical Case: We present a 48-year-old woman, referred for hospital care, due to dysarthria, diplopia and ataxia. She had a previous history of an episode of sudden dysarthria and ataxia,about 10 years earlier without any other relevant symptoms. At the time she did not seek medical care and had partial recovery, maintaining ataxia.

A thorough clinical history confirmed the presence of xerostomia. The patient denied xerophthalmia or any other relevant symptoms. Neurological examination revealed cerebellar ataxia, with a predominance of the lower limbs, multidirectional nystagmus and marked cerebellar dysarthria. The brain magnetic imaging revealed bilateral cerebellar atrophy and the brain positron emission tomography (PET) scan demonstrated cerebellar glycolytic hypometabolism. She had antibody positivity for anti-SSA and anti-SSB and minor salivary glands biopsy revealed lymphocytic infiltration. Other causes of ataxia were ruled out (e.g neoplastic, toxic, immune-mediated, vitamin deficiency, infectious diseases, degenerative disorders and genetic conditions). Considering the 2016 ACR/EULAR classification criteria for PSS, and the absence of other causes, the patient was diagnosed with SS with cerebellar degeneration. She started pulses of methylprednisolone (1g/day) for 3 days followed by prednisolone tapering, in association with 3 cycles of cyclophosphamide 750 mg/m2 up to 1 g and after this intravenous immunoglobulin, without sustained results. Therefore, rituximab was initiated with improvement in dysarthria and coordination. Since then. the patient has been treated with rituximab twice yearly (2 infusions of 1g) and the disease is stable.

FIGURE I,II, III: IMAGES 1, 2 AND 3 - MRI OF THE BRAIN WITH SIGNS OF BILATERAL CEREBELLAR ATROPHY



Discussion: There were only about five cases described in the literature with PSS patients who presented with cerebellar degeneration and ataxia. The pathophysiology of this condition still remains unclear. There is no consensus on specific therapy for the management of PSS with CNS involvement. In these cases reports, methylprednisolone, cyclophosphamide and rituximab were used with different efficacies.

Conclusion: Ataxia caused by cerebellar atrophy due to PSS is a very rare entity. The diagnosis can sometimes be challenging especially when the neurological abnormalities precede the classic glandular involvement. In our clinical case, rituximab was the only treatment that achieved a clinical response.

REFERENCES:

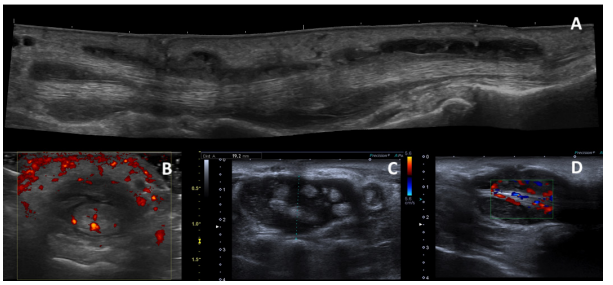
Heidary M, Alesaeidi S, Afshari K. Cerebellar degeneration in primary Sjögren syndrome. BMJ Case Rep. 2018;2018:1–4

126 - *CANDIDA ALBICANS* TENOSYNOVITIS OF THE HAND

Catarina Tenazinha[1, 2, 3], Rita Barros[1], Vasco C Romão[1, 2, 3]

[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Rheumatology Research Unit, Instituto de Medicina Molecular, Lisboa, Portugal, [3]Faculdade de Medicina da Universidade de Lisboa, Lisboa, Portugal

FIGURE 1. RIGHT HAND THIRD DIGIT FLEXOR TENOSYNOVITIS (A AND B) AND LEFT HAND COMMON FLEXOR TENOSYNOVITIS (C AND D)



A 72-year-old woman with Systemic Lupus Erythematosus, secondary Sjögren's Syndrome, secondary Antiphospholipid Antibody Syndrome and Miastenia Gravis presented with painful swelling of the 3 rd right and 1 st to 4 th left hand digits that she had had for the last 2 months. She was under Prednisolone 15mg id and monthly IV immunoglobulin 2g/kg with disease remission for the previous year. Clinical examination was suggestive of hand flexor tenosynovitis, which was confirmed by ultrasound and CT-scan. Direct exam of the synovial fluid revealed many polymorphonuclear cells and cultural exam grew *Candida albicans*. Treatment with oral fluconazole 400mg id was effective.

Candida albicans is a rare cause of infectious tenosynovitis associated to immunocompromised states. This is the first case reporting bilateral tenosynovitis of the hand, highlighting the possible role of concealed hematogenous spread of opportunistic microorganisms in atypical clinical manifestations in immunocompromised patients.

168 - ESCLEROSE SISTÉMICA COM ENVOLVIMENTO GASTROINTESTINAL EXUBERANTE – O PAPEL DA IMUNOGLOBULINA ENDOVENOSA

Ana Bento da Silva[1, 2], Maria Helena Lourenço[1, 2], Laura Gago[1, 2], Rita Pinheiro Torres [1, 2], Nuno Pina Gonçalves[1, 2, 3], Ana Lúcia Fernandes[4], José Adriano Oliveira Marona[1, 2], Manuela Costa[1], Maria João Gonçalves[1, 2, 5], Jaime C. Branco[1, 2, 6]

[1]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [2]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [3]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [4]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, [5]Instituto Politécnico de Lisboa, Escola Superior de Tecnologia da Saúde de Lisboa, Lisboa, Portugal, [6]Comprehensive Health Research Center – CHRC, Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, Lisboa, Portugal

Introdução: A Esclerose Sistémica (ES) é uma doença autoimune com envolvimento multiorgânico, sendo o sistema digestivo o mais frequentemente abrangido e associado a morbilidade significativa1.

Caso Clínico: Mulher, 69 anos, fumadora, com antecedentes de osteoporose fraturária, hipertensão arterial e doença arterial periférica; tinha diagnóstico de ES limitada com 6 meses de evolução, com esclerose cutânea (score de Rodnan 30/51 pontos), artrite, miosite e hipertensão arterial pulmonar provável. Imunologicamente, apresentava anticorpos antinucleares em título de 1/640, anticorpo anti-SSA positivo e anti-th/to duvidoso. Foi medicada com prednisolona 7.5mg/dia e metotrexato subcutâneo 15mg/semana. Inicia quadro de enfartamento precoce, disfagia para líquidos, distensão e dor abdominal tipo cólica, dejeções diarreicas abundantes, incontinência fecal e perda ponderal de 10Kg em 2 meses, tendo sido internada no Serviço de Reumatologia para investigação. Do estudo destaca-se: exame bacteriológico e parasitológico das fezes e pesquisa de Clostridium difficile negativos; tomografia computorizada (TC) abdominopélvica com evidência de pequena quantidade de ascite com predomínio na cavidade pélvica e em topografia sub-hepática, sem outras alterações; trânsito esofágico compatível com diminuição da motilidade esofágica. Admitido envolvimento gastrointestinal pela ES; iniciou terapêutica com rifaximina, esomeprazol, domperidona e suplementos hiperproteicos e hipercalóricos. Por melhoria clínica inicial, teve alta hospitalar ao 9º dia de internamento.

Posteriormente, apresentou novo agravamento do quadro abdominal, com perda ponderal e desnutrição grave rapidamente progressiva, motivando reinternamento em setembro de 2020. Analiticamente, apresentava múltiplos distúrbios hidro-eletrolíticos (hipocaliémia, hipomagnesémia e hipocalcémia). O estudo endoscópico evidenciou esofagite de refluxo, candidíase esofágica, gastrite associada a Helicobacter pylori; a colonoscopia revelou hiperemia da mucosa do reto e sigmoide com algum exsudado ade-

rente e prolapso rectal. O internamento foi marcado por episódios recorrentes de suboclusão intestinal, apresentando dor abdominal agravada, paragem de emissão de fezes e vómitos. A radiografia abdominal revelou exuberantes níveis hidroaéreos e a TC uma distensão marcada de ansas intestinais. O parecer da Cirurgia Geral foi realizar uma abordagem conservadora (instituição de dieta zero e colocação de sonda nasogástrica), por ausência de oclusão mecânica. Realizou terapêutica procinética e laxante, ciclos rotativos de antibioterapia e octreótico sem benefício. Manteve episódios recorrentes de suboclusão intestinal, impossibilitando a reinstituição de dieta e condicionando desnutrição grave, o que motivou início de nutrição parentérica em outubro. Iniciou ciclos de imunoglobulina endovenosa 2g/Kg mensal com melhoria progressiva do quadro abdominal, permitindo reintrodução gradual da dieta com tolerância e recuperação ponderal. Suspendeu a nutrição parentérica em junho de 2021, encontrando-se atualmente sob dieta mole.

Conclusão: O envolvimento gastrointestinal grave é um marcador de mau prognóstico e mortalidade na ES, sendo a terapêutica limitada e dirigida ao tratamento sintomático1. Este caso vem realçar o papel da imunoglobulina endovenosa como terapêutica promissora na abordagem destes doentes.

REFERÊNCIAS

1. Miller, J. B., Gandhi, N., Clarke, J., & McMahan, Z. (2018). Gastrointestinal involvement in systemic sclerosis: an update. Journal of Clinical Rheumatology, 24(6), 328.

170 - UM CASO ATÍPICO DE MIOSITE FOCAL

Ana Bento da Silva[1, 2], Rita Pinheiro Torres [1, 2], Maria Helena Lourenço[1, 2], Catarina Cabral[3], Rita Reis[3], Jaime C. Branco[1, 2, 4], Maria João Gonçalves[1, 2, 5]

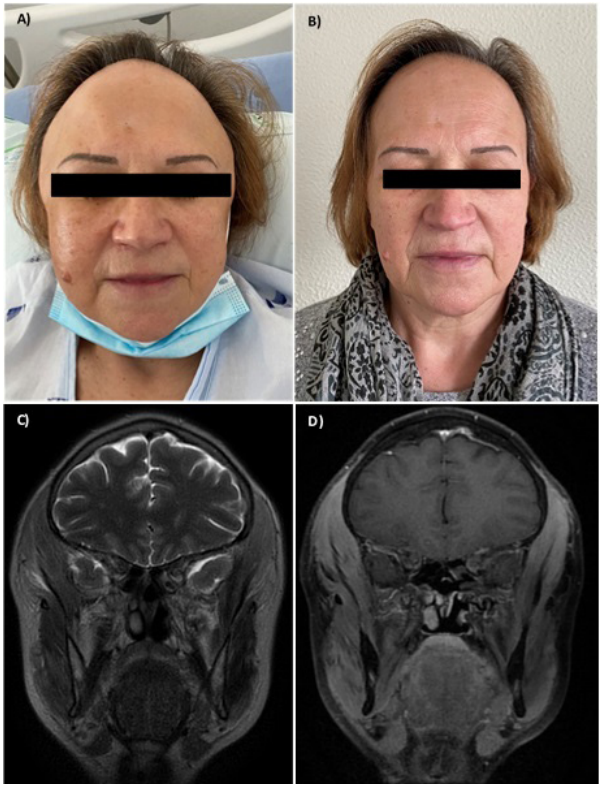
[1]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [2]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [3]Medicina Interna, Centro Hospitalar de Lisboa Ocidental EPE, Hospital de Egas Moniz, Lisboa, Portugal, [4]Comprehensive Health Research Center – CHRC, Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, Lisboa, Portugal, [5]Instituto Politécnico de Lisboa, Escola Superior de Tecnologia da Saúde de Lisboa, Lisboa, Portugal

Introdução: A miosite focal é uma doença rara que afeta tipicamente um músculo, mais frequentemente ao nível dos membros inferiores, e pode estar associada a patologia radicular, doenças infeciosas ou autoimunes, neoplasias e trauma.

Caso Clínico: Senhora de 64 anos de idade, não fumadora, com antecedentes de dislipidemia sob fluvastatina 80mg/dia e perturbação depressiva, medicada com sertralina 50mg/dia e loflazepato de etilo 2mg em SOS. Dirige-se ao Serviço de Urgência (SU) por um quadro com quatro dias de evolução de tumefação da região temporal direita, associada a dor local e disfonia; apresentava-se também subfebril, com uma temperatura axilar de 37.5ºC. Realizou estudo analítico, sendo de destacar proteína C reativa (PCR) de 1mg/dL. Perante suspeita de abcesso dentário, iniciou antibioterapia empírica com Amoxicilina/Ácido Clavulânico 875mg/125mg de 12/12 horas e Metronidazol 250mg de 8/8 horas, mas com agravamento progressivo, com extensão da tumefação para a região retroauricular direita e temporal esquerda, o que motivou retorno ao SU ao 10º dia de evolução do quadro. Não manifestava outros sinais ou sintomas e o restante exame objetivo era inocente. O estudo analítico de reavaliação revelou elevação dos valores da PCR (11.7mg/dL), velocidade de sedimentação (VS 52mm/h), creatina quinase (CK 623 U/L), mioglobina (83 U/L), aspartato aminotransferase (AST 56 U/L) e alanina aminotransferase (ALT 69 U/L). As hemoculturas e as serologias para Treponema pallidum, Vírus da Imunodeficiência Humana, Vírus da Hepatite B e C e Epstein-Barr foram negativas. O estudo imunológico, incluindo os anticorpos antinucleares e os associados a miopatias inflamatórias, foi negativo. Realizou uma ressonância magnética facial que demonstrou espessamento e hipersinal T2/FLAIR dos músculos temporal, masséter e pterigoides bilateralmente (mais notório à direita), com realce após a administração endovenosa de gadolinium, sugerindo miosite. Procedeu-se a biópsia do músculo temporal direito, que revelou uma marcada infiltração linfocítica (predominantemente de linfócitos T CD3), assim como marcação de múltiplas fibras com produtos dos Complexos de Histocompatibilidade Major classe I, corroborando o diagnóstico de miopatia inflamatória.

A ressonância magnética cranioencefálica, tomografia computorizada torácica, abdominal e pélvica, mamografia, ecografia pélvica endovaginal, endoscopia digestiva alta e colonoscopia não demonstraram quaisquer sinais sugestivos de processo neoplásico.

FIGURA 1: FOTOGRAFIAS DA DOENTE ANTES E APÓS TRATAMENTO COM PREDNISOLONA E IMAGENS DE RESSONÂNCIA MAGNÉTICA FACIAL



Iniciou terapêutica com prednisolona 0.5mg/Kg/dia (30mg/dia), com redução marcada da tumefação temporal e normalização dos valores da PCR, VS e CK.
Conclusão: contrariamente à sua apresentação mais típica, apresenta-se um caso de miosite focal dos músculos mastigatórios, uma forma raramente documentada em humanos. Investigações recentes sugerem uma distinta origem embrionária para os músculos estriados craniofasciais e músculos dos membros, levantando a hipótese de que um diferente mecanismo de doença possa estar envolvido na miosite dos músculos mastigatórios.

172 - MESENTERIC VASCULITIS IN SYSTEMIC ERYTHEMATOUS LUPUS: A RARE AND SEVERE COMPLICATION

Maria Helena Lourenço[1, 2], Ana Bento da Silva[1, 2], Rita Pinheiro Torres [1, 2], Nuno Pina Gonçalves[1, 2, 3], Santiago Andres Rodrigues Manica[1, 2], MJ Gonçalves [1, 2], Inês Silva[1, 4], Maria Manuela Costa[1], Jaime C. Branco[1, 2, 4]
[1]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [2]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [3]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [4]NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

Introduction: Systemic erythematous lupus (SLE) is a multisystemic autoimmune disease. 50% of the patients experience gastrointestinal (GI) symptoms (mainly related to medication or infection): the most common being nausea, vomiting, anorexia, and abdominal pain1. Mesenteric vasculitis is a severe and rare complication of SLE (0.2-9.7%) and one of the most frequent causes of severe acute abdominal pain[2]. We present a case of a woman with a 20-year-diagnosis of SLE admitted to our inward department following a vascular flare of the disease and GI symptoms, with a mesenteric vasculitis diagnosis after a thorough investigation.
Clinical case: A 57-year-old Colombian female previously diagnosed with SLE (positive anti-double stranded DNA antibodies (anti-dsDNA), high titres of antinuclear antibodies, low levels of complement fraction (C) 3 and 4, normocytic and normochromic anaemia, malar rash, polyarthralgias, Raynaud phenomena, pleurisy, chronic kidney disease (stage III of the Kidney Disease: Improving Global Outcomes (KDIGO) classification)3 and peripheral nervous system involvement) was admitted to the Rheumatology inward department in March 2020, due to a vascular and articular flare of the disease (Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) 34). This followed a septic shock with origin in a urinary tract infection, worsened by a hypovolemic shock due to dehydration. The patient was previously medicated with prednisolone (PDN) 15mg/day, having multiple drugs intolerances. At admission, she presented bloody diarrhoea and diffuse abdominal pain, digital ulcers of both feet and hands, with necrosis of several toes, with palpable arterial pulses (worsened by Raynaud phenomena and vasopressor therapy) and polyarthralgias without arthritis. Blood analysis showed low levels of both C3 and C4, high titres of anti-dsDNA, anaemia and leukopenia. Due to the GI symptoms, an abdominal computed tomography (CT) scan was performed and revealed colonic thickening, especially in the rectosigmoid junction; colonoscopy showed ulcered areas mainly located in the

sigmoid and rectum and biopsy presented a necrotizing inflammatory process with large ulceration and fibrinous-granulocyte exudate; the immunochemistry was negative for both cytomegalovirus and herpes simplex infection. Stool sample culture tests were negative. Gastroenterology evaluation was in favour of a mesenteric vasculitis due to SLE. Pulses of methylprednisolone were administered (1000mg/day; 3 days), followed by PDN 1mg/kg/day and hydroxychloroquine 400mg/day. Cyclophosphamide was also started (NIH regimen)4. CT scan was repeated with resolution of the colonic thickening. The digital ulcers and necrosis of the toes also improved with therapy. Considering her stability (SLEDAI 12), the patient was discharged and followed in the outpatient clinic with a scheduled cyclophosphamide scheme. During a 9-month period of follow-up (rituximab as maintenance therapy), there was no recurrence of the GI symptoms and there was a complete resolution of the digital ulcers and necrosis.

Discussion: Mesenteric vasculitis is a rare manifestation of SLE; it leads to ischemia of the superficial layers, which can progress to deeper ulcers, causing perforation, haemorrhage, and eventually death5. Corticotherapy is the primary treatment, although cyclophosphamide may be needed. This patient responded well to the therapy and there were no more GI relapses. This case enhances the importance of being aware of all new symptoms during the course of the disease, allowing a fast and appropriate treatment.

202 - MYOCARDITIS IN ADULT STILL DISEASE: A RARE OCCURRENCE IN A RARE DISEASE

Nuno Pina Gonçalves[1, 2, 3], Rita Pinheiro Torres [2, 3], Agna Neto[1, 3], Santiago Andres Rodrigues Manica[2, 3], Maria João Gonçalves[2, 3], Maria Manuela Costa[3], Nelson Vale[4], Sandra Falcao[3], Jaime C. Branco[2, 3]

[1]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [2]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [3]Serviço de Reumatologia, Centro Hospitalar Lisboa Ocidental | Hospital Egas Moniz, Lisboa, Portugal, [4]Department of Cardiology, Hospital de Santa Cruz, Centro Hospitalar Lisboa Ocidental, Lisboa, Portugal

Introduction: Adult Still's disease (ASD) is a rare inflammatory disease mainly manifested by fever, arthritis and evanescent rash. Although rare, myocarditis is a potentially fatal complication that can lead to arrhythmias, heart failure and cardiac tamponade. Clinical case: Male, 39 years old, with no relevant medical history, started a clinical setting of high fever (42.5ºC), headache, sore throat, myalgia and polyarthralgia (shoulders, wrists and knees), pink non-pruritic rash (initially on the trunk with progression to the arms and legs that got worse with the fever spikes) and abdominal discomfort. He was admitted suspecting of Lyme disease. Blood cultures were negative but two cycles of antibiotics were made (ceftriaxone and doxycycline followed by ciprofloxacin and metronidazole) along with hydrocortisone 200mg/day. A colonoscopy with biopsies was performed with no findings. A presumptive diagnosis of Crohn's disease was made, being discharged with messalazine and corticosteroids. Two weeks after he was readmitted due to new onset chest pain and worsening of joint pain. Electrocardiogram showed an ST elevation in I, II, AvL, AvF, and V2-V6 leads. Cardiac magnetic resonance showed acute myopericarditis evidenced by oedema and interstitial fibrosis of the myocardium. An extensive work-up was made, with the following remarkable results: leucocytosis, high ferritin (>10000µg/L), C-reactive protein (VALOR) and sedimentation rate (120mm/h). After excluding coronary heart disease, the patient was transferred to the rheumatology ward. At admission polyarthritis of the shoulders, elbows, wrists and knees was present, although with no major functional limitation, along with bilateral basal crackles. Laboratory work-up showed marked neutrophilic leucocytosis (19,9x103/L), normochromic, normocytic anaemia (10.5g/L), elevation of liver enzymes (AST 44U/L, ALT 33U/L), nt-proBNP (528 pg/mL), CRP (26.1 mg/dL) and ESR (120mm/h). A diagnosis of ASD was made and prednisolone 1mg/Kg/day was started. However, fever recurred along with incapacitating polyarthritis of the shoulders, wrists, knees and ankles and a new onset of resting dyspnoea. A chest x-ray showed a worsening of the pleural effusion. Meropenem was started along with 3 consecutive pulses of 1000mg of methylprednisolone. A thoracentesis was performed and 100mL of inflammatory sterile pleural effusion were collected. On day 6, the patient experienced palpitations with a heart rate of 250bpm. A supraventricular tachycardia is diagnosed and reversed. Anakinra 100mg/day and methotrexate 15mg/week were started, with resolution of dyspnoea, pleural effusion and polyarthritis. At three months follow-up oligoar-

thritis recurred (wrists, right knee) with high ferritin (>500µg/L). Due to secondary failure to anakinra, synoviorthesis of the joints was performed and tocilizumab 162mg was prescribed. After 2 months of tocilizumab treatment, persistent arthralgia (wrists and knees) with morning stiffness and maintained elevated ferritin prompted switch to canakinumab 4mg/Kg/dose every 4 weeks with a complete symptom remission and ferritin levels returning to normal range values.

Discussion: The occurrence of pleuritis, acute respiratory syndrome and myocarditis prompted quick therapeutic initiation that was shown to be life-saving. Despite the patient not having developed macrophage activation syndrome (MAS), close monitoring is warranted since there is no evidence on how to predict ASD course, MAS can occur at any time and severe disease manifestations are related with high morbidity and mortality rate.

245 - CALCINOSE PARAVERTEBRAL NA ESCLEROSE SISTÉMICA – UMA LOCALIZAÇÃO ATÍPICA

Maria Seabra Rato[1], Filipe Oliveira Pinheiro[1], Alexandra Bernardo[1], Teresa Martins-Rocha[1], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal

Introdução: A calcinose é uma manifestação bem conhecida da esclerose sistémica (ES), afetando cerca de 1 em cada 3 doentes. A sua fisiopatologia não é completamente conhecida embora alguns estudos apontem o stress mecânico, a hipoxia tecidular e a inflamação crónica como os principais mecanismos envolvidos. As localizações mais frequentes são os dedos das mãos, os antebraços e os cotovelos. A forma paravertebral é uma localização rara, com apenas alguns casos descritos na literatura, podendo condicionar uma importante limitação funcional.

Caso clínico: Doente do sexo feminino, 60 anos de idade, seguida em consulta por ES forma cutânea limitada com 6 anos de evolução apresentando envolvimento microvascular, articular e cutâneo, medicada com nifedipina 60mg id e pentoxifilina 400mg tid. Apresentava positividade para anticorpos antinucleares (no título de 1/1000) e para o anticorpo anti-centrómero. Sem calcinose objetivada anteriormente ou envolvimento cardiopulmonar documentado. Tinha como antecedentes pessoais de relevo osteoporose fraturária, estando sob terapêutica com ácido zoledrónico desde há 4 anos, assim como colangite biliar primária medicada com ácido ursodesoxicólico. Em consulta de rotina, referiu dorso-lombalgia de ritmo mecânico e tumefação na região paravertebral esquerda com 4 meses de evolução. Negava a existência de traumatismo prévio, mas relacionava o início da dor com a execução de um movimento súbito de flexão lombar. Ao exame objetivo, apresentava uma tumefação paravertebral esquerda, ao nível da transição dorsolombar, de consistência pétrea, imóvel, com aproximadamente 6 cm de maior diâmetro e dolorosa à palpação. Não se observava limitação da amplitude dos movimentos, diminuição de força muscular ou sensibilidade dos dermátomos torácicos e dos membros inferiores. Realizou radiografia e, posteriormente, tomografia (TC) da coluna dorso-lombar, que evidenciaram uma lesão hiperdensa nos músculos paravertebrais da região posterior esquerda com extensão da 12º vértebra dorsal à 3ª vértebra lombar, sem osteólise ou compromisso neurológico associados. Estes achados foram compatíveis com o diagnóstico de calcinose paravertebral. Analiticamente, apresentava doseamento de creatinina sérica, cálcio ionizado, fósforo, paratormona e enzimas musculares dentro dos limites da normalidade. Revendo o processo clínico da doente, verificou-se que já apresentava em TC torácico realizado 5 anos antes, microcalcificações dos músculos paravertebrais esquerdos, embora não tenham sido relatadas. Como plano terapêutico, foi instituída otimização da terapêutica analgésica, com boa resposta clínica. A doente mantém seguimento regular em consulta de Reumatologia.

Conclusão: A calcinose paravertebral na região dorsolombar é uma manifestação rara da esclerose sistémica que pode ser causa de dor local, destruição óssea e complicações neurológicas por compressão da medula espinhal ou de raízes nervosas. O seu tratamento é desafiante porque não existe nenhuma terapêutica com eficácia comprovada na redução das lesões, estando muitas vezes limitado à terapêutica analgésica, ou em casos mais graves, à remoção cirúrgica. No caso descrito, a doente estava medicada com bifosfonato pela osteoporose que não impediu a progressão das lesões de calcinose. A documentação deste caso clínico alerta para a possibilidade de localizações pouco usuais da calcinose na ES e para a necessidade de identificação de terapêuticas farmacológicas dirigidas capazes de prevenir ou reduzir as lesões de calcinose.

256 - O PAPEL DA BIÓPSIA SINOVIAL POR AGULHA ECO-GUIADA, NO DIAGNÓSTICO DE INFECÇÕES OSTEOARTICULARES

Hugo Parente[1], Catarina Soares[1], Francisca Guimarães[1], Soraia Azevedo[1], Carmo Afonso[1], Filipa Teixeira[1]

[1]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal

Introdução: O diagnóstico diferencial de artropatias crónicas com manifestações atípicas pode ser um desafio. A biópsia sinovial por agulha é um procedimento bem tolerado e seguro1 que pode ser a chave diagnóstica no caso de infeções menos comuns (fungos, micobactérias, espiroquetas, etc), seja pela sua identificação direta e in situ, ou pela identificação de alterações microscópicas úteis no diagnóstico: granulomas, infiltração neutrofílica perivascular, etc. O uso adjuvante da ecografia minimiza as constrições técnicas, com taxas de sucesso de 82-96% na obtenção de amostras[2].

Objetivo: Apresentação de quatro casos de artropatia crónica cuja biópsia sinovial revelou infecção.

Casos clínicos:

A) Mulher de 57 anos, com monoartrite do punho, com 5 meses de evolução. Na ecografia, apresentava sinovite de grau 3, com sinal de power-doppler. O estudo microbiológico da membrana sinovial foi negativo, mas o exame histológico identificou estruturas sugestivas de esporos fúngicos que, juntamente com altos níveis séricos de antigénio de Aspergillus, apoiaram o diagnóstico de artrite fúngica. Foi medicada com voriconazol, com melhoria clínica gradual.

B) Homem de 44 anos, sem história prévia de artrite, apirético, com monoartrite crónica do joelho. O líquido sinovial era hipercelular, sem cristais, mas com isolamento de Staphylococcus Aureus. Efectuou antibioterapia dirigida, sem qualquer melhoria. A histologia da membrana sinovial revelou esporos fúngicos e, tal como no caso anterior, foram identificados níveis séricos elevados do antigénio de Aspergillus. Iniciou terapêutica com voriconizol, com resolução do quadro.

C) Homem de 79 anos, diabético, com monoartrite do punho com 3 meses de evolução. Dada a condrocalcinose radiográfica, assumiu-se inicialmente o diagnóstico de doença por deposição de cristais de pirofosfato de cálcio e foi medicado com anti-inflamatórios, sem melhoria. Ecograficamente, apresentava sinovite do punho, tenossinovite dos flexores e 4º compartimento. No estudo da membrana sinovial, isolou-se Staphylococcus Aureus pelo que iniciou antibioterapia dirigida, durante 3 meses, com resolução completa.

D) Homem de 79 anos, com oligoartrite dos punhos com 8 anos de evolução. O envolvimento inicial era monoarticular, com melhoria parcial com anti-inflamatórios e tratamento local. Nos últimos 3 anos, iniciou envolvimento bilateral dos punhos, sem outros sintomas. O estudo analítico apresentava elevação dos parâmetros inflamatórios e factores reumatóides negativos. No líquido sinovial, nunca se identificaram cristais. A ressonância magnética mostrou sinovite extensa dos punhos bem como alterações erosivas e ainda tenossinovite dos flexores e extensores do punho. O estudo da membrana sinovial foi negativo para micobactérias e Tropheryma Whipplei, identificando, porém, Klebsiella Pneumoniae e Staphylococcus Caprae. Foi medicado com trimetropim/sulfametoxazol, que se encontra em curso.

Discussão/conclusão: Estes casos sublinham o potencial diagnóstico da biópsia sinovial por agulha. Dado que uma infecção de baixo grau se manifesta, comumente, por artrite crónica e destruição articular, é fundamental suspeitar da presença de agentes atípicos (fungos, micobactérias, Borrelia Burgdorferi, Tropheryma Whipplei) nestes casos. A biópsia sinovial permite a sua identificação através de coloração direta, cultural e análise por PCR, bem como estudo histopatológico. Pode complementar ou até substituir procedimentos menos invasivos, integrando dados microbiológicos e histopatológicos, essenciais ao diagnóstico de artropatias de origem indeterminada.

269 - SÍNDROME DE BUSCHKE-OLLENDORFF – A RESPEITO DE UM CASO CLÍNICO

Diogo Guimarães da Fonseca[1], Beatriz Samões[1], Tiago Beirão[1], David Afonso João[2], Antónia Furtado[2], Romana Vieira[1], P Pinto[1]

[1]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [2]Serviço de Anatomia Patológica, Centro Hospitalar de Vila Nova de Gaia / Espinho, Vila Nova de Gaia, Portugal

Introdução: A síndrome de Buschke-Ollendorff (SBO) é uma doença rara, autossómica dominante, caracterizada pela presença de lesões ósseas escleróticas, nomeadamente osteopoiquilose, localizadas principalmente nas regiões epifisárias dos ossos tubulares. A presença de outro tipo de lesões escleró-

ticas, mais concretamente melorreostose, também se encontra descrita. Esta síndrome caracteriza-se ainda pela presença de nevos cutâneos, que do ponto de vista histopatológico afetam principalmente o colagénio e as fibras elásticas, com feixes de colagénio espessos e largos e fibras elásticas aumentadas, irregulares ou fragmentadas. A sua identificação obriga, no entanto, a recurso a colorações específicas, nomeadamente tricrómico de Masson ou orceína. Clinicamente surgem como pequenas pápulas disseminadas com coloração igual à da pele ou amarelada. A mutação frequentemente encontrada consiste numa perda de função, em heterozigotia, no gene LEMD3, que codifica para a proteína da membrana nuclear interna, LEMD3, que interage com as vias de sinalização de BMP e TGF-beta.

Caso clínico: Homem de 49 anos de idade, encaminhado para consulta de Reumatologia por dor isquiática direita associada a lesões blásticas a nível ilíaco direito e já submetido a amplo estudo etiológico nos últimos dois anos, nomeadamente com biópsia óssea dirigida sem qualquer evidência de processo neoplásico. Clinicamente o doente apresentava dor palpação da tuberosidade isquiática direita, sem qualquer relação com o local de descrição da lesão óssea biopsada, e apresentava ainda lesões cutâneas papulares a nível das nádegas e face posterior da coxa esquerda. A avaliação do estudo radiográfico previamente realizado, revelou a presença de múltiplas lesões escleróticas, arredondadas, infracentimétricas a sugerir osteopoiquilose a nível da extremidade proximal dos fémures. O estudo radiográfico foi alargado e revelou a presença destas mesmas lesões a nível das epífises dos ossos longos, cabeças dos metacarpos e carpo, cabeças dos metatarsos e tarso, glenoide e acrómio, acetábulo, ossos da pelve e epífise femoral. Por suspeita de SBO foi pedida revisão da histologia cutânea que após a realização de técnicas histoquímicas específicas, mais concretamente orceína, evidenciou uma derme com espessamento significativo de feixes de colagénio, rodeados por fibras elásticas. De realçar inexistência de envolvimento axial e distribuição bilateral. Tendo em conta a suspeita clínica de Síndrome de Buschke-Ollendorff, foi realizado estudo genético, tendo sido identificado uma variante em heterozigotia no gene LEMD3, corroborando dessa forma o diagnóstico. De realçar que a queixa loco-regional do doente estava relacionada com uma bursite isquiática, tendo sido alvo de tratamento dirigido com melhoria clínica.

Conclusão: Este caso reporta um diagnóstico raro, mas que deve ser considerado no diagnóstico diferencial de lesões ósseas escleróticas, muitas vezes identificadas acidentalmente em exames de radiografia convencional. A referenciação tardia deste doente à consulta de Reumatologia, fez com que fosse alvo de múltiplos exames complementares de diagnóstico pela suspeita única de etiologia neoplásica. A identificação precoce desta síndrome bem como a respetiva tranquilização do doente no que concerne ao seu carácter benigno é importante e realça o papel da Reumatologia no estudo das diversas doenças ósseas metabólicas e hereditárias.

CASOS CLÍNICOS (COM EXPOSIÇÃO)

019 - CHRONIC CHIKUNGUNYA DISEASE: A CASE OF VIRUS-INDUCED EROSIVE AND DEFORMING POLYARTHRITIS

Matilde Bandeira[1, 2], Eduardo Dourado[1, 2], JE Fonseca[1, 2]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introduction: Several viral infections cause musculoskeletal symptoms ranging from self-limited arthralgia to chronic arthritis. Severe musculoskeletal symptoms are typical of Chikungunya virus infection. In the acute phase, many patients develop disabling polyarthralgia or polyarthritis. Chronic Chikungunya is defined by persistent symptoms for over three months after infection and may mimic rheumatoid arthritis or peripheral spondylarthritis.

Outbreaks of Chikungunya virus took place mainly in African and Asian countries until 2013, after which local transmission began in the American continent. There are reports of local transmission in Europe (France and Spain) but none in Portugal. Reported cases of Chikungunya in Portugal between 2014 and 2018 were less than five, according to the European Centre for Disease Prevention and Control, all of which imported cases. Most cases were imported from Brazil and Portuguese-speaking African countries.

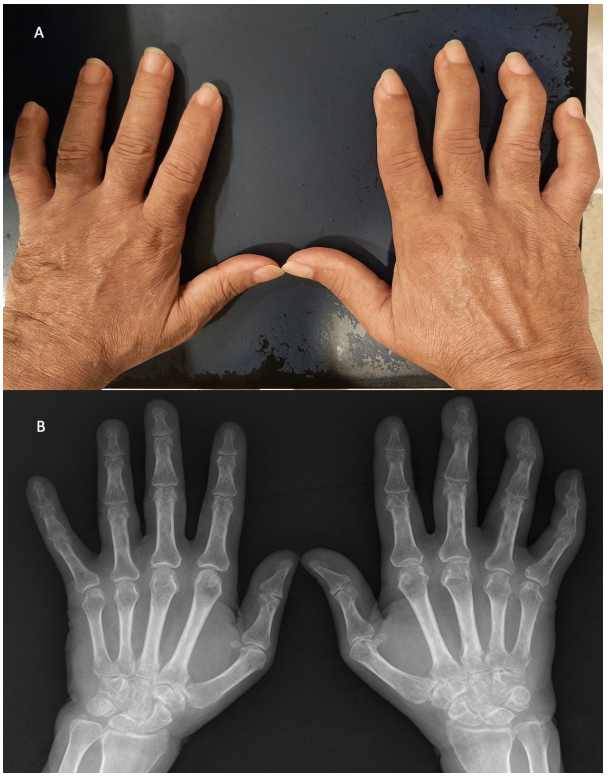
Clinical vignette: A 61 years-old man presented to

the Rheumatology clinic in January 2021 complaining of inflammatory polyarthralgia and morning stiffness of up to three hours since March 2020. At the onset of symptoms, during a trip to Angola, the patient also had a high fever and generalized myalgia that lasted two weeks. At the time of presentation, a weight loss of 14Kg in the previous nine months was reported (initial weight of 85Kg). The patient is of Indian descent, was born in Mozambique, lives in Portugal for over 40 years, and had a history of type 2 diabetes mellitus, traumatic subluxation of the left shoulder and past smoking habits.

There were 18 tender and 7 swollen joints on clinical examination, particularly the metacarpophalangeal (MCP) and proximal interphalangeal (PIP) joints. Swan neck deformity was noted on the third and fourth right fingers, and Boutonnière deformity on the fifth fingers bilaterally (figure 1).

Blood workup had no significant increase of inflammatory markers, rheumatoid factor and anti-citrullinated peptide antibody were negative, but positive IgG and negative IgM against Chikungunya virus were present. Hand ultrasonography documented synovitis with Doppler sign in the wrists, multiple MCP and PIP joints. Hand radiographs revealed exuberant osteopenia and erosions, mainly on right hand PIP joints.

FIGURE. A. PATIENT'S HANDS, AFTER DMARD TREATMENT; B. HAND RADIOGRAPH WITH EXUBERANT OSTEOPENIA AND EROSIONS



Treatment was started with 10 mg of prednisolone, hydroxychloroquine and methotrexate, but methotrexate dose optimization and the addition of sulfasalazine were necessary to maintain clinical response as the prednisolone was tapered.

A remarkable clinical improvement was noted with total remission of synovitis. However, fatigue and anorexia persisted.

Discussion: We present one of the few confirmed Chikungunya cases in Portugal with a more severe disease course than other published reports.

Chronic Chikungunya disease with polyarthritis is reported in 14% of Chikungunya infections. The mechanism by which the disease evolves to chronic arthritis in some but not all patients is still uncertain. However, known risk factors for chronicity include diabetes mellitus, osteoarthritis, severe immunological response during the subacute phase and age over 45 years. Chikungunya virus-related disease should be a part of the differential diagnosis of a seronegative rheumatoid arthritis-like clinical picture. Inquiry over recent or past travels and acute infection symptoms before the onset of the articular disease is essential to diagnose the disease.

021 - NEITHER ARMS NOR LEGS WERE FEELING WELL: A CASE OF ANTI-NOR-90 LUPUS MYOSITIS

Eduardo Dourado[1, 2], Filipe Cunha Santos[3], Inês Cordeiro[1, 2]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal

Introduction: The differential diagnosis of diminished muscle strength and myalgia is vast and includes neurologic diseases, fibromyalgia and myositis. When myositis is associated with a malar rash, dermatomyositis (DM) and systemic lupus erythe-

matosus (SLE) may be hard to distinguish. A careful clinical evaluation and selected immunological tests help to confirm a diagnosis.

Clinical vignette: A 50-year-old black female presented to the Rheumatology clinic with bilaterally diminished proximal strength and proximal myalgia of the four limbs. She had a 4-year history of arthralgia of the hands (morning stiffness longer than one hour), painless oral ulcers (more than twice a week), asthenia and increased hair loss. She had no history of dyspnea, dysphagia, dysphonia, telangiectasia, Raynaud phenomenon, digital ulcers, pitting scars, calcinosis, Gottron's papules or heliotrope rash. She was previously diagnosed with fibromyalgia by her assisting General Practitioner. No treatment had been started. On physical examination, the patient was walking with crutches and presented a malar rash sparing the nasolabial folds, bilateral sclerodactyly (mRSS 4), slight microstomy, manual muscle testing (MMT) 37/50 (right), 59/70 (left), and 4/18 fibromyalgia tender points.

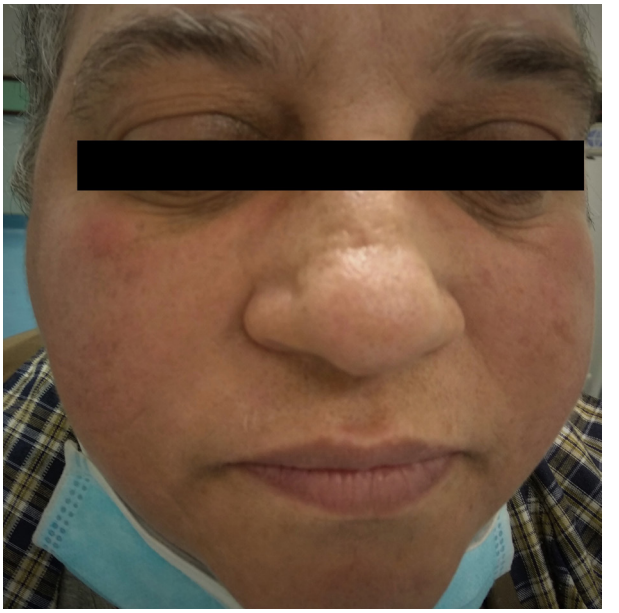
She had elevated inflammatory markers (erythrocyte sedimentation rate 80 mm/h, C-reactive protein 2.06 mg/dL), a normocytic normochromic anaemia (haemoglobin 10.8 mg/dL), slightly elevated creatine kinase (282 U/L) and normal aldolase (4.3 U/L).

Anti-nuclear antibodies (ANA) were positive up to the 1:160 titer and presented a homogeneous pattern (AC-1). The extractable nuclear antigen panel (including anti-Sm, anti-RNP, anti-SSA and anti-SSB) was negative, and there was no hypocomplementemia. Chemiluminescent immunoassay for double-stranded DNA (dsDNA) was positive (118 UI/mL, N<26), and immunoblot assay for systemic sclerosis (SSc)-associated antibodies revealed anti-NOR90 positivity. Thigh magnetic resonance imaging (MRI) and ultrasound (US) documented semimembranosus and semitendinosus myositis on the right thigh and biceps femoris myositis on the left thigh. Myositis was identified by STIR-sequence hyperintensity and asymmetrical volume of contralateral muscles in MRI and hyperechogenicity and asymmetrical volume of contralateral muscles in the US.

A diagnosis of SLE with mucocutaneous, articular and muscular involvements was assumed. The patient fulfils the 2019 EULAR/ACR classification criteria for SLE (ANA and dsDNA positivity, malar rash, inflammatory arthralgia) and has several non-criteria manifestations.

The patient is now under treatment with hydroxychloroquine and methotrexate and improved significantly. In the last appointment, the patient had no painful nor swollen joints, MMT was 142/150 and the skin had no lesions.

FIGURE. FIXED MACULAR ERYTHEMA OVER THE MALAR EMINENCES SPARING THE NASOLABIAL FOLDS FAVOURING THE DIAGNOSIS OF LUPUS-ASSOCIATED RASH



Discussion: Lupus myositis affects less than 5% of SLE patients. The risk is highest for black women. About half the patients with lupus myositis present with systemic sclerosis overlap features, and myositis may become clinically apparent late in the disease course. The characteristics of the malar rash are essential in the clinical distinction between SLE and DM. Most patients with lupus myositis have no myositis-associated autoantibodies identified. Anti-NOR90 is a rare autoantibody that binds the nucleolus-organizing regions and can be present in the serum of SLE, SSc or primary Sjögren's syndrome patients.

028 - RESPOSTA A ANAKINRA APÓS FALÊNCIA DE BDMARDS EM DOENTES COM ARTRITE REUMATOIDE E GOTA CONCOMITANTE – DESCRIÇÃO DE 2 CASOS

Filipe Oliveira Pinheiro[1], Maria Seabra Rato[1], Pedro Madureira[1], Georgina Terroso[1], Miguel Bernardes[1, 2], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar

Universitário de São João, Porto, Portugal, [2]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

A gota e a artrite reumatóide (AR) são duas artropatias inflamatórias comuns, com prevalências estimadas de 3% e 0,5-1%, respetivamente. As duas doenças podem coexistir no mesmo doente, embora de forma muito rara, com impacto no diagnóstico e tratamento. Curiosamente, a literatura diz-nos que a prevalência de gota nos doentes com AR é inferior à da prevalência na população geral, com diversas teorias postuladas para explicar esse fenómeno, entre as quais a de que o uso de anti-inflamatórios não-esteróides e corticoterapia poder mascarar as manifestações de gota, ou de a IL-6 ter propriedades uricosúricas. Clinicamente as duas doenças podem ser difíceis de distinguir, podendo apresentar doença poliarticular (mais comum com a gota de longa evolução), ou nódulos nas superfícies extensoras, que podem ser interpretados como nódulos reumatóides ou tofos gotosos. Até hoje, nenhum estudo avaliou o uso de anakinra e a resposta ao mesmo em doentes com gota e AR concomitante.

Os autores apresentam de seguida dois casos de doentes com AR cumprindo critérios de classificação ACR-EULAR 2010, com doença ativa apesar de instituição de csDMARD, e que permaneceram com atividade de doença após instituição de bDMARD não-antagonistas da IL-1, tendo apenas atingido target terapêutico após iniciação de anakinra.

O primeiro caso reporta-se a um indivíduo do sexo masculino, com AR diagnosticada aos 27 anos, e diagnosticado com gota aos 57 anos após biópsia de nódulo compatível com tofo gotoso, e com hiperuricemia de longa data; medicado com sulfassalazina (história de hepatotoxicidade ao metotrexato) e com falência primária ao etanercept, certolizumab e tocilizumab, apresentando DAS28 4V-PCR de 5,33 em julho de 2016, altura em que se decide iniciar anakinra, com ótima resposta, apresentando atualmente a doença em remissão (última avaliação em abril de 2020 com DAS28 4V-PCR de 1,42).

O segundo caso diz respeito a um indivíduo do sexo feminino, com AR desde os 52 anos, diagnosticada com gota aos 53 anos após apresentar hiperuricemia, tofos gotosos e erosões em saca-bocados em diversas metacarpo-falângicas e interfalângicas proximais e distais; por manter atividade de doença elevada e toxicidade hepática ao metotrexato, decidido iniciar tocilizumab (suspenso 3 meses após por toxicidade hepática), feito posteriormente switch para etanercept, com resposta modesta, apresentando após 1 ano DAS28 4V-PCR de 3,70; decidido nessa altura realizar terapêutica com anakinra, com ótima resposta, apresentando após um ano de terapêutica DAS28 4V-PCR de 2,17.

Assim, torna-se importante avaliar de forma cuidada a coexistência de artrite reumatóide e de gota, sobretudo em doentes com doença poliarticular e refratária ao tratamento instituído, pela sua importância clínica, mas sobretudo no que respeita à seleção do tratamento mais adequado para os doentes.

Até à data, este é o primeiro relato sobre resposta a anakinra em doentes com AR e gota com história de ineficácia a outros bDMARDs.

033 - PARAPARÉSIA NUM DOENTE COM ARTRITE PSORIÁTICA – UM DESAFIO DIAGNÓSTICO

Filipe Oliveira Pinheiro[1], Maria Seabra Rato[1], Pedro Madureira[1], Iva Brito[2, 3]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal, [3]Unidade de Reumatologia Pediátrica e do Jovem Adulto, Centro Hospitalar e Universitário de São João, Porto, Portugal

Introdução: A síndrome de Guillain-Barré (SGB) é uma polineuropatia desmielinizante imunomediada aguda com apresentação heterogénea, surgindo geralmente após um evento infecioso. A sua existência em doentes com artrite psoriática é limitada a casos isolados, nomeadamente em associação com agentes farmacológicos, particularmente os anti-TNFα.

Descrição do Caso Clínico: Homem de 41 anos, com hipertensão arterial, diabetes mellitus tipo 2, dislipidemia e artrite psoriática com 7 anos de evolução, atualmente sob terapêutica com metotrexato subcutâneo 20mg/semana. Recorreu ao Serviço de Urgência com quadro de 2 dias de evolução de febre, mialgias e diarreia, associado a fraqueza dos membros inferiores de intensidade progressiva. À admissão apresentava paraparésia grau 2 e reflexos osteotendinosos abolidos nos membros inferiores, sem alterações evidenciáveis nos membros superiores. Analiticamente sem aumento dos marcadores inflamatórios sistémicos, tomografia cerebral e coluna sem alterações de relevo. Estudo do líquido cefalorraquidiano (LCR) com pleocitose linfocítica com proteinorráquia normal.

Apresentou evolução desfavorável com progressão ascendente dos défices neurológicos, com necessidade de admissão na Unidade de Cuidados Intermédios. Após exclusão de outras causas, realizou estudo serológico vírico com positividade inaugural para HIV1, com documentação de cópias de RNA viral no LCR. Fez ressonância magnética da coluna sem evidência de mielite. Assumida SGB em contexto pós-infecioso (primoinfeção a HIV), tendo cumprido 5 dias de imunoglobulina endovenosa. Iniciou posteriormente terapêutica antirretroviral com FTC/TDF/DTG. Teve uma evolução favorável, com melhoria dos défices neurológicos presentes à admissão, e com recuperação sem sequelas.

Conclusões: A SGB foi raramente descrita em associação ao HIV e à artrite psoriática, o que torna este um caso particularmente relevante e desafiante, sobretudo em formas com apresentação clínica menos típica e na ausência de infeção por HIV previamente conhecida.

037 - A CASE OF RHUPUS DIAGNOSED IN PREGNANCY IN A PATIENT WITH RHEUMATOID ARTHRITIS AND ANTIPHOSPHOLIPID SYNDROME

Ana Margarida Duarte-Monteiro[1, 2], Fernando Saraiva[1, 2], JE Fonseca[1, 2]

[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

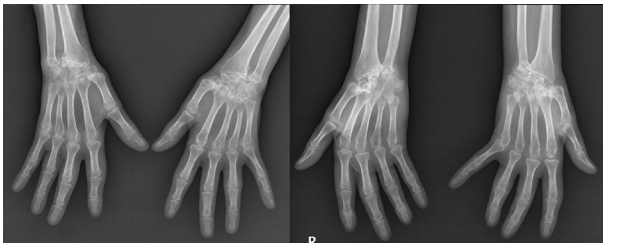
Introduction: While overlap syndromes are relatively common, Rhupus is a rare and underdiagnosed entity, with a reported prevalence in Systemic Lupus Erythematosus (SLE) patients between 0.09 and 9.7%. The mean age of diagnosis is 45 years, and it is more prevalent in women. The diagnosis is considered when there are characteristics and manifestations of SLE and rheumatoid arthritis (RA), and the most agreed diagnostic definition consists in the fulfillment of both the classification criteria for SLE (SCLICC, ACR) and RA (ACR, ACR/EULAR). To date, no universally accepted diagnostic criteria for Rhupus have been developed, which creates barriers to the determination of the real prevalence of this entity. Consequently, in clinical practice, there are often doubts about the classification of patients with both SLE and RA manifestations, with consequences in defining the best follow-up and management strategies.

Antiphospholipid syndrome (AFS) can be secondary to other systemic autoimmune diseases, predominantly SLE, or a primary entity. Nevertheless, it is not clear if a diagnosis of AFS is a predictor of the later development of SLE, with some limited retrospective and prospective studies suggesting no actual relation. There seem to be no significant differences in the prevalence of antiphospholipid antibodies between rhupus and SLE patients.

Clinical vignette: We present the case of a 40-year-old woman diagnosed with RA at 22 years old. Rheumatoid factor (RF) and anti-citrullinated peptide antibodies (ACPA) were negative, and she had erosive disease involving the metacarpophalangeal joints, wrists, elbows and knees, fulfilling the RA ACR/EULAR 2010 classification criteria. At this time, Antinuclear Antibodies (ANAs) were also found to be negative. The patient had a history of infertility and later obstetric morbidity (1st pregnancy at 35 years old with fetal loss at 25 weeks). During a second pregnancy a year later, lupus anticoagulant and anticardiolipin were found to be positive in two separate determinations, and she fulfilled AFS criteria. At 29 weeks, the patient developed subacute cutaneous lupus, and immunological disturbances appeared de novo (ANA + 1/640, anti-dsDNA + 577). Serum complement was normal. At this time, SLE criteria (SLICC 2012) were met. Thus, the diagnosis of Rhupus was made.

Conclusions: We present a case in which a diagnosis of Rhupus was made during pregnancy in a patient with a previous diagnosis of RA and AFS. In 2/3 of Rhupus patients, RA is the first diagnosis, as was the case in our patient. Rhupus patients with an initial RA diagnosis are usually younger than patients with isolated RA. SLE involvement is usually manifested

FIGURE. HAND X-RAY OF A RHUPUS PATIENT WITH EROSIVE DISEASE



by hematologic, skin, serous and/or kidney alterations, while arthritis usually has an RA like pattern and is often erosive and deformative. During pregnancy, increase in estrogen and progesterone levels may induce a switch in cytokine polarization from Th1 to Th2, which can have a role in modulation of SLE and RA characteristics in Rhupus patients. We found only another case report of transition of RA manifestations to SLE during pregnancy. While no definite conclusions can be made, it is also worth to note the development of SLE after a previous diagnosis of AFS in a patient with RA.

046 - PSORIASIS AND PSORIATIC ARTHRITIS – A STELLAR RESPONSE TO USTEKINUMAB

Ana Margarida Duarte-Monteiro[1, 2], Eduardo Dourado[1, 2], Ana Valido[3], Ortins-Pina, Ana[4]

[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Serviço de Reumatologia, Unidade Local de Saúde do Litoral Alentejano, Santiago do Cacém, Portugal, [4]Zentrum für Dermatopathologie Freiburg, Freiburg, Germany

Introduction: Psoriatic arthritis affects around 30% of psoriasis patients. While cutaneous and joint disease activity are frequently not parallel, simultaneous severe and extensive involvement is not rare. Ustekinumab is approved for the treatment of both psoriasis and psoriatic arthritis.

Clinical Vignette: A 23 years-old black female was transferred from a Guinea-Bissau hospital after one year of hospitalization due to extensive dermatosis and deforming arthritis. Symptom onset had happened five years earlier and were progressively worsening.

In Portugal, the patient was admitted to the Dermatology ward and was jointly evaluated by dermatologists and rheumatologists. The diagnosis of psoriasis was histologically confirmed. The patient had severe and extensive involvement of the whole tegument, with a Psoriasis Area Severity Index (PASI) of 26/72. The scalp and nails were severely affected. The patient also had peripheral arthritis affecting the metacarpophalangeal, proximal and distal interphalangeal joints, knees and ankles. The arthritis was clinically active with a DAS28-PCR of 4.94, and significant structural damage was already present, with severe functional impact (HAQ-DI 2.125). Imaging evaluation with both conventional radiographs and musculoskeletal ultrasound confirmed the presence of advanced erosive disease with joint deformity.

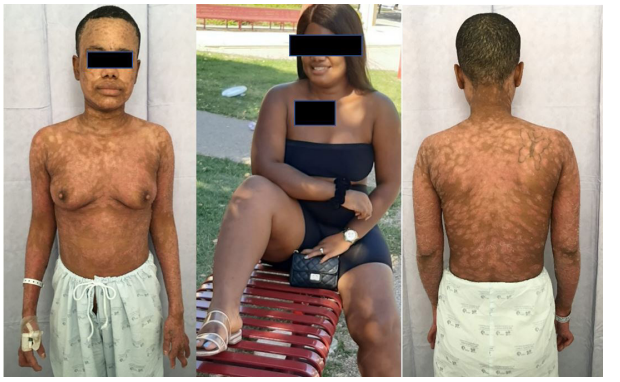
At this point, the patient fulfilled the criteria for the diagnosis of psoriatic arthritis. Additionally, the patient had a substantial weight loss, night sweats and bilateral opacities on chest x-ray. She was diagnosed with miliary tuberculosis and was treated with tuberculostatic therapy for six months with a good outcome.

The patient was initially treated with conventional synthetic disease-modifying anti-rheumatic drugs (csDMARDs) with no skin or joint improvement. Considering that the patient had a miliary tuberculosis history, the multidisciplinary decision was to start biological DMARD (bDMARD) therapy with ustekinumab. At six months, the patient had achieved a cutaneous complete clinical response, with a PASI of 0/72 and a good articular response (DAS 28-PCR 2.46), positively impacting function (HAQ-DI 1.000), quality of life and self-image.

Three years after starting ustekinumab, the patient maintained the cutaneous remission and had also achieved joint remission. There were no reported adverse events of therapy, including no tuberculosis reactivation.

Conclusions: We report the case of a patient with concomitant severe psoriasis and incapacitating psoriatic arthritis. This case highlights the importance of multidisciplinary care, facilitating timely access to specialized therapeutic techniques, such as skin biopsy and musculoskeletal ultrasound. Defining a diag-

FIGURE. CUTANEOUS RESPONSE TO USTEKINUMAB



nosis and performing the pre-bDMARD safety checklist was essential to a safe and effective therapeutic decision. This case further supports the reported efficacy of ustekinumab in both skin and joint involvements and its documented safety in patients with a history of tuberculosis. Despite the late initiation of effective therapy, it is worth noting the profound and long-lasting impact caused by ustekinumab.

048 - OSTEOMYELITIS CAUSED BY CLOSTRIDIUM PERFRINGENS IN A PATIENT WITH RHEUMATOID ARTHRITIS

Rita Cunha[1], Catarina Ambrósio[1], Nuno Neves[1], Renata Aguiar[1], Anabela Barcelos[1, 2, 3]

[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, EPE, Aveiro, Aveiro, Portugal, [2]CHRC, Comprehensive Health Research Center, CEDOC, NOVA Medical School, Universidade Nova de Lisboa, Lisboa, Portugal, [3]Public Health Research Centre, NOVA National School of Public Health, Public Health Research Centre, Universidade NOVA de Lisboa, Portugal, Lisboa, Portugal

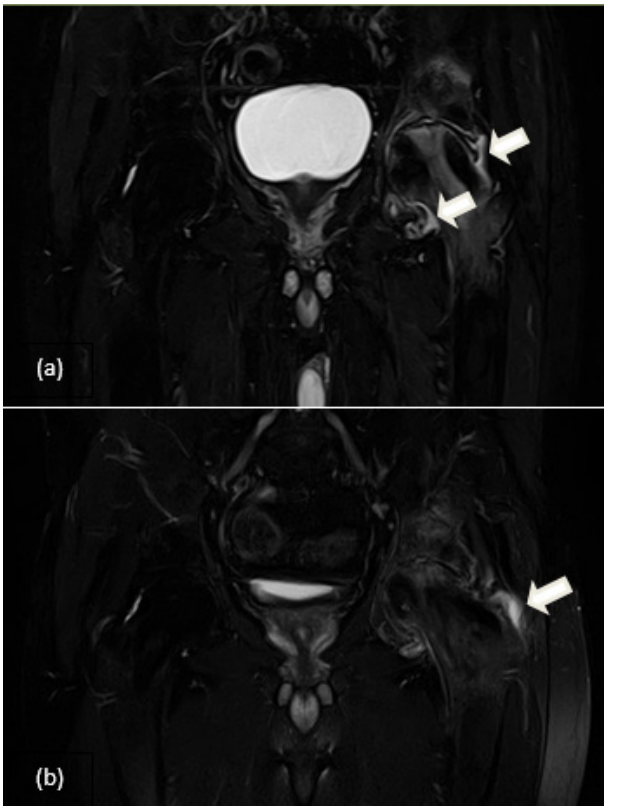
Introduction: Osteomyelitis caused by anaerobic bacteria is very rare. There are some reports of spondylodiscitis caused by Clostridium perfringens, but we are not aware of any previous reports of osteomyelitis caused by this bacterium affecting femur and acetabulum.

Clinical Case: We describe a case of a 48-year-old male with rheumatoid arthritis under intravenous (iv) monthly tocilizumab and bilateral hip osteoarthritis presenting with new onset severe left hip pain and fatigue. Laboratory tests showed leucocytosis with neutrophilia and elevation of inflammatory markers. A magnetic resonance imaging (MRI) revealed increased acetabulum and femoral head bone marrow intensity that reflects bone oedema and effusion. Bone culture was positive for Clostridium perfringens. A diagnosis of osteomyelitis, caused by Clostridium perfringens, affecting femur and acetabulum was established. The patient initiated treatment with iv G penicillin and iv clindamycin. Clindamycin was switched for cotrimoxazole due to a cutaneous reaction. The treatment was shifted to oral amoxicillin and clavulanic acid that was maintained continuously. A follow-up MRI, 2 months after iv antibiotic therapy, demonstrated reduction of bone marrow oedema and effusion and, despite the functional limitation, the patient has controlled pain and refuses surgery.

Discussion: Clostridium perfringens is a Gram-positive, anaerobic, spore-forming bacillus, ubiquitous in the soil and water and it is also considered a commensal of the gastrointestinal tract. Septic arthritis and osteomyelitis due to Clostridium perfringens is an unusual clinical occurrence that is rarely documented, and most cases were associated with traumatic penetrating injury, which wasn't present in our case or haematogenous route. However, in some cases, the aetiology is unknown, as in this patient.

Conclusion: This case emphasizes the importance of considering alternative explanations for new onset of hip pain, even in patients with osteoarthritis, especially if immunocompromised. A high level of clinical suspicion is required to a correct diagnosis, early treatment and to achieve a better outcome.

FIGURE I: CORONAL T2 -WEIGHTED IMAGE WITH FAT SUPPRESSION AT BASELINE AND AT FOLLOW-UP



REFERENCES:

Yong CH, Lam M. Discitis and Clostridium perfringens bacteraemia. BMJ Case Rep. 2017;2017:bcr2017220646.

Fauser DJ, Zuckerman JD. Clostridial septic arthritis: case report and review of the literature. Arthritis and rheumatism. 1988;31(2):295-8

053 - DOENÇA DE STILL DO ADULTO INDUZIDA POR PEGINTERFERÃO BETA EM DOENTE COM ESCLEROSE MÚLTIPLA

Bruno Miguel Fernandes[1], Miguel Bernardes[1], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal

Introdução: A Doença de Still do Adulto (SOAD) é uma doença inflamatória multissistémica que afeta principalmente jovens adultos. A tríade clássica inclui picos febris diários, artrite e rash cutâneo evanescente e a elevação da ferritina é o achado laboratorial mais característico. A fisiopatologia não está totalmente esclarecida, sendo que alguns dados apontam para que a doença surja após a ativação de uma cascata inflamatória dominada pelo sistema imune inato por um determinado trigger.
Caso clínico: Doente do sexo feminino, 51 anos, com esclerose múltipla e dislipidemia. Em janeiro de 2018 foi enviada à Reumatologia por quadro de rash cutâneo pruriginoso afetando a região do decote com posterior generalização para o dorso e membros após a terceira administração de peginterferão-beta1a (subcutâneo a cada 2 semanas). O fármaco foi suspenso e, posteriormente, apresentou poliartrite simétrica das interfalângicas proximais das mãos e artralgias de ritmo inflamatório nos tornozelos. Associadamente, referia odinofagia, disfagia para sólidos, mialgias e "arrepios de frio" sem objetivação de febre. Negava fenómeno de Raynaud ou outras queixas sistémicas. O exame objetivo mostrou tenossinovite dos flexores de ambas as mãos, blefarite e rash cutâneo eritematoso com predomínio no tronco. O estudo complementar mostrou: anemia (hemoglobina 10.4 g/dL; normocítica, normocrómica), leucocitose com neutrofilia (leucócitos 18360/mm3 e neutrófilos 16140/mm3), elevação dos parâmetros inflamatórios (velocidade de sedimentação 96mm/h e proteína c-reativa 50.9 mg/L), elevação da aminotransferase (45 U/L) e da ferritina (4906.7 ng/mL), eletroforese de proteínas séricas normal, anticorpos antinucleares positivos (>1/1000, padrão mosqueado), com restante estudo imunológico negativo e serologias víricas todas negativas. Iniciou prednisolona 15mg/dia, com melhoria inicial das artralgias e do rash cutâneo mas com recrudescência das queixas sempre que era iniciado o desmame da corticoterapia. Em maio/2018 surgiram de novo adenomegalias axilares e cervicais dolorosas; realizou biópsia cirúrgica de uma adenopatia axilar cuja histologia mostrou um padrão reativo misto (hiperplasia folicular e parafolicular). No final de 2018, além do recrudescimento das queixas articulares e do rash cutâneo, a doente referia astenia e picos febris noturnos, com o estudo laboratorial a mostrar nova elevação dos parâmetros inflamatórios. Foi realizada biópsia cutânea cuja histologia monstrou alterações inespecíficas, enquadráveis em reação de hipersensibilidade (toxicodermia/urticária). Também foi excluída a síndrome periódica associada ao recetor do fator de necrose tumoral (TNF) através de estudo genético. Em janeiro/2019, foi assumido o diagnóstico de SOAD e iniciou-se tratamento com metotrexato subcutâneo até 20mg/semana. Em junho/2019, dado manter as queixas articulares, o rash, a odinofagia e os picos febris diários, foi iniciado o tocilizumab subcutâneo (162mg/semana) em monoterapia. Observou-se franca melhoria das queixas articulares, do rash cutâneo e dos picos febris, com normalização progressiva das alterações laboratoriais aos 6 meses de tratamento, permitindo um desmame gradual da corticoterapia, estando atualmente sob prednisolona 5mg/dia.
Conclusão: A importância do interferão na resposta hiperinflamatória da SOAD tem vindo a ser salientada na literatura mas o papel específico e a resposta observada aos diferentes tipos de interferão ainda não estão completamente esclarecidos. Descrevemos um caso de SOAD que se manifestou após tratamento com peginterferão-beta1a.

061 - EFFICACY OF INTRAVENOUS IMMUNOGLOBULIN IN SHRINKING LUNG SYNDROME ASSOCIATED WITH MIXED CONNECTIVE TISSUE DISEASE

Manuel Silvério-António[1, 2], Nikita Khmelinskii[1, 2]
[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Hospital de Santa Maria, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Shrinking lung syndrome (SLS) is a rare manifestation of connective tissue diseases (CTDs), namely systemic lupus erythematosus (SLE). It is characterized by reduced lung volumes and extra-pulmonary restrictive ventilatory pattern. We report a case of SLS associated with mixed connective tissue disease (MCTD) treated with intravenous immunoglobulin

(IVIg).

A 42-year-old woman presented with a six-month history of symmetric polyarthritis. Associated symptoms included puffy hands, triphasic Raynaud's phenomenon, fatigue and mild exertional dyspnea. Laboratory results showed normocytic anemia, elevated C-reactive protein and erythrocyte sedimentation rate and positive antinuclear antibodies with a speckled pattern by immunofluorescence assay and anti-U1RNP specificity (> 6438 U/L). Chest and joints radiographs were normal and fingernail capillaroscopy excluded a scleroderma pattern. A diagnosis of MCTD was made and she started treatment with prednisolone 10 mg daily and methotrexate up to 20 mg weekly. Arthritis remission was achieved after eight weeks, but dyspnea was worsening and associated with episodic pleuritic chest pain. A chest computed tomography revealed left diaphragmatic elevation and left basal atelectasis with no parenchymal abnormalities. Pulmonary function tests (PFT) revealed a severe restrictive pattern with a decreased forced vital capacity (37%), maximal inspiratory pressure (55%) and diffusing capacity for carbon monoxide (63%) with normal DLCO corrected for alveolar volume. Clinical, analytical and electromyogram studies excluded myopathic involvement. Rituximab was the first-line treatment decided. However, concerns related to serologic evidence of a hepatitis B infection and the threat of COVID-19 infection delayed treatment initiation. Meanwhile, she was treated with a 3 months course of IVIg 2g/Kg/month, which was associated with clinical improvement and almost complete normalization of the PFT. After six months, the patient is stable, with minimal exertional dyspnoea. Rituximab is being re-evaluated depending on PFT results.

SLS treatment is derived from the limited evidence of its management in the context of SLE, which includes moderate to high dose corticosteroids alone or in combination with a second immunosuppressive agent. Recently, rituximab has been used in refractory SLS.

SLS mechanisms are yet to be elucidated, but several studies suggest the contribution of myopathic damage. An extra-pulmonary restrictive pattern with decreased respiratory pressures is seen in most patients without other clinical or enzymatic evidence of myopathy, suggesting the involvement of respiratory muscles. Having this rationale in mind and considering the efficacy of IVIg in idiopathic inflammatory myopathies, we added IVIg to the patient's treatment strategy with an excellent response. To the best of our knowledge, this is the first report of IVIg use in SLS.

064 - REACTIVE ARTHRITIS AS A RARE COMPLICATION OF INTRAVESICAL BACILLUS CALMETTE–GUÉRIN TREATMENT

Matilde Bandeira[1, 2], Eduardo Dourado[1, 2], Filipe Lopes[3], Sofia C Barreira[1, 2]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Serviço de Urologia, Centro Hospitalar Universitário de Lisboa Norte, Lisboa, Portugal

Introduction: Intravesical bacillus Calmette–Guérin (BCG) immunotherapy is a recommended treatment for non-muscle-invasive bladder cancer, typically employed after transurethral resection of bladder tumours. Musculoskeletal consequences of this treatment are rare and can occur several months after treatment onset.

Clinical vignette: A 68 years-old man was referred to the Rheumatology clinic in May 2020 because of low back and bilateral shoulder and ankle pain since 2017. The pain was worse in the morning, but there was no morning stiffness or swelling. The patient had a history of bladder cancer, with two previous surgeries (both in 2017) and intravesical BCG administration (from 2017 to July 2019). Symptom onset occurred 4 to 6 months after the BCG treatment initiation.

On clinical examination, the patient had polyarthritis, mainly affecting the small joints of the hands and tender ankles and sacroiliac joints. Right peroneal tenosynovitis was also clinically evident. He displayed no extraarticular symptoms.

The patient had no elevation of inflammatory markers and was HLA-B27 negative. Hand ultrasonography (US) documented erosions of all distal interphalangeal joints with grey-scale synovitis graded up to grade 3 with Doppler sign. Bilateral shoulder US displayed bilateral glenohumeral synovitis and tenosynovitis of the long head of the biceps brachii, with an evident proliferation of the synovial sheath (Figure 1 A and B). Synovitis of the right tibiotarsal joint and tenosynovitis of multiple tendons in the

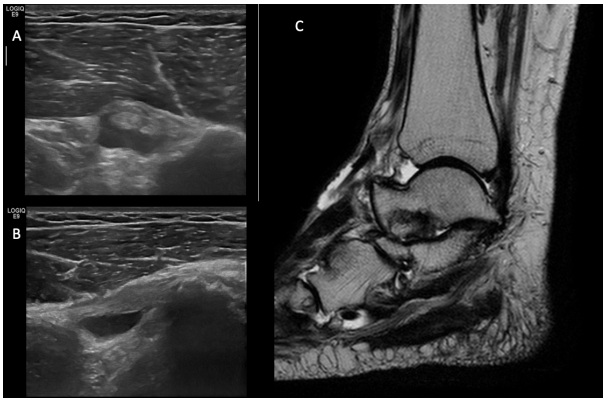
ankles were also documented. A magnetic resonance imaging of the sacroiliac joints was performed, showing degenerative changes with no inflammatory aspects (Figure 1 C).

Nonsteroidal anti-inflammatory drugs (NSAIDs) improved the patient's articular complaints only slightly. The addition of 5mg of prednisolone and a tibiotarsal joint local glucocorticoid injection led to a remission of the articular and periarticular complaints. Slow tapering of the oral GC ensued, and the patient remained clinically asymptomatic even with GC suspension.

Discussion: Reactive arthritis after intravesical BCG is more common in men and can present as large and small joints polyarthritis or asymmetrical mono-oligoarthritis involving the ankles and knees, frequently associated with tenosynovitis and enthesitis. Less than half of patients are HLA-B27 positive, with no prognostic value. It is frequently self-limited, occurring typically after a mean of 5.8 instillations with a duration of up to six months. Persistent arthritis after intravesical BCG is rare but can occur in up to 10% of the cases, with a higher likelihood of chronicity in the polyarticular clinical pictures.

Reactive arthritis induced by intravesical BCG should be considered in the differential diagnosis of arthritis and refractory tenosynovitis in BCG exposed patients. The typical delay between BCG administration and symptom onset should be considered. Treatment with NSAIDs or GCs is generally effective, and isoniazid is a treatment alternative for refractory cases. The latter seems to increase the risk of cancer progression and recurrence since it decreases the effectiveness of BCG immunotherapy.

FIGURE. A/B. RIGHT SHOULDER US TRANSVERSE/ LONGITUDINAL - LHBB TENOSYNOVITIS. C. RIGHT ANKLE MRI - TIBIOTARSAL SYNOVITIS



065 - ACIDENTE VASCULAR CEREBRAL – UMA APRESENTAÇÃO ATÍPICA DE ARTRITE DE CÉLULAS GIGANTES

Inês Genrinho[1], Ana Viegas[2], João Tavares[2], Paulo Monteiro[1], Telma Santos[3]

[1]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [2]Medicinina interna, Centro Hospitalar Tondela Viseu, Viseu, Portugal, [3]Neurologia, Centro Hospitalar Tondela Viseu, Viseu, Portugal

Introdução: A Artrite de Células Gigantes (ACG) é uma vasculite granulomatosa que atinge as artérias de médio e grande calibre. Surge habitualmente após os 50 anos, sendo mais comum nas mulheres e caracteriza-se classicamente por cefaleia persistente, claudicação mandibular e alterações visuais. Cerca de 50% dos casos de ACG podem ainda apresentar-se com clínica típica de Polimialgia Reumática (PMR), encontrando-se estas duas patologias intimamente relacionadas. A elevação marcada da velocidade de sedimentação (VS) associada a uma clínica típica, deve fazer suspeitar o seu diagnóstico. A ocorrência de AVC é uma complicação rara, com uma prevalência de 2-7%. Os fatores de risco para AVC mais descritos são o sexo masculino, idade avançada, Diabetes Mellitus e neuropatia óptica isquémica anterior. O tratamento do AVC nestes casos assenta na corticoterapia, podendo ser útil a combinação com antiagregantes plaquetares.

Caso clínico: Homem, 76 anos, com cefaleia holocraniana, ataxia e tonturas desde há 2 meses, com posterior surgimento de dor temporal bilateral, miodesópsias e fotópsias no olho direito, assim como noção de perda ponderal e fadiga generalizada. Antecedentes pessoais de hipertensão arterial e tabagismo, medicado com irbesartan. Exame objetivo com discreto desvio em pronação do membro superior direito, dismetria na prova calcanhar-joelho à direita e marcha atáxica com desequilíbrio sustentado para a direita. Analiticamente: anemia normocítica e normocrómica ligeira (Hb 13.7g/dL), VS 51mm/h e PCR 4.62mg/dL. Angio-RM CE com lesões isquémicas agudas a nível do lobo occipital esquerdo e hemisférios cerebelosos; Ecodoppler das artérias temporais com halo hiperecóico dos ramos frontais, parietais e tronco comum bilateralmente; e biópsia das artérias temporais compatível com ACG. Assim, foi confirmada a suspeita diagnóstica de ACG com AVC posterior. Iniciou terapêutica com pulsos de metilprednisolona e posteriormente prednisolona 1mg/kg/dia, apresentando diminuição dos parâmetros inflamatórios e recuperação

gradual dos défices neurológicos.
Discussão: A paucidade de sintomas constitucionais e escassa clinica típica, torna o diagnóstico de ACG desafiante. Apesar de constituir uma complicação rara da ACG, a presença de AVC está associada a maior mortalidade e maior recidiva desta entidade. A distinção clínica e imagiológica entre os AVC de etiologia vasculítica e aterosclerótica é difícil, no entanto o envolvimento das artérias vertebro-basilares é mais comum na ACG (40-60%), pelo que deve ser considerada no diagnóstico diferencial em AVCs com esta localização.
Conclusão: A ACG tem uma apresentação clássica, contudo existem casos atípicos, como os AVC, que podem gerar atraso diagnóstico e início da terapêutica. A ACG deve ser ponderada em doentes com AVC, sobretudo acima dos 50 anos, na presença de VS aumentada, clínica típica e atingimento vertebrobasilar.

070 - CONSEQUÊNCIAS TARDIAS COMO APRESENTAÇÕES INICIAIS DE ARTRITE REUMATÓIDE

Diogo Esperança Almeida[1], Emanuel Costa[1], Ana Margarida Correia[1], Joana Leite Silva[1], Joana Sousa-Neves[1], Ana Ribeiro[1], Marcos Cerqueira[1]
[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

Introdução: A artrite reumatóide (AR) é uma artropatia inflamatória com potenciais manifestações extra-articulares. Algumas destas manifestações apresentam-se tipicamente no caso de um insuficiente controlo da actividade inflamatória sistémica por períodos prolongados ou em fases tardias de doença. A amiloidose AA sistémica e a leucemia linfocítica de grandes células granulares (LGL) são, respectivamente, exemplos destas manifestações. A primeira pode ocorrer associada a qualquer condição que provoque inflamação persistente e, actualmente, estima-se que surja em menos de 1% dos doentes com AR, graças ao alargamento das opções terapêuticas e consequente melhoria do prognóstico. A LGL é uma doença hematológica maioritariamente associada a doenças imunomediadas e que se manifesta com esplenomegalia, citopenias (em particular, neutropenia,) e intercorrências infecciosas, podendo estar no espectro clínico da síndrome de Felty, uma consequência tardia de AR. Os autores apresentam o relato de dois casos em que a manifestação extra-articular conduziu ao diagnóstico de AR.
Casos clínicos: Caso 1: Doente do sexo masculino de 38 anos de idade, com seguimento em hemato-oncologia desde 2017 por neutropenia, linfocitose e hipergamaglobulinemia detectadas em análises de rotina, sem queixas associadas. Da investigação, destacava-se expansão clonal de células CD8+ no sangue periférico e medula óssea, que conduziu ao diagnóstico de LGL. Não foi instituída terapêutica, mantendo apenas vigilância clínica e analítica. Em 2019, desenvolveu sintomas e sinais inflamatórios articulares palindrómicos a envolver joelhos, cotovelos, punhos e pequenas articulações das mãos. A avaliação complementar mostrou positividade em títulos elevados de factor reumatoide (FR) e anticorpos anti-peptídeos citrulinados (AC anti-CCP). Foi estabelecido o diagnóstico de AR e iniciou tratamento com metotrexato, com boa resposta clínica articular e também dos parâmetros hematológicos.
Caso 2: Doente do sexo feminino de 61 anos de idade, agricultora, com antecedentes pessoais de hipertensão arterial, dislipidemia e gonartrose bilateral com necessidade de artroplastia total do joelho bilateral. Foi encaminhada à consulta de Nefrologia por depressão da função renal e proteinúria detectadas em rastreio cardiovascular nos cuidados de saúde primários. Foi submetida a biópsia renal que revelou depósitos de amiloide A; o estudo subsequente, mostrou elevação de parâmetros inflamatórios e positividade em título elevado de FR. Nesse contexto, foi avaliada por Reumatologia e, apesar de nunca ter valorizado as suas frequentes queixas músculo-esqueléticas, apresentava poliartrite e evolução radiográfica típica de AR. Encontra-se actualmente em fase inicial de tratamento dirigido.
Conclusões: As consequências extra-articulares são um elemento fundamental no diagnóstico diferencial de manifestações sistémicas em doente com diagnóstico de AR previamente estabelecido. Nos casos apresentados, pelo contrário, a manifestação extra-articular constituiu a chave para o diagnóstico primário. No caso 1, destaca-se ainda a atipia da LGL preceder, em 2 anos, o surgimento das queixas decorrentes da AR. Apesar de constituírem consequências habitualmente tardias, o controlo da actividade inflamatória da AR é a base do tratamento e melhoria do prognóstico de qualquer uma das situações.

071 - TRATAMENTO DE ESPONDILARTRITES DE DIFÍCIL CONTROLO COM UPADACITINIB - EXPERIÊNCIA DE UM CENTRO

Diogo Esperança Almeida[1], Emanuel Costa[1], Ana

Margarida Correia[1], José Redondo Costa[1], Joana Leite Silva[1], Joana Sousa-Neves[1], Marcos Cerqueira[1], Ana Ribeiro[1]
[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

Introdução: As espondilartrites são um grupo heterogéneo de doenças reumáticas inflamatórias crónicas, com potencial envolvimento axial e periférico, cuja fisiopatiologia se baseia na inflamação da entese. O upadacitinib (UPA) é o primeiro inibidor da janus-kinase (JAK) de segunda geração, com actividade selectiva de inibição da JAK1 e aprovação recente para o tratamento de AP e EA na Europa. Os autores descrevem a experiência do seu centro com este fármaco no tratamento de doentes com falência ou intolerência prévias a fármacos com diversos mecanismos de acção estabelecidos no tratamento de espondilartrites.
Casos clínicos: Caso 1: Doente do sexo feminino, de 61 anos de idade com diagnóstico de EA desde os 50 anos, mas com lombalgia inflamatória típica desde os 20 anos; apresenta ainda envolvimento periférico simétrico de pequenas articulações desde há cerca de 2 anos, refractário a disease-modifying anti-rheumatic drugs (DMARD) clássicos. Apresentou, de forma sucessiva, falência secundária a 3 fármacos antiTNFa (golimumab, adalimumab e infliximab) e falência primária a secucinumab. Por manter actividade elevada dos componentes axial e periférico (DAS28-PCR 4.67, ASDAS-PCR 4.9), iniciou UPA 15mg diário. Após 1 mês de tratamento, com boa tolerância, reporta melhoria franca de ambas as componentes de doença (DAS28-PCR 2.61, ASDAS-PCR 1.8) com suspensão de toma contínua de anti-inflamatório não-esteróide (AINE) e redução significativa da dose de corticosteróides.
Caso 2: Doente do sexo masculino, de 55 anos de idade com EA desde os 40 anos a cumprir terapêutica biotecnológica por insuficiente controlo de doença com AINE. Apesar de boa resposta inicial com fármaco antiTNFa (adalimumab), desenvolveu, além de falência secundária ao fármaco, sarcoidose como provável reação paradoxal ao fármaco (resolvida após suspensão). Iniciou, nesse contexto, terapêutica com secucinumab, tendo tido falência primária (PCR 19.60mg/L, BASDAI 7.5, ASDAS-PCR 4.6). Por este motivo, iniciou UPA e, após 1 mês de terapêutica, apresenta melhoria global mas incompleta (PCR 7.20mg/L, BASDAI 6.2, ASDAS-PCR 3.2), tendo também boa tolerância até à data.
Caso 3: Doente do sexo masculino, 31 anos de idade com espondilartrite indiferenciada com envolvimento axial e periférico de grandes e médias articulações desde os 20 anos, tratado inicialmente com metotrexato, sulfassalazina e AINE. Por insuficiente controlo de actividade periférica, iniciou terapêutica com adalimumab, com boa resposta inicial mas falência secundária aos 2 anos; posteriormente, cumpriu infliximab durante 7 anos, tendo tido também falência secundária após período de suspensão temporária por toxicidade hepática. Teve ainda falência primária a secucinumab (ASDAS-PCR 5.2, artrite de cotovelos e tornozelos). Por este motivo, iniciou UPA e, com 3 meses de tratamento, tem resposta clínica muito significativa de ambas as componentes de doença (ASDAS-PCR 1.8, sem artrite periférica).
Conclusões: Na ainda curta experiência do centro com UPA no tratamento de espondilartrites, o fármaco parece constituir uma opção viável para doentes de difícil controlo e com experiência prévia com outros fármacos dirigidos. Destaca-se a rapidez de resposta terapêutica nos casos 1 e 3, o aparente bom perfil de segurança e a acção nas componentes axial e periférica. Serão necessários dados de "vida real" mais extensos (em número de doentes tratados e tempo de seguimento) para corroborar os dados dos ensaios clínicos que promoveram a aprovação do fármaco nesta indicação.

072 - TENOSSINOVITE DO EXTENSOR COMUM DOS DEDOS - UM QUADRO INAUGURAL DE ARTRITE REUMATÓIDE

Rafaela Nicolau[1], Nádia Martins[1], Paulo Monteiro[1]
[1]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Introdução: A tenossinovite do extensor comum dos dedos é frequente nos doentes com Artrite Reumatoide (AR) e pode constituir forma de apresentação da doença. Caracteriza-se por uma massa localizada na bainha dos tendões que se torna mais evidente através da extensão ativa do punho e dedos ao gerar um deslizamento proximal da massa sob o retináculo – Tuck sign.
Caso clínico: Doente do sexo feminino de 69 anos, agricultora, sem antecedentes pessoais relevantes e com antecedentes familiares de AR, referenciada à consulta de Reumatologia por quadro de poliatralgias de ritmo inflamatório com noção de tumefação envolvendo mãos, pés, joelhos e tibio-társicas associado a

massa nodular da superfície extensora de ambos os punhos com cerca de 7 meses de evolução. Sem outras queixas além das descritas, nomeadamente febre, clínica sética, lesões cutâneas ou urogenitais.

Além da tumefação descrita mais exuberante no lado direito, não apresentava outras alterações ao exame objetivo. (Figura 1)

Realizou ecografia do punho direito que revelou a presença de tenossinovite do extensor comum dos dedos com Power Doppler. As radiografias das mãos, joelhos e pés não mostraram alterações.

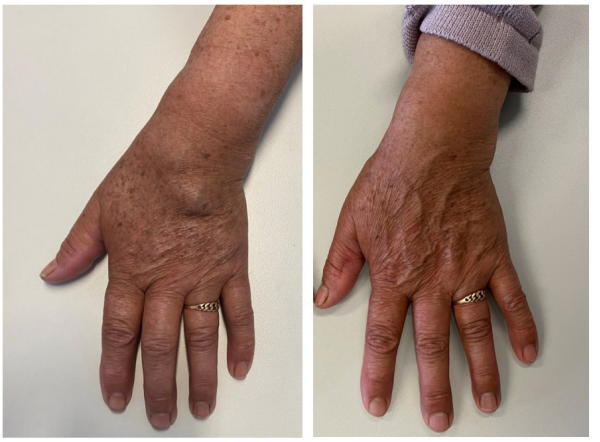
À exceção de um aumento da velocidade de sedimentação (48 mm/h), não apresentava outras alterações analíticas hemograma e proteína C reativa normais, anticorpos antinucleares, fator reumatoide, anticorpo anti-péptido cíclico citrulinado negativos, enzima de conversão da angiotensina e uricemia normais, IGRA negativo, serologias para VIH, VHB, VHC, VDRL, Brucelose e doença de Lyme não reativas.

Foi efetuada infiltração com 40 mg de acetato de metilprednisolona da tenossinovite do extensor comum dos dedos da mão direita com resolução do quadro. (Figura 1)

Durante o seguimento desta doente foi objetivado quadro de poliartrite envolvendo metacarpofalângicas e interfalângicas proximais de ambas as mãos, cerca de 12 meses após o quadro inaugural.

Foi admitido o diagnóstico de AR seronegativa e iniciada terapêutica com metotrexato na dose inicial de 10 mg/semana, folicil 10 mg/semana e metilprednisolona na dose de 6 mg id com redução posterior em associação a suplementação com cálcio e vitamina D, verificando-se uma evolução favorável. Foi ainda iniciada terapêutica antirreabsortiva com bifosfonato semanal.

FIGURA 1- SUPERFÍCIE EXTENSORA DA MÃO DIREITA ANTES E APÓS INFILTRAÇÃO COM METILPREDNISOLONA



Conclusão: A tenossinovite é uma manifestação frequente na AR inicial, descrita em cerca de 50% dos casos, percentagem que aumenta até aos 80% quando o diagnóstico é feito com recurso à ressonância magnética. Qualquer tendão pode ser envolvido, embora seja particularmente frequente a inflamação dos flexores, extensor comum dos dedos e extensor ulnar do carpo. A abordagem terapêutica consiste no tratamento da doença articular com ou sem recurso à infiltração local.

077 - PACHYDERMODACTYLY: THE ROLE OF ULTRASONOGRAPHY, SUPERB MICROVASCULAR IMAGING AND ELASTOGRAPHY IN DIAGNOSIS

Ana Cláudia Martins Novais[1], L Soares-de-Almeida[2, 3], Jorge Garcia[4], João Madruga Dias[4, 5]

[1]Unidade de Saúde Familiar Locomotiva, ACeS Médio Tejo, Entroncamento, Portugal, [2]Clínica Universitária de Dermatologia de Lisboa, Hospital de Santa Maria, Centro Hospitalar Universitário de Lisboa Norte, Lisboa, Portugal, [3]Faculdade de Medicina da Universidade de Lisboa, Lisboa, Portugal, [4]Serviço de Reumatologia, Centro Hospitalar Médio Tejo, Torres Novas, Portugal, [5]EpiDoc Research Unit, CEDOC, NOVA Medical School, Lisboa, Portugal

Pachydermodactyly is a rare and benign superficial fibromatosis characterized by painless and progressive swelling of periarticular soft tissues of the proximal interphalangeal (PIP) joints, most commonly of both hands. There is no tenderness, warmth, morning stiffness, or reduced range of motion associated. Our purpose is to highlight the diagnostic utility of ultrasonography, superb microvascular imaging (SMI) and elastography in pachydermodactyly.

We report the case of a 15-year-old adolescent white boy, with a 6-month history of insidious and progressive, asymptomatic swelling of the lateral and dorsal regions of the metacarpophalangeal (MCP) and PIP joints of both hands. Articular ultrasonography showed thickening of the skin around the lateral regions of the PIP and MCP joints, with no synovitis, hydrarthrosis or muscle, tendon, or bone changes. Elastography revealed lower elasticity in the aforementioned skin regions, corresponding to increased tissue hardness due to hyperkeratosis. No SMI or Doppler signals were detected in epidermal or dermal

tissues, as well as in tendons, joints, and bone.

This case report shows that ultrasonography, SMI and elastography may play a significant role in the accurate diagnosis of pachydermodactyly and exclusion of alternative conditions. These imaging modalities have no ionizing radiation, they are fast, inexpensive, and performed on site. They do not require usage of contrast agents and thus can eliminate the need of invasive procedures such as skin biopsy. They also contribute to reduce health care costs with unnecessary complementary tests and inappropriate treatment.

096 - GASTROINTESTINAL INVOLVEMENT IN ANTI-NEUTROPHIL CYTOPLASMIC ANTIBODIES-ASSOCIATED VASCULITIS

Nuno Pina Gonçalves[1, 2, 3], Sandra Falcao[2], Maria Manuela Costa[2], Jaime C. Branco[2, 3]

[1]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Lisboa Ocidental | Hospital Egas Moniz, Lisboa, Portugal, [3]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

Introduction: Granulomatosis with polyangiitis (GPA) is a necrotizing vasculitis that most commonly involves the lungs and the kidneys. The gastrointestinal tract is less commonly involved but can be the presenting feature and lead to serious complications.
Clinical case: Male, 77 years old, with a history of hypertension, atrial fibrillation and a inguinal hernia submitted to laparoscopic hernioplasty, presents to the emergency department with malaise, anorexia and weight loss (10kg) in association with diffuse abdominal pain and vomiting. Bowel movements were reportedly maintained, with no blood on the stools (although rectorrhagia was later noted but attributed to haemorrhoids). Laboratory work-up showed anaemia (Hb 11.9g/L), leucocytosis (11.800x103U/L) and creatinine 2.26mg/dL (compatible with acute kidney injury). Na abdominal computed tomography (CT) showed a small intestine obstruction (jejunoileal) in "closed-loop". The patient was submitted to a segmental enterectomy (surgical report mentioned a small intestine volvulus due to a bridle) and the pathology report described areas of ischemic necrosis, with a mixed inflammatory infiltrate. The patient was admitted to the surgery ward. Kidney function kept worsening and at day five post-op, diuresis was minimal. The patient had to start haemodialysis. Dyspnoea and hypoxemia ensued and at day 8 the patient was admitted to the intensive care unit due to type II respiratory failure with concurrent alveolar haemorrhage. Endotracheal intubation was needed. Epistaxis was also noted but attributed to nasogastric tube. An extensive work-up ensued with the following remarkable results: haemoglobin 8.3g/L, creatinine 7.8mg/dL (with oliguria); Erythrocyte sedimentation rate 102mm/h; anti-neutrophil cytoplasmic antibodies (ANCA) positive for PR3 with a high titter (>10.000U/L); negative blood cultures; HIV negative; kidney biopsy revealed extracapillary glomerular proliferation with crescent formation, focal segmental glomerulosclerosis with a collapsing variant and acute tubular necrosis in 80% of the tubules. Body CT scan showed bilateral alveolar condensation and pleural effusion with no pathological intestinal findings post-op. Collaboration of the rheumatology service was solicited, a diagnosis of granulomatosis with polyangiitis was proposed and the following treatment was instituted: 3 consecutive pulses of 1g methylprednisolone followed by prednisolone 1mg/Kg/day, 3 cycles of plasmapheresis immediately followed by cyclophosphamide (CYCLOPS protocol). There was a gradual clinical and laboratorial improvement and the patient was discharged with no respiratory symptoms and a normalized kidney function. At 4 month follow-up, arthritis was noted (ankles and left knee) and a rise in PR3 titters was observed (at prednisolone 30mg/day). Maintenance therapy with methotrexate 15mg/week and rituximab 500mg was proposed. No new relapses were noted until the present day.
Discussion: Although the intestinal involvement was attributed to the presence of a bridle, an acute kidney injury and pulmonary haemorrhage with positive ANCA is very suggestive of ANCA- associated vasculitis. Gastrointestinal involvement usually manifests as abdominal pain and vomiting. The histologic findings of the small bowel sample were inconclusive, as in most literature case reports. In this patient, the sequence of events cannot exclude gastrointestinal involvement in GPA (e.g mesenteric artery vasculitis), and may even suggest it as the presenting feature, usually meaning severe disease.

113 - EFICÁCIA DO METOTREXATO NO TRATAMENTO DE ARTRITE PSORIÁTICA COM ENVOLVIMENTO UNGUEAL GRAVE

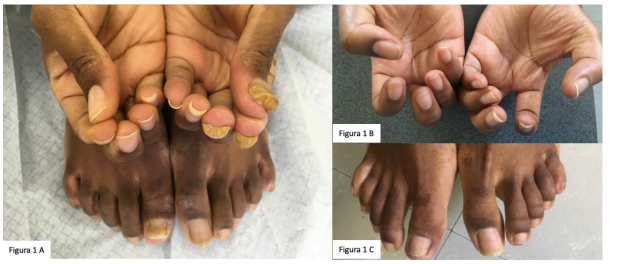
Ana Lúcia Fernandes[1], Jaime C. Branco[2], Alexandre Sepriano[2]

[1]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, [2]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal

Introdução: A artrite psoriática (AP) é uma doença reumática crónica caracterizada por inflamação articular, e pela coexistência de manifestações extra-músculo-esqueléticas (EME) envolvendo, entre outros, a pele, unhas e os olhos. Em alguns casos as manifestações EME podem dominar o quadro clínico e representar importantes desafios terapêuticos. No presente caso descrevemos a evolução clínica de uma doente com AP com envolvimento EME grave, detalhando a sua abordagem diagnóstica, bem como a abordagem terapêutica que permitiu o controlo bem-sucedido da doença.

Descrição: Doente do sexo feminino, de 41 anos, sem antecedentes pessoais ou familiares de relevo. Apresenta lombalgia baixa de ritmo mecânico desde há 5 anos e, desde há 1 ano, artralgias assimétricas, de ritmo misto, e rigidez a nível das articulações interfalângicas do 4º e 5ª dedos da mão esquerda e mais recentemente do ombro esquerdo. Além do quadro articular, refere desde há 2 anos, desenvolvimento de alterações ungueais nas mãos e pés de agravamento progressivo. Fez sucessivas terapêuticas antifúngicas orais e tópicas, sem melhoria. O exame micológico direto e histopatológico ungueal feitos na consulta de dermatologia foram ambos negativos. Ao exame objetivo, na consulta de reumatologia, mostrava onicodistrofia e onicogrifose exuberantes de várias unhas da mão e do pé esquerdos (Figura 1A). Não tinha lesões cutâneas sugestivas de psoríase. Apresentava oligoartrite das pequenas articulações da mão e pé esquerdos, com limitação pronunciada da flexão dos dedos da mão. Tinha ainda mobilização ativa dolorosa do ombro esquerdo acima dos 90º e dor à palpação da inserção supra-espinhoso. Não apresentava tenosinovite/entesite noutras localizações. Dos exames complementares de diagnóstico, destaca-se velocidade de sedimentação normal, PCR negativa, HLA-B27 negativo e estudo radiográfico das mãos, pés, coluna vertebral e bacia sem alterações de relevo. Assumiu-se artrite psoriática com envolvimento articular periférico e ungueal. A doente foi inicialmente medicada com prednisolona 15 mg/dia e, posteriormente, com metotrexato 15mg/semana, com franca melhoria das artralgias e rigidez. Ocorreu ainda regressão assinalável da onicodistrofia e onicogrifose apenas após 7 semanas do início do metotrexato (Figura 1B, C).

FIGURA 1A: PSORÍASE UNGUEAL NA PRIMEIRA CONSULTA. FIGURA 1B E 1C: EVOLUÇÃO APÓS 7 SEMANAS DE TERAPÊUTICA COM METOTREXATO.



Conclusão: A evidência actual demonstra de forma robusta a superioridade de agentes biológicos sobre DMARDs de síntese convencionais, como o metotrexato, no tratamento da psoríase ungueal. Contudo os efeitos de grupo observados em estudos não se traduzem necessariamente a todos os casos de doentes individuais. Este caso demonstra a eficácia do metotrexato no controlo da psoríase ungueal, mesmo nas suas formas mais graves.

114 - O PAPEL DA TERAPÊUTICA MÉDICA E DO REUMATOLOGISTA NUM CASO REFRATÁRIO DE SINOVITE VILONODULAR PIGMENTADA

Bruno Miguel Fernandes[1], Inês M Rodrigues[2], Mariana M Chaves[3], Madalena Pimenta[2], Francisco Rego Costa[2], Miguel Bernardes[1], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Serviço de Radiologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [3]Serviço de Radiologia, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal

Introdução: A sinovite vilonodular pigmentada (SVNP) é uma doença neoplásica benigna incomum que se caracteriza pela proliferação excessiva da membrana sinovial, afetando principalmente jovens adultos. O atingimento é tipicamente monoarticular, sendo o joelho a articulação mais frequentemente atingida. A sua etiologia não está completamente esclarecida mas foi possível demonstrar a presença de uma translocação envolvendo os genes do fator estimulador de macrófagos (M-CSF) e o do colagéneo 6A3, resultando num aumento da expressão aumentada do M-CFS.

Caso clínico: Doente do sexo masculino, de 38 anos, foi encaminhado de Ortopedia para consulta de Reu-

matologia por SVNP do joelho direito refratária a tratamento cirúrgico. Tratava-se de um doente com início da sintomatologia quinze anos antes, referindo gonalgia de ritmo mecânico, assim como "sensação de falha" do joelho ao correr. A ressonância magnética (RM) inicial mostrou achados compatíveis com sinovite vilonodular difusa do joelho direito, tendo sido submetido a três sinovectomias por artrocoscopia e a uma sinovectomia aberta. O diagnóstico foi corroborado pela anatomia patológica da primeira cirurgia. Apresentou sucessivas recidivas após todas as intervenções cirúrgicas e, à observação por Reumatologia, referia gonalgia de ritmo mecânico e noção de tumefação articular a espaços, tendo realizado várias sinoviorteses químicas nos últimos anos (uma a duas vezes por ano), com melhoria muito discreta da sintomatologia. Ao exame objetivo apresentava crepitações patelofemorais, sinal da tecla positivo e dor à flexão máxima do joelho (115º). O estudo radiográfico mostrava já gonartrose femurotibial à direita e a ressonância magnética evidenciava extensas alterações de SVNP, com envolvimento articular difuso.

Tendo em conta a refratoriedade às cirurgias e após discussão em reunião de serviço, optou-se pelo tratamento off-label com imatinib 400mg por dia. Aos 12 meses de tratamento, o doente apresentava melhoria clínica marcada, sem necessidade de novos gestos locais, com melhoria da mobilidade do joelho e sem dor à flexão máxima, referindo apenas gonalgia mecânica a espaços, controlada com medicação analgésica. A RM do joelho não demonstrou progressão da extensão da SVNP.

Conclusão: A história natural da sinovite vilonodular pigmentada conduz à destruição progressiva da articulação afetada. A sinovectomia cirúrgica é o tratamento de escolha, no entanto, a recidiva é frequente. A sinoviortese com radioisótopos pode ser utilizada em casos refratários, muitas vezes em associação à cirurgia. A descoberta do papel do M-CSF na fisiopatologia da doença levantou a hipótese do uso terapêutico dos inibidores da tirosina cinase com ação inibitória do M-CSF neste contexto de doença. Tal como neste caso clínico, alguns casos descritos na literatura mostraram resposta sintomática e imagiológica ao imatinib em doentes com sinovite vilonodular pigmentada refratária a terapêutica cirúrgica.

117 - TENDINOPATIA CALCIFICANTE INDUZIDA POR ESTATINAS: UM CASO CLÍNICO COM ROSUVASTATINA

Bruno Miguel Fernandes[1], Miguel Bernardes[1], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal

Introdução: A tendinopatia calcificante caracteriza-se pela formação de depósitos de hidroxiapatite de cálcio nos tendões, afetando mais frequentemente os ombros, em particular o tendão do supraespinhoso. O quadro clínico mais frequente é uma apresentação dolorosa aguda e intensa, com resolução espontânea em dias ou semanas. É mais frequente em mulheres de meia idade, sendo que a fisiopatologia não se encontra totalmente conhecida.

Caso clínico: Doente do sexo masculino, 54 anos de idade, com antecedentes de alopécia androgénica tratada com finasterida 1mg id, hiperuricemia discreta e dislipidemia sob rosuvastatina 20mg id desde há cerca de 2 meses, com prática regular de exercício físico aeróbio (30-45 minutos, 3 vezes por semana). Iniciou dor mecânica na região do grande trocanter esquerdo, de início agudo, tornando-se constante, irradiada para a virilha e incapacitante para a marcha 5 dias depois, tendo sido submetido a betametasona 14mg intramuscular e ibuprofeno 600mg 3id, sem melhoria inicialmente. Por esse motivo, pediram-lhe ressonância magnética (RM) das ancas e da coluna lombar. Oito dias após o início das queixas iniciou omalgia esquerda incapacitante, motivo pelo qual recorreu à Reumatologia. Tinha mudado o anti-inflamatório não esteroide para acemetacina 60mg+90mg/dia há 3 dias e, à observação, apresentava já melhoria das queixas no trocanter esquerdo, observando-se ainda dor à palpação local assim como limitação parcial e dor na excursão ativa e passiva do ombro esquerdo nos vários planos de movimento. Não apresentava outras alterações ao exame reumatológico. Negava episódio infecioso prévio ao início das queixas, alterações gastrointestinais, história de uveíte, psoríase cutânea ou lítiase renal. Nesta data, na primeira consulta de Reumatologia, optou-se por suspender a rosuvastatina 20mg id, iniciada dois meses antes. O estudo laboratorial não revelou quaisquer alterações, nomeadamente no metabolismo fosfocálcico, proteinograma ou valores séricos de paratormona. A RM da coluna lombar não mostrou alterações de relevo e a RM das ancas mostrou sinais de entesopatia calcificante, com várias calcificações adjacentes ao grande trocânter esquerdo, superficialmente à inserção do tendão do glúteo médio, a maior com 12mm de diâmetro (figura 1) e a ecografia do ombro esquerdo revelou volumosa

FIGURA 1. SINAIS DE TENDINOPATIA CALCIFICANTE NO TENDÃO GLÚTEO MÉDIO POR RESSONÂNCIA MAGNÉTICA.

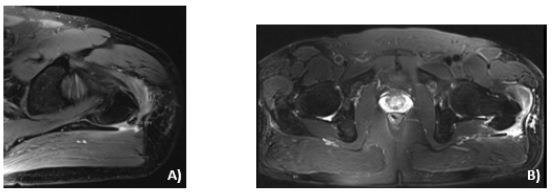


Figura 1. Ressonância magnética das ancas: Observam-se sinais de tendinopatia calcificante com várias calcificações na inserção do tendão glúteo médio no grande trocânter e exuberantes alterações inflamatórias circundantes, nomeadamente edema interesticial no tecido adjacente (A) corte coronal *fat-saturated* coronal da região trocantérica esquerda; B) corte transversal *fat-saturated* T2).

calcificação (2x1.5cm) na superfície da inserção do tendão infra-espinhoso, com espessamento dos planos adjacentes da bursa subacromial e calcificação com 0.4cm na inserção do supra-espinhoso. O doente apresentou melhoria, retomando plenamente a sua atividade laboral e o exercício físico aos 16 dias.

Conclusão: Na maioria dos casos não é possível identificar uma causa subjacente à tendinopatia calcificante. Por vezes, esta associa-se a patologias sistémicas, principalmente à diabetes mellitus. Alguns dados têm demostrado uma associação rara das estatinas a tendinopatia, incluindo tendinopatia calcificante e roturas tendinosas, cujo mecanismo não está totalmente esclarecido. Este evento adverso é ainda mais raro que a fraqueza muscular com elevação da CPK, rabdomiólise ou miopatia inflamatória. Os casos de tendinopatia associada a estatinas verificam-se normalmente no primeiro ano de tratamento, são mais frequentes em indivíduos do sexo masculino não sedentários (desportistas) e melhoram com a suspensão do fármaco, sendo o tendão de Aquiles o mais frequentemente afetado. Apresentamos um caso curioso de tendinopatia calcificante hiperálgica múltipla após início de rosuvastatina, alertando o Reumatologista para esta causa rara de tendinopatia.

123 - MYOCARDIAL INVOLVEMENT IN SYSTEMIC SCLEROSIS – A CASE REPORT

Salomé Garcia[1], Bruno Miguel Fernandes[1], Daniela Santos Oliveira[1], Ana Martins[1], Diogo Guimarães da Fonseca[2], Frederico Rajão Martins[3], Beatriz Samões[2], Georgina Terroso[1], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/ Espinho, Vila Nova de Gaia, Portugal, [3]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal

Background: Cardiac involvement is rare in patients with collagen diseases but a serious condition when it does occur. We report a rare case of myocarditis in a patient with systemic sclerosis (SSc) who presented with sudden chest pain and heart faillure (HF) signs and symptoms.

Clinical case: We report the case of a 29-year-old female with the diagnosis of a diffuse cutaneous form of SSc since 2019 with immunological (anti-nuclear antibodies 1/1000 with a homogeneous pattern, positivity for anti-SCL70 and anti-PM Scl 100) cutaneous, articular, vascular, gastrointestinal and muscle involvement with elevation of muscle enzymes and proximal muscle strength deficit of the lower limbs at the time of diagnosis. Currently, she was taking nifedipine 30 mg/day, methotrexate 15 mg/week, pentoxifylline 400 mg three times a day and prednisolone 7.5 mg daily. She presented to the emergency department with a tight squeezing chest pain, palpitations and shortness of breath and the clinical evaluation revealed arrhythmic cardiac auscultation and bilateral lower third pulmonary crackles. Upon admission, we observed the following vital signs: blood pressure of 116/65 mmHg, heart rate of 150 beats per minute, respiratory rate of 18 breaths per minute and oxygen saturation of 99% on room air with a pulse oximeter. Laboratory investigations revealed the following results: thyroid function and cortisol levels were normal, the serum troponin T level was 0.015 ng/mL (normal value <0.016 ng/mL) and serum brain natriuretic peptide (BNP) was 1002.3 pg/mL (< 100.0 pg/mL). Her electrocardiogram (ECG) showed an atrial flutter with rapid ventricular response (heart rate 150 bpm). Previous ECG was unremarkable. The transthoracic echocardiogram (ETT) revealed moderate dilatation of the four cardiac chambers with biventricular dysfunction (left ventricular ejection fraction 22%). Cardiac catheterization was performed, demonstrating normal coronary arteries. She was submitted to electrical cardioversion with incomplete response. Chemical cardioversion with amiodarone was performed with restoration of sinus rythm. She initiated hypocoagulation with low molecular weight heparin and nebivolol 2.5 mg/daily. One week later, she performed a cardiac magnetic ressonance which showed dilatation of the four cardiac chambers with improving biventricular dysfunction (left ventricular ejection fraction of 41% and 39% of the right ventricle) and moderate pericardial effusion; there was no evidence of myocardial oedema or myocardial fibro-

sis. Myocardial biopsy showed interstitial fibrosis and oedema, capillary wall thickening and mild intensity lymphocytic infiltrate and the PCR and RT-PCR analysis for cardiotropic viruses (coxsackievirus, adenovirus, influenza A–B virus, parvovirus-B19, human herpesvirus type 6and 8 and herpes simplex virus type 1 and 2, human cytomegalovirus, varicella–zoster virus, Epstein–Barr virus and COVID-19) were negative. The diagnosis of a virus-negative myocarditis was made and the patient started treatment with monthly cyclophosphamide (CYC) 1000 mg with good clinical and analytical response. Currently she is on her fourth administration without any signs or symptoms of HF and her BNP is 108 pg/mL.

Conclusions: Clinically active myocarditis, despite uncommon, causes significant morbi-mortality and is typically associated with myositis and overlap syndromes, but also with diffuse cutaneous forms. Immunossupressive drugs such as CYC have been used with some benefit in these patients, despite no evidence of prolonged survival.

129 - LESÕES CUTÂNEAS NUM DOENTE COM ESPONDILARTRITE AXIAL SOB AGENTE ANTI-TNF-α DE INÍCIO RECENTE – CASO CLÍNICO

Ana Martins[1], Daniela Santos Oliveira[1], Frederico Rajão Martins[2], Filipe Oliveira Pinheiro[1], Maria Seabra Rato[1], Diogo Guimarães da Fonseca[3], Sofia Pimenta[1, 4], Miguel Bernardes[1, 4], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [3]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [4]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Introdução: Os inibidores do fator de necrose tumoral alfa (anti-TNF-α) são fármacos essenciais no tratamento de inúmeras doenças reumáticas. Geralmente, são bem tolerados, no entanto, podem causar efeitos adversos, nomeadamente a nível cutâneo. O espectro das lesões cutâneas induzidas pelo anti-TNF-α inclui a psoríase, lesões semelhantes ao lúpus cutâneo, eczema, hidradenite supurativa, síndrome de Sweet, pioderma gangrenoso, vasculite, entre outras. A vasculite induzida por anti-TNF-α é uma entidade rara, cuja incidência é inferior a 0,5%. De seguida, é apresentado o caso clínico de um doente com vasculite cutânea provável induzida por anti-TNF-α.

Caso clínico: Doente do sexo masculino de 58 anos de idade, com espondilartrite axial radiográfica diagnosticada há 3 anos, medicado com AINE em dose terapêutica e, desde há 4 meses, sob anti-TNF-α (adalimumab 40 mg s.c, quinzenal), recorreu ao hospital de dia por uma intercorrência. Referia, desde há 2 semanas, o aparecimento de lesões escuras violáceas, não pruriginosas, na face lateral da perna esquerda, com posterior progressão para a contralateral. Negava sintomas constitucionais, queixas respiratórias, gastrointestinais, geniturinárias, alterações da sensibilidade ou força muscular. Sem história de fenómeno de Raynaud, fotossensibilidade, alopécia, queixas secas, artralgias periféricas, úlceras orais, genitais ou serosite. Sem infeção recente ou uso de novos fármacos ou alimentos.

Ao exame objetivo, apresentava lesões cutâneas violáceas, não pruriginosas e com relevo, na face lateral da perna esquerda, a maior com 2 cm de diâmetro, e na face anterolateral da perna direita, sugestivas de púrpura palpável (figura 1). Sem outras alterações no exame por aparelhos e sistemas.

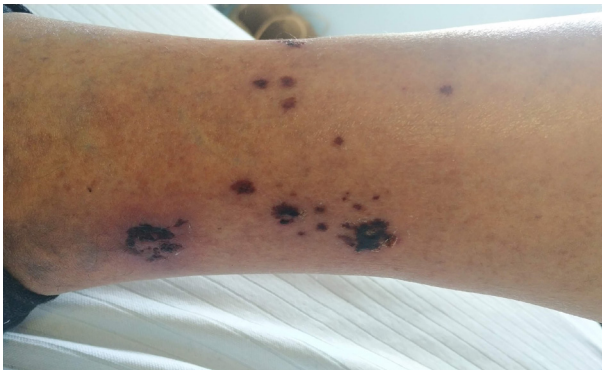
Analiticamente, apresentava-se sem anemia, leucocitose ou trombocitopenia, com PCR discretamente elevada (14.1 mg/L) e sem alterações nas funções renal e hepática. As serologias para borrelia, CMV, parvovirus B19, EBV e HSV tipo 1 e 2 excluíram infeção aguda. O TPPA foi negativo, bem como as hemoculturas. Apresentava ANA positivos (1/1000, padrão mosqueado), com restante estudo imunológico negativo.

Foi discutido o caso com dermatologia e, tendo em conta as características das lesões e do último fármaco introduzido, foi assumido o diagnóstico de provável vasculite cutânea de pequenos vasos secundária ao adalimumab. O doente iniciou terapêutica com betametasona tópica e prednisolona oral (10 mg/dia). Em simultâneo, procedeu-se à suspensão da terapêutica com adalimumab e, tendo em conta a necessidade de bioterapia para controlo da doença reumática inflamatória de base, foi pedido switch para secucinumab.

Após a suspensão do adalimumab, verificou-se uma resolução completa das lesões cutâneas. Aos 6 meses de seguimento, o doente mantém-se sem lesões cutâneas e apresenta boa resposta clínica ao secucinumab.

Conclusão: O aparecimento de lesões cutâneas num doente sob anti-TNF-α é um desafio diagnóstico e implica uma investigação etiológica alargada e, sem-

FIGURA. LESÕES CUTÂNEAS VIOLÁCEAS COM RELEVO NA FACE LATERAL DA PERNA ESQUERDA, SUGESTIVAS DE PÚRPURA PALPÁVEL.



pre que possível, uma avaliação histológica para confirmação do diagnóstico. Na literatura, estão descritos poucos casos de vasculite cutânea de pequenos vasos associada ao anti-TNF-α e é rara a sua relação com o adalimumab. Nos doentes com vasculite cutânea induzida por anti-TNF-α, o tratamento com um segundo anti-TNF-α pode ser equacionado. No entanto, sempre que possível, deve-se optar por um agente biotecnológico de outra classe, ou seja, com outro mecanismo de ação, uma vez que isso se associa a menor probabilidade de recorrência.

133 - UM CASO DE SÍNDROME DE SCHNITZLER ATÍPICO OU UMA NOVA ENTIDADE NOSOLÓGICA?

Maria Seabra Rato[1], Filipe Oliveira Pinheiro[1], Diogo Guimarães da Fonseca[2], Daniela Santos Oliveira[1], Ana Martins[1], Frederico Rajão Martins[3], Miguel Bernardes[1, 4], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [3]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal, [4]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Introdução: A síndrome de Schnitzler (SSch) é uma doença autoinflamatória caracterizada por rash urticariforme e gamapatia monoclonal, acompanhados por febre, dor articular ou óssea, adenopatias e elevação de parâmetros inflamatórios. O diagnóstico de SSch é um desafio, não só pela sua raridade, mas também devido à ausência de testes de diagnóstico específicos e ao amplo espetro de diagnósticos diferenciais.

Caso clínico: Doente do sexo feminino, de 37 anos de idade, enviada à consulta por história com 2 anos de evolução de poliartralgias de ritmo inflamatório e lesões urticariformes dispersas pelo tegumento, não pruriginosas, com duração inferior a 24 horas, sem lesão residual associada. A doente já teria sido medicada com vários anti-histamínicos sem benefício, notando uma melhoria parcial sob prednisolona oral (10-20mg/dia). Como antecedentes pessoais salientava-se uma hemitiroidectomia direita há 8 anos por carcinoma folicular da tiróide. Ao exame objetivo eram evidentes máculas e placas urticariformes dispersas pelo tronco e membros, artrite das interfalângicas proximais e adenopatias axilares bilaterais palpáveis, indolores e de consistência duro-elástica. Analiticamente apresentava elevação da velocidade de sedimentação (40mm, 1ªh) e da proteína C reativa (13mg/L). Os níveis de imunoglobulina (Ig) M estavam ligeiramente elevados (418mg/dL; N 55-300) enquanto a IgA, IgE e a IgG apresentavam valores séricos dentro da normalidade. A eletroforese de proteínas séricas evidenciou um pico de base larga na região gama. Contudo, nem a imunofixação sérica, nem a urinária, mostraram qualquer gamapatia monoclonal. Os anticorpos antinucleares foram positivos em título baixo (1/80, padrão homogéneo), com anti-ds-DNA, anti-ENA, ANCA e crioglobulinas negativos. A biópsia da pele mostrou lesão com características de dermatite granulocítica com leucocitoclasia, sem depósitos imunes. Apesar da diminuição dos níveis séricos de C1q, C3c e C4, a ausência de vasculite de pequenos vasos com necrose fibrinóide na histologia da pele e a negatividade dos anticorpos anti-C1q permitiram excluir uma vasculite urticariforme. A TC tóraco-abdomino-pélvica mostrou adenomegalias cervicais, axilares, mediastínicas e inguinais e hepatomegalia discreta. A tomografia por emissão de positrões com FDG-18 só evidenciou gânglios linfáticos hipermetabólicos supra e infradiafragmáticos. A biópsia aspirativa de uma das adenopatias axilares revelou histologia e imunofenotipagem compatíveis com gânglio reativo. A apresentação clínica e a histologia cutânea sugeriam um SSch, pelo que o diagnóstico foi assumido mesmo na ausência de gamapatia monoclonal. A doente iniciou tratamento com anacinra (100mg s.c./dia) com remissão da urticária e das artralgias em 48h, outro dado a favor do diag-

nóstico. No entanto, por reação sistemática no local da picada, fez switch para tocilizumab (162mg s.c./semana), apresentando boa resposta clínica aos 3 meses de terapêutica. Mantém-se em seguimento, sem aumento progressivo da IgM sérica e sem evolução para gamapatia monoclonal.

Conclusão: Apesar da ausência de gamapatia monoclonal, o que impede o cumprimento dos critérios de diagnóstico de SSch, este é um caso compatível com esta entidade em vários aspetos: as manifestações clínicas, os achados histológicos e a resposta à terapêutica. Da revisão da literatura, encontrámos apenas 8 casos descritos de SSch sem gamapatia monoclonal, que correspondem a 2-3% do total. Permanece por esclarecer se esta é uma entidade diferente ou se o espetro da doença deveria ser alargado para incluir este subtipo.

138 - ESPONDILARTRITE ASSOCIADA À DOENÇA INFLAMATÓRIA INTESTINAL – A IMPORTÂNCIA DA DISCUSSÃO MULTIDISCIPLINAR

Francisca Guimarães[1], Catarina Soares[1], Hugo Parente[1], Emanuel Costa[2], Daniela Santos-Faria[1], Joana Ramos Rodrigues[1], Filipa Teixeira[1], José Tavares-Costa[1], Carmo Afonso[1], Daniela Peixoto[1]
[1]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [2]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

Introdução: As manifestações reumáticas são o principal envolvimento extraintestinal da doença inflamatória intestinal (DII), sendo que cerca de 10% dos doentes desenvolve uma espondilartrite. Os anti-TNF são a base do tratamento biotecnológico das espondilartrites associadas à DII, contudo, perante a sua ineficácia ou toxicidades, são escassas as opções terapêuticas que demonstrem benefício em todas as manifestações desta patologia. Por este motivo, é crucial a abordagem multidisciplinar.

Caso Clínico: Homem de 32 anos, seguido em Reumatologia desde 2018 por espondilartrite axial associada a DII (colite ulcerosa), sob adalimumab 40 mg/quinzenal, desde junho de 2018, com boa resposta. Em março de 2019, por agravamento da doença intestinal, a posologia foi ajustada para 80 mg/quinzenal. Contudo, apesar de melhoria clínica, constatou-se o surgimento de psoríase palmo-plantar exuberante, refratária a tratamento tópico, com posterior sobreinfeção bacteriana, que motivou a suspensão deste fármaco. Em julho de 2019, iniciou infliximab 5mg/kg de 8/8 semanas, com melhoria progressiva da psoríase, mantendo-se assintomático do ponto de vista axial e intestinal até abril de 2020, altura em que teve agravamento da lombalgia inflamatória e iniciou oligoartrite dos membros inferiores e entesite (aquiliana e fáscia plantar). Neste contexto, iniciou metotrexato, prednisolona e, posteriormente, sulfassalazina, com melhoria da artrite periférica, mas persistência das queixas axiais e entesopáticas. Assim, foi ajustada a posologia do infliximab, inicialmente para 5mg/kg de 6/6 semanas e, posteriormente, para 10 mg/kg de 6/6 semanas, porém, sem benefício clínico evidente ao fim de 6 meses.

Face à refratariedade terapêutica, em março de 2021, após discussão com Gastroenterologia, foi efetuado switch para tofacitinib 5mg 2x/dia, mantendo os restantes imunomoduladores clássicos. Às 12 semanas após início de tofacitinib, o doente mantinha sacralgia inflamatória, entesopatia e reiniciou dor abdominal e diarreia, motivo pelo qual foi aumentada posologia para tofacitinib 10 mg bid, foi optimizada dose de salazopirina para 3 g/dia e iniciou messalazina 1000g/dia em supositório. Na avaliação após 4 semanas, o doente apresentava melhoria franca das queixas axiais, entesopáticas e gastrointestinais, com um BASDAI de 0.8 e um ASDAS PCR de 1.8, com diminuição dos parâmetros inflamatórios.

Conclusão: Este caso retrata a complexidade da espondilartrite associada à DII, a diversidade do envolvimento musculoesquelético e o desafio terapêutico na doença refratária ou perante efeitos adversos dos anti-TNF, onde as opções terapêuticas são parcas. Nestas situações, os inibidores da JAK surgem como uma opção com resultados promissores. No caso clínico descrito, o tofacinib na posologia 10 mg bid demonstrou benefício, não só nas queixas gastrointestinais, mas também axiais e entesopáticas. Salienta-se também a importância da multidisciplinaridade e decisão partilhada entre a Reumatologia e a Gastroenterologia na orientação destes doentes.

139 - COEXISTENCE OF GOUT, SEPTIC ARTHRITIS AND SUBACUTE INFECTIVE ENDOCARDITIS: A CASE REPORT

Francisca Guimarães[1], Miguel Costa[2], Hugo Parente[1], Diogo Esperança Almeida[1, 3], Ana Sofia Pinto[1, 4], Carmo Afonso[1], Filipa Teixeira[1], Joana Urbano[2], Joana Leite Silva[1, 3]
[1]Rheumatology Department, Unidade Local de Saúde do

Alto Minho, Ponte de Lima, Portugal, [2]Internal Medicine Department, Unidade Local de Saúde do Alto Minho, Viana do Castelo, Portugal, [3]Rheumatology Department, Hospital de Braga, Braga, Portugal, [4]Rheumatology Department, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal

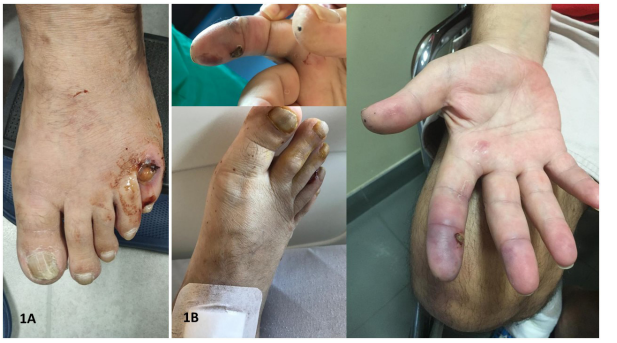
Background: Infective endocarditis (IE) usually presents with fever and heart murmur. It can affects every organ system1, with 15 to 30% of the patients reporting musculoskeletal symptoms. Septic arthritis due to septic embolization is a rare form of presentation. We report a 63-year-old male presenting with oligoarthritis in whom gout, septic arthritis and IE coexisted.

Clinical case: A 63-year-old male presented in our Rheumatology department with acute oligoarthritis involving the left wrist, and both tibiotarsal (TT) joints. The patient reported previous episodes of podagra and history of hyperuricemia. Additionally, he had personal history of prosthetic aortic and mitral mechanic valves, atrial fibrillation, schizophrenia, and drug and alcohol abuse in the past. He was diagnosed with gout and started on prednisolone, colchicine and non-steroid anti-inflammatory drug.

Three weeks later, he referred significant improvement of the articular complains except for the right TT joint. Also, he reported a traumatic wound in the left foot with purulent exudate in the previous two weeks and skin lesions in the previous week. On the physical examination, the patient presented monoarthritis of the right TT with exuberant inflammatory signs, an infected wound in the left foot (Figure 1A), Osler nodes and Janeway lesions (Figure 1B). He had no fever, dyspnea, heart murmurs or peripheral edema. Laboratory tests identified mild normocytic anemia, leukocytosis with neutrophilia, as well as elevated erythrocyte sedimentation rate (99 mm/min), and C-reactive protein (16.76 mg/dL), acute renal lesion (serum creatinine level of 2.0 mg/dL) and hyperuricemia (8.20 mg/dL). A diagnostic arthrocentesis was performed and allowed the diagnose of septic arthritis. Hemocultures, synovial fluid and the exudate from the right foot wound were sent to microbiology. The case was discussed with Internal Medicine, and due to the suspicion of IE, he started on empiric ceftriaxone, vancomycin, imipenem, which were then changed to vancomycin, gentamicin and imipenem.

He was transferred to the Orthopedic department where he was submitted to surgical TT arthrotomy and then transferred to the department of Internal Medicine. A transthoracic echocardiogram (TTE) was performed which showed

FIGURE 1



no evidence of vegetations.

Seven days after admission the patient suffered an embolic stroke with hemorrhagic transformation and was transferred to Neurosurgery department to be submitted to surgical drainage. Due to the high suspicion of IE of the mechanic valve and the low sensibility of TTE in these cases, a transesophageal echocardiogram (TEE) was performed which confirmed the diagnose of subacute IE. Later on, the blood cultures isolated the bacteria Kocuria Rosea.

This patient completed 7 weeks of antibiotics with clinical and analytic improvement. However, unfortunately, the patient suffered neurological and was referred to a rehabilitation unit.

Discussion: This case highlights the need for a high suspicion for IE and the early start of empiric antibiotic, since in the reported case the patient didn't present with fever and the TTE was normal. Yet, the presence of Osler and Janeway lesions associated with the septic embolization made the diagnose of IE the most probable. Also, it alerts that in a high-risk patient such as with prosthetic heart valves, the diagnosis of septic arthritis should alert for the presence of IE, especially in the presence of bacteriemia. Finally, it also reports an atypical agent for IE, Kocuria Rosea, with only few reported cases in literature.

143 - MERALGIA PARESTÉSICA - A PROPÓSITO DE UM CASO CLÍNICO

Cristina Prata[1], Patricia Ribeiro Santos[1], Rute Nascimento[2], Luis Gonçalves[1], Patricia Figueiredo[1]
[1]USF Sobreda, ACeS Almada-Seixal, Almada, Portugal, [2]USF Arandis, ACeS Oeste Sul, Torres Vedras, Portugal

A meralgia parestésica (MP), ou síndrome Bernhardt–Roth, consiste numa mononeuropatia compres-

siva manifestada por disestesia na distribuição do nervo cutâneo femoral lateral (ncfl). Estes doentes tipicamente descrevem sensação de queimadura, frio, choque, parestesia ou alteração da sensibilidade cutânea na face antero-lateral da coxa. Não têm alterações motoras ou da força muscular e os reflexos estão preservados. Os sintomas podem ser leves e fugazes, desaparecendo espontaneamente, ou intensos, limitando o doente durante anos. Pode haver queixas de dor na face lateral da coxa até ao joelho que podem agravar com a extensão da perna e, por isso, estes doentes evitam estar muito tempo de pé. Sentar pode aliviar. Em alguns casos não há posição de conforto. Frequentemente, os doentes são tratados para patologias da coxa ou joelho, ou para lombalgia antes do diagnóstico de MP. Como os doentes têm dificuldade em descrever os sintomas, não é incomum pensar em distúrbio psiquiátrico.

Ao tornar-se superficial o ncfl é suscetível a trauma e compressão por causas como: gravidez, órtese, cinto apertado, escoliose e espasmo muscular. Pode haver lesão por incisões abdominais ou pélvicas, tais como na apendicectomia. A compressão também pode ocorrer dentro da pélvis, em casos de mioma uterino, abcesso e tumor retroperitoneal.

O diagnóstico de MP é essencialmente clínico. A eletroneuromiografia (EMG) pode ser utilizada, porém há dificuldade na obtenção de potenciais sensitivos, o que prejudica a análise do mesmo. Ressonância magnética e tomografia computadorizada são ineficazes para a localização da compressão, mas devem ser utilizadas na determinação da sua causa e no diagnóstico diferencial.

Trata-se de uma mulher de 35 anos, que recorre à consulta no centro de saúde a 2/6/2021 por queixas de parestesia na face antero-lateral da coxa direita, sem perda de força, com cerca de 3 meses de evolução. Nega trauma. Antecedentes pessoais irrelevantes, à exceção do parto a 27/2/2021 às 39 semanas por ventosa. Havia já recorrido à urgência hospitalar por duas vezes, realizou radiografia da coluna lombar e anca, tendo sido medicada com AINE, sem melhoria.

Na consulta foi solicitado EMG que revelou "ausência do potencial de ação sensitivo do nervo femoro-cutâneo externo direito. Condução nervosa normal. Condução nervosa dos nervos peroneal comum (motora) e sural (sensitiva), dentro da normalidade. Nos músculos tibial anterior, gémeo interno, adutor longo, psoas-iliaco e quadricipete femoral observam-se potenciais de unidade motora de características normais."

Perante a história clínica, exame objetivo e resultado do EMG, foi colocada a hipótese de MP e foi referenciada à consulta de Reumatologia, onde realizou infiltração local com corticoide, com melhoria significativa das queixas.

A MP poucas vezes é considerada como diagnóstico diferencial, visto a sintomatologia se sobrepor à de outros, mais comuns, como a lombalgia. A avaliação clinica é feita através do mapeamento cuidadoso da área de disestesia, aumentando o grau de suspeição para MP e confirmando o envolvimento do ncfl.

O diagnóstico de MP é essencialmente clínico, não tendo sempre a EMG um resultado esclarecedor. O tratamento inicial é conservador: analgésicos, anti-inflamatórios não esteroides, uso de roupas largas, perda de peso, corticóides e anestésicos locais, consoante a causa da compressão. Quando falha, considerar cirurgia, não havendo consenso sobre o melhor procedimento: ressecção ou descompressão do nervo com excisão da porção lateral do ligamento inguinal.

144 - AUTOLOGOUS HEMATOPOIETIC STEM CELL TRANSPLANTATION IN A PATIENT WITH PROGRESSIVE LIMITED SYSTEMIC SCLEROSIS

Raquel Freitas[1], Carlos Pinho Vaz[2], António Campos[2], Ana Cordeiro[1]

[1]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [2]Serviço de Transplantação de Medula Óssea, Instituto Português de Oncologia (IPO) Porto, Porto, Portugal

Background: Systemic sclerosis (SSc) is a rare connective tissue disease with a broad spectrum of clinical and immunologic features. Treatment in SSc is aimed at symptom control and management of organ complications. Off-label use of immunomodulators and/or immunosuppressants is frequent in clinical practice. The use of autologous hematopoietic stem cell transplantation (HSCT) for severe or rapidly progressive SSc has been used and recent randomized controlled trials proved its efficacy in SSc (1-3).

Case report: A 44 year-old woman was diagnosed with limited cutaneous SSc in our center. At SSc diagnosis, she had Raynaud phenomenon for 6 years, peripheral polyarthralgia, severe gastrointestinal (GI) complains in the last 2 years, distal skin thickening and microstomia. At that time point there was evidence of myositis and interstitial lung disease (non-specific interstitial pneumonia involving more

than 20% of lung parenchyma). She was started on low dose prednisolone, mycophenolate mofetil (MMF) up to 2000 mg/day, hydroxychloroquine 400mg/day, proton pump inhibitor, prokinetics, mebeverine and pentoxifylline. Despite this, our patient maintained with severe GI complains, no improvement in functional respiratory tests and had major impairment on her quality of life. Furthermore, MMF had to be suspended due to leucopenia and hepatic liver enzymes elevation. Consequently, she was proposed and accepted to autologous hematopoietic stem cell transplantation (HSCT) in Instituto Português de Oncologia (IPO) in Porto city.

She had some complications after the procedure that were easily management during hospital stay, namely an acute pulmonary edema and fever (microbiologic cultures negative), but she had no further complications in the following months.

After 8 months of HSCT the patient maintains without pharmacologic immunosuppression. Skin thickening slightly improved, CK levels normalized, without muscle pain nor weakness and pulmonary function tests also improved (Table 1). Nevertheless, she maintains with severe GI complains.

We decided to describe the first case of SSc in our rheumatology department submitted to HSTC. Despite remaining with disease activity 8 months after HSCT, specially with GI complains, our patient had no further progression of the disease and even improved her functional respiratory tests, skin thickening and myositis. Our patient had at diagnosis a severe and progressive SSc and had hepatic liver enzymes elevation with MMF. As so, HSCT offered the possibility of an immunosuppression-free period, no worsening of a progressive and severe disease, and improvement of organ manifestations that are associated with worst outcome such as ILD.

TABLE 1 – PARAMETERS EVALUATED AT DIAGNOSIS AND 8 MONTHS AFTER HSCT

	At diagnosis	8 months after HSCT
Skin thickening (mRSS)	9	7
FVC	60	64.7
DLCO	32	42.4
PASP	23	22
Hemoglobin	12.8	13.1
CK	700	122
UCLA STC GIT 2.0	1.24	0.84

CK - creatine kinase; DLCO – diffusing capacity for carbon monoxide; FVC – forced vital capacity; UCLA STC GIT 2.0 – UCLA Scleroderma Clinical Trial Consortium Gastrointestinal Tract 2.0; mRSS - modified Rodnan skin score; PASP – pulmonary artery systolic pressure; SF-36 – 36-Item Short Form Survey.

REFERENCES:

1. Burt RK, Shah SJ, Dill K, et al. Autologous non-myeloablative haemopoietic stem-cell transplantation compared with pulse cyclophosphamide once per month for systemic sclerosis (ASSIST): an open-label, randomised phase 2 trial. Lancet 2011; 378: 498-506
2. Van Laar JM, Farge D, Sont JK, et al. Autologous hematopoietic stem cell transplantation vs intravenous pulse cyclophosphmide in diffuse cutaneous systemic sclerosis: a randomized clinical trial. JAMA 2014; 311: 2490-2498
3. Sullivan KM, Goldmuntz EA, Keyes-Elstein L, et al. Myeloablative autologous stem-cell transplantation for severe scleroderma. N Engl J Med. 2018;378(1):35-47.

158 - NEGLIGENCIAR A ARTRITE REUMATÓIDE – QUAIS AS CONSEQUÊNCIAS?

Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Tiago Beirão[1], Flávio Campos Costa[1], Romana Vieira[1], Taciana Videira[1], Joana Aleixo[1], P Pinto[1]
[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

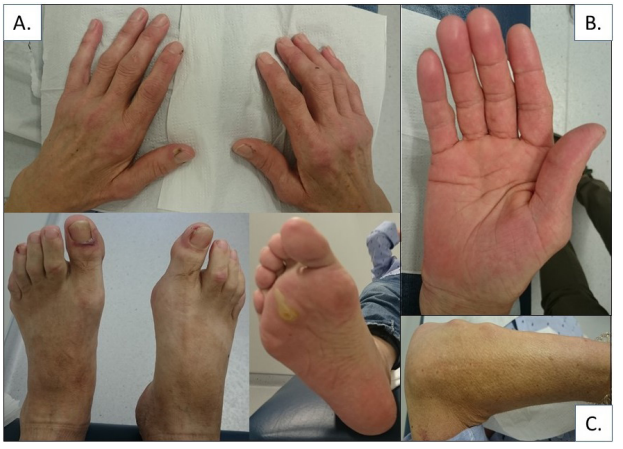
Introdução: A crioglobulinemia mista é uma vasculite de pequenos vasos mediada por imunocomplexos, classicamente associada a infeção pelo vírus da Hepatite C, mas que pode estar associada a doenças do tecido conjuntivo.

Descrição: Mulher de 54 anos com osteoporose (OP) fraturária sob bifosfonato oral, enviada à consulta por poliartrite com atingimento aditivo e simétrico de pequenas e médias articulações desde há 10 anos, sob prednisolona (PDN) em SOS. Referia emagrecimento acentuado e aparecimento no último ano de lesões nodulares na superfície extensora dos antebraços e de lesões cutâneas periungueais e na planta dos pés. Negava outros sintomas constitucionais e sugestivos de conectivite. À primeira observação, apresentava marcha com apoio em canadiana, caquexia, telangiectasias faciais, esclerodactilia, múltiplas fissuras e eritema periungueais nas mãos e pés e ulcerações punctiformes da planta do pé direito (fig. 1A), desvio cubital dos dedos das mãos, polegar em Z (fig. 1B), nódulos reumatóides nos antebraços (fig. 1C) e mobilidade passiva limitada e dolorosa da articulação coxo-femural (CF) direita. Por diagnóstico clínico de artrite reumatóide (AR) e enquanto se procedia ao estudo de vasculite/sobreposição com esclerose sistémica (ES) iniciou metotrexato (MTX) 10mg/sem, PDN 15 mg/dia e nifedipina 30mg/d. Analiticamente

com velocidade de sedimentação (VS) de 80 mm/h, proteína C reactiva (PCR) de 1.14 mg/dL, FR de 74 UI/mL, ACCP de >340 U/mL, ANA de 1/640 com padrão homogéneo e imunoblot de anticorpos anti--nucleares, citoplasmáticos e miopatias negativos, anticorpos específicos de ES, do síndrome antifosfolipídico e ANCA MPO e PR3 negativos, enzimas musculares normais, IgG de 1490 mg/dL (680-1450) e IgM normal, eletroforese das proteínas com faixa gama de base larga de 1.60 g/dL e imunofixação negativa, marcadores víricos negativos e proteinúria de 667,20 mg/24h. O estudo radiográfico mostrou artrite erosiva das mãos e pés e necrose avascular (NAV) da CF direita. A capilaroscopia não mostrou padrão esclerodérmico. Foram excluídas causas paraneoplásicas e infeciosas. Por manter poliartrite e intolerância ao MTX, iniciou leflunomida 10 mg/d, também suspensa por diarreia. Realizou pulso de metilprednisolona (MTP) 500mg sem melhoria clínica ou analítica (VS=86 mm/h e PCR=8.50 mg/dL com proteinúria de 1788,70 mg/d, sem alteração da função renal). Pela presença de crioglobulinas positivas do tipo II efetuou-se o diagnóstico de crioglobulinemia mista do tipo II com envolvimento cutâneo e renal em associação com AR de longa evolução não tratada. Realizou 3 pulsos de MTP 1g e iniciou rituximab (RTX) 1g semestral com melhoria clínica e analítica. Aquando do 1º ciclo de RTX em TC tórax verificou-se a presença de tromboembolismo (TEP) da artéria lobar inferior direita pelo que iniciou hipocoagulação. Questiona--se se o TEP foi uma manifestação da vasculite ou paraneoplásica, dado que, 3 meses após, teve o diagnóstico de adenocarcinoma do pulmão (T1N0M0), tendo sido tratada com radioterapia localizada com intuito curativo. Realizou artroplastia total da anca direita com resolução das queixas álgicas e atualmente caminha sem apoio.

Conclusão: A crioglobulinemia mista tipo II é uma consequência infrequente da AR e raramente se associa a tumores sólidos. Este caso ilustra como a perpetuação de um estado inflamatório pode culminar num quadro vasculítico ampliando a gravidade do prognóstico com dano direto de órgãos nobres como o rim. Acrescem ainda as sequelas de anos de corticoterapia sem supervisão como a OP e a NAV.

FIGURE. : EXAME FÍSICO: (A) LESÕES PERI-UNGUEAIS E PLANTARES; (B) DESVIO CUBITAL DOS DEDOS; (C) NÓDULOS REUMATÓIDES



159 - LOMBALGIA - ANALISAR COM SENSIBILIDADE E PERSPICÁCIA

Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Tiago Beirão[1], Flávio Campos Costa[1], Romana Vieira[1], Taciana Videira[1], Joana Aleixo[1], P Pinto[1]
[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

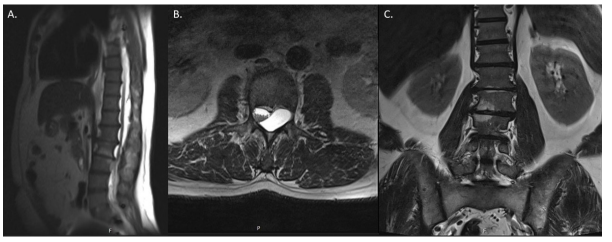
Introdução: O envolvimento axial pela artrite psoriática é uma manifestação relativamente comum, ocorrendo isoladamente em cerca de 5% dos casos e em associação a outros padrões em até 40% dos doentes.

Caso Clínico: Homem de 55 anos seguido em consulta de Reumatologia por Artrite Psoriática com envolvimento periférico articular e ocular, sob metotrexato 20 mg/semana e prednisolona 10 mg/dia, inicia quadro de lombalgia aguda, persistente ao longo do dia, agravada em ortostatismo mas também com despertar noturno e rigidez prolongada, com irradiação para a região nadegueira e face posterior da coxa bilateralmente associado a mialgias e disestesias inespecíficas, sem alterações ao exame neurológico. Analiticamente apresentava elevação dos parâmetros inflamatórios (VS 47 mm/h e PCR 9.23 mg/dL). O estudo radiológico mostrava ligeira esclerose do bordo ilíaco das articulações sacroilíacas (SI) e na coluna dorso-lombar ligeira escoliose com retilinização lombar, osteofitose e discopatias das últimas vértebras dorsais até L3. Iniciou acemetacina 180mg/dia mas por agravamento progressivo com limitação nas atividades diárias ao longo de 1 mês associou--se sucessivamente diazepam 10mg/dia, gabapentina em aumento até 900 mg/dia, tapentadol em aumento até 200mg/dia e inclusivamente prednisolona 20mg/dia mas sem resposta clínica. Na última observação vinha em cadeira de rodas e depressivo por quadro

álgico intenso e incapacidade funcional. Realizou RM das SI que não mostrou sacroileite. A EMG apresentou radiculopatia de L3 e L4 bilateral. A RM lombar relatava uma volumosa formação quística intracanalar no espaço epidural, com extensão craniocaudal de D11 até L3, medindo cerca de 14,5 cm x 2,5 cm de diâmetros crânio-caudal e transverso, com algumas septações internas, condicionando moldagem óssea, com retro-escavação da vertente posterior dos corpos vertebrais, sugestiva de quisto meníngeo tipo Ia (fig. 1-A). O quisto condicionava marcada deformação do saco dural, desviando-o anterior e lateralmente para a direita, propiciando compressão plurirradicular em D12-L1, L1-L2 e L2-L3, bem como moldagem e deformação do cone medular provocando provável mielopatia (fig. 1-B). Existiam ainda sinais de dificuldade no retorno venoso associado a estenose do canal raquidiano nos níveis superiores a L3. Discutidas as imagens com a Imagiologia colocou-se ainda a hipótese da presença de lesões inflamatórias de Anderrson (fig. 1-C). O doente fez prednisolona 40mg/dia durante 3 dias no pré-operatório e foi submetido a intervenção cirúrgica com recuperação motora mantendo lombalgia ligeira. Pela presença provável das lesões de Anderrson e episódios de repetição de episclerite foi pedido adalimumab e irá repetir posteriormente RM lombar.

Conclusão: Este caso retrata a dificuldade no diagnóstico diferencial de uma lombalgia mista num doente com artrite psoriática. As lesões de Anderrson surgem em cerca de 8% dos doentes com espondilartrite, podendo ser detetadas precocemente por RM. Os quistos meníngeos subaracnoideus são incomuns e, quando surgem, habitualmente são assintomáticos surgindo como achados incidentais em RM. Neste caso, havendo melhoria significativa com a cirurgia, parece tratar-se de um caso peculiar de quisto meníngeo subaracnoideu sintomático.

FIGURE: RM LOMBAR - (A, B) QUISTO SUBARACNOIDEU; (C) LESÕES DE ANDERRSON



167 - SÍNDROME DO CONFLITO ISQUIOFEMORAL – UMA CAUSA RARA DE COXALGIA

Maria Seabra Rato[1], Raquel Miriam Ferreira[1], Filipe Oliveira Pinheiro[1], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal

Introdução: A dor na anca é uma queixa músculo-esquelética frequente e muitas vezes de diagnóstico difícil, devido à complexidade anatómica loco-regional e assim ao amplo espectro de possíveis etiologias. Uma das hipóteses a considerar é a síndrome do conflito isquiofemoral. Inicialmente descrita apenas em mulheres pós-artroplastia da anca foi posteriormente também observada em indivíduos sem história de cirurgia ou traumatismo. Trata-se de uma condição pouco reconhecida, dada inespecificidade dos sintomas, requerendo realização de ressonância magnética (RM) para o adequado diagnóstico.

Caso clínico: Doente do sexo masculino, de 73 anos, é enviado à consulta de Reumatologia por coxalgia esquerda. Como antecedentes pessoais de relevo destacavam-se adenocarcinoma do cólon ascendente tratado cirurgicamente há 13 anos e diagnóstico recente de linfoma folicular, sem evidência de infiltração medular, tendo feito 6 ciclos de quimioterapia com utilização de doses elevadas de corticoterapia. Apresentava também múltiplos fatores de risco cardiovascular: hipertensão arterial, dislipidemia e tabagismo. O doente referia uma dor na anca esquerda na região póstero-lateral, de ritmo mecânico, com 6 meses de evolução, ocasionalmente com irradiação para o membro inferior esquerdo, limitante para a marcha e de agravamento progressivo. Negava dor noturna ou sintomas constitucionais. Ao exame objetivo, apresentava bom estado geral, sem atrofias ou deformidades visíveis; dor à palpação do quadrante inferior externo da região glútea esquerda e na extensão passiva da anca em adução; palpação do grande trocânter e manobra de abdução resistida da anca indolores e sem limitação da mobilidade da articulação coxofemoral; marcha antálgica à esquerda. A radiografia da bacia mostrou ligeiras alterações degenerativas das coxofemorais bilateralmente, sem sinal de crescente ou perda da esfericidade femoral; calcificações heterotópicas projetadas acima da grande tuberosidade do fémur esquerdo e uma assimetria com substancial diminuição do espaço entre o pequeno trocânter do fémur e a tuberosidade isquiática à esquerda compa-

rativamente ao lado contra-lateral. A radiografia da coluna lombar evidenciou discopatia L5-S1. Analiticamente, apresentava hemograma, fosfatase alcalina, cálcio e fósforo, lactato desidrogenase láctica e marcadores inflamatórios dentro dos parâmetros de normalidade. A RM da anca esquerda evidenciou aumento de sinal na sequência T2 do músculo quadrado femoral sugestiva de edema intramuscular associado a uma redução da amplitude do espaço isquiofemoral. Não eram evidentes sinais de necrose avascular ou de lesões tumorais. A RM da coluna lombar não mostrou compressão radicular. As manifestações clínicas, o exame objetivo e os achados dos exames complementares de diagnóstico foram compatíveis com síndrome de conflito isquiofemoral. Foi instituída terapêutica analgésica e o doente foi proposto para tratamento fisiátrico com melhoria das queixas ao fim de 3 meses de tratamento.

Conclusão: Este caso exemplifica a importância do diagnóstico diferencial na avaliação de um doente com dor na anca. Embora a história clínica pudesse inicialmente sugerir outros diagnósticos mais prováveis, a elevada suspeição clínica permitiu o diagnóstico de uma condição menos frequente. O reconhecimento desta entidade e a exemplificação deste caso poderão contribuir para a identificação de situações semelhantes muitas vezes subdiagnosticadas.

169 - SEVERE CYTOMEGALOVIRUS COLITIS IN A PATIENT WITH SYSTEMIC VASCULITIS: A RARE COMPLICATION

Maria Helena Lourenço[1, 2], João Borralho[3, 4], Inês Silva[1, 5], João Alves[3, 5], Rita Sampaio[6], Kamal Mansinho[3, 7], Jaime C. Branco[1, 2, 5]

[1]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [2]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [3]Infectious Diseases and Tropical Medicine, Centro Hospitalar de Lisboa Ocidental, Lisboa, Portugal, [4]Institute of Microbiology, Faculdade de Medicina da Universidade de Lisboa, Lisboa, Portugal, [5]NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [6]Pathological Anatomy Department, Centro Hospitalar de Lisboa Ocidental - Hospital Egas Moniz, Lisboa, Portugal, [7]Institute of Hygiene and Tropical Medicine, Universidade NOVA de Lisboa, Lisboa, Portugal

Introduction: Eosinophilic granulomatosis with polyangiitis (EGPA) is a vasculitis of small- and medium-sized vessels characterized by lung, paranasal sinus, skin, kidney, nervous system and joints involvement, associated with peripheral eosinophilia. Immunosuppressive drugs are the mainstay of treatment, but their use increases the risk of opportunistic infections.

Cytomegalovirus (CMV) infection is common and usually benign. However, in immunocompromised patients, reactivation can occur, carrying poorer outcomes. Colitis is a rare complication mainly reported in patients with human immunodeficiency virus and transplant recipients, with scarce data in ANCA-associated vasculitis (AAV).

We report a case of colonic perforation due to confirmed CMV infection in a patient with EGPA previously treated with high-dose immunosuppressants.

Case Report: A 78-year-old woman diagnosed in May 2020 with EGPA (asthma, sinusitis, bullous hemorrhagic vasculitis, symmetric and additive polyarthritis of hands, elbows and tibiotarsal joints, sensorimotor polyneuropathy and peripheral eosinophilia), previously treated with prednisolone 1mg/kg/day, methotrexate 10mg/week and cyclophosphamide (2 cycles; CYCLOPS protocol), in clinical remission, was admitted to the emergency room due to abdominal pain, nausea and vomiting. Abdominal computed tomography scan revealed colonic thickening, with perforation of the anorectal transition and extraperitoneal air. The patient underwent surgery, during which a bleeding rectal ulcer was detected. Biopsies were performed, followed by a terminal colostomy and a course of broad-spectrum antibiotics.

Immunohistochemical examination of the ulcer was positive to CMV infection. CMV viral load (VL) was 254 IU/mL, compatible with disseminated CMV disease. Two months before, CMV serologies had revealed positive M and G immunoglobulins, with CMV VL <178 IU/mL. No signs or symptoms of other CMV organic disease were documented. After 21 days of treatment with ganciclovir, CMV VL was undetectable. Secondary prophylaxis with valganciclovir was started in conjunction with the vasculitis treatment (prednisolone 20mg/day), with no CMV disease recurrence.

Discussion: EGPA is a potential life-threatening disease. Glucocorticoids, cyclophosphamide and rituximab induce remission of the severe forms of disease, predisposing patients to opportunistic infections, such as disseminated CMV disease.

There is sparse data regarding prophylactic or pre-emptive treatment of CMV infection in patients with AAV. This case reinforces the need of guidelines concerning surveillance and prophylaxis of CMV infection in these patients.

171 - PAQUIDERMODACTILIA: O IMITADOR DA ARTRITE IDIOPÁTICA JUVENIL

Ana Bento da Silva[1, 2], Maria Helena Lourenço[1, 2], Maria João Gonçalves[1, 2, 3], Jaime C. Branco[1, 2, 4], Manuela Costa[1], Ana Filipa Mourão[1, 2]

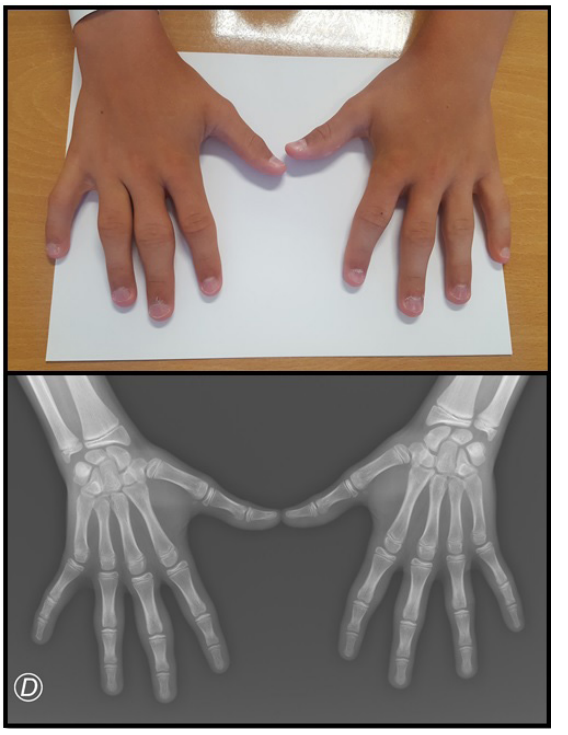
[1]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [2]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [3]Instituto Politécnico de Lisboa, Escola Superior de Tecnologia da Saúde de Lisboa, Lisboa, Portugal, [4]Comprehensive Health Research Center – CHRC, Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, Lisboa, Portugal

Introdução: A Paquidermodactilia é uma forma rara benigna de fibromatose digital, caracterizada por um edema de partes moles dos dedos das mãos, assintomático1. Geralmente, o envolvimento é bilateral e simétrico, com predileção pelas articulações interfalângicas proximais (IFPs) do segundo, terceiro e quarto dedos2. É mais prevalente em adolescentes do sexo masculino e a sua etiologia é desconhecida, apesar de se admitir associação com estimulação mecânica repetitiva2. O diagnóstico diferencial é extenso, destacando-se a artrite idiopática juvenil2.

Caso Clínico: Jovem do sexo masculino, 12 anos de idade, com antecedentes de eczema atópico. Apresenta-se em consulta de Reumatologia por quadro de início insidioso e lentamente progressivo com 1 ano de evolução de tumefação indolor das IFPs de ambas as mãos, sem limitação da mobilidade associada. Negava afeção de outras articulações, psoríase ou outras manifestações cutâneas, alterações do trânsito intestinal, fenómeno de Raynaud, sintomatologia respiratória ou cardiovascular ou trauma prévio. Objetivamente, apresentava deformação e tumefação das IFPs do segundo, terceiro, quarto e quinto dedos de ambas as mãos, mais notório à direita, mas sem dor ou limitação de movimentos; sem outros achados a destacar. Realizou terapêutica com ibuprofeno, sem benefício. Analiticamente: sem elevação de parâmetros inflamatórios; hemograma, função renal e hepática e ionograma incluindo cálcio sem alterações; anticorpos antinucleares com título de 1/160, HLA B27 positivo e elevação da enzima de conversão da angiotensina (143 U/L); restante estudo imunológico negativo, incluindo fator reumatoide, anticorpo anti-CCP, anticorpo anti-dsDNA, anti-antigénios nucleares solúveis, anti-antigénios hepáticos, anti-Saccharomyces cerevisiae e anti-citoplasma do neutrófilo; serologias para Rubéola, Toxoplasma, Citomegalovírus, Vírus Epstein-Barr, SARS-CoV2, Vírus da Imunodeficiência Humana e Vírus da Hepatite B e C negativas. A radiografia das mãos evidenciou edema de partes moles, sem envolvimento ósseo ou articular e a ecografia não revelou quaisquer alterações sugestivas de atividade inflamatória articular ou peri-articular. Apesar dos achados analíticos, o doente mantem-se assintomático e sem evidência de outras manifestações, razão pela qual foi admitido o diagnóstico de Paquidermodactilia. Pela natureza benigna desta entidade e ausência de sintomas, não foi introduzida terapêutica dirigida. Mantém, contudo, vigilância regular em consulta de Reumatologia.

Conclusão: A Paquidermodactilia é considerada o

FIGURE. PAQUIDERMODACTILIA: TUMEFACÇÃO DAS ARTICULAÇÕES INTERFALÂNGICAS PROXIMAIS 2 A 5 DE AMBAS AS MÃOS; RADIOGRAFIA DAS MÃOS DO DOENTE



imitador de muitas doenças osteoarticulares, particularmente a artrite idiopática juvenil, tornando o seu diagnóstico um grande desafio e conduzindo muitas vezes à realização desnecessária de múltiplos exames complementares2,3. Contudo, é prudente a vigilância regular em consulta de Reumatologia.

REFERÊNCIAS:

1. Chu, H., Song, J., & DO YOUNG, K. I. M. (2016). Pachydermodactyly: a benign cutaneous condition that may be misdiagnosed as a joint disorder. The Journal of rheumatology, 43(8), 1615-1616.
2. Naviglio, S., Mazzolai, M., & Ventura, A. (2017). Bilateral Finger Swelling in an Adolescent. The Journal of pediatrics, 188, 299.
3. Agudo-Mena, J. L., Buedo-Rubio, M. I., García-Atienza, E. M., & Escario-Travesedo, E. (2017). Pachydermodactyly: The great mimicker. Reumatologia clinica, 15(6), e156-e157.

173 - HEADACHE IN SYSTEMIC ERYTHEMATOUS LUPUS: A TRICKY DIAGNOSIS

Maria Helena Lourenço[1, 2], Ana Bento da Silva[1, 2], Rita Pinheiro Torres [1, 2], Nuno Pina Gonçalves[1, 2, 3], MJ Gonçalves [1, 2], Inês Silva[1, 4], Ana Filipa Mourão[1, 2, 4], Jaime C. Branco[1, 2, 4]

[1]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [2]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [3]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [4]NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

Introduction: Systemic Lupus Erythematosus (SLE) is a chronic and heterogenous autoimmune disease. Neuropsychiatry manifestations are variable and common (14-95%) but associated with a worse prognosis1. Headache may be a symptom of the disease itself or may have other aetiologies as infection, medication, or others (like hypertension or sinusitis).

We present a case report of a woman previously diagnosed with SLE who suffered from headache and fever for one week until being admitted in our inward department; after a thorough investigation, she was diagnosed with cytomegalovirus (CMV) meningitis.

Clinical case: A 61-year-old female diagnosed in 2016 with SLE (titres of antinuclear antibodies of 1/160, leukopenia, polyarthralgias without arthritis, oral ulcers, and non-scarring alopecia), and medicated with azathioprine (AZA) 150mg/day and deflazacort 3mg/day, was admitted to the Rheumatology inward department in August 2020, due to a holocranial headache, recurrent fever, asthenia, anorexia, night sweats, and oligoarthralgias, lasting from one week. At admission, the patient was febrile (38.4ºC) and a systolic murmur was audible on the aortic focus; neurological and articular exam were normal. Blood analysis revealed normocytic normochromic anaemia (9,8 g/dL), leukopenia (3100/µL) and lymphopenia (840/µL), moderately high inflammatory markers (erythrocyte sedimentation rate 55 mm/h; C-reactive protein 3mg/dL), hepatic cytolysis (aspartate aminotransferase 61U/L, alanine aminotransferase 63U/L; lactate dehydrogenase 555U/L), normal levels of complement fraction 3 and 4, negative titres of anti-double stranded DNA antibodies and absence of antiphospholipid antibodies; blood cultures were negative. The brain tomography scan was normal, as also as the brain magnetic resonance, excluding typical images from neurological involvement of SLE. Bone marrow biopsy excluded a myeloproliferative syndrome. Due to sustained fever and heart murmur, a heart ultrasound was performed to exclude endocarditis; the exam showed no intra-cavitary mass or thrombi. In the meantime, viral serologies revealed a positive CMV immunoglobulin M and G in the serum, with a high viral load (VL) (10442UI/mL) and the following lumbar punction showed a cerebrospinal fluid (CSF) positive for CMV (100 copies/mL) and negative for other agents. CMV meningitis was assumed as a primoinfection of a disseminated CMV disease and ganciclovir was started (5mg/kg 12/12h); the patient was transferred to the Infectious Diseases department. Endoscopic studies excluded gastrointestinal involvement and ophthalmological evaluation excluded retinitis. After a 17-day course of therapy, there was no recurrence of headache or fever; inflammatory markers were negative, as well as serum CMV VL. Lumbar puncture was repeated and did not detect CMV VL in the CSF. The patient was discharged with no need of further therapy (Systemic Lupus Erythematosus Disease Activity Index 1); AZA was suspended and hydroxychloroquine started (400mg/day).

Discussion: Neurological manifestations of disseminated CMV disease include meningitis, encephalitis, and retinitis; they are usually associated to an immunocompromised status2. In SLE, CMV meningitis is rarely described, with only one case reported after a brief review in literature3. The patient described in

this case was immunocompromised due to therapy and persistent lymphopenia. The investigation until the final diagnosis was long and not always obvious, which enhances the importance of a thorough investigation when facing a SLE patient with new-onset headache.

174 - A OUTRA FACE DOS FÁRMACOS BIOTECNOLÓGICOS: UM CASO DE TOXIDERMIA A ADALIMUMAB

Ana Bento da Silva[1, 2], Maria Helena Lourenço[1, 2], João Teles de Sousa[3], Rita Sampaio[4], Jaime C. Branco[1, 2, 5], Maria João Gonçalves[1, 2, 6]

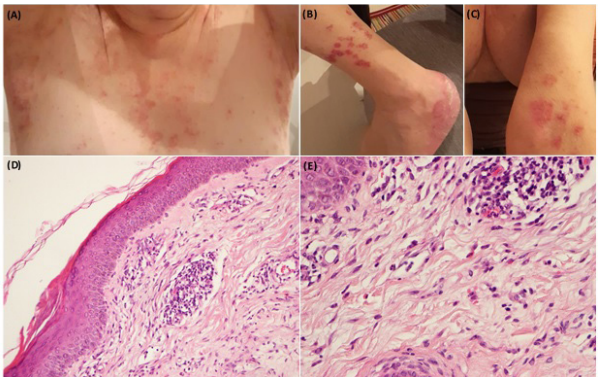
[1]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [2]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [3]Dermatology, Centro Hospitalar de Lisboa Ocidental EPE, Hospital de Egas Moniz, Lisboa, Portugal, [4]Pathological Anatomy Department, Centro Hospitalar de Lisboa Ocidental - Hospital Egas Moniz, Lisboa, Portugal, [5]Comprehensive Health Research Center – CHRC, Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, Lisboa, Portugal, [6]Instituto Politécnico de Lisboa, Escola Superior de Tecnologia da Saúde de Lisboa, Lisboa, Portugal

Introdução: O adalimumab é um anticorpo monoclonal recombinante humano anti Fator de Necrose Tumoral (TNF), aprovado para o tratamento de diversas doenças reumatológicas, incluindo a artrite psoriática1,2. A sua eficácia já foi largamente comprovada, mas apesar da sua boa tolerância, os agentes anti-TNF não são desprovidos de eventos adversos. Estudos prévios reportam o surgimento de lesões cutâneas em cerca de 25% dos doentes (maioritariamente na forma de reações no local da injeção, infeções cutâneas, dermatite eczematosa, psoríase e tumor cutâneo não melanoma), porém, são geralmente ligeiras e raramente conduzem à suspensão da terapêutica3,4.

Caso Clínico: Mulher, 69 anos, com história de psoríase palmo-plantar, osteoartrose nodal, síndrome do túnel cárpico e perturbação depressiva. Apresenta diagnóstico de Artrite Psoriática desde 2019, medicada inicialmente com metotrexato subcutâneo 15mg/semana com boa resposta clínica, mas posteriormente substituído por sulfassalazina até dose de 3g/dia por intolerância gastrointestinal ao primeiro agente. Após switch terapêutico, desenvolve novo flare articular e cutâneo, com poliartrite (punhos, metacarpofalângicas e interfalângicas proximais das mãos), bursite subacromiodeltoideia esquerda recorrente e agravamento das lesões psoriáticas palmo-plantares. Foi proposto início de terapêutica biológica com adalimumab, que iniciou em outubro de 2020, com melhoria considerável tanto do ponto de vista articular como cutâneo. Contudo, após a terceira administração, desenvolve agravamento marcado das lesões psoriáticas e rash cutâneo generalizado eritematoso e pruriginoso. A biópsia cutânea revelou paraqueratose e espongiose focais e, na derme superficial, ectasia vascular e infiltrado linfocitário perivascular discreto com alguns eosinófilos, aspetos compatíveis com toxidermia. Suspendeu adalimumab em janeiro de 2021 com melhoria progressiva do rash cutâneo, mas reativação da doença de base, razão pela qual iniciou secucinumab 150mg/mês em março de 2021, com necessidade posterior de incremento da dosagem para 300mg/mês por resposta insatisfatória. Mantém vigilância regular em consulta de Reumatologia e Dermatologia, aguardando-se resposta terapêutica.

Conclusão: Os fármacos biotecnológicos vieram revolucionar a abordagem de muitas doenças reumatológicas, mas os eventos adversos a eles associados não são desprezáveis, principalmente numa época de grande disseminação destes agentes. Este caso vem alertar para a necessidade de reconhecer estas raras manifestações cutâneas, que tornam inevitável a suspensão do fármaco, impondo grandes desafios na abordagem terapêutica destes doentes.

FIGURA 1: (A-C) LESÕES CUTÂNEAS APRESENTADAS PELA DOENTE APÓS INÍCIO DE ADALIMUMAB. (D-E) BIÓPSIA CUTÂNEA



REFERÊNCIAS:

1. https://www.ema.europa.eu/en/documents/product-information/humira-epar-product-information_pt.pdf

2. https://reference.medscape.com/drug/amjevita-humira-adalimumab-343187#10
3. Atzori, L., Mantovani, L., Pinna, A. L., & Usai, P. (2015). Cutaneous Adverse Reactions during Anti-Tnf Alpha Treatment for Inflammatory Bowel Diseases: The Experience of the Dermatology Clinic of Cagliari. Journal of Pharmacovigilance.
4. Kerbleski, J. F., & Gottlieb, A. B. (2009). Dermatological complications and safety of anti-TNF treatments. Gut, 58(8), 1033-1039.

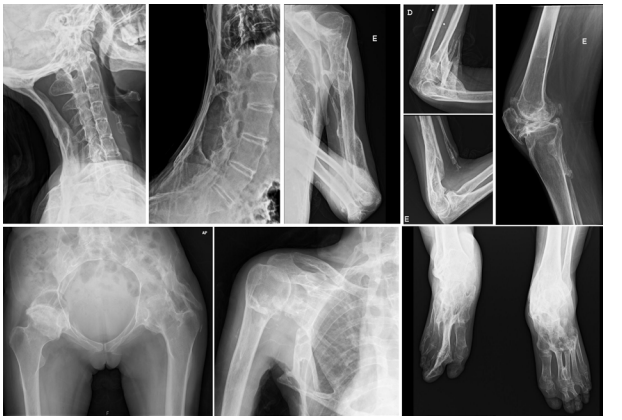
176 - "A MULHER DE PEDRA": UM CASO DE FIBRODISPLASIA OSSIFICANTE PROGRESSIVA

Ana Isabel Maduro[1], Beatriz Mendes[1], André Saraiva[1], Adriana Carones[1], Maria João Salvador[1, 2], Armando Malcata[1, 2], Sara Serra[1]

[1]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [2]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal

Introdução: A fibrodisplasia (ou miosite) ossificante progressiva (FOP) é uma doença genética muito rara (1:2000000), autossómica dominante, causada por uma mutação de ganho de função no gene ACVR1. Caracteriza-se por ossificação heterotópica progressiva dos músculos e outros tecidos moles como fáscias e tendões; e malformações esqueléticas, incluindo má formação do hálux bilateral (presente em 95% dos casos à nascença). A neoformação óssea extra-esquelética origina pontes rígidas, podendo levar a limitação da mobilidade (quando envolve tecidos moles próximo das articulações), complicações odontológicas (por anquilose da mandíbula) e insuficiência cardiorrespiratória (devido ao envolvimento da caixa torácica), principal causa de morte nestes doentes.
Caso Clínico: Doente do sexo feminino de 55 anos, com diagnóstico de FOP diagnosticada na infância, tendo perdido seguimento pouco tempo depois. Referenciada, recentemente, à consulta de Reumatologia. Trata-se de uma doente com um quadro progressivo de limitação da mobilidade articular e dependência para as atividades de vida diárias. Estas queixas terão começado ainda na infância, inicialmente com rigidez e limitação da mobilização da coluna cervical e lombar, seguindo-se as articulações dos ombros, cotovelos e coxofemorais. Atualmente, apresentava incapacidade marcada para a marcha, estando praticamente acamada e só fazendo levante para tarefas mínimas e sempre com ajuda de terceiros. Negava dor e tumefação articular. Negava queixas sistémicas. Sem antecedentes pessoais ou familiares relevantes. Era nulípara. Ao exame objetivo, destacava-se grande limitação funcional global, com ausência de mobilidade do esqueleto axial e redução muito significativa da mobilidade das articulações periféricas. A doente apresentava marcha em pequenos passos com postura fixa em bicos de pés à esquerda, movimentando-se em bloco, sem mobilidade nas cinturas escapular e pélvica; postura rígida com incapacidade de mobilizar a coluna em todos os seus segmentos; flexo dos cotovelos 90º e limitação da flexão dos punhos; coxofemoral esquerda em extensão mantida à direita e flexão à esquerda. Apresentava, ainda, microdactilia do hálux bilateralmente e atrofia muscular generalizada. Sem artrite ou deformação articular na avaliação individual. Radiograficamente, verificou-se a presença de ossificação heterotópica dos tecidos moles em diferentes localizações, formando pontes ósseas junto a várias articulações (imagens) e condicionando rigidez e limitação da mobilidade das articulações envolvidas (algumas já com anquilose). Durante estes anos apenas foi medicada com anti-inflamatório não esteróide (AINE) para as crises. Recentemente referenciada para consulta de Medicina Física e Reabilitação e consulta de Genética. A doente foi, ainda, aconselhada a evitar traumas e procedimentos médicos invasivos, já que podem exacerbar a progressão da FOP.
Conclusão: Apresentámos um caso de FOP com manifestações clínicas e radiográficas características e com um longo tempo de evolução, já com grande incapacidade funcional. O caso ilustra uma doença muito rara e extremamente debilitante, sem tratamento específico ou modificador da doença, cuja evolução é, muitas vezes, inevitável. De facto, a esperança média de vida nestes doentes é de aproximadamente 40 anos e, maioria deles, fica confinado

FIGURE. MIOSITE OSSIFICANTE



a uma cadeira de rodas no final da segunda década. O tratamento recomendado consiste na prevenção e alívio sintomático de surtos, medidas de suporte e reabilitação funcional e aconselhamento genético.

178 - POST-VACCINATION DIAGNOSIS OF SYSTEMIC LUPUS ERYTHEMATOSUS

Jorge Pestana Lopes[1], Raquel Roque[1], Maria José Santos[1, 2]

[1]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introduction: Systemic Lupus Erythematosus (SLE) is a complex chronic inflammatory immune mediated disease. The exact interaction between genetic and environmental factors that influence the loss of immunological tolerance against self-antigens remains mostly elusive. One of the consequences of SLE and of the immunosuppressive drugs used in the treatment of SLE is a heightened risk of infection with increased mortality. In this patient population vaccination plays an important role in the prevention of serious disease and several high-quality systematic reviews have proven the safety and efficacy of most vaccines in SLE patients.

Despite the undeniable importance of vaccination, vaccines act as a source of immune stimulation. There are some reports in the medical literature of various autoimmune diseases diagnosed following vaccination including SLE. Most of these cases are associated with hepatitis B and human papillomavirus vaccines, but on a populational level there is no proven increase in the risk of SLE following vaccination.

Herein, we report a case of SLE diagnosed after receiving three different vaccines.

Case Report: A 45-year-old Caucasian male patient with unremarkable medical history, not under any medication and no known familial history of autoimmune diseases.

In preparation for a trip to Zanzibar the patient was prescribed three different vaccines that were all administered in the same day: inactivated poliomyelitis (IPV) , typhoid fever (TCV) and hepatitis A (HAV).

One week later he developed fatigue and severe migratory arthritis, involving the hands, elbows, knees and feet. The initial laboratory tests ordered at a private hospital showed a normal blood count and renal function with an elevated ESR (120 mm/1st h). He was medicated with 40 mg prednisolone/day, tapering to 5 mg after 3 weeks, with a good response.

The patient was referred to our Rheumatology department nearly six months after the initial symptoms, due to complaints of inflammatory, migratory arthralgia. At our observation he presented with arthritis of the left elbow and left wrist. The laboratory evaluation revealed elevated ESR and CRP, low C3 and C4 levels, ANA titer of 1/2560, high anti-dsDNA antibody titer (234.0) with positive anti-RNP antibody. A diagnosis of SLE was made and medication was started with hydroxychloroquine 400 mg/day. Six months later due to persistent inflammatory arthralgia and the appearance of a photosensitive, diffusely erythematous rash the patient was started on methotrexate with a favorable response.

After 1 year of follow-up there are no other manifestations besides skin and articular involvement. The patient remains at low disease activity, currently being medicated with hydroxychloroquine 400 mg/day, methotrexate 20 mg/weekly and prednisolone 5 mg/day.

Discussion: We report a case of SLE diagnosed after simultaneous administration of three different vaccines, including a live attenuated vaccine.

While IPV has been implicated in flares of SLE in a retrospective study, none of the described vaccines is typically associated with the onset of SLE.

Nevertheless, to date, there is no proven causal association between vaccination and autoimmune diseases, and vaccines are warranted, effective and safe in SLE patients.

179 - MIOSITE DE CORPOS DE INCLUSÃO – A IMPORTÂNCIA DA SUSPEIÇÃO DIAGNÓSTICA

Marlene Sousa[1], Liliana Saraiva[1], Ana Isabel Maduro[1], André Saraiva[1], Tânia Santiago[1, 2]

[1]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [2]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal

Introdução: A miosite de corpos de inclusão (MCI), apesar de ser uma entidade rara, constitui a miopatia inflamatória mais frequente depois dos 50 anos. Caracteriza-se pela diminuição assimétrica da força muscular mais frequentemente nos quadricípites femorais e flexores dos dedos. (1) Pelo facto de ter uma evolução insidiosa (2) pode ser subdiagnosticada e

confundir-se com alterações próprias da idade. Este caso pretende alertar para a importância de considerar esta hipótese diagnóstica em doentes acima dos 50 anos com sintomas musculares.

Caso clínico: Um homem de 75 anos, com artrite reumatoide seropositiva em remissão, hipertensão arterial, hiperplasia benigna da próstata, gastrite crónica, e enxaqueca, medicado com perindopril, indapamida, amlodipina e propanolol.

Referia diminuição da força muscular de predomínio nos segmentos proximais e distais dos membros superiores (MS) e proximais dos membros inferiores (MI), de agravamento lentamente progressivo nos 3 anos anteriores que ele desvalorizou durante muito tempo. Na primeira observação por Reumatologia apresentava atrofia dos braços, antebraços, interósseos das mãos e coxas. Tinha diminuição da força muscular nos segmentos proximais (grau II/V) e distais (grau III/V) dos MS e proximais dos MI (grau III/V à esquerda e IV/V à direita). Realizou eletromiografia (EMG) que revelou sinais de lesão da fibra muscular dos músculos proximais dos MS e MI. Realizou biópsia do músculo deltoide com presença de fibras anguladas, grupos de fibras atróficas, necróticas e com presença de vacúolos sarcoplasmáticos. O estudo histo-enzimológico evidenciou aumento difuso da atividade em fibras atrofiadas com a nicotinamida adenina dinuclotideo-tetrazólio redutase (NADH-TRase) e succinato desidrogenase (SDHase) e ainda fibras com aumento sarcolémico e fibras citocromo C oxidase (COX) negativas em número superior ao esperado para a idade; o infiltrado linfocitário era constituído por linfócitos CD8+ e CD4+ e raros linfócitos B, CD20+ perivasculares. Estas alterações foram compatíveis com uma MCI. Analiticamente apresentava fator reumatoide (FR) e anti-CCP positivos (6730 IU/ml e 340 IU/ml respetivamente), com ANAs, ENAs e anticorpos específicos de miosites negativos. O hemograma e a bioquímica não tinham alterações de relevo, incluindo a creatinina quinase e aldolase dentro dos valores de referência.

O doente tem mantido seguimento em consultas de Reumatologia, Neurologia e programa de Fisioterapia. Dois anos após o diagnóstico mantém as alterações acima descritas no mesmo grau de défice. Tem dificuldade na marcha e alguma incapacidade na realização das atividades de vida diárias.

Conclusão: A MCI é subdiagnosticada porque os sintomas podem confundir-se com outras doenças neuromusculares e da idade. Sintomas típicos como as quedas frequentes, a dificuldade de preensão nas mãos e a disfagia podem passar despercebidos. A MCI carece de tratamento eficaz e o seu diagnóstico precoce permitirão a previsão das necessidades de fisioterapia, reabilitação da marcha, prevenção de quedas e melhoria das atividades de vida diária. Para além disso, evita o diagnóstico errado de outras miopatias inflamatórias e doenças neuromusculares que pode ser motivo de tratamentos inapropriados com corticoides em altas doses ou imunossupressores.

REFERÊNCIAS:

1. Elie Naddaf, Richard J. Barohn, Mazen M Dimachkie, Inclusion Body Myositis: Update on Pathogenesis and Treatment, Neurotherapeutics (2018) 15:995–1005
2. Mazen M. Dimachkie, Richard J. Barohn, Inclusion Body Myositis, Neurologic Clinics, Volume 32, Issue 3, August 2014, 32 (3): 629-646

182 - LÚPUS E PIODERMA GANGRENOSO INDUZIDOS PELO SECUKINUMAB

Maria Seabra Rato[1], Nuno Gomes[2], Sofia Magina[2, 3], Filipe Oliveira Pinheiro[1], Alexandra Bernardo[1], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Serviço de Dermatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [3]Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Introdução: O lúpus induzido (LI) por fármacos é uma entidade que partilha manifestações clínicas e imunológicas com o lúpus eritematoso sistémico (LES) idiopático e que está temporalmente associado à exposição a determinados fármacos. Ao contrário da relação já conhecida do LI com os antagonistas do fator de necrose tumoral, essa associação não é reconhecida para o secukinumab, um inibidor da interleucina 17A. O pioderma gangrenoso (PG) é uma dermatose neutrofílica caracterizada por ulceração necrotizante e dolorosa da pele. A sua etiologia e patogénese permanecem desconhecidas. Contudo, muitos autores acreditam numa possível etiologia imune devido à associação, em 30 a 70% dos casos, do PG com doenças auto-imunes, embora a associação com LES seja rara. Existem casos de PG induzido por fármacos, particularmente em relação com infliximab, adalimumab e rituximab.

Caso clínico: Doente do sexo feminino, de 67 anos, com antecedentes de psoríase e de artrite psoriática com 18 anos de evolução, medicada com adalimu-

mab 40mg subcutâneo quinzenal desde há 4 anos, em remissão clínica. Por iniciativa própria suspendeu o adalimumab, com reaparecimento da psoríase e da artrite 3 meses após a interrupção do mesmo. Por esse motivo, reiniciou o tratamento biotecnológico, com resolução da artrite, mas com escassa resposta do ponto de vista cutâneo (PASI > 10), razão pela qual fez switch terapêutico para secukinumab 300mg subcutâneo. Seis semanas após o início do secukinumab recorreu à consulta por febre, odinofagia, poliartralgias e lesões ulceradas da pele com duas semanas de evolução. Ao exame objetivo, apresentava lesões aftosas na cavidade oral, nódulos ulcerados com drenagem purulenta na perna esquerda, mão esquerda e dorso e poliartrite. Analiticamente apresentava elevação da velocidade de sedimentação (54mm/h) e da proteína C reativa (124mg/L), anticorpos antinucleares positivos (1/1000 padrão homogéneo) e anti-ds-DNA elevado (483UI/mL). Apresentava também consumo de complemento (C4). Os anticorpos anti-histonas, anti-SSA, anti-SSB, anti-SM, anti-RNP e fator reumatóide foram negativos. Foi realizada biópsia do bordo de uma das úlceras que evidenciou na derme profunda e no tecido subcutâneo um infiltrado inflamatório com polimorfonucleares neutrófilos, sinais de cariorréxis e constituição de abcesso compatíveis com PG. Foi feito o diagnóstico de PG e de LI por secukinumab. A doente suspendeu o secukinumab e iniciou tratamento com prednisolona 0.5mg/kg/dia em esquema de desmame com melhoria clínica. Ao fim de 10 semanas de tratamento apresentou resolução das lesões de pioderma, mas com agravamento da psoríase pelo que foi proposta para início de brodalumab.

Conclusão: Na literatura são raros os casos descritos de pioderma gangrenoso e de lúpus induzidos pelo secukinumab. A associação das duas entidades no mesmo doente, induzida por este fármaco, não se encontra previamente descrita. Este caso ilustra a importância da necessidade de compreensão dos mecanismos fisiopatológicos destas reações adversas.

183 - HIPEROSTOSE ESQUELÉTICA IDIOPÁTICA DIFUSA - UMA CAUSA RARA DE DISFAGIA

Maria Seabra Rato[1], Raquel Miriam Ferreira[1], Filipe Oliveira Pinheiro[1], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal

Introdução: A hiperostose esquelética idiopática difusa (DISH) é uma doença não inflamatória, de etiologia desconhecida, caracterizada por calcificação e ossificação dos ligamentos vertebrais. Afeta mais frequentemente os segmentos torácicos e lombares, sendo o envolvimento da coluna cervical mais raro. Embora na maioria das vezes a DISH seja assintomática e encontrada acidentalmente em exames de imagem, pode ser uma causa de disfagia e de comprometimento da via aérea.

Caso clínico: Doente do sexo masculino, de 81 anos, é enviado à consulta de Reumatologia por cervicalgia de ritmo mecânico com mais de 10 anos de evolução. Referia também disfonia e disfagia progressiva para líquidos e sólidos, nos últimos 6 meses, principalmente aquando da hiperextensão cervical, nomeadamente com o movimento exigido pela deglutição de comprimidos, respeitantes à sua medicação habitual. Negava odinofagia e dispneia. Sem perda ponderal significativa. Negava hábitos alcoólicos ou tabágicos. Apresentava como antecedentes pessoais apneia obstrutiva do sono e excesso ponderal. Ao exame objetivo observava-se limitação da amplitude cervical em todos os planos mas principalmente na extensão. Não eram evidentes massas ou adenopatias cervicais palpáveis. Foi solicitada a realização de endoscopia digestiva alta que não revelou alterações. A avaliação por Otorrinolaringologia mostrou um abaulamento da parede posterior da faringe que dificultava a completa avaliação das estruturas faringo-laríngeas. No entanto, não foram observadas aparentes alterações estruturais ou diminuição da sensibilidade laríngea. A radiografia e posteriormente a tomografia computorizada cervical evidenciaram ossificação do ligamento longitudinal anterior desde a 2ª à 7ª vértebra cervical (C2 a C7) com preservação dos espaços intervertebrais e osteofitose exuberante, sobretudo em C2-C3 e C4-C5 que causavam compressão sobre as estruturas áreas e digestivas cervicais com desvio das cartilagens laríngeas para a esquerda. A ressonância magnética do pescoço não evidenciou massas, áreas de anormal captação na faringe ou na cavidade oral ou alteração de sinal medular. Estes achados foram compatíveis com o diagnóstico de DISH como causa da disfagia e disfonia. O doente foi medicado com nimesulida orodispersível, iniciou reabilitação da deglutição e foi encaminhado para consulta de Ortopedia, tendo sido proposto para cirurgia de excisão de osteófitos na ausência de melhoria com o tratamento médico instituído.

Conclusão: Apesar da DISH cervical ser na maioria

dos casos uma condição assintomática, em situações excecionais de ossificações exuberantes e avançadas pode ocasionar complicações graves, como compressão da faringe, estenose esofágica e distúrbios neurológicos. Assim, embora seja uma causa rara de disfagia a DISH deve ser considerada no diagnóstico diferencial principalmente em doentes idosos, do sexo masculino e com alguns fatores de risco como a obesidade. Desta forma, o seu reconhecimento é de extrema importância, podendo ser facilmente diagnosticada com exames de imagem. O tratamento passa por medidas conservadoras, estando a cirurgia reservada para casos de doença mais grave.

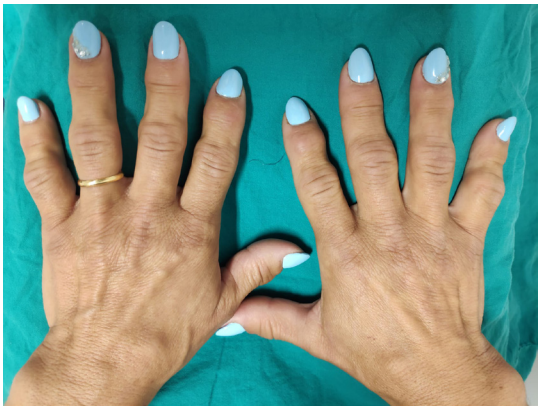
187 - ARTROPATIA ACROMEGÁLICA: A PROPÓSITO DE UM CASO CLÍNICO

Ana Filipa Gonçalves[1], Xénia Verraest[2], Flávio Campos Costa[3], Romana Vieira[3], Joana Aleixo[3], Taciana Videira[3], P Pinto[3]

[1]Medicina Física e de Reabilitação, Centro de Reabilitação do Norte - CHVNG/E, Vila Nova de Gaia, Portugal, [2]Medicina Física e de Reabilitação, Centro de Medicina de Reabilitação da Região Centro – Rovisco Pais, Coimbra, Portugal, [3]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Introdução: A Acromegalia é uma doença rara caracterizada pela produção excessiva de hormona do crescimento (GH) e do seu principal mediador, o fator de crescimento semelhante à insulina tipo 1 (IFG-1). Ambas são essenciais para o normal crescimento, diferenciação e reparação do osso e cartilagem, mas, quando em excesso, podem provocar uma artropatia semelhante à osteoartrose primária (OA), mas com características distintas como hipertrofia da cartilagem articular, aumento do espaço articular, hipertrofia sinovial e dos tecidos moles peri-articulares. A prevalência desta artropatia varia entre 30-70%. A sua fisiopatologia não é totalmente conhecida, mas pressupõe-se existirem duas fases: numa 1ª fase o excesso de GH e IGF-1 provoca hipertrofia da cartilagem e aumento do espaço interarticular; na 2ª fase, quando ocorre dano na cartilagem, mecanismos anormais de reparação levam à produção excessiva de fibrocartilagem, podendo ocorrer calcificação e formação de osteófitos. Nesta fase, a hipertrofia da cartilagem pode persistir ou progredir com perda da mesma e diminuição da interlinha articular. A artropatia acromegálica manifesta-se predominantemente por artralgias de ritmo mecânico. As alterações radiográficas podem não corresponder à clínica apresentada pelo doente e variar consoante as fases da artropatia.
Caso clínico: Mulher de 55 anos com antecedentes de HTA e síndrome do túnel cárpico que foi resolvido, foi encaminhada por alterações articulares degenerativas graves no raio x das mãos. A doente refere notar aumento da espessura dos dedos das mãos e alargamento dos pés nos últimos 10 anos, com necessidade de aumentar o tamanho da aliança e o número do calçado. Nega artralgias, alterações menstruais, galactorreia, cefaleias e alterações da visão. Sem história de polipose intestinal e antecedentes de insuficiência cardíaca. No entanto, refere alterações semelhantes no pai. Ao exame objetivo observa-se aumento da dimensão dos dedos das mãos e dos pés, fácies retangular e alargamento da base do nariz. Sem artrite periférica nem outros sinais de relevo. Analiticamente apresenta apenas anticorpos antinucleares positivos com padrão mosqueado fino denso (1/160) e aumento da velocidade de sedimentação (47mm/h). No raio x das mãos observam-se alterações típicas como tufting das falanges distais, alargamento da base das falanges e ligeiro aumento da interlinha articular ao nível da 1ª carpometacarpiana da mão direita, assim como alterações degenerativas. Por suspeita de Acromegalia foi encaminhada para a consulta de Endocrinologia, onde se confirmou o diagnóstico, apresentando uma GH de 110,0 ng/ml e IGF-1 758 ng/ml. Aguarda RMN cerebral para iniciar tratamento adequado.
Conclusão: Neste caso clínico foram as alterações fenotípicas que levaram à suspeita de Acromegalia e não os sintomas de artropatia ou alterações características da mesma nos raio x. No entanto, os sin-

FIGURE: MÃOS



tomas associados a esta artropatia e o espessamento da cartilagem articular podem melhorar com o tratamento e controlo da hipersecreção de GH, quando na primeira fase da doença. Contudo, muitos doentes mantêm a sintomatologia, pois na segunda fase, após ocorrer dano da cartilagem, as alterações articulares decorrentes tornam-se irreversíveis e a OA tende a progredir apesar do controlo adequado da produção hormonal, sendo que o tratamento nesta fase segue as orientações da OA na população geral. Como se trata de uma condição que tem impacto na qualidade de vida dos doentes, é importante a avaliação músculo-esquelética para diagnóstico e tratamento multidisciplinar apropriado.

192 - ESCLEROSE SISTÉMICA E ESPONDILARTRITE AXIAL RADIOGRÁFICA: UMA ASSOCIAÇÃO RARA

Agna Neto[1, 2], Margarida Faria[1], Ricardo Figueira[1], Mário Rodrigues[1]

[1]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [2]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

Introdução: A co-existência de esclerose sistémica (ES) e Espondilartrite axial radiográfica (r-axSpA) é excepcionalmente rara. Desconhecem-se mecanismos etiopatogénicos comuns claros. Os escassos casos descritos na literatura sugerem uma possível causa genética com presença de alelos de susceptibilidade para ambas as entidades; ou uma associação entre ocorrência de ES e a terapêutica prévia com anti-TNF-α. Descreve-se um caso com diagnósticos simultâneos de ES e r-axSpA.

Caso Clínico: Doente do sexo masculino, de 50 anos, fumador (20 UMA), com antecedentes de dislipidémia, recorreu à consulta de Reumatologia por cervicodorsolombalgia de ritmo inflamatório, associada a rigidez matinal de 60 minutos, com 6 anos de evolução. Referia alívio apenas parcial das queixas álgicas com anti-inflamatórios não esteróides (AINEs). Sem história de artralgias periféricas, dactilite, entesite, psoríase, uveíte ou alterações gastrointestinais. Adicionalmente, referia extremidades frias (mãos e pés) desde há 2 anos, sem evidente Fenómeno de Raynaud, perda de peso ou outros sintomas constitucionais. Ao exame objetivo, salientava-se teste de FABER positivo bilateralmente, franca limitação da mobilidade da coluna dorsolombar e, particularmente, da coluna cervical. Destacava-se ainda esclerodactilia, puffy hands e pitting scars nas polpas digitais, sem úlceras ativas e sem artrite periférica. Da avaliação laboratorial, refira-se hemograma, função renal e hepática, parâmetros inflamatórios e urina II normais; HLA-B27 positivo, anticorpos antinucleares positivos (1/640, padrão AC-29) e anti-Scl-70 positivo. Os estudos de radiologia convencional revelaram sacroiliíte de grau IV bilateral, anquilose total da coluna e reabsorção das falanges distais do 2.º dedo da mão esquerda e do 2.º e 3.º dedos da mão direita. A videocapilaroscopia evidenciou um padrão esclerodérmico precoce. Assumiram-se os diagnósticos de Esclerose Sistémica limitada e Espondilartrite axial radiográfica. À presente data, aguarda realização de ecocardiograma, tomografia computorizada de tórax e provas de função respiratórias. Iniciou pentoxifilina 400mg/2x dia, manteve acemetacina 90mg/dia e atualmente, por manter doença axial ativa (BASDAI 5.4, ASDAS-PCR 2.1) sob AINE, está a aguardar início de DMARD.

Discussão/Conclusão: Este caso retrata a presença incomum de ES e r-axSpA no mesmo doente, cumprindo inclusive critérios de classificação para ambas as entidades (critérios ASAS para axSpA e critérios ACR/EULAR para ES). Constitui ainda um desafio terapêutico: após falência dos tratamentos convencionais, os anti-TNF-α são habitualmente a primeira opção terapêutica na r-axSpA, embora o seu papel na ES seja controverso, existindo descrições na literatura que sugerem que possam conduzir a uma progressão da fibrose. Os inibidores de JAK, pelo seu potencial antifibrótico, podem ser uma alternativa promissora na abordagem a ambas as condições.

194 - ENVOLVIMENTO DO SISTEMA NERVOSO PERIFÉRICO DE APRESENTAÇÃO POUCO COMUM: A PROPÓSITO DE UM CASO CLÍNICO

Ana Martins[1], Daniela Santos Oliveira[1], Filipe Oliveira Pinheiro[1], Maria Seabra Rato[1], Diogo Guimarães da Fonseca[2], Sofia Pimenta[1, 3], Pedro Madureira[1], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [3]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Introdução: As principais manifestações clínicas

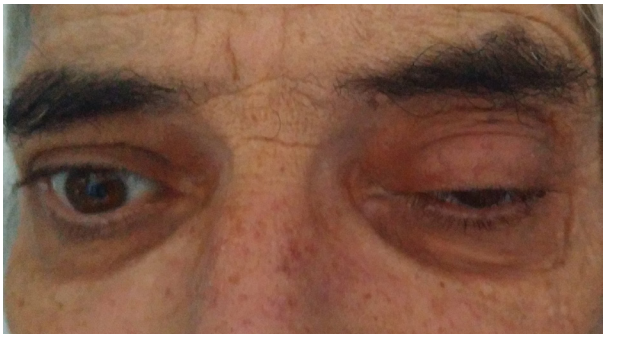
das vasculites sistémicas são as mioartralgias, a febre, a perda ponderal, a anorexia, o envolvimento renal, cutâneo e neurológico. Relativamente às manifestações neurológicas, o envolvimento do sistema nervoso periférico (SNP) é frequente, ocorrendo em até 70% dos pacientes. Já o envolvimento do sistema nervoso central é mais raro, ocorrendo em menos de 15% dos pacientes e o envolvimento dos nervos cranianos é, ainda, mais raro, estimando-se que a sua prevalência seja inferior a 5%.

Caso clínico: Homem de 61 anos, com hipertensão arterial (HTA), foi internado por hipoestesia a nível da face plantar dos pés, sintomas constitucionais (perda ponderal de 13 kg e anorexia) e elevação marcada dos parâmetros inflamatórios. Realizou eletromiografia que documentou uma polineuropatia axonal sensitivo-motora simétrica grave. Para despiste de etiologia paraneoplásica, realizou TC toraco-abdomino-pélvico, EDA, colonoscopia, PET, doseamento de PSA livre e total, lavado bronco-alveolar, citologia urinária e imunofixação sérica que excluiram neoplasia. Do estudo infecioso, a destacar infeção pelo VHB e VHC no passado e reatividade para TPPA/TPHA e VDRL. Realizou punção lombar e o VDRL e FTA/Abs no líquido cefalorraquidiano foram negativos, pelo que se excluiu neurossífilis. Cumpriu 3 tomas semanais de penicilina G benzatina para tratamento de sífilis latente. Foi excluído infeção por micobactérias, CMV, HSV tipo 1 e 2, EBV, parvovírus b12 e VIH. O estudo imunológico foi negativo (ANA, ANCA, crioglobulinas, anti-DNAds, anti-ENA, AAF, anti-neuronais) e os níveis de complemento foram normais. Os níveis da ECA e ceruloplasmina foram, também, normais.

Durante o internamento, apresentou diplopia, ptose palpebral e alterações da oculomotricidade de forma súbita à esquerda (figura 1). Realizou RMN cerebral e do neuro-eixo que não revelaram alterações. Realizou angioTC dos troncos supra-aórticos e intracraniano que documentou lesão aneurismática da artéria comunicante posterior esquerda. Posteriormente, realizou angio-RMN que revelou espessamento do nervo oculomotor à esquerda, aspeto sugestivo de neuropatia do III par craniano, não se verificando compressão do nervo oculomotor pela lesão aneurismática.

Tendo em conta os achados clínicos, analíticos e imagiológicos, foi colocada a suspeita diagnóstica de Poliarterite Nodosa (PAN). Realizou pulsos de metilprednisolona 1g durante 3 dias e, posteriormente, prednisolona 0.5mg/kg/dia. Objetivou-se melhoria

FIGURA 1. ENVOLVIMENTO DO III PAR CRANIANO ESQUERDO COM PTOSE ASSOCIADA.



dos parâmetros inflamatórios, resolução da diplopia, melhoria da oculomotricidade e da hipostesia. A biópsia de nervo sural documentou, posteriormente, uma neuropatia axonal grave e espessamento hialino de alguns vasos do endonervo, o que pode ser enquadrado num processo vasculítico em fase tardia. Manteve-se sob corticoterapia em desmame e iniciou azatioprina com ótima evolução clínica, tendo ocorrido reversão da neuropatia do nervo oculomotor e regressão parcial da hipostesia dos pés.

Conclusão: A neuropatia periférica tem um diagnóstico diferencial abrangente, onde as vasculites sistémicas se incluem. Embora neste caso clínico não se verificassem algumas das manifestações mais frequentes, a presença de perda ponderal, síndrome biológico inflamatório, polineuropatia periférica, HTA, aneurisma cerebral, infeção por VHB no passado, histologia compatível e ótima resposta à terapêutica favorece o diagnóstico de PAN. Um reconhecimento precoce da etiologia da neuropatia é fundamental, uma vez que permite o tratamento adequado e melhora o prognóstico do doente.

195 - PHALANGEAL MICROGEODIC SYNDROME – A RARE VASCULAR ACROSYNDROME IN CHILDHOOD

Maria Seabra Rato[1], Bernardo Santos[2], Filipe Oliveira Pinheiro[1], Mariana Rodrigues[3], Francisca Aguiar[3], Iva Brito[3, 4]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar do Baixo Vouga, E.P.E., Aveiro, Portugal, [3]Unidade de Reumatologia Pediátrica e do Jovem Adulto, Centro Hospitalar e Universitário de São João, Porto, Portugal, [4]Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Introduction: Phalangeal microgeodic syndrome (PMS) is a rare disease of unknown etiology, which affects fingers and/or toes of children.
Clinical case: The authors describe the case of a 9-year-old girl, who presented to the Pediatric Rheumatology outpatient clinic in November 2020 with painful swelling and cyanosis of all fingers for 3 weeks which had not improved with oral ibuprofen. She had no history of previous COVID-19 and, besides mild IgA vasculitis in 2015, was otherwise healthy. On physical examination, painful spindle-shaped swelling with functional limitation and cyanosis of fingers, associated with chilblain-like lesions were observed. She was started on pentoxifylline 400 + 200 mg/day and, later, nifedipine 0.5 mg/kg/day, and advised to avoid exposure to cold. Five months later she had marked improvement of pain and swelling, maintaining mild cyanosis. Laboratory workup revealed normal complete blood count, normal erythrocyte sedimentation and C-reactive protein, normal biochemical liver and renal parameters. Immunology tests showed normal IgG, IgA and IgM, positive antinuclear antibodies 1/100 and negative antidsDNA, ANCA, ACPA, rheumatoid factor, anti-ENA, anticentromere and antiphospholipd antibodies. Hand radiographs showed multiple small, round radiolucent images at the edge of the metaphysis of several phalanges. Hand MRI showed bone marrow edema in middle and distal phalanges of all fingers, and distal part of proximal phalanges (more diffuse and prominent in the second and fifth, bilaterally); there were no relevant changes in joints or tendons. These findings are compatible with PMS.
Conclusion: This is a rare disease that should be included in the differential diagnosis of patients presenting with vascular acrosyndromes complaints. It can be misinterpreted as an infectious or post-infectious (namely COVID-19-related), inflammatory, or even malignant condition. It usually has a benign course and resolves with conservative treatment. Pediatric rheumatologists should be aware of this entity, as a timely and precise diagnosis prevents further investigations and complications.

196 - NEFROPATIA DE IGA E SARCOIDOSE – QUADRO RENAL ATÍPICO NO JOVEM ADULTO

Frederico Rajão Martins[1], Joana Morais Romano[2], João Frazão[3], Iva Brito[4, 5]
[1]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal, [2]Serviço de Medicina Física e de Reabilitação, Unidade Local de Saúde de Matosinhos, Hospital de Pedro Hispano, Matosinhos, Portugal, [3]Serviço de Nefrologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [4]Unidade de Reumatologia Pediátrica e do Jovem Adulto, Centro Hospitalar e Universitário de São João, Porto, Portugal, [5]Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Introdução: A sarcoidose é uma doença inflamatória sistémica rara, com incidência estimada de 1,3 a 3,7/100 000 na Europa. Caracteriza-se pela presença de granulomas não caseosos em qualquer órgão ou sistema, sendo o envolvimento pulmonar o mais descrito. O diagnóstico assenta sobretudo em aspetos radiográficos sugestivos e evidência histopatológica de granulomas epitelioides não caseosos, após exclusão de outras doenças granulomatosas. O envolvimento renal é pouco frequente, sendo a nefrite intersticial granulomatosa a entidade nosológica mais comum. No entanto, glomerulonefrite secundária associada à sarcoidose, nomeadamente nefropatia de IgA e glomerulonefrite membranosa, pode também ocorrer.
Caso clínico: Jovem de 17 anos do sexo masculino, referenciado a consulta de Reumatologia Pediátrica por quadro de oligoartrite periférica simétrica de instalação subaguda, com envolvimento de joelhos e tornozelos, tendo sido previamente medicado com naproxeno 500 mg bid, com alívio parcial do quadro álgico. Não apresentava antecedentes pessoais ou familiares de psoríase, aftose oral ou genital recorrente, história de trauma ou infeção recentes. Do estudo complementar efetuado, salientava-se elevação ligeira de parâmetros inflamatórios (VS 16 mm/h e PCR 5,6 mg/L), elevação marcada do valor sérico da enzima de conversão de angiotensina (307 U/L) e subida progressiva do nível de cálcio ionizado sérico (valor máximo 2.68 mEq/L). O estudo imunológico era negativo - fator reumatoide, anti dsDNA, anticorpos anti-citoplasma do neutrófilo, alelo HLA B27 -, C3 e C4, observando-se ligeira hipergamaglobulinemia policlonal. Foi efetuado estudo serológico viral e ecocardiograma transtorácico com exclusão de quadro infecioso. Realizou radiografia de tórax que não revelou achados de relevo, documentando-se posteriormente em tomografia computadorizada a presença de pequenas adenopatias mediastínicas, com ligeira densificação intersticial subpleural a nível apical, achados compatíveis com sarcoidose pulmonar.

Por leucoeritrocitúria sustentada e proteinúria

subnefrótica (230 mg/L), com creatinina máxima de 1 mg/dL, realizou ecografia renal que não revelava alterações, e após observação por Nefrologia é orientado para realização de biópsia renal que revelou glomérulos com discreto e segmentar aumento de matriz mesangial e discretas lesões de dano tubular agudo, com positividade a IgA e C3c na imunofluorescência, tendo-se documentado em exame ultra-estrutural muito raros depósitos no mesângio e transição mesangiocapilar, achados enquadráveis em glomerulonefrite mesangioproliferativa focal (provável nefropatia por IgA).

Estabeleceu-se então o diagnóstico de sarcoidose com envolvimento pulmonar (estadio II), articular e glomerulonefrite secundária associada.

Conclusão: Alterações no sedimento urinário, em doente com sarcoidose, devem sempre obrigar a um despiste de glomerulonefrite secundária, mesmo considerando este atingimento raro. O envolvimento articular sarcoidótico, embora pouco comum, deve ser também considerado em doente com artrite inclassificável.

199 - WHIPPLE'S DISEASE: AN ATYPICAL PRESENTATION MIMICKING RHEUMATOID ARTHRITIS

Frederico Rajão Martins[1], Raquel Miriam Ferreira[2], Ana Martins[2], Daniela Santos Oliveira[2, 3], Maria Seabra Rato[2], Filipe Oliveira Pinheiro[2], Diogo Guimarães da Fonseca[4], Lúcia Costa[2]

[1]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [3]Center for Health Technology and Services Research (CINTESIS),, Faculdade de Medicina da Universidade do Porto, Porto, Portugal, Porto, Portugal, [4]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Introduction: Whipple's disease is a rare chronic multi-systemic entity caused by the gram positive actinobacteria Tropheryma whipplei, with a yearly estimated incidence of 1 to 6 cases per 10 000 000 persons. Musculoskeletal manifestations range from relapsing oligo or polyarthritis affecting middle and large joints, mostly knees, wrists and tibiotarsal joints, and to a lesser extent the small joints of the hands. Other frequent symptoms are malabsorption syndrome and central nervous system manifestations, with a classical triad of dementia, supranuclear ophthalmoplegia and myoclonus being highly specific for this condition. However, subtle neurological symptoms such as progressive cognitive impairment and psychiatric manifestations may also be seen. In a clinical suspicion setting, the gold standard for diagnosis is the demonstration of the organism in the involved tissue (usually the small bowel) by at least two methods – periodic acid-Schiff stain, polymerase chain reaction (PCR) or immunohistochemical stain - or a positive PCR testing of two involved sites. Lumbar puncture should always be executed as PCR detection of T.whipplei's DNA in cerebrospinal fluid is possible in up to 50% of cases.

Clinical case: A seventy-three-year-old man with a previous diagnosis of seronegative polyarthritis recurred to a Rheumatology consultation with a 2-year clinical picture of persistent polyarthritis of the wrists, metacarpophalangeal and tibiotarsal joints, with mild improvement under prednisolone 5mg id and methotrexate 10mg weekly. With increasing immunosuppression, the articular component improved but an insidious worsening of a previously described steatorrhea, new onset weight loss, gait ataxia and major depressive disorder was noted, leading to endoscopic studies that were inconclusive. During follow-up, blood analysis revealed sustained leukocytosis, elevated erythrocyte sedimentation rate (80 mm/hr) and C-reactive protein (74 mg/L) with negative results for rheumatoid factor, anti-nuclear, anti-citrulinated peptide and anti-tranglutaminase antibodies. Diverse infectious aetiologies were excluded with negative bacteriologic, virologic, parasitologic and mycobacteriologic faecal exams. Neoplasic entities were also excluded with thoraco-abdominal-pelvic CT, and vasculitides with positron emission tomography. New upper digestive endoscopy showed no macroscopic changes and histological findings weren't supportive of the over-cited hypothesis, although the duodenal PCR was strongly positive for T.Whipplei DNA. The patient was admitted in the Rheumatology department for repetition of endoscopic study and lumbar puncture execution, with a new positive PCR for T.Whipplei in the duodenal tissue, as well as in cerebrospinal fluid and faeces.

Whipple's disease with articular and neurologic involvement was diagnosed, methotrexate was suspended and the patient treated with ceftriaxone 2 gr iv id in a total of 4 weeks, followed by sulfamethoxazole trimethoprim 800 / 120 mg bid. Significant clinical and laboratorial improvement was observed, with

complete resolution of arthritis, diarrhoea, increased weight and better motor coordination by 1 month of treatment.

Conclusion: Strong clinical suspicion is crucial for the diagnosis of Whipple's disease, as the diagnosis of this entity may prove to be very challenging. Small bowel mucosa biopsy is warranted, even in the absence of macroscopic changes, and lumbar puncture must be executed even in the absence of neurological symptoms.

203 - GIANT CELL ARTERITIS: A POSSIBLE PRESENTATION OF A PARANEOPLASTIC SYNDROME

Nuno Pina Gonçalves[1, 2, 3], Maria Manuela Costa[2], Sandra Falcao[2], Jaime C. Branco[2, 3]

[1]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [2]Serviço de Reumatologia, Hospital de Egas Moniz, Centro Hospitalar de Lisboa Ocidental, Lisboa, Portugal, [3]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

Introduction: Giant cell arteritis (GCA) is the most common idiopathic vasculitis and almost always occurs after age 50 years, with a peak incidence in patients older than 70 years old. There is no established cause, but senesce, sex and genetic factors have all been shown to contribute to the occurrence of this disease. However, concurrent malignancy is not rarely present and may be the precipitating factor for the presentation of GCA.

Clinical Case: Male, 74 years old, with medical history of hypertension, presents to the emergency room with a clinical onset of sudden vision loss on the right eye (amaurosis fugax) and loss visual acuity on the left eye, along with a pulsatile type occipital headache and jaw claudication. Marked pain and morning stiffness of the shoulders and hip girdle were also present for over a month. Optic evaluation show marked oedema of the optic nerve on the right and intraretinal oedema and anterior ischemic optic neuropathy was assumed in the context of giant cell arteritis. A collaboration to the rheumatology service was solicited. Physical examination showed a marked reduction of the shoulder and pelvic girdle and a diagnosis of polymyalgia rheumatica was made. Laboratory work-up was unremarkable with no elevation of inflammation markers. Ultrasound of the temporal arteries was performed, showing an anechoic halo in the common trunk, frontal and parietal branches if the temporal artery, compatible with arteritis, on both sides, supporting the diagnosis of GCA. Physical examination showed a marked reduction of the shoulder and pelvic girdle and a concomitant diagnosis of polymyalgia rheumatica was made. Treatment was started with salicylic acid 150mg/day, 3 consecutive pulses of 1000mg of metilprednisolone, followed by prednisolone 60mg/day with a progressive tapering. A diagnosis of diabetes mellitus secondary to corticoid therapy was made, which prompted the introduction of methotrexate 15mg/week as a corticoid sparing agent. At follow-up, although there was a complete resolution of the articular symptoms, loss of vision on the left eye was permanent and there were still complaints of occipital headache. A Computed tomography body scan was performed which documented a heterogeneous and enlarged prostate gland with marked irregular contour coexisting an abnormal densification of the surrounding fat tissue, suggesting a proliferative process. Prostate-specific antigen was within normal range. A prostate biopsy was scheduled and tissue pathology report showed a small cells neuroendocrine carcinoma of the prostate. The patient was referred to the oncology department and a staging positron emission tomography scan revealed an extensive malignant neoplasm of the prostate, extended to the surrounding soft tissues, with metastatic extension to the pelvic and mediastinal ganglions, liver, osteomedullary tissues and pulmonary parenchyma. Chemotherapy was started with cisplatin/etoposide, but rapid decline in health status occurred and after a month and a half the patient died.

Discussion: Malignant neoplasms can present as paraneoplastic syndromes that may be the precipitating factor for GCA. Although the association between ACG and cancer is not fully understood, several clinical cases have been reported. Atypical ACG presentations may signal a concurrent neoplasm, but a high suspicion index should always be present.

205 - MIOPATIA POR SARCOIDOSE: O DESAFIO DIAGNÓSTICO

Ana Isabel Maduro[1], André Saraiva[1], Marlene Sousa[1], Liliana Saraiva[1], Armando Malcata[1, 2], Sara Serra[1]

[1]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [2]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal

Introdução: A sarcoidose é uma doença granulomatosa inflamatória multissistémica de etiologia desco-

nhecida que pode envolver praticamente todos os sistemas de órgãos. A apresentação neurológica ocorre apenas em cerca de 5% dos doentes. O envolvimento muscular sintomático é raro e a apresentação como miopatia é ainda mais incomum.

Caso Clínico: Doente do sexo masculino de 53 anos com antecedentes de sarcoidose pulmonar e ganglionar, diagnosticada em 2007, no contexto de alterações analíticas (cálcio e enzima de conversão da angiotensina francamente aumentados) e TC tórax com adenomegalias mediastínicas. O diagnóstico foi posteriormente confirmado por histologia do parênquima pulmonar, encontrando-se desde então sob corticoterapia crónica em doses médias a altas. Recentemente, apresentava-se em remissão clínica, a cumprir tratamento com metilprednisolona 8 mg, em dias alternados. Referenciado à consulta de Reumatologia por quadro de défice de força muscular do membro inferior direito com cerca de 6 meses de evolução e agravamento progressivo. Referia, ainda, mialgias na região proximal de ambos os membros inferiores e limitação funcional para subir escadas e levantar-se de uma cadeira sem apoio. Ao exame físico objetivou-se uma diminuição da força muscular proximal dos membros inferiores (grau 4/5 na flexão da coxa direita e grau 4+/5 na flexão da coxa esquerda), com atrofia dos músculos quadríceps, bilateralmente. Apresentava também, estigmas de síndrome de cushing secundário, nomeadamente obesidade central e fácies pletórica, tradutores de utilização prolongada de glucocorticóides (GCT). Analiticamente, verificou-se um aumento da CK (914 U/L), da aldolase (11.1U/L) e da mioglobina (183 ng/mL). Sem outras alterações relevantes. O EMG revelou sinais miopáticos, com características agudas de atividade. Neste contexto, realizou biópsia muscular que mostrou infiltrado de células mononucleares com formação de granulomas não caseosos, compatível com miopatia por sarcoidose (imagem). Feito o diagnóstico e, após ponderação de risco-benefício, optou-se por iniciar prednisolona 1mg/kg/dia. O início da corticoterapia em doses terapêuticas tinha sido protelado dado não se poder descartar a hipótese de miopatia associada aos GCT, algo que só foi possível após obtenção do resultado da biópsia. Devido ao risco de efeitos secundários associados ao CGT (num doente já com estigmas de utilização excessiva de corticoterapia), foi considerada a associação de um DMARD, tendo-se optado pela azatioprina (em detrimento do metotrexato), pelo facto do doente apresentar antecedentes de alcoolismo e manter consumos excessivos. Na consulta de reavaliação (6 semanas após início da prednisolona), o doente apresentava já normalização das alterações analíticas e melhoria da força muscular.

Conclusão: Este caso ilustra a dificuldade diagnóstica que pode surgir quando um quadro de fraqueza muscular ocorre num doente com sarcoidose a cumprir corticoterapia crónica e em doses apreciáveis. De facto, a doença muscular sintomática na sarcoidose é rara e é importante considerar outras causas de miopatia, mesmo num paciente com sarcoidose estabelecida, nomeadamente a hipótese de miopatia induzida por GCT. Neste sentido, a biópsia poderá desempenhar um importante papel no diagnóstico diferencial. Este caso demonstra, ainda, a dificuldade na gestão terapêutica quando a corticoterapia pode ser a solução, mas também um problema. A introdução de um DMARD como poupador de GCT, poderá permitir minimizar a ocorrência de outros possíveis efeitos adversos.

206 - SÍNDROME ANTICOAGULANTE LÚPICO-HIPOPROTROMBINEMIA: UM CASO CLÍNICO DESAFIANTE

Laura Gago[1], Rita Pinheiro Torres [1, 2], Maria Helena Lourenço[1, 2], Ana Bento da Silva[1, 2], Ana Lúcia Fernandes[3], Nuno Pina Gonçalves[1, 2, 4], Maria João Gonçalves[1], Ana Filipa Mourão[1, 2], Maria Manuela Costa[1], Jaime C. Branco[1, 2]

[1]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [2]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [3]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, Faro, Portugal, [4]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal

Introdução: O Síndrome Anticoagulante Lúpico-Hipoprotrombinemia está associado à presença de anticorpos contra a protrombina (fator II). Trata-se de um distúrbio raro estando clinicamente associado a infeções virais e a Lúpus Eritematoso Sistémico (LES). Deve suspeitar-se deste síndrome na presença de aumento dos tempos de coagulação (tempo de protrombina (TP) e tempo de tromboplastina parcial ativada (aPTT)) em associação com a presença de anticoagulante lúpico.

Caso Clínico: Mulher de 58 anos com diagnóstico de LES e Síndrome Anti-Fosfolípido (SAAF) secundário

foi admitida no serviço de internamento para investigação de aumento dos tempos de coagulação. Trata-se de doente com história de metrorragias abundantes em idade fértil, 5 abortos espontâneos após as 12 semanas, trombose em vasos do olho esquerdo, acidente vascular cerebral (AVC) isquémico, e Enfarte Agudo do Miocárdio (EAM) extenso, não tendo sido colocado cardio-desfibrilhador implantável (CDI) ou iniciada anticoagulação por aumento dos tempos de coagulação.

Analiticamente apresentava trombocitopenia (plaquetas 143 x109/L), aumento do TP (18.8 segundos), prolongamento do aPTT (>120 segundos) e tempo de trombina normal (21 segundos). Procedeu-se ao teste de mistura do plasma da doente com plasma normal não se verificando correção dos valores de tempo de coagulação. Os fatores II, V, VII e X encontravam-se reduzidos, não havendo correção com plasma normal. Os fatores IX, XI e XIII encontravam-se também reduzidos, porém após correção por saturação do anticorpo com plasma normal verificou-se correção dos mesmos. Procedeu-se ao doseamento do fator VIII e IX pelo método cromogénico, estando os seus valores dentro do intervalo de normalidade. Realizou-se também tromboelastrograma, compatível com estado de hipercoagubilidade. Para além disso apresentava pesquisa de anticoagulante Lúpico positivo, anticorpo anti-Cardiolipina IgG > 640 GPL/mL e anticorpo anti-Beta-2-Glicoproteina IgG > 867 UA/mL. Iniciou-se tratamento com azatioprina 25 mg (doente não podia realizar hidroxicloroquina por aumento do intervalo QT). Dada a ocorrência de múltiplos eventos trombóticos no passado foi iniciada anticoagulação com enoxaparina em doses progressivamente maiores até estar em dose terapêutica, posteriormente com switch para varfarina 5mg/dia, estando até à data sem ocorrência de novos eventos trombóticos ou hemorrágicos.

Foi proposta também para terapêutica com rituximab que ainda não realizou.

Discussão: Apesar de estar associado a maior risco de hemorragias, já foram descritos casos de trombose associados ao Síndrome Anticoagulante Lúpico-Hipoprotrombinemia, uma vez que a ligação anticorpo antifosfolipido/protrombina aumenta o risco de trombose em doentes com SAAF. Não existem recomendações para o tratamento desta síndrome, porém é recomendado o início de imunossupressão no caso de não haver resolução espontânea dos tempos de coagulação.

207 - REFRACTORY EOSINOPHILIC FASCIITIS: CASE REPORT

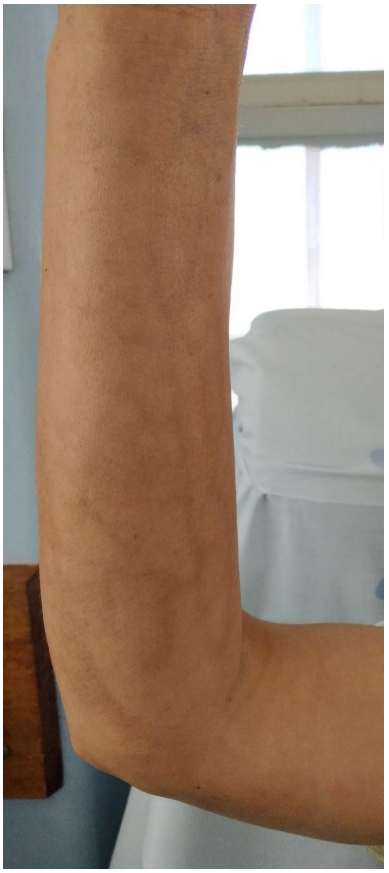
Daniela Santos Oliveira[1, 2], Ana Martins[1], Filipe Oliveira Pinheiro[1], Maria Seabra Rato[1], Diogo Guimarães da Fonseca[3], Carlos Vaz[1, 4], Pedro Madureira[1], Lúcia Costa[1]

[1]Rheumatology Department, Centro Hospitalar de São João, Porto, Portugal, [2]Center for Health Technology and Services Research (CINTESIS), Faculty of Medicine, University of Porto, Porto, Portugal, Porto, Portugal, [3]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [4]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal

Introduction: Eosinophilic fasciitis (EF) is a rare connective tissue disease, with unclear etiology. This condition is characterized by hardening and thickening of the skin, mainly affecting the extremities and associated with peripheral eosinophilia, elevated erythrocyte sedimentation rate (ESR) and hypergammaglobulinemia. The majority of patients respond to high-dose corticosteroids. Thus, a case of EF is being reported for its rarity and partial response to prednisolone. Case report: We report a case of a 47-year-old female, with a personal history of multinodular goiter and no usual medication. This patient was admitted to the rheumatology service due to pain and skin hardening of right upper and lower limbs for the last 5 months. There were no systemic complaints, skin rash, Raynaud's Phenomenon, genital or oral ulcers, respiratory, gastrointestinal or genitourinary manifestations. No trauma or exacerbated physical activity were reported. During these months, the patient was medicated with an anti-inflammatory drug and low-dose corticosteroids without relief. General physical examination was normal. On physical examination, skin thickening on right leg and forearm were observed. On the forearm, the groove sign when patient elevating the limb was visible (Figure 1). Left limbs, hands and fingers were unaffected. There were no other mucocutaneous changes or peripheral arthritis. On investigation, she had peripheral eosinophilia (8.6%), an elevated ESR (43 mm in the first hour) and polyclonal hypergammaglobulinemia (gamma region). Hemogram, platelet, hepatic parameters, renal function and muscle enzymes were normal. Rheumatoid factor (38.3 UI/mL) and antinuclear antibody (1/640, homogeneous pattern) were positive. Remaining immunological study was negative, name-

ly complement levels. Osteoarticular (elbows, fists, knees, ankles) radiographs and right forearm and leg ultrasounds were normal. Magnetic resonance imaging of right forearm documented thickening, edema and hyper-uptake of fascial planes, without significant muscle involvement. Biopsy of deep skin and muscle showing fibrinous exudate, marked lymphoplasmacytic inflammatory infiltrate and occasional eosinophils, findings compatible with EF. Prednisolone 1mg/kg/day treatment was instituted with partial clinical and analytical response. Methotrexate was initiated early, with favorable pain response and sustained decline of peripheral eosinophilia and ESR. After one year of follow-up in rheumatology consultation, under methotrexate 25 mg/week and prednisolone 5 mg/day, the patient maintains skin thickening and hardening. Thus, in the future the possibility of introducing another immunosuppressive therapy namely mycophenolate mofetil will be discussed. Conclusion: EF is a rare clinical condition with delayed diagnosis. A high index of clinical suspicion of EF is needed to establish an early diagnosis and to target treatment preventing unnecessary invasive exams. Treatment of EF remains poorly understood: although most cases respond well to high doses of oral corticosteroids, some patients may be refractory. Therefore, more studies are needed to explore other therapeutic options.

FIGURE 1: GROOVE SIGN.



209 - WUNDERLICH SYNDROME AS A RARE CLINICAL MANIFESTATION OF POLYARTERITIS NODOSA

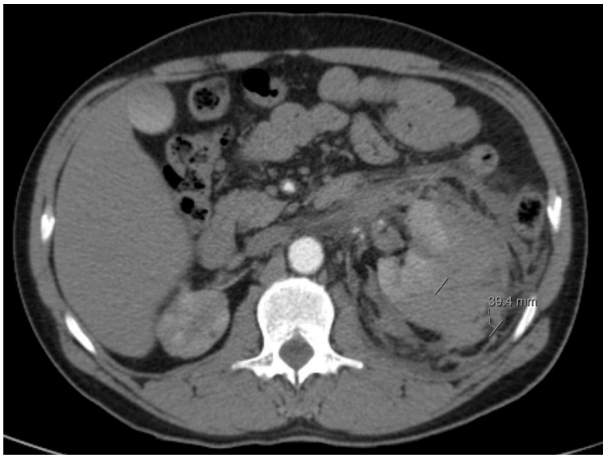
Daniela Santos Oliveira[1, 2], Ana Martins[1], Frederico Rajão Martins[3], Maria Seabra Rato[1], Filipe Oliveira Pinheiro[1], Diogo Guimarães da Fonseca[4], Carlos Vaz[1, 5], Eva Mariz[1, 5], Lúcia Costa[1]

[1]Rheumatology Department, Centro Hospitalar de São João, Porto, Portugal, [2]Center for Health Technology and Services Research (CINTESIS), Faculty of Medicine, University of Porto, Porto, Portugal, Porto, Portugal, [3]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [4]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [5]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal

Introduction: Renal involvement is the most common complication of polyarteritis nodosa (PAN) and usually presents with arterial hypertension, proteinuria and renal insufficiency. Spontaneous subcapsular and perirenal hematoma (Wunderlich syndrome -WS) in PAN is rare and its diagnosis can be confused with acute pyelonephritis resulting in delayed treatment. Thus, a case of WS in PAN is being reported for its rarity and complexity of diagnosis. Case report: We report a case of a 48-year-old male, smoker (27 pack-year) with a history of recurrent episodes of bilateral flank pain, painful nodular skin lesions in the lower limbs and weight loss of 10 kg with 1 year of evolution. Further, the patient reported recent onset of difficult-to-control arterial hypertension medicated with perindopril, indapamide and amlodipine. On several visits to the emergency department, he was diagnosed with acute pyelonephritis. In the last visit, the patient additionally reported abdominal and chest pain with refractory hypertension. There were no arthralgias, fever, mucocutaneous changes, respiratory and genitourinary manifestations. At physical exam, the patient was pale, hypertensive (252/145 mmHg) and with abdomen diffusely distended and painful during deep palpation. On investigation, he had hy-

pochromic microcytic anemia (hemoglobin (Hb) 7.5 g/dL), elevation of C-reactive protein (CRP 120 mg/L) and creatinine 1.2 mg/dL. Angio-CT-abdominopelvic showed a large left perirenal hematoma with a small active hemorrhagic focus and images suggestive of pseudoaneurysms with 6 mm (Figure 1). The patient was admitted to the intensive care unit and transfused with 1 unit of packed red blood cells. After hemodynamic stabilization, he was referred to the Urology service where he completed 7 days of antibiotic therapy with ceftriaxone. An arteriography, after 24 hours, documented a slight irregularity in the contours of the renal artery without evident pseudoaneurysms or active hemorrhage foci. Due to the persistence of pain and elevation of CRP with no infectious or neoplastic foci, the patient was transferred to Rheumatology service for further study. In our service, patient remained with mild pain and controlled blood pressure with amlodipine, metoprolol and lisinopril. The analytical study revealed Hb stabilization (9.7 g/dL) with persistent elevation of CRP (100 mg/L) and creatinine 1.7 mg/dL. Platelet, hepatic parameters, erythrocyte sedimentation rate and muscle enzymes were normal. 24-hour urine collection revealed proteinuria (0.46 mg/24h). Further evaluation revealed normal protein electrophoresis and complement; negative serologies for hepatitis B and HIV and negative blood and urine cultures. Rheumatoid factor, antinuclear antibodies, extractable nuclear antigen antibodies and Antineutrophil Cytoplasmic Antibodies were negative. Lower limb electromyogram was normal. Lower limb biopsy showed muscle with mild changes in the neurogenic profile. For suspicion of PAN patient started pulses of methylprednisolone 1g/day (3 days) with clinical and analytical improvement. The Angio-CT reassessment reported a significant reduction in the size of peri-renal hematoma, with no active hemorrhage. The patient was referred to the Rheumatology consultation with prednisolone 0.5 mg/kg/day and indication to initiate intravenous cyclophosphamide. Conclusion: WS in PAN is a rare life-threatening complication whose diagnosis can be a complex challenge. Thus, a high index of clinical suspicion is needed to obtain an early accurate diagnosis preventing serious complications.

FIGURE 1- ANGIO-CT-ABDOMINOPELVIC SHOWING LARGE LEFT PERIRENAL HEMATOMA WITH A SMALL ACTIVE HEMORRHAGIC FOCUS.



212 - MANTLED IN THE BLOOD

Margarida Lucas Rocha[1], Ana Teodósio Chícharo[1], Vítor Teixeira[1], Graça Sequeira[1]
[1]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal

A caucasian 46-years-old man with previous history of ulcerative colitis and transient interstitial pneumonia secondary to either mesalazine treatment or cockatoo feathers dust, was referred to Rheumatology due to positive antinuclear antibodies (ANA) and anti-dsDNA antibodies. He complained of night sweats and fatigue, but denied joint pain, oral and genital ulcers, alopecia, photosensitivity, rash, purpura, xerostomy, xerophthalmia, Raynaud's phenomenon and neurologic complains. Persistent thrombocytopenia for at least 3 years was noted, and for the last year anaemia, lymphopenia and neutropenia were also evident. On physical examination, we noted pale mucous membranes and splenomegaly was clearly evident on abdominal palpation.

Iron levels, vitamin B12 and folic acid were normal. There was no serological evidence of HIV 1/2, viral hepatitis or Treponema pallidum. The patient had an immunological scar for Epstein-Barr virus (EBV), Parvovirus and Cytomegalovirus infection.

Apart from positive ANA and high titre anti-dsDNA, the patient also presented with severe complement C4 consumption and positive lupus anticoagulant, anti-cardiolipin IgM/IgG and anti-beta2-gp1 IgM on more than one occasion. A chest and abdomen CT showed multiple axillary, mediastinal and retroperitoneal adenopathies and confirmed a significant splenomegaly (> 25 cm).

The bone marrow biopsy revealed trilinear hematopoietic tissue with reactive changes and infiltration

of a diffuse pattern by small B-cell lymphoma CD20+, CD5+, CyclinD1-, SOX11+/-, LEF1-, CD23-, compatible with mantle cell lymphoma (MCL). A lymph node biopsy of the internal jugular chain was performed and showed a classical morphology of MCL, with a nodular and mantle pattern, p53<10% (wild type) and a Ki67 of 25%. The patient was referred to the Instituto Português de Oncologia for follow-up and treatment.

Despite several immunological features of Systemic Erythematosus Lupus (SLE) in this case, we believe they were driven by the MCL.

The interplay between the immune system and cancer cells is known to be very complex and bidirectional. Of note, a higher risk of non-Hodgkin Lymphoma (NHL) has been previously reported in many autoimmune diseases, including SLE.

Recently in a Taiwan study, Wang L. et. al published the first nationwide population based study to report a bidirectional relationship between SLE and NHL. The study revealed that the patients with SLE had a slightly higher risk of overall cancer, and that the highest risk was for NHL. Several hypotheses can corroborate this association, namely a possible common genetic basis, B cell and T cell activation in the pathogenesis of both autoimmune diseases and NHL and probably the same trigger factors. In this case, the patient had an immunological scar from past EBV infection, which has been described as important in the etiopathogenesis of both SLE and NHL.

213 - JACCOUD´S ARTHROPATHY IN A PATIENT WITH OSTEOGENESIS IMPERFECTA

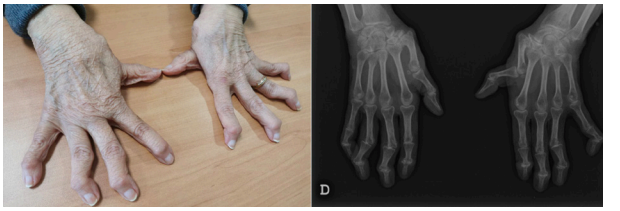
Margarida Lucas Rocha[1], Ana Teodósio Chícharo[1], Vítor Teixeira[1], Graça Sequeira[1]

[1]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal

Osteogenesis imperfecta (OI) is a rare congenital disorder of the collagen production that results in brittle bones and affects other body organs and systems containing collagen, with an estimated prevalence of 660 cases in Portugal (APOI, 2020). Joint mobility caused primarily by ligamentous laxity may also be present in OI.

Recently, we diagnosed a type I OI in a 66 years-old portuguese woman who suffered from osteoporosis at the level of the femoral neck and osteopenia at the lumbar spine, had a previous low-impact radial head fracture, low stature, lumbar scoliosis, multiple teeth loss and blue scleras. She also had a family history of OI (son). Her son´s genetic analysis identified a heterozygous variant, c.2032>T; p.(Glu678*), in COL1A1 exon 31, not present in population databases but that had been previously reported in association with osteogenesis imperfecta by Benusiené and Kucinskas, 2003. This nonsense variant is predicted to produce a prematurely truncated protein or the transcript is degraded by nonsense mediated decay.

FIGURE: PHOTOGRAPH OF THE PATIENT'S HANDS WITH JACCOUD´S ARTHROPATHY (ON THE LEFT) AND RADIOGRAPHY OF BOTH HANDS (ON THE RIGHT)



In addition to typical features of OI, our patient also complained of mild joint pain of the fingers and presented severe progressive rheumatoid arthritis-like hand deformities, including swan neck deformities and Z-thumb in both hands. The deformities were reducible and arthritis was absent, suggesting a Jaccoud's arthropathy. There were no clinical symptoms suggestive of systemic lupus erythematosus (SLE) nor a past history of rheumatic fever. There was also no evidence of skin hyperextensibility and vascular fragility.

An ultrasound of the hands confirmed absent synovitis, with exception of low-grade synovitis in the fifth left proximal interphalangeal, radiocarpal and right mid-carpal region, which were considered normal. There was also no evidence of bone erosions or tenosynovitis. Rheumatoid factor, anti-citrulinated protein and antinuclear antibodies were negative. Therefore, we ruled out Rheumatoid Arthritis (RA) and SLE. She was treated for OI with zoledronic acid, calcium and vitamin D.

We did not find any association between Jaccoud's arthropathy and OI in the English or Portuguese literature, so we believe this is the first report of JA associated with OI. JA was traditionally described as occurring post-rheumatic fever. Nowadays, it's seen more often in association with SLE and RA. Although, JA is not recognized as a characteristic feature of OI, it is associated with ligamentous and capsular hyperlaxity which are also present in many OI patients and

other collagen disorders. Joint hypermobility as a cardinal feature of OI has a variable incidence of 34 to 100% in pediatric patients [Engelbert RH et al.; 1998; Brizola et al., 2014]. Interestingly, the prevalence of joint hypermobility seems to be independent of the severity of OI.

In conclusion, we suggest that JA should be recognized as a feature of OI, as a result of the well-known ligament laxity and joint hypermobility of collagen disorders.

214 - ARTRITE SÉPTICA DO JOELHO POR MICRORGANISMO ATÍPICO

Ana Margarida Correia[1], Emanuel Costa[1], Diogo Esperança Almeida[1], Joana Leite Silva[1], Joana Sousa-Neves[1], Ana Ribeiro[1], Marcos Cerqueira[1]
[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

Introdução: O *Propionibacterium acnes* é um bacilo Gram positivo, anaeróbio, com baixa patogenicidade, que está presente na flora da pele. Está associado, embora pouco frequentemente, a infeções de articulações protésicas do ombro, mas também, mais raramente, a quadros de artrite séptica de articulações nativas, como o joelho. A artrite por *P. acnes* tem geralmente uma apresentação clínica subtil e indolente, sendo comum os exames culturais do líquido sinovial serem negativos, dado o seu crescimento fastidioso em meio de cultura, requerendo mais que os habituais 5 dias de incubação.

Caso clínico: Apresenta-se o caso de um doente do sexo masculino com 34 anos, previamente saudável, com artrite do joelho direito desde 2017, 2 semanas após traumatismo contundente do joelho direito. Foi avaliado inicialmente em França e submetido a várias artrocenteses e injeções intra-articulares de corticoesteróide, com melhoria clínica apenas parcial e recidiva do quadro. No final de 2019, por manter dor e derrame do joelho direito, foi submetido a artroscopia com realização de sinovectomia e meniscectomia, sem resolução das queixas, com limitação funcional progressiva e necessidade diária de anti-inflamatório e analgésico. Em 2020 foi referenciado à consulta de Reumatologia para exclusão de outras causas de monoartrite crónica e recidivante do joelho direito. Não se constatavam outros sinais e sintomas associados, nomeadamente febre. Realizada artrocentese, com aspiração de líquido sinovial de características inflamatórias (52 000 células, 95% neutrófilos), sendo o exame microbiológico do líquido negativo. Pela manutenção da sintomatologia e progressiva redução da mobilidade articular do joelho direito, foi realizada biópsia sinovial para esclarecimento etiológico. Histologicamente, a membrana sinovial não exibia alterações, mas no exame microbiológico foi isolado *P. acnes*. Analiticamente apresentava subida dos parâmetros inflamatórios, com velocidade de sedimentação de 45 mm/h e proteína C reativa de 59,7 mg/L, sem leucocitose. Radiograficamente, verificava-se redução uniforme da entrelinha articular e múltiplas irregularidades corticais, a sugerir gonartrose secundária. Assumido como diagnóstico mais provável uma artrite séptica. Foi submetido a artroscopia para confirmação diagnóstica e lavagem intra-articular. Foram colhidas hemoculturas em aerobiose e anaerobiose, todas negativas. Na membrana sinovial colhida intra-operatoriamente foi isolado novamente *P. acnes*, estabelecendo-se o diagnóstico de artrite séptica. Foi instituída antibioterapia endovenosa com vancomicina e, posteriormente, penicilina, que cumpriu durante 3 semanas, seguida de 6 semanas de amoxicilina oral. O doente apresentou franca melhoria clínica e analítica, com controlo da dor, redução dos sinais inflamatórios articulares e recuperação, ainda que parcial, da mobilidade articular.

Conclusão: O caso clínico apresentado pretende demonstrar a importância de excluir etiologia infeciosa em casos de monoartrite crónica, destacando ainda a valorização de microrganismos como o *P. acnes*, muitas vezes assumidos como contaminação, conduzindo ao atraso diagnóstico e terapêutico, com eventual prejuízo funcional e estrutural. Salienta-se ainda o papel da biópsia sinovial para auxílio diagnóstico em casos atípicos e em que os restantes meios complementares são negativos ou dúbios. Apesar da história de traumatismo e manipulação da articulação, não se pode firmar a etiologia da infeção, contudo, ressalva-se, também, a importância da assepsia na realização de qualquer procedimento invasivo.

217 - TIME AND TIDE WAIT FOR NO MAN

Margarida Lucas Rocha[1], Ana Teodósio Chícharo[1], Vítor Teixeira[1], Graça Sequeira[1]
[1]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal

A 39-year-old Portuguese women was referred for observation in the rheumatology clinic due to scleroderma facies with microstomy and beaked nose, that

were apparent since late childhood. There was no other clinical feature suggestive of systemic sclerosis. The patient had a set of comorbidities typical of older age such as type 2 diabetes mellitus, dyslipidaemia/hypertriglyceridemia, early-onset menopause and, bilateral cataract surgery before the age of 30. She had also removed a chest melanoma from a non-sun exposed area. She had family history of leukemia and breast cancer, parental consanguinity and a healthy sibling.

During physical examination, short stature, exceptionally low-weight, sarcopenia, greying of the hair and premature loss of scalp hair stood out. Cutaneous xerosis was also evident, especially on the face with hyperpigmentation spots and a hyperkeratosic ulcer of the right elbow. Another unexpected finding was osteoarthritic deformations of both hands.

Laboratory tests showed negative antinuclear antibodies and diabetes mellitus autoantibodies. ESR and CRP were within the reference values. A bone densitometry revealed low bone mass density for age at the spine and hip level.

After ruling out systemic sclerosis, a clinical diagnosis of Werner syndrome was established taking into consideration the International Registry of Werner Syndrome criteria. A molecular diagnosis further confirmed the diagnosis, showing a mutation of the WRN gene, with a newly described variant: c.1127del, p. (Asp376Valfs * 3).

Werner Syndrome is also called Adult Progeria and is a rare autosomal recessive disorder characterized by telomere dysfunction leading to low life expectancy and early appearance of characteristics and comorbidities typically found in elderly individuals. This disease is associated several rheumatic conditions such as early osteoarthritis, early osteoporosis, sarcopenia, soft tissue calcifications and scleroderma facial features.

221 - OLIGOARTRITE DAS ARTICULAÇÕES TRAPÉZIO-METACÁRPICAS: ARTRITE PARANEOPLÁSICA OU IATROGENIA MEDICAMENTOSA?

Ana Lúcia Fernandes[1], Susana Fernandes[2]
[1]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, [2]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal

Introdução: A patologia reumática paraneoplásica é definida por sintomas musculo-esqueléticos secundários a uma doença maligna subjacente, mas que não estão diretamente relacionados com a massa tumoral ou metástases. A gestão sintomática pode consistir num desafio terapêutico.

Caso clínico: sexo masculino, 61 anos, com antecedentes pessoais de adenocarcinoma da próstata, com metastização óssea extensa. Completou 8 ciclos de quimioterapia (docetaxel, leuprorrelina e prednisolona) e, por progressão bioquímica e imagiológica iniciou terapêutica com acetato de abiraterona, grosserrelina e prednisolona 10 mg/dia.

Descreve quadro com 1 mês de evolução, de início subagudo de dor e tumefação da base de D1 da mão esquerda e, mais recentemente, também da direita, inicialmente com ritmo mecânico e com progressão para ritmo misto. Negava trauma, envolvimento de outras articulações e queixas sistémicas. Ao exame objetivo, apresentava franca tumefação da base de D1 bilateralmente, com rubor, dor e calor associados e grande limitação funcional. Analiticamente, tinha VS 26-43 mm/h, PCR 0.9-3.3 mg/dL, FR e ac anti-CPP negativos, ácido úrico dentro dos valores normais (valor confirmado posteriormente) e serologias virais negativas. Na radiografia das mãos evidenciava apenas discreta rizartrose bilateral, sem sinais de condrocalcinose. Realizou ecografia dos punhos e mãos, que mostrou na articulação trapézio-metacárpica bilateralmente sinovite grau III, derrame articular grau II e sinal doppler grau III, sem outras alterações de relevo. Foi feita infiltração bilateral com 40 mg de acetato de metilprednisolona, com melhoria. Após discussão com Oncologia, optou-se por iniciar metotrexato 20 mg/semana. Dois meses depois, teve novo agravamento sintomático, desta vez acompanhado de agravamento bioquímico do PSA. A cintigrafia de corpo inteiro mostrou novas lesões metastáticas no osso ilíaco. Equacionou-se a hipótese de oligoartrite paraneoplásica. Realizou biópsia sinovial, que revelou apenas infiltrado inflamatório inespecífico e foi negativo para pesquisa de células neoplásicas. Aumentou-se dose de metotrexato para 25 mg/semana, trocou-se prednisolona 10 mg/dia para metilprednisolona 8 mg/dia, cumpriu ciclo de 15 dias de acemetacina 120 mg/dia associado a inibidor da bomba de protões e realizou nova infiltração intra-articular bilateral com 20 mg de hexacetonido de triamcinolona. Realizou ainda radioterapia no local das novas lesões metastáticas. Após 6 meses de seguimento, o doente encontra-se estabilizado do ponto de vista oncológico

e sem novos episódios de dor ou tumefação articular.
Conclusão: o desafio diagnóstico do presente caso prendeu-se com as hipóteses diagnósticas de iatrogenia medicamentosa e artrite paraneoplásica. Estão descritos diversos sintomas musculo-esqueléticos como efeitos adversos secundários à terapêutica com abiraterona, embora casos de artrite franca não estejam descritos na literatura. A artropatia paraneoplásica encontra-se descrita em vários tipos de tumores, e pode mimetizar outras patologias reumatológicas inflamatórias, como a artropatia microcristalina ou a artrite reumatóide. Numa série de 65 pacientes (Kisacik B et al, 2017), nas apresentações paraneoplásicas predominam o género masculino, início assimétrico e marcadores inflamatórios elevados. A evolução clínica destas manifestações, geralmente refratárias à terapêutica convencional, habitualmente segue um curso paralelo à evolução da neoplasia.

222 - ARTERITE DE CÉLULAS GIGANTES COM ENVOLVIMENTO EXTENSO DE GRANDES VASOS

Ana Lúcia Fernandes[1], Susana Fernandes[2]
[1]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, [2]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal

Introdução: A arterite de células gigantes (ACG) é uma vasculite sistêmica de grandes e médios vasos, com um pico de incidência entre os 70 e 79 anos. Pode ser responsável por um envolvimento extenso, com acometimento da aorta e dos seus ramos proximais.
Caso clínico: mulher, 72 anos, sem antecedentes pessoais ou familiares de relevo. Apresentou-se na consulta de Reumatologia com queixas de cefaleia temporal bilateral com irradiação holocraneana e cervical com pelo menos 1 mê de evolução. Negava alterações visuais, claudicação mandibular, hipersensibilidade do coro cabeludo e dor ou rigidez das cinturas escapulares. Negava também queixas sistémicas. Tinha sido previamente medicada em cuidados primários com prednisolona (PDN) 30 mg/dia e subsequente desmame até 6 mg/dia, com melhoria transitória e posterior recidiva dos sintomas. Analiticamente, a realçar aumento de VS (90 mm/h) e PCR (5.877 mg/dL), ANA positivo 1/160 padrão mosqueado e ANCA MPO/PR3 negativos. Fez ecodoppler das artérias temporais, que mostrou sinal do halo bilateralmente. Foi medicada novamente com PDN 30 mg/dia e metotrexato (MTX) 15 mg/dia, com melhoria das queixas. Já após 1 mês sob esta terapêutica, realizou PET-scan, que mostrou captação aumentada de radiofármaco em grandes vasos: artérias carotídeas, vertebrais, subclávias, aorta torácica e abdominal, ilíacas e femoral. O controlo analítico mostrou normalização dos valores anteriormente alterados. Foi assumido diagnóstico de arterite de células gigantes com envolvimento de grandes vasos e a doente foi submetida a 3 pulsos de 1g de metilprednisolona, seguido de PDN 40 mg/dia e MTX 20 mg/semana. Manteve seguimento em consulta, realizando desmame gradual de PDN, com resolução completa dos sintomas e sem recidiva clínica e analítica até à data.
Conclusão: Na ACG o envolvimento subclínico de grandes vasos está presente numa percentagem significativa dos pacientes, embora a sua repercussão sintomática seja observada menos frequentemente.

A ecografia com estudo colour-doppler dos vasos da cabeça, pescoço e membros suberiores, quando realizada por operador experiente, pode ser utilizada em alternativa à biópsia, como procedimento diagnóstico inicial. Contudo, o envolvimento aórtico abdominal é documentado com maior sensibilidade através de PET-scan, que apresenta ainda vantagem na investigação de eventual processo neoplásico concomitante.

O desmame da corticoterapia deve ser progressivo para evitar recidivas, sobretudo em casos com envolvimento vasculítico extenso.

O follow-up destes doentes deve considerar a vigilância e rastreio de potenciais aneurismas/dissecção da aorta, uma vez que estes estão associados ao aumento da mortalidade nesta patologia.

223 - DOENÇA MISTA DO TECIDO CONJUNTIVO – EVOLUÇÃO PARA LÚPUS ERITEMATOSO SISTÉMICO COM MANIFESTAÇÕES GRAVES

Maria Margarida Cunha[1], Jorge Pestana Lopes[1], Ana Catarina Duarte[1], Filipe Vinagre[1]
[1]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal

A doença mista do tecido conjuntivo (DMTC) é classicamente descrita como um síndrome de sobreposição de lúpus eritematoso sistémico (LES), polimiosite/dermatomiosite, e esclerose sistémica, manifestando-se mais frequentemente por fenómeno de Raynaud,

puffy hands, sinovite e miosite em doentes com títulos altos de anticorpos anti-U1 RNP. O envolvimento renal é pouco frequente, sendo particularmente incomum a glomerulonefrite proliferativa. O envolvimento grave do sistema nervoso central (SNC) é também raro.

Caso Clínico: homem de 33 anos, melanodérmico, que recorre à Consulta de Reumatologia por um fenómeno de Raynaud complicado de úlceras digitais, artralgias de ritmo misto dos pés e mãos, diarreia, anorexia e perda ponderal de 19% do peso corporal com 7 meses de evolução. Objectivamente apresentava esclerodactilia, pitting scars de várias polpas digitais das mãos e artrite das tibiotársicas e metatarsofalângicas. Analiticamente destacava-se hemograma sem citopénias, velocidade de sedimentação (VS) 34mm/1ªh, proteína C reactiva <0.1mg/dL, hipergamaglobulinémia policlonal, diminuição da fracção C3 do complemento, ANA 1/1280 e anticorpos anti-RNP positivos; padrão capilaroscópico com redução da densidade capilar e dilatações capilares, sem outras alterações. Admitiu-se o diagnóstico de DMTC e foi medicado com nifedipina 30mg/dia, prednisolona 15mg/dia, hidroxicloroquina 400mg/dia, rifaximina e azatioprina 2mg/Kg/dia; esta última suspensa posteriormente por leucopénia com neutropenia e substituida por metotrexato 10mg/semana. Verificou-se melhoria do fenómeno de Raynaud e da artrite e resolução da diarreia. Posteriormente por agravamento do fenómeno de Raynaud complicado de úlcera digital, fez iloprost endovenoso e iniciou sildenafil, com melhoria. Dois anos após o diagnóstico apresentou mucosite oral associada ao metotrexato que motivou a sua suspensão e fez nesse contexto um ciclo de rituximab, com melhoria clínica.

Três anos após o diagnóstico verifica-se, laboratorialmente, subida persistente da VS para 120mm/1ªh, hipocomplementémia de C3 e também C4, e surgimento de anticorpos anti-Sm, dsDNA 510UI/mL, nucleossomas, proteína P ribossomal, histonas, Ku e centrómero positivos, cumprindo então critérios de LES de acordo com os critérios ACR/EULAR. Um ano mais tarde admitido por edema periférico e hematoproteinúria de novo, com proteinúria de 24h de 2785mg. Realizou biópsia renal, que demonstrou glomerulonefrite membranoproliferativa, focal e segmentar, compatível com nefrite lúpica classe III. Fez pulsos de metilprednisolona 500 mg durante 3 dias, iniciando posteriormente prednisolona 1mg/Kg, micofenolato de mofetil 500mg/dia com subida da dose até 2 gr/dia e lisinopril 2.5mg/dia. No entanto, após alta, abandonou toda a terapêutica e é admitido por psicose, com ideação delirante. Realizou Ressonância magnética crânio-encefálica e punção lombar sem alterações. Admitiu-se envolvimento neuropsiquiátrico pela doença de base, tendo iniciado prednisolona 1mg/Kg e ciclofosfamida mensal, bem como olanzapina e lorazepam, com resolução do quadro psicótico em cerca de 2 semanas.

Neste doente houve uma evolução das manifestações clínicas de DMTC para um quadro de LES, com envolvimento renal e do SNG grave, incomuns na DMTC. Este caso é um exemplo de como o diagnóstico de DMTC pode não ser estanque, podendo assistir-se à sua evolução para outra forma de doença do tecido conjuntivo.

226 - SUBCUTANEOUS SARCOIDOSIS IN-PATIENT WITH PSORIATIC ARTHRITIS: A CASE REPORT

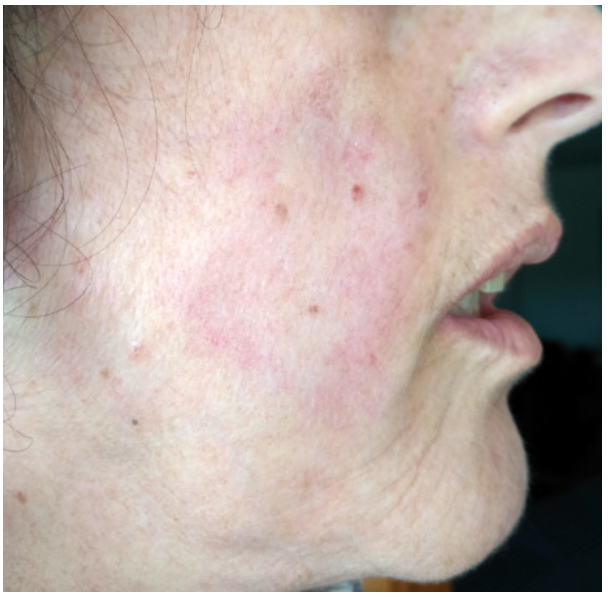
Bernardo Santos[1], Inês Cunha[1], Manuela Loureiro[2], Anabela Barcelos[1]

[1]Serviço de Reumatologia, Centro Hospitalar do Baixo Vouga, E.P.E., Aveiro, Portugal, [2]Dermatologia, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal

Introduction: Sarcoidosis is a multisystem chronic inflammatory disorder of unknown etiology characterized by the presence of noncaseating granulomas. It mainly affects the lungs, lymph nodes, skin and eyes. Excluding intrathoracic sarcoidosis, the incidence of isolated forms is rare and of these, the skin is the most common organ involved. Skin involvement may be the initial presentation. Given this challenge, though the biopsy specimen demonstrating non-caseating granulomas as the cornerstone diagnostic procedure, recent imaging modalities such as ultrasound and positron-emission tomography (PET) are being increasingly used for diagnosis.

Case Presentation: A caucasian women in their early 50s with axial and peripheral psoriatic arthritis, with 16 years of evolution, presented in the follow up visit at the rheumatology department, complaining of inflammatory pain on some small joints of the hands and painful subcutaneous nodules around the face and in the arms. The physical examination showed arthritis at the 4th and 5th proximal interphalangeal joints in the right hand, four palpable subcutaneous nodules: two at the frontal and submandibular region measured 1,7cm x 1cm and 0,5cm x 0,8cm, respec-

FIGURE: PALPABLE SUBCUTANEOUS NODULE AT RIGHT SIDE OF FACE MEASURING 2,5CM X 3CM.



tively; one at right side of face measured 2,5cm x 3cm (Figure 1) and one at the proximal extensor surface of the right elbow. It was decided to perform a biopsy of the frontal subcutaneous nodule that revealed dermohypodermic granulomatous infiltrate, represented by large histiocytes and multinucleated cells, resembling sarcoid type (Figure 2). Facing these findings, it was requested an 18-FDG-PET that documented subcutaneous thick tracer all over the body, more concentrated to the upper limbs and axillar lymph nodes correlating with the suspected hypodermic sarcoid diagnosis. She started prednisone dose of 60 mg daily and hydroxychloroquine 6,5mg/kg/day and repeated 18-FDG-PET 12 weeks after the beginning of the treatment which revealed absence of hypodermic sarcoid activity.

Conclusion: We emphasize the importance of exploring all the new signs and symptoms of each disease during follow up, especially when an immune inflammatory condition like psoriatic arthritis is present. Further research is needed to create new tools for evaluation, treatment and long-term follow-up of these isolated cutaneous presentations.

233 - ARTROPATIA DE JACCOUD E GOTA POLIARTICULAR- UMA ASSOCIAÇÃO INCOMUM

André Saraiva[1], Ana Isabel Maduro[1], Liliana Saraiva[1], Marlene Sousa[1], Margarida Coutinho[1]
[1]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

Introdução: A artropatia de Jaccoud é caracterizada por deformações severas redutíveis e não erosivas das articulações. Pode envolver qualquer articulação, mas afeta primariamente as metacarpo-falângicas (MCF). Inicialmente descrita em doentes com febre reumática, tem sido associada a várias doenças reumáticas crónicas, especialmente Lúpus eritematoso sistémico (LES). Pode facilmente ser confundida com as deformações objetivadas na artrite reumatoide (AR). A sua associação com gota é muito incomum.

Caso Clínico: Os autores descrevem o caso clínico de um doente do sexo masculino de 52 anos, referenciado à consulta de Reumatologia por poliartralgias de ritmo misto, migratórias, recorrentes, de instalação aguda e com sinais inflamatórios exuberantes. As queixas envolviam as mãos, punhos e antepés, evoluindo por crises desencadeadas pelo consumo de alimentos ou bebidas ricas em purinas. Encontrava-se medicado com alopurinol 300mg id e colquicina 1mg, os quais cumpria apenas em crise. Ao exame objetivo era evidente a presença de nódulos duros, indolores, de coloração esbranquiçada sobre a superfície extensora dos cotovelos. Apresentava, também, deformações redutíveis em colo de cisne, desvio cubital da mão direita, polegar em Z, hallux valgus bilateral e calosidades atípicas (alterações sugestivas de artropatia de Jaccoud). Analiticamente apresentava reagentes de fase aguda e hiperuricemia normal, bem como ausência de fator reumatoide (FR) e anticorpo anti- péptido citrulinado cíclico (anti-CCP). Foi realizada aspiração de um dos nódulos que revelou a presença de cristais de urato monossódico, confirmando tratar-se de um tofo gotoso. Radiologicamente era evidente a presença de subluxações das MCF à direita e de osteoartrose secundária das radiocárpicas e de algumas MCF's (Figura 1). A radiografia dos pés evidenciou lesões líticas tipo "saca-bocado" na 1ª, 4ª e 5ª metatarso-falângicas (MFT's) direitas (Figura 2). Perante o quadro foi assumido o diagnóstico de gota tofácea poliarticular com artropatia de Jaccoud associada. O doente foi aconselhado a cumprir alopurinol regularmente, bem como dieta pobre em purinas

Discussão: A evolução clínica por crises, de instalação aguda e evolução recorrente, desencadeadas pelo consumo de alimentos ricos em purinas sugeria tratar-se de um quadro de gota. Por outro lado, a presen-

FIGURA 1. MÃOS E RADIOGRAFIA DAS MÃOS (AP) – ALTERAÇÕES COMPATÍVEIS COM OSTEOARTROSE DE AMBAS AS RADIOCÁRPICAS, CARPOS E DE ALGUMAS MCF. DESVIO CUBITAL DA MÃO DIREITA E POLEGAR EM Z BILATERAL

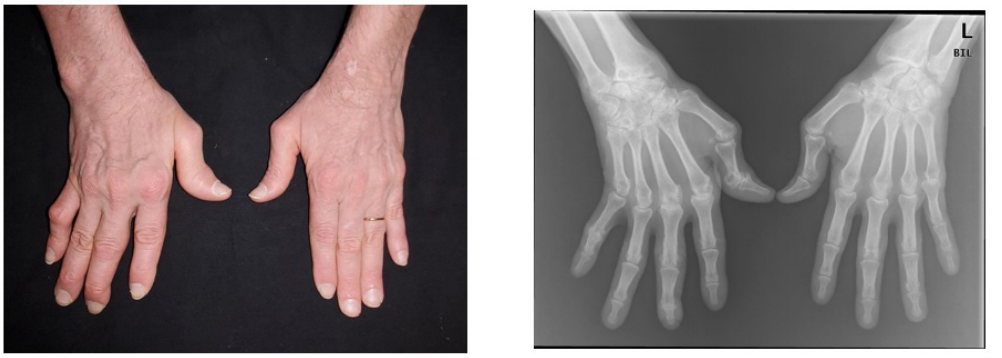
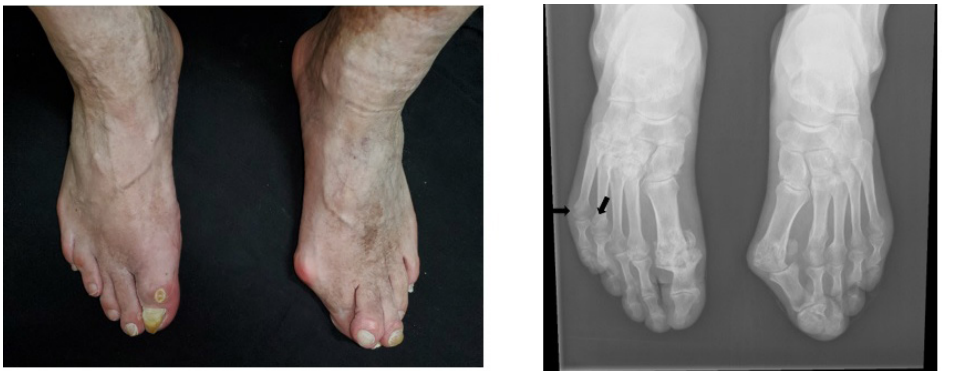


FIGURA 2. PÉS E RADIOGRAFIA DOS PÉS (AP) – ALTERAÇÕES COMPATÍVEIS COM OSTEOARTROSE DE DA 1ª MTF (BILATERALMENTE) E LESÕES TIPO "SACA-BOCADO" NAS 4ª E 5ª MTF (SETA)



ça de artropatia de Jaccoud assimétrica e a ausência de FR e anti-CCP, bem como a presença de lesões radiológicas tipo " saca-bocado" afastaram a hipótese de Artrite Reumatoide. A identificação de cristais de urato monossódico numa das lesões nodulares permitiu estabelecer o diagnóstico de gota tofácea.

A terapêutica e adequado controlo da doença de base, neste caso, foram fundamentais, permitindo a prevenção de novas crises

Conclusão: A artropatia de Jaccoud é caracterizada pela reversibilidade das deformações, ao contrário do que se verifica na Artrite Reumatoide. Embora esteja mais comummente associada a LES pode, raramente, surgir em associação com artrites microcristalinas, tal como descrito neste caso.

235 - LESÃO PSEUDOTUMORAL NA OROFARINGE EM DOENTE COM ESCLEROSE SISTÉMICA: UM DESAFIO DIAGNÓSTICO

Bruno Miguel Fernandes[1], Miguel Bernardes[1], Candida Abreu[2], Gil Coutinho[3], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Serviço de Infecciologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [3]Serviço de Otorrinolaringologia, Centro Hospitalar Universitário de São João, Porto, Portugal

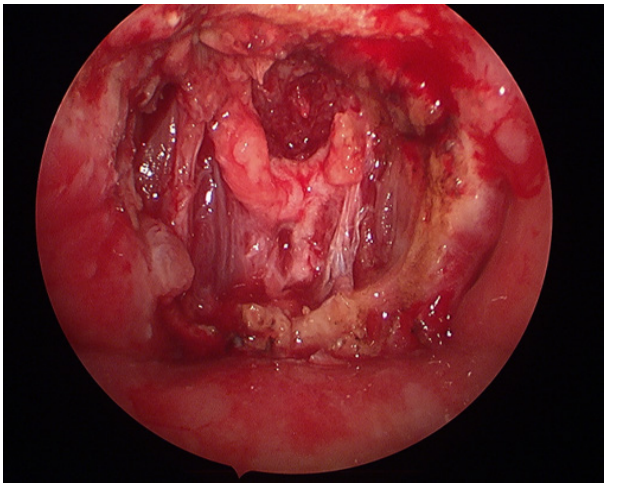
Introdução: A esclerose sistémica é uma doença do tecido conjuntivo que se caracteriza pela fibrose da pele e de orgãos internos, alterações na microvasculatura e anomalias na imunidade celular e humoral. O risco de neoplasias está aumentado nos doentes com esclerose sistémica, assim como o risco infecioso, tanto por fatores relacionados com a doença como com o seu tratamento.

Caso clínico: Doente do sexo feminino, 66 anos, com antecedentes de esclerose sistémica forma cutânea limitada com 20 anos de evolução. Como manifestações clinico-laboratoriais cumulativas da doença apresentava: anticorpos anti-nucleares (>1/1000, padrão centromérico) e anti-centrómero positivos, telangiectasias, microstomia, fenómeno de Raynaud das mãos, esclerodactilia, úlceras digitais recorren-

tes com necessidade de terapêutica com alprostadilo endovenoso, calcinose das extremidades dos 4º e 5º dedos das mãos e disfagia com ausência de peristaltismo na parte inferior do esófago e hipotomia do esfíncter esofágico inferior. Encontrava-se medicada com metotrexato 10mg/semana oral, nifedipina 60mg id, pentoxifilina 400mg tid, losartan 100mg id, naproxeno 500mg bid e omeprazol 40mg id. Em agosto de 2020 iniciou quadro de hipoacúsia e otalgia bilaterais com predomínio à esquerda e, um mês depois, disfagia em agravamento progressivo - inclusivé na toma dos comprimidos - e disfonia, assim como diarreia e cólicas abdominais. Associadamente, apresentava emagrecimento de 4kg nos últimos 6 meses. Inicialmente foi tratada com azitromicina, sem melhoria. Realizou tomografia computorizada cervical que revelou lesão infiltrativa e expansiva com aparente origem no cavum faríngeo e extensão amigdalina. Nesta altura, o metotrexato foi suspenso. Em consulta de Otorrinolaringologia (ORL), foi observada na loca amigdalina esquerda da orofaringe, uma lesão infiltrativa e aparentemente necrosada; a fibroscopia mostrou aparente lesão neoplásica da nasofaringe com continuidade com a lesão observada na loca amigdalina, tendo sido realizadas biópsias (figura 1). A histologia mostrou processo inflamatório agudo e presença de exsudato fibrino-leucocitário - a traduzir ulceração - sem sinais de malignidade. A biópsia cirúrgica da lesão corroborou os resultados do exame prévio, observando-se também infiltrado inflamatório mononucleado em algumas áreas. A endoscopia digestiva alta mostrou úlcera com 3cm na junção esofagogástrica, que foi submetida a várias biópsias, tendo a histologia mostrou um processo reativo e o estudo microbiológico revelou forte positividade na PCR de DNA de citomegalovirus (CMV), assim como positividade fraca para HSV 1, 2 e 7. A doente apresentava serologia IgM para CMV duvidosa e PCR de CMV positivo no sangue. Por este motivo, a doente foi observada por Infecciologia, que assumiu a possibilidade de infeção por CMV, tendo a doente iniciado valganciclovir 45mg/dia durante 21 dias. Após o tratamento, a doente apresentou melhoria clara da disfagia e resolução das cólicas e da diarreia, com a observação por ORL a mostrar completa resolução das lesões à observação da orofaringe.

Conclusão: As infeções sintomáticas por CMV são mais comuns em doentes imunodeprimidos. Diarreia e dor abdominal são os sintomas gastrointestinais mais comuns destas infeções, no entanto, ainda que incomum, o atingimento da parte superior do tubo digestivo é possível. Este caso salienta a importância do reumatologista estar alerta para a possibilidade de infeções menos comuns em doentes com doenças inflamatórias reumáticas sob imunossupressão.

FIGURA 1. IMAGEM DE FIBROSCOPIA A MOSTRAR LESÃO ULCERATIVA/FIBRINOSA DA PAREDE POSTERIOR DA NASOFARINGE.



239 - SINOVITE VILONODULAR PIGMENTADA – DO TÍPICO AO ATÍPICO!

André Saraiva[1], Ana Isabel Maduro[1], Adriana Carones[1], Beatriz Mendes[1], Margarida Coutinho[1]
[1]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

Introdução: As articulações e a sinovial podem ser acometidas por tumores benignos e malignos. Os tumores benignos mais comuns são a sinovite vilonodular pigmentada (SNVP) e a condromatose sinovial. A SNVP afeta ambos os sexos com igual prevalência, surgindo comumente entre os 20 e 50 anos. É geralmente monoarticular e as localizações mais comuns são as grandes articulações (joelho (80%) e a coxo-femoral (15%).

Caso Clínico: Os autores reportam 2 casos de SNVP

Caso 1: Doente do sexo masculino de 24 anos com tumefação articular isolada e permanente do joelho esquerdo (figura 1) com 8 anos de evolução, associado a dor de ritmo mecânico. Negava queixas axiais ou sistémicas. Ao exame objetivo (EO) o joelho esquerdo apresentava-se tumefacto, indolor à mobilização passiva, mas com limitação da mobilidade. O estudo

analítico pautava pela normalidade. A ecografia revelou acentuado espessamento sinovial em todos os recessos articulares e moderado derrame articular, tendo a artrocentese revelado um derrame hemático (figura 2). A ressonância magnética nuclear (RMN) documentava alterações compatíveis com SNVP (figura 3). A biópsia da membrana sinovial foi inconclusiva. Foi submetido a sinovectomia total tendo o estudo anatomo-patológico da peça cirúrgica confirmado o diagnóstico

Caso Clínico 2: Doente do sexo feminino de 62 anos, com história de tumefação progressiva da 2ª Metatarso falângica (MTF) direita, com 5 anos de evolução. Nos últimos 2 anos havia iniciado um quadro de poliartralgias de ritmo misto, envolvendo os punhos, tibiotársicas e antepé, o que motivou referenciação para a consulta de Reumatologia. Sem queixas sistémicas.

Ao EO apresentava tumefação e limitação dolorosa dos punhos e tibiotársicas, moderada tumefação duro-elástica da 2ª MTF direita e squezze teste positivo das MTF's. O estudo laboratorial, não revelou alterações de destaque. O estudo ecográfico comprovou a presença de sinovite ligeira dos punhos e tibiotársicas, bem como a presença de exuberante distensão da 2ª MTF direita, contrastando com a presença de sinovite ligeira de algumas das restantes MTF. A RMN (figura 4) articular das articulações sintomáticas comprovou os achados e revelou erosões articulares nos carpos, metacarpos e 1ª MTF direita. Na 2º MTF direita, identificou acentuado espessamento da sinovial, com hipossinal em T2, alterações sugestivas

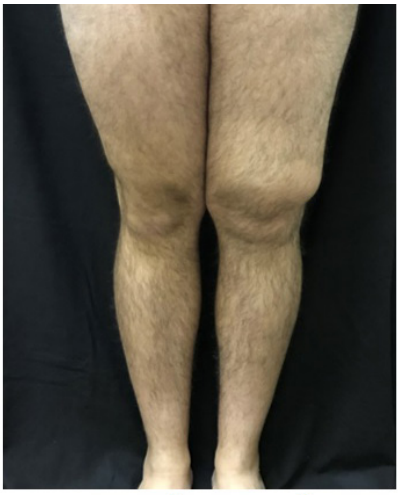


FIGURA 1: TUMEFAÇÃO ISOLADA CRÓNICA DO JOELHO ESQUERDO

FIGURA 2: LÍQUIDO SINOVIAL HEMÁTICO

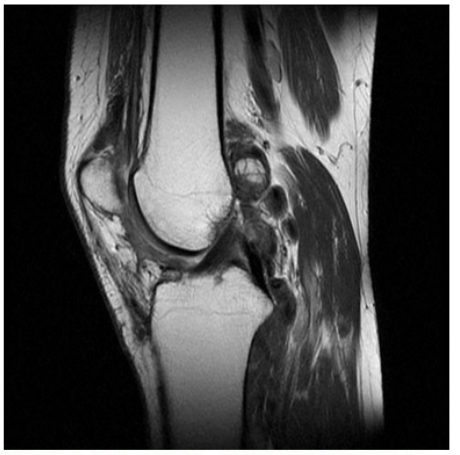


FIGURA 3: RMN JOELHO – SINOVITE VILONODULAR PIGMENTADA: ESPESSAMENTO IRREGULAR E HETEROGÉNEO DA SINOVIAL, COM EXTENSO COMPONENTE SÓLIDO NO ESPAÇO FEMORO-PATELAR, COM AGREGADOS SÓLIDOS

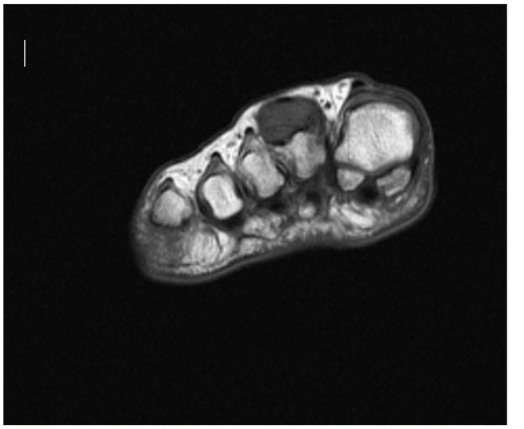


FIGURA 4: SINOVITE VILONODULAR PIGMENTADA DA 2ª METATARSO-FALÂNGICA; MARCADO ESPESSAMENTO SINOVIAL COM HIPOSSINAL EM T2 E EFEITO DE "BLOOMING"

de SNVP. Foi referenciada à consulta de Ortopedia, encontrando-se em lista de espera para cirurgia da 2ª MTF direita.

Discussão: O primeiro caso representa um caso típico de SNVP pela sua localização, duração do quadro e idade de apresentação. Já o segundo representa uma situação atípica, não só pela idade da apresentação, mas também porque envolve uma pequena articulação e porque se apresenta sobreposta a segundo quadro de patologia reumática, tendo sido objetivado de forma quase acidental. A Biópsia da membrana sinovial é o Gold Standard para o diagnóstico, mas a RMN pode sugerir o diagnóstico, revelando alterações quase patognomónicas

O tratamento cirúrgico é a primeira linha. A recorrência não é incomum. Em casos refratários a radiosinoviortese ou radioterapia são alternativas.

Conclusão: A SNVP é uma neoplasia benigna, mas que pode ser localmente destrutiva. A apresentação clínica pode ser diversa. O diagnóstico diferencial com artropatias inflamatórias, infeciosas e outros tumores é essencial. Apesar de se apresentar, na maioria das vezes, como uma monoartrite crónica de grande articulação, pode envolver, menos frequentemente, pequenas articulações e ocorrer em concomitância com outros diagnósticos reumatológicos.

241 - UMA CAUSA INCOMUM DE CERVICALGIA NO IDOSO

Margarida Faria[1], Duarte André Ferreira[2], Lídia Teixeira[1], Daniel Carvalho[1], Agna Neto[1], Nuno Pina Gonçalves[1], Ricardo Figueira[1], Alberto Quintal[1], Mário Rodrigues[1]

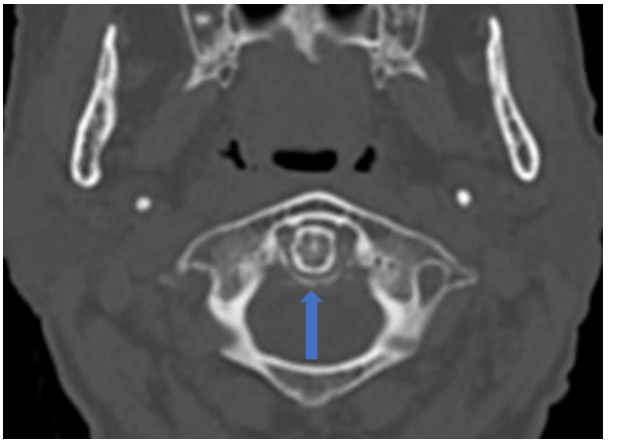
[1]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [2]Serviço de Medicina Interna, Hospital Central do Funchal, Funchal, Portugal

Introdução: A crowned dens syndrome (CDS) é uma forma de apresentação de Doença por Deposição de Cristais de Pirofosfato de Cálcio (DPPC), resultante da deposição destes cristais nos ligamentos cruciforme e alar, em torno do dente do áxis. Clinicamente, carateriza-se por cervicalgia aguda, rigidez cervical e febre. A identificação de calcificação em torno no processo odontóide, que se traduz por uma imagem em coroa, através de tomografia computorizada (TC), é o gold standard para o diagnóstico. O tratamento consiste no uso de anti-inflamatórios não esteróides, baixas doses de colchicina ou de corticosteroides.

Caso clínico: Mulher de 87 anos, internada na unidade de Acidente Vascular Cerebral (AVC) por AVC isquémico submetido a trombólise, é observada pela Reumatologia por quadro de cervicalgia aguda e gonalgia direita de ritmo inflamatório. Na anamnese, foi possível apurar história de gonalgia recidivante com vários anos de evolução. Ao exame objetivo, a destacar rigidez e dor na rotação cervical e choque de rótula positivo no joelho direito. Foi feita artrocentese do joelho direito com saída de líquido sinovial amarelo opaco de caraterísticas inflamatórias e hipercelularidade (2000 células/mm3 com predomínio de polimorfonucleares) e com presença de cristais rombóides e com fraca birrifrigência à microscopia ótica. O estudo radiográfico demonstrava condrocalcinose na sínfise púbica, joelhos e ligamento triangular do carpo. Por fim, a observação das imagens da TC crânio-encefálica revelou calcificação do ligamento transverso do atlas, sugestiva de CDS (imagem 1). Deste modo, foi assumido o diagnóstico de DDPPC e iniciada colchicina, 1 mg bidiário com melhoria significativa das queixas álgicas e de rigidez cervical passados 2 dias. Um mês mais tarde, foi observada na consulta de Reumatologia, com resolução completa das queixas álgicas, mobilidade cervical com amplitudes preservadas e ausência de sinais de derrame articular. O estudo complementar de possíveis causas de DDPPC permitiu a identificação de hiperparatiroidismo secundário a deficiência de vitamina D.

Conclusão: Uma vez que constitui uma causa rara de cervicalgia aguda, com uma incidência de 2% nestes doentes, a CDS é frequentemente equivocada como meningite, abcesso ou polimialgia reumática e con-

FIGURA: CALCIFICAÇÃO DO LIGAMENTO TRANSVERSO DO ATLAS (SETA) NA TC CRÂNIO-ENCEFÁLICA EM CORTE TRANSVERSAL, NA DOENTE DO CASO CLÍNICO.



sequentemente, o doente é submetido a investigação invasiva e/ou a tratamento com antibióticos de largo espectro desnecessários. Deste modo, o clínico deverá manter elevada suspeição para esta patologia, sobretudo em doentes com idades avançadas. Além disso, neste caso clínico, o diagnóstico de CDS no contexto de DDPPC conduziu ao diagnóstico de hiperparatiroidismo secundário.

REFERÊNCIAS:

1. Ledingham D, Cappelen-Smith C, Cordato D: Crowned dens syndrome. Pract Neurol. 2018, 18:57-59.
2. Siau K, Lee M and Laversuch CJ: Acute pseudogout of the neck-the crowned dens syndrome: 2 case reports and review of the literature. Rheumatol Int. 31:85–88. 2011.

242 - AFINAL HAVIA OUTROS (CRISTAIS): UM CASO INCOMUM DE DOR CERVICAL

Beatriz Mendes[1], Helena Assunção[1], Ana Isabel Maduro[1], Adriana Carones[1], Liliana Saraiva[1], Mariana Luis[1], Armando Malcata[1, 2]

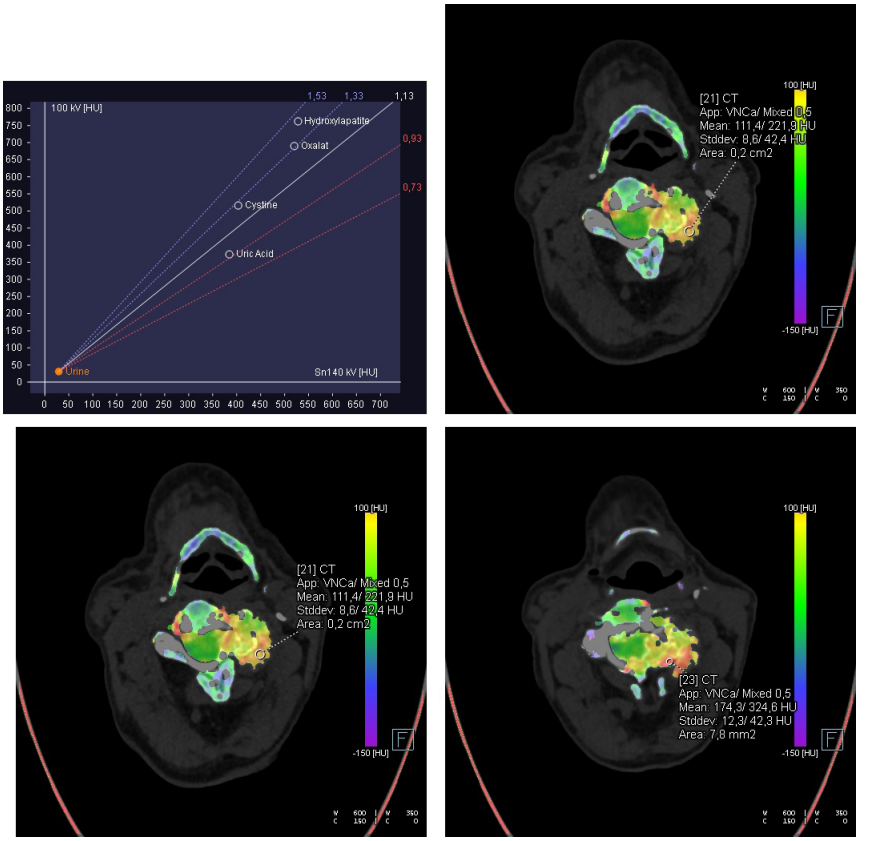
[1]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [2]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal

Introdução: As artropatias microcristalinas são uma causa comum de artrite periférica. Os cristais mais frequentemente identificados são os de monourato de sódio, pirofosfato de cálcio diidratado e fosfato básico de cálcio (incluindo hidroxiapatite). Embora descrito, o envolvimento axial em qualquer uma destas artropatias metabólicas é raro.

Caso Clínico: Doente do sexo masculino, de 56 anos de idade, orientado para consulta de Reumatologia por gota tofácea poliarticular. Tinha antecedentes pessoais de carcinoma espinocelular da língua em 2014 (ressecado); tumor neuroendócrino gástrico diagnosticado em Novembro de 2020 (ainda em estudo); hipertensão arterial mal controlada; doença renal crónica estádio 3a; etilismo crónico (75 g/dia de álcool); e tabagismo no passado (50 UMA).

À observação na consulta de Reumatologia referia

FIGURA 1 - TC DE DUPLA ENERGIA DA COLUNA CERVICAL



queixas de cervicalgia predominantemente mecânica, com ocasionais despertares nocturnos por dor, sem irradiação, sem carácter disestésico, sem noção de défices de força ou alterações sensitivas. Negava sintomas constitucionais ou história de traumatismo local recente. Ao exame objectivo apresentava dor e limitação da mobilidade da coluna cervical em todos os planos e dor à palpação das últimas apófises espinhosas cervicais. Nas pequenas articulações de ambas as mãos, punhos, cotovelos e joelhos apresentava tofos gotosos. Analiticamente destacava-se hiperuricémia de 7,9 mg/dL. Os parâmetros inflamatórios eram normais. Hemoculturas, serologias infecciosas (nomeadamente para Brucella) e IGRA foram negativos. Radiograficamente apresentava erosões em "saca bocado" em várias articulações metacarpofalângicas, interfalângicas proximais e distais de ambas as mãos; condrocalcinose de ambos os joelhos; e lesões erosivas envolvendo os corpos vertebrais de C5 e C6. A tomografia computorizada (TC) da coluna cervical revelou lesão óssea expansiva, de limites irregulares, com disrupção da cortical óssea, envolvendo os elementos posteriores de C3-C5, com marcada expansão óssea para os tecidos paravertebrais, de densidade heterogénea, com áreas hipodensas e áreas de densidade cálcica, sugerindo lesão neoplásica. A ressonância magnética da coluna cervical não mostrou achados sugestivos de osteomielite, espondilodiscite ou neoplasia. A biópsia guiada por TC não foi realizada por incapacidade do doente em permanecer em decúbito ventral. De modo a avaliar a presença de envolvimento axial por doença microcristalina em doente com gota tofácea e condrocalcinose foi solicitada TC de dupla energia (TC-DE), que revelou calcificações da metade esquerda das vértebras C3 e C4, compostas por cristais de hidroxiapatite e oxalato de cálcio (figura 1).

Conclusão: Este caso ilustra a necessidade de considerar a hipótese, embora rara, de envolvimento axial por artropatia microcristalina no diagnóstico diferencial de cervicalgia. Demonstra ainda o desafio diagnóstico neste doente. A TC-DE tem um papel decisivo, uma vez que esta entidade pode assumir aspectos radiográficos semelhantes aos de lesões neoplásicas e infecciosas. A peculiaridade do caso decorre igualmente da coexistência de diferentes tipos de cristais.

243 - GRANULOMATOSE EOSINOFÍLICA COM POLIANGEÍTE - CASO CLÍNICO COM APRESENTAÇÃO COM MONONEURITE MÚLTIPLA

Diogo Guimarães da Fonseca[1], Maria Seabra Rato[2], Filipe Oliveira Pinheiro[2], Ana Martins[2], Daniela Santos Oliveira[2, 3], Frederico Rajão Martins[4], Miguel Bernardes[2, 5], Lúcia Costa[2]

[1]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [3]Center for Health Technology and Services Research (CINTESIS),, Faculdade de Medicina da Universidade do Porto, Porto, Portugal, Porto, Portugal, [4]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [5]Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Introdução: A Granulomatose eosinofílica com poliangeíte (EGPA), previamente designada de Síndrome de Churg-Strauss, consiste numa vasculite de pequenos vasos que se associa à presença de granulomas extra-vasculares, eosinofilia, e, em cerca de metade dos doentes, de anticorpos anti-citoplasma do neutrófilo (ANCA), com predomínio de anti-mieloperoxidase (anti-MPO). Ocorre habitualmente em doentes com asma de início tardio e rinite alérgica. As principais manifestações são: febre, artralgias, mialgias, infiltrados pulmonares fugazes, afeção cardíaca e do sistema nervoso periférico.

Caso clínico: Apresenta-se o caso clínico de um indivíduo do sexo masculino, de 48 anos de idade, com antecedentes de polipose nasal e asma com início aos 35 anos. Nos últimos dois meses que antecederam o seu internamento hospitalar, as exacerbações da asma eram recorrentes e de difícil controlo, com necessidade de recurso a corticoterapia oral em doses crescentes. No último mês prévio à admissão passou a apresentar de novo os seguintes sintomas: febre, mialgias de predomínio proximal e hipostesia em meia no pé direito e com irradiação proximal até ao joelho. Analiticamente apresentava marcada elevação das proteínas de fase aguda, eosinofilia periférica [2000/mm3 (28,5%)], ANCA e anti-MPO positivos (68 UI/mL). A eletromiografia revelou lesão axonal muito recente dos nervos peroneal profundo direito, peroneal superficial direito e tibial direito, corroborando o diagnóstico de mononeuropatias múltiplas. No decurso do internamento, apresentou ainda elevação das troponinas, sem alterações da função sistólica biventricular no ecocardiograma, tendo-se optado, posteriormente, pela realização de ressonância magnética cardíaca que não evidenciou áreas de edema miocárdio ou realce tardio sugestiva de fibrose. O doente foi tratado com pulsos de metilprednisolona

1000mg durante 3 dias, com subsequente corticoterapia oral em alta dose e ciclofosfamida endovenosa como tratamento de indução e segundo o protocolo do ensaio CYCLOPS.

Conclusão: Este caso reporta um diagnóstico de novo de EGPA, realçando a sua associação frequente com asma de início tardio e com concomitante agravamento clínico, com necessidade frequente de corticoterapia oral e, por vezes, internamento hospitalar nas semanas prévias ao início da vasculite. Revisita ainda a possibilidade de envolvimento do sistema nervoso periférico nestes doentes, com desenvolvimento de mononeurite múltipla, bem como destaca a importância da ressonância magnética cardíaca na avaliação de um eventual envolvimento cardíaco pela EGPA que pode impactar fortemente no prognóstico vital do doente.

247 - UM OLHAR REUMATOLÓGICO SOBRE ÚLCERAS CUTÂNEAS - A PROPÓSITO DE UM CASO CLÍNICO

Diogo Guimarães da Fonseca[1], Filipe Oliveira Pinheiro[2], Maria Seabra Rato[2], Bruno Miguel Fernandes[2], Frederico Rajão Martins[3], Ana Martins[2], Daniela Santos Oliveira[2, 4], José Pinto[2], Lúcia Costa[2]
[1]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [3]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [4]Center for Health Technology and Services Research (CINTESIS), Faculty of Medicine, University of Porto, Porto, Portugal, Porto, Portugal

Introdução: A Esclerose sistémica é uma doença do tecido conjuntivo que se caracteriza por vasculopatia proliferativa, fibrose e disfunção imunológica. As principais manifestações são resultado dessas mesmas alterações, nomeadamente o fenómeno de Raynaud e as consequências vasculares subsequentes, com particular foco no desenvolvimento de lesões ulcerativas, bem como a progressão para fibrose e atrofia da pele. As úlceras no fenómeno de Raynaud habitualmente atingem as extremidades digitais com posterior progressão proximal e que pode evoluir para necrose e gangrena. Outras localizações suscetíveis são as proeminências ósseas, sendo a retração mecânica da pele um importante fator adicional. A face dorsal das articulações metacarpofalângicas (MCF) e interfalângicas proximais (IFP) é uma localização atípica, mas descrita na literatura e habitualmente de difícil controlo terapêutico.

Caso clínico: Apresenta-se o caso clínico de um indivíduo do sexo masculino, de 76 anos de idade, que recorreu ao Serviço de Urgência por lesões ulcerativas envolvendo a face dorsal da 2ª MCF direita, 2ª e 3ª IFP direita com cerca de 3 anos de evolução, sempre em tratamento de penso nos Cuidados de Saúde Primários e no domicilio. Na anamnese, o doente descreve poliartralgias de ritmo inflamatório dos punhos e pequenas articulações das mãos bilateralmente, fenómeno de Raynaud com mais de 20 anos de evolução, e episódios pregressos de úlceras a nível das extremidades digitais no período de Inverno. Ao exame objetivo apresenta múltiplas telangiectasias na região da face e mãos, as respetivas lesões ulcerativas com cerca de 1cm de maior diâmetro, sem exsudato purulento, sem exposição óssea ou tendinosa e poliartrite simétrica com envolvimento de articulações MCF e IFP. Destaca-se ainda, esclerose cutânea distalmente aos cotovelos e contractura em flexo das mãos bilateralmente. No estudo subsequente, a capilaroscopia evidenciou diversos megacapilares e hemorragias ativas ,sem áreas avasculares, compatível com padrão esclerodérmico em fase precoce. Os anticorpos anti-nucleares encontravam-se negativos, bem como o respetivo painel de anticorpos específicos de esclerose sistémica. Foi submetido a angio-ressonância do membro superior direito que não revelou etiologia macrovascular. Foi assumido o diagnóstico de esclerose sistémica, tendo iniciado terapêutica vasodilatadora com nifedipina e iloprost no internamento. Iniciou igualmente terapêutica imunossupressora com metotrexato e corticoterapia em baixa dose. Na consulta de reavaliação, 3 meses após alta do internamento, o doente apresentava cicatrização completa das úlceras, sem evidência de recidiva e sem artrite periférica.

Conclusão: Este caso reporta um diagnóstico tardio de esclerose sistémica, com a particularidade de apresentar lesões ulcerativas numa localização atípica, com um atraso diagnóstico e de referenciação considerável apesar das lesões tróficas evidentes. O diagnóstico foi corroborado com as restantes manifestações clínicas, os achados na capilaroscopia e a exclusão de etiologia macrovascular. Realça-se, portanto, o papel da Reumatologia no diagnóstico diferencial das lesões ulcerativas, particularmente se associadas a outras manifestações sugestivas de conectivite. Por fim destacamos ainda, a excelente resposta ao início

da terapêutica vasodilatadora, particularmente relevante por se tratar de uma localização descrita na literatura de difícil controlo terapêutico.

248 - DESSATURAÇÃO E TENSÃO ARTERIAL IMENSURÁVEL NOS MEMBROS SUPERIORES – UMA APRESENTAÇÃO INICIAL DE ARTERITE DE CÉLULAS GIGANTES COM OCLUSÃO BILATERAL DAS ARTÉRIAS AXILARES

Filipe Oliveira Pinheiro[1], Pedro Madureira[1], Maria Seabra Rato[1], Diogo Guimarães da Fonseca[2], Ana Martins[1], Daniela Santos Oliveira[1, 3], Frederico Rajão Martins[4], Bruno Miguel Fernandes[1], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [3]Center for Health Technology and Services Research (CINTESIS), Faculdade de Medicina da Universidade do Porto, Porto, Portugal, Porto, Portugal, [4]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal

Introdução: A arterite de células gigantes (ACG) é uma vasculite que afeta vasos de grande e médio calibre em doentes com idade mais avançada, sendo muito rara antes dos 50 anos. Afeta sobretudo artérias cranianas, mas o envolvimento extra-craniano tem vindo a ser cada vez mais reconhecido, acometendo o arco aórtico, artérias subclávias e axilares. As complicações vasculares descritas mais frequentemente são as estenoses e os aneurismas, mas a oclusão de grande vaso como forma de apresentação inicial é rara.
Caso Clínico: Doente do sexo feminino, de 64 anos, com antecedentes de artroplastia total da anca esquerda, que recorreu ao SU por um quadro de astenia, anorexia, perda ponderal e cefaleia holocraniana com hipersensibilidade do couro cabeludo, com cerca de 1 mês de evolução; ao exame objetivo apresentava à avaliação tensões arteriais dos membros superiores não mensuráveis e dessaturação avaliada em oxímetro periférico (SpO2 80% em ar ambiente) e parestesias com a elevação dos mesmos, com pulsos temporais superficiais palpáveis mas filiformes. Era portadora de TC cervico-toraco-abdomino-pélvico, realizado previamente para estudo de síndrome constitucional, que mostrava "espessamento parietal difuso de praticamente todos os eixos arteriais abrangidos no presente exame, nomeadamente dos vasos supra-aórticos, de toda aorta e seus ramos ilíacos comum, achados que sugerem vasculite com atingimento dos vasos de grande calibre". Foi internada para estudo e tratamento. No internamento realizou estudo analítico que mostrou elevação marcada dos parâmetros inflamatórios (PCR 132mg/L e VS 115 mm/1ªh) e anemia; angioTC dos membros superiores que demonstrou "espessamento parietal circunferencial do arco aórtico e dos troncos arteriais supra-aórticos, aspeto mas evidente nas artérias subclávias e particularmente expressivo nas artérias axilares, onde se verifica oclusão quase completa; ecodoppler cervical e temporais que revelou "eixos carotídeos cervicais acessíveis com espessamento concêntrico e hipoecogénico nas artérias carótidas comuns e bifurcação da carótida comum direita, condicionando estenoses luminais de cerca de 40-50% à esquerda e cerca de 30-40% à direita, com aceleração da velocidade de fluxo na carótida comum esquerda, (…) sem sinal do halo nas artérias temporais superficiais (…) e artérias axilares com espessamento vasculítico em toda a sua extensão acessível, condicionando estenoses luminais de pelo menos 75% à esquerda e de pelo menos 80% à direita, ambas com aceleração significativa da velocidade de fluxo." . Foi avaliada por Oftalmologia que não detetou alterações ao exame oftalmológico. Perante hipótese à admissão mais provável de arterite de células gigantes, iniciou terapêutica com metilprednisolona 1g 3 dias e posteriormente 60 mg/dia (1 mg/kg/dia), com melhoria sintomática expressiva e pressão arterial detetável em ambos os membros superiores. Teve alta com esquema de desmame de corticoterapia.
Conclusão: Os autores visam com este caso descrever uma apresentação inicial rara, com oclusão bilateral de vaso de grande calibre, a necessitar de instituição terapêutica célere, e que respondeu à corticoterapia em dose elevada, sem necessidade de procedimentos endovasculares.

249 - SÍNDROME NEFRÓTICO NA DOENÇA MISTA DO TECIDO CONJUNTIVO - UM APARENTE CONTRASSENSO

Filipe Oliveira Pinheiro[1], Pedro Madureira[1], Maria Seabra Rato[1], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal

Introdução: A doença mista do tecido conjuntivo (DMTC) é uma doença do tecido conjuntivo caraterizada pela presença de anticorpos anti-RNP (anti-U1 ribonucleoprotein) e por um conjuntivo de sinais e

sintomas que partilha com outras conectivites, como o lúpus eritematoso sistémico e a esclerose sistémica, entre outras. No entanto, um dos fatores que caratériza a DMTC é a ausência de atingimento renal grave, sendo este considerado um ponto-chave da doença; na verdade, os títulos elevados de anti-RNP parecem ser protetores contra o envolvimento renal, mesmo quando ocorrem noutras conectivites.

Caso Clínico: Doente do sexo feminino, de 32 anos, com diagnóstico de DMTC há cerca de 1 ano, com atingimento articular (artrite), muscular (enzimas musculares elevadas), hematológico (anemia) e cutâneo/vascular (fenómeno de Raynaud + puffy hands), com ANA em título 1/1000 mosqueado e com anti-RNP positivo forte, medicada com prednisolona 5mg/dia. Apresentou em estudo analítico de rotina proteinúria na faixa nefrótica, acompanhada de hematúria microscópica, que motivou a realização de biópsia renal para estudo do quadro. No internamento realizou estudo analítico que mostrou anemia (Hb 11.4g/dL, normocítica), com estudo imunológico e trombótico sem alterações; biópsia renal mostrou: "Biópsia renal com lesões de esclerose segmentar (3/36) e presença de depósitos de tipo imune mesangiais e membranares por imunofluorescência." Iniciou terapêutica com prednisolona 1 mg/kg/dia em esquema de redução de dose progressiva e hidroxicloroquina, com boa resposta clínica, tendo-se associado posteriormente micofenolato de mofetil, com manutenção de resposta e regressão das alterações do sedimento urinário verificadas previamente.

Conclusão: Este caso representa um possível atingimento renal grave, excecionalmente descrito em associação à DMTC. O atingimento renal na DMTC deve ser confirmado com a realização de biópsia renal e exclusão de outras causas, mas é possível e deve ser tratado de forma intensiva, ainda que os anticorpos anti-RNP possam ser protetores contra o desenvolvimento de doença renal grave.

250 - LIPOMA ARBORESCENTE DA SINOVIAL EM DOENTE COM ARTRITE PSORIÁTICA – CASO CLÍNICO

Diogo Guimarães da Fonseca[1], Beatriz Samões[1], Maria Seabra Rato[2], Filipe Oliveira Pinheiro[2], Daniela Santos Oliveira[2, 3], Ana Martins[2], Frederico Rajão Martins[4], Teresa Martins-Rocha[2], Lúcia Costa[2]
[1]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [3]Center for Health Technology and Services Research (CINTESIS),, Faculdade de Medicina da Universidade do Porto, Porto, Portugal, Porto, Portugal, [4]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal

Introdução: O lipoma arborescente da sinovial é uma lesão intra-articular, rara, benigna, caracterizada pela substituição difusa do tecido sinovial por adipócitos maduros, que origina uma proliferação vilosa lipomatosa da membrana sinovial. A sua etiologia é desconhecida, no entanto, tem sido sugerida a associação com a doença articular degenerativa e inflamatória, nomeadamente artrite reumatoide e artrite psoriática, colocando a hipótese de se tratar de um processo reativo. O quadro clínico habitual compreende uma monoartrite crónica, acompanhada de derrame articular com exacerbações intermitentes. O líquido sinovial habitualmente apresenta características macroscópicas normais. A ecografia permite documentar projeções vilosas hiperecogénicas ("frond-like") associadas a derrame articular. A ressonância magnética é, no entanto, o exame diagnóstico principal, tendo como achados patognomónicos a presença de uma massa sinovial de arquitetura vilositária e de isointensidade com a gordura subcutânea. O tratamento recomendado é a sinovectomia cirúrgica, com raríssimos casos de recorrência da patologia.

Caso clínico: Apresenta-se o caso clínico de uma doente do sexo feminino, de 53 anos de idade, com diagnóstico de artrite psoriática há mais de 15 anos, com atingimento poliarticular, sob metotrexato 25mg/semana subcutâneo e certolizumab pegol 400mg subcutâneo quinzenal. Desde o último ano, apresentava gonalgia direita com noção de tumefação recorrente e limitação acentuada da mobilidade. Ao exame objetivo, apresentava franca tumefação do joelho direito, sem calor ou rubor associados, com incapacidade de extensão completa e flexão limitada a 60º. Não apresentava mais nenhuma articulação periférica dolorosa ou tumefacta. Foi realizada ecografia, que revelou extensa hipertrofia sinovial nos recessos supra-patelar e para-patelares associada a projeções vilosas hiperecogénicas e lobuladas, bem como derrame articular moderado. A artrocentese revelou líquido sinovial de aspeto límpido, cor cítrica e viscosidade normal. O exame citológico revelou contagem leucocitária e de polimorfonucleares compatível com líquido inflamatório (30.000/mm3 e 60% respetivamente). A coloração Gram e o exame cultural foram

compatíveis com líquido sinovial amicrobiano. Foi ainda realizada ressonância magnética que demonstrou exuberante sinovite, com proliferação adiposa de arquitetura vilositária. Paralelamente, apresentava erosões ósseas articulares e periarticulares. Foi assumido o diagnóstico de lipoma arborescente da sinovial, sendo referenciada para consulta de Ortopedia, aguardando orientação.

Conclusão: Este caso reporta um diagnóstico raro de lipoma arborescente da sinovial, destacando a sua possível associação a patologias inflamatórias, as suas características imagiológicas típicas e o seu carácter crónico e recidivante. Corrobora outros casos descritos de lipoma arborescente em doentes com artrite psoriática, destacando a possível etiologia reativa.

251 - COVID-19: TRIGGER IMUNOLÓGICO PARA MIOPATIA INFLAMATÓRIA?

Margarida Faria[1], Agna Neto[1], Ricardo Figueira[1], Lídia Teixeira[1], Inês Margarido[2], Daniel Carvalho[1], Mário Rodrigues[1]

[1]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [2]Serviço de Medicina Interna, Hospital Central do Funchal, Funchal, Portugal

As miopatias inflamatórias idiopáticas (IIM) correspondem a um grupo heterogéneo de doenças musculares imunomediadas caraterizadas por diminuição da força muscular proximal e envolvimento frequente de outros órgãos. A infeção pelo vírus Severe Acute Respiratory Coronavírus 2 (SARS-CoV-2) tem demonstrado potencial de desencadear patologias imunomediadas. Descrevemos o caso clínico de um doente que desenvolveu IIM cerca de 2 meses após infeção COVID-19.

Homem de 68 anos com antecedentes de hipertensão arterial e de tabagismo (40 UMA), com história de infeção assintomática pelo SARS-CoV-2 2 meses antes, inicia quadro de mialgias acometendo os membros superiores e inferiores acompanhadas de diminuição da força muscular proximal, o que impedia a abdução acima dos 90º e dificultava a marcha. Apresentava CK 1596 U/L, CK-mB 33 U/L e VS 35 mm/h. Neste contexto, foi internado no serviço de Medicina Interna, tendo iniciado 40 mg/dia de prednisolona (PDN) com melhoria das queixas e consequente alta. No entanto, aquando de descontinuação da corticoterapia, apresentou recidiva das queixas musculares e ainda disfagia de novo, inicialmente para sólidos e depois para líquidos, com posterior reinternamento e transferência para o serviço de Reumatologia. Ao exame objetivo, a salientar ausência de lesões cutâneas ou artrite e diminuição da força muscular, de predomínio proximal (MMT-8 119). Analiticamente, apresentava leucocitose (15.2x103/µL) com neutrofilia (12.4 x103/µL), trombocitose (471 x103/µL) e anemia (10.1 g/dl) microcítica e hipocrómica, VS de 40 mm/h, PCR de 18 mg/L, CK 3860 U/L com isoenzima CK-mB 60 U/L, troponina T 0.185 ng/mL, ANAs positivos (1/160) e anticorpos anti-SRP, anti-NXP2 e anti-Ro52 positivos. No estudo eletromiográfico destacavam-se achados compatíveis com miopatia inflamatória e neuropatia sensitiva axonal ligeira nos membros inferiores. Paralelamente, foram realizados exames de imagem e endoscópicos para pesquisa de neoplasia, e eletrocardiograma e a ecografia transtorácica, não tendo sido identificada qualquer alteração de relevo nestes exames. Apesar da indisponibilidade de realização de biópsia muscular e de RMN cardíaca e muscular, foi assumido o diagnóstico IIM com envolvimento muscular, cardíaco e esofágico. Foi iniciada terapêutica com pulsos intravenosos de metilprednisolona, 1000 mg/dia durante 3 dias, com importante melhoria das queixas álgicas e descida da CK, seguido de PDN 1 mg/kg/dia e azatioprina (AZT) com escalada da dose até 2 mg/kg/dia. Por manutenção das queixas de disfagia, realizou terapêutica com imunoglobulina humana, iv, 1 g/kg/dia durante 2 dias, com resolução da disfagia, melhoria da força muscular (MMT-8 126) e normalização dos valores de CK. Passados 2 meses, encontra-se assintomático, com melhoria progressiva da força muscular, cumprindo fisioterapia, AZT 2 mg/kg/dia e PDN em desmame.

Neste caso clínico, o estudo de um quadro de mialgias e diminuição da força muscular associados a valores de CK elevados, num doente com infeção recente por SARS-CoV-2, conduziu ao diagnóstico de uma miopatia inflamatória. Por sua vez, este diagnóstico num doente idoso e com positividade para o anticorpo anti-NXP2 impôs a pesquisa de uma doença neoplásica associada que, após investigação exaustiva, não foi identificada, embora a investigação se mantenha durante 2 a 3 anos. Atendendo à existência de outros casos na literatura, foi colocada a hipótese de a recente infeção pelo vírus SARS-CoV-2 ter sido o desencadeante imunológico para o desenvolvimento da IIM, ainda que uma relação causal não possa ser demonstrada.

252 - QUANDO AS DOENÇAS DAS CRIANÇAS ATINGEM OS ADULTOS – UMA APRESENTAÇÃO GRAVE DE VASCULITE POR IgA

Filipe Oliveira Pinheiro[1], Pedro Madureira[1], Maria Seabra Rato[1], Diogo Guimarães da Fonseca[2], Ana

Martins[1], Daniela Santos Oliveira[1, 3], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/ Espinho, Vila Nova de Gaia, Portugal, [3]Faculdade de Medicina da Universidade do Porto, Departamento de Medicina e CINTESIS, Porto, Portugal

Introdução: A vasculite por IgA, anteriormente denominada de Púrpura de Henoch-Schonlein, é uma doença que afeta primariamente doentes em idade pediátrica, sobretudo entre os 3 e os 15 anos, sendo muito rara a sua incidência acima dos 20 anos de idade, estimada em 1-5 casos por 100.000 habituantes. Curiosamente, é nos adultos que a doença pode ser significativamente mais grave, sobretudo no que diz respeito ao atingimento renal, com taxas de progressão para doença renal crónica e terminal superiores às das crianças, e gastrointestinal, com inflamação do intestino delgado marcada que pode causar dor abdominal intensa e hemorragias abundantes.
Caso Clínico: Doente do sexo masculino, de 66 anos, com antecedentes de pancreatites litiásicas de repetição e psicose delirante crónica com ideação de ciúme e défice cognitivo ligeiro, que recorreu ao SU por quadro com 12 horas de evolução de dor abdominal, diarreia e exantema purpúrico; à avaliação médica inicial apresentava um desconforto ligeiro à palpação abdominal, com lesões purpúricas dispersas pelos membros. Realizou estudo analítico que não evidenciou alterações de relevo, à exceção de proteinúria discreta em exame sumário de urina, de 0.30g/L; ecografia abdominal que mostrou "segmento de delgado de paredes espessadas e estratificadas, com fluxo parietal presente, sem distensão luminal anómala, podendo traduzir alteração inflamatória (infeção/doença inflamatória intestinal/"ansa sentinela" no contexto de pancreatite?)". Neste contexto foi decidido estudo por angio-TC abdominal que mostrou "espessamento parietal estratificado e captante de segmento ileal localizado no quadrante inferior direito, com cerca de 25 cm de extensão, com algum edema do respectivo meso, alterações que mais provavelmente estão em relação com processo inflamatório". Durante a permanência no SU desenvolveu quadro de oligoartrite dos joelhos e dor abdominal intensa, com hematoquézias abundantes, tendo sido avaliado por Reumatologia e internado neste contexto. No internamento a destacar inicialmente quadro de dor abdominal intensa, sem resposta satisfatória a analgesia, e manutenção de hematoquézias. Realizou estudo imunológico sem alterações, estudo bacteriológico e parasitológico de fezes negativo, colonoscopia com áreas focais de eritema com biópsia a demonstrar apenas alterações inflamatórias inespecíficas; foi evidenciada hematúria e proteinúria nefrótica (9g/24h) que motivaram o estudo por biópsia renal, que mostrou necrose fibrinóide, fibrose intersticial e atrofia tubular ligeiras, com estudo por imunoflorescência indireta a demonstrar positividade membranar, mesangial e glomerular para IgA, compatível com nefropatia de IgA. Iniciou terapêutica com metilprednisolona 500mg 3 dias e uma perfusão de ciclofosfamida com reversão quase completa do quadro clínico e alterações laboratoriais. Teve alta com o diagnóstico de Púrpura de Henoch-Schonlein do adulto com envolvimento articular (oligoartrite + tenossinovite), cutâneo (púrpura palpável), intestinal (dor abdominal + diarreia sanguinolenta + espessamento ileal) e renal (hematúria + proteinúria nefrótica). Após 18 meses de seguimento, encontra-se em remissão da doença, sem registo de recidiva até ao momento.
Conclusão: Os autores realçam a importância do adequado reconhecimento deste diagnóstico na idade adulta, uma vez que a doença assume uma maior gravidade, com a necessidade mais frequente de terapêutica imunossupressora e citotóxica agressiva, e cujas sequelas para o doente podem ser críticas.

254 - OVERLAPS EM REUMATOLOGIA – UM DESAFIO NO DIAGNÓSTICO!

Ana Aguiar[1], Rita Fonseca[2]
[1]Serviço de Patologia Clínica, Centro Hospitalar Entre Douro e Vouga, Aveiro, Portugal, [2]Rheumatology Department, Centro Hospitalar Entre Douro e Vouga, Aveiro, Portugal

Introdução: As síndromes de sobreposição em Reumatologia são um grupo diverso que inclui doentes com manifestações clínicas bem definidas de várias doenças do tecido conjuntivo. Por vezes o diagnóstico constitui um desafio pela heterogeneidade de apresentações clínicas.
Caso-clínico: Mulher de 49 anos sem antecedentes relevantes, encaminhada à consulta de Reumatologia por poliartralgias das mãos de ritmo inflamatório, rigidez matinal superior a uma hora e Raynaud de início recente.

Ao exame físico, de salientar poliartrite das mãos, acrocianose e ligeira esclerodactilia.

Analiticamente destacava-se: elevação de velocidade de sedimentação (VS) (68 mm/1ªh) e da proteína C reativa (PCR) (11.5 mg/L), fator reumatoide positivo, anti-CCP negativo, consumo de complemento (C4 com C3 normal), electroforese das proteínas séricas com aumento policlonal das imunoglobulinas, anticorpos anti-nucleares e anticorpos contra antigénios extraíveis do núcleo (anti-ENA) ambos negativos.

A radiografia das mãos não mostrou erosões. A capilaroscopia do leito ungueal evidenciou padrão esclerodérmico em fase activa.

Foram pedidas as criglobulinas, que se revelaram positivas para o tipo IgA, IgG e IgM, com características policlonais e com frações monoclonais com atividade factor reumatóide. As serologias para os vírus da hepatite B, C e imunodeficiência humana (HIV) foram negativas.

Perante esta investigação diagnóstica, tínhamos uma doente com poliartrite, fator reumatóide positivo, crioglobulinemia mista e clínica muito sugestiva de esclerose sistémica, contudo com estudo imunológico negativo.

Após alguns dias, a doente volta à consulta, com queixas de fraqueza muscular. No novo estudo analítico verificou-se elevação da creatina cinase (CK 2029U/L), mioglobina (805.7 ng/mL) e aldolase (49.8U/L).

Realizou-se biópsia muscular que mostrou infiltrados inflamatórios mononucleares preferencialmente localizados ao perimísio e em posição perivasculares, identificando-se também pequenos infiltrados localizados ao endomísio. Os infiltrados inflamatórios descritos eram compostos por população mista de linfócitos T CD4 e CD8, linfócitos B e macrófagos.

O painel de immunoblotting para as miosites revelou positividade para o anticorpo anti-Ku.

Estes novos dados, permitiram confirmar a presença de síndrome de sobreposição esclerose sistémica e polimiosite com crioglobulinemia mista secundária.

A paciente iniciou tratamento com prednisolona 1 mg/kg e imunoglobulina humana endovenosa (2g/kg dividido em 4 dias durante 6 meses), observando-se melhoria clínica. Após este período, suspendeu a imunoglobulina e iniciou metotrexato. Atualmente encontra-se assintomática, CK normal e crioglobulinas negativas.

Conclusão: O caso apresentado sobressai pela raridade e pelo extremo desafio diagnóstico que constituiu. Perante um doente com artrite e fator reumatóide positivo, com restante imunologia negativa, facilmente se assumiria o diagnóstico de artrite reumatóide. Contudo, a presença de outras manifestações clínicas e analíticas altamente sugestivas de outras patologias deve sempre criar a suspeição da presença de sindromes de sobreposição. Neste caso a abordagem multidisplinar entre a reumatologia e a patologia clínica foram cruciais para o diagnóstico final que permitiu um tratamento atempado e com sucesso.

259 - SHRINKING LUNG SYNDROME (SLS): A RARE MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS

Vanessa Fraga[1], Ana Catarina Duarte[1], Sandra Sousa[1], Maria José Santos[1, 2]

[1]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introduction: Shrinking lung syndrome (SLS) is an uncommon pulmonary manifestation of systemic lupus erythematosus (SLE), characterized by progressive dyspnea, chest pain, elevated hemidiaphragm and a restrictive pattern on pulmonary function tests, without evidence of interstitial disease or significant pleural disease on chest computed tomography (CT).
Case presentation: A 21-year-old Caucasian female with a 6-year history of SLE with renal and articular involvement treated with mycophenolate mofetil (750+500mg/day), was admitted with fever, shortness of breath, fatigue, right-sided pleuritic chest pain, intermittent arthralgias, increased hair loss and persistent sinus tachycardia, worsening over the last 2 months. Approximately 5 months before she had a lupus flare, manifested by right-sided pleural effusion, low C4 and elevated anti-dsDNA that was treated with glucocorticoids (prednisolone 40mg/day) and increased immunosuppression (mycophenolate mofetil 2g/day) with complete clinical recover and radiological remission. However, elevated right hemidiaphragm persisted.

Two months before admission she was diagnosed with pulmonary thromboembolism and deep vein thrombosis and started oral anticoagulants. Nevertheless, shortness of breath on exertion and sinus tachycardia worsened and new-onset fever developed. Antiphospholipid antibodies were persistently negative.

At admission, she was normotensive, tachycardic

(heart rate 150bpm), febrile, with polypnea in room air (SpO2 96% at rest and 88% on exertion). Breath sounds were absent bilaterally over the posterior basal chest wall. Chest X-ray showed normal lung parenchyma and elevated right hemidiaphragm. The pulmonary function tests showed markedly decreased lung volumes with a restrictive pattern (FEV1 17.9% FVC 16.2% FEV1/CVF 96%; unable to perform DLCO).

A thorough investigation, which included blood and urine cultures, thoraco-abdomino-pelvic CT scan, echocardiography, PET-scan, bone marrow biopsy, lymph node biopsy, ventilation/perfusion scintigraphy, excluded "de novo" pulmonary thromboembolism, lung parenchyma/pleural involvement, malignancy and infectious diseases. The diagnosis of SLS was made and the patient treated accordingly with glucocorticoids and rituximab.

Conclusion: SLS is a rare complication of SLE scarcely described in medical literature and probably underdiagnosed.

The presence of dyspnea, decreased lung volumes with normal lung parenchyma, and elevated diaphragm in SLE patients should alert clinicians for the diagnosis of SLS.

Despite the optimal therapy for SLS being unknown, glucocorticoids and immunosuppressive therapy, such as rituximab, seem to improve both symptoms and pulmonary function.

260 - UMA CAUSA RARA DA ELEVAÇÃO DA CREATINA FOSFOQUINASE: A IMPORTÂNCIA DO EXAME FÍSICO

Hugo Parente[1], Catarina Soares[1], Francisca Guimarães[1], Soraia Azevedo[1], Carmo Afonso[1], Filipa Teixeira[1]

[1]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal

Introdução: A miosite de causa infecciosa é incomum, estando frequentemente associada a infecção contígua, trauma penetrante, cirurgia, corpos estranhos, insuficiência vascular/isquemia ou disseminação hematogénea. O curso clínico e o grau de cronicidade dependem da subjacente etiologia, sendo a bacteriana tipicamente mais focal, ao invés da vírica e parasitária, mais difusas e com mialgias generalizadas. A imunossupressão, a doença hepática crónica, a infecção pelo vírus da imunodeficiência humama (HIV) e a diabetes mellitus tipo 2 (DM2) associam-se a uma maior incidência de piomiosite, definida como uma infecção intramuscular aguda secundária a disseminação hematogénea. Surge, na sua maioria, por *Staphyloccocus aureus*[2] e menos comummente, por bactérias gram negativas. A musculatura mais afectada engloba os quadricípites e os glúteos, possivelmente por um atingimento mais extenuante do exercício físico, a este nível. A patogénese implícita pressupõe bacteriémia transitória (assintomática) necessariamente associada a lesão muscular (por exemplo, sobreuso), sendo as hemoculturas positivas em apenas 5-35% dos casos.

Objetivo: Apresentação de um caso de miosite infecciosa bacteriana.

Homem de 69 anos, com antecedentes de DM2, enviado a consulta de Reumatologia por febre e elevação da creatina fosfoquinase (573 UI/L). Na avaliação em consulta de Reumatologia, apenas havia a salientar tumefacção na região mamária direita onde, posteriormente, iniciou dor e sinais inflamatórios locais. Não havia história conhecida de infecção prévia, uso de drogas injectáveis, trauma ou cirurgia local. Na investigação diagnóstica, foi constatada elevação da VS e PCR, e realizada ecografia que demonstrou edema muscular do grande peitoral, compatível com miosite e alterações inflamatórias na articulação condro-costal. A tomografia axial computorizada (TAC), confirmou processo inflamatório centrado à articulação condro-esternal com envolvimento do 4º arco costal anterior direito, e extensão aos tecidos moles adjacentes, incluindo músculo grande peitoral, transverso torácico e ligamento esternocostal radial. Fez inicialmente tratamento anti-inflamatório, com melhoria parcial, mas agravamento posterior com drenagem purulenta. Foi colhida zaragatoa do exsudado purulento, que demonstrou a presença de *Escherichia coli* multissensível. Realizou-se biópsia local incisional para exame histológico, estudo bacteriológico, micobacteriológico e micológico, tendo confirmado o patogéneo isolado em zaragatoa prévia. Não se identificaram fungos ou micobactérias. As serologias para HIV e os vírus de hepatite B e C foram negativas, assim como o estudo de etiologia paraneoplásica. O doente foi medicado com os antibióticos flucloxacilina e ciprofloxacina por 3 meses, com resolução clínica e analítica do quadro.

Discussão/conclusão: Este caso evidencia uma patologia rara, de uma etiologia bacteriana menos típica, sem inequívoco factor desencadeante e com atingimento destructivo numa localização pouco comum. A sintomatologia inflamatória, a febre, a elevação de

enzimas musculares, o antecedente de DM2 e a imagiologia explícita definiram um percurso diagnóstico que culminou na imprescindível documentação bacteriológica do agente envolvido. A pesquisa por estados de imunodepressão e neoplasia são essenciais.

268 - ENVOLVIMENTO CARDÍACO NAS MIOPATIAS INFLAMATÓRIAS: SÉRIE DE CASOS

Laura Gago[1], Maria Helena Lourenço[1, 2], Ana Bento da Silva[1, 2], Rita Pinheiro Torres [1, 2], Ana Lúcia Fernandes[3], Nuno Pina Gonçalves[1, 2, 4], Maria João Gonçalves[1], Ana Filipa Mourão[1, 2], Maria Manuela Costa[1], Jaime C. Branco[1, 2]

[1]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [2]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [3]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, Faro, Portugal, [4]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal

Introdução: As miopatias inflamatórias caracterizam-se por fraqueza muscular proximal, aumento de enzimas musculares, anormalidades na eletromiografia e biópsia muscular. Pode também existir envolvimento pulmonar e cardíaco. Descrevem-se 3 casos clínicos de doentes com miopatias inflamatórias com envolvimento miocárdico.

Caso clínico 1: Mulher, 49 anos, saudável. Apresentava quadro de fraqueza muscular proximal associada a disfagia para sólidos e fadiga com limitação funcional grave (Manual muscle testing – MMT 27) com 2 semanas de evolução. Analiticamente, apresentava creatinina quinase (CK) 12300 U/L, mioglobina 6456ug/dL, lactato desidrogenase (LDH) 2245 U/L, troponina T 2639 ng/L, proteína C reativa (PCR) 2 mg/dL e anticorpo anti-SRP positivo forte. Os exames complementares de diagnóstico eram compatíveis com miopatia necrotizante imunomediada. A Ressonância Magnética (RM) cardíaca apresentava alterações compatíveis com miocardite. Foi medicada com imunoglobulina humana intravenosa (IvIg) e metilprednisolona 1000 mg durante três dias, posteriormente iniciou micofenolato de mofetil na dose de 1000mg de 12/12h e por manter um quadro clínico com alguma gravidade iniciou-se rituximab (RTX) uma semana depois. Apresentou agravamento do quadro com insuficiência respiratória tipo I e disfagia grave para sólidos e líquidos com necessidade de internamento em unidade de cuidados intensivos, de onde teve alta cerca de 2 meses depois, melhorada clinicamente.

Caso clínico 2: Homem, 30 anos, previamente saudável, com quadro de disfagia para sólidos, disfonia, fraqueza muscular proximal, rash heliótropo periorbitário, cervical, torácico e dorsal com dois meses de evolução, com grande incapacidade funcional associada (MMT 88). Analiticamente apresentava: aspartato aminotransferase (AST) 215 U/L; alanina aminotransferase (ALT) 101 U/L; CK 2588 U/L; Mioglobina 505 U/L; LDH 647 U/L; troponina T 364 ng/L; anticorpo anti-NXP2 positivo. Os exames complementares de diagnóstico e a clínica eram compatíveis com dermatomiosite. A RM cardíaca não apresentava miocardite aguda, porém existiam duvidas quanto à existência de lesões de miocardite subaguda. Apresentava também derrame pericárdico de localização inferior. Iniciou-se tratamento com IgIV 2g/kg durante 5 dias e RTX, com franca melhoria do quadro clinico.

Caso clínico 3: Sexo feminino, 55 anos, saudável, com quadro de 7 meses de evolução de poliartralgias, fraqueza muscular proximal, dispneia e toracalgia associadas a hiperpigmentação dos cotovelos e lesões esfoliativas das mãos. Ao exame objetivo era evidente poliartrite, sinal de Gottron nos cotovelos, esboço de mãos de mecânico e um MMT15 135/150. Analiticamente apresentava AST 275 U/L; ALT 269 U/L; CK 8248 U/L; mioglobina 1218 ug/L; LDH 633 U/L; troponina T 1376 ng/L; anticorpo anti-Jo1 positivo. Os exames complementares de diagnóstico e a clínica eram compatíveis com Síndrome Anti-Sintetase. Na RM era evidente miopericardite em fase subaguda e ligeiro derrame pericárdico (3 mm). Iniciou terapêutica com IvIg 2g/kg durante 5 dias, prednisolona 60 mg e metotrexato 25 mg. Irá iniciar tratamento com RTX.

Discussão/Conclusão: O envolvimento cardíaco nas miosites inflamatórias é fator de mau prognóstico, sendo a principal causa de morte. Sabe-se que a idade, anticorpos anti-nucleares positivos, ratio AST/CK e a presença de doença pulmonar intersticial são fatores preditores de envolvimento cardíaco. É importante o diagnóstico precoce para que o tratamento seja adaptado, muitas vezes com necessidade de otimizar a terapêutica imunossupressora.

271 - DERMATOMIOSITE PARANEOPLÁSICA ANTI-MDA5/CADM-140 POSITIVA – CASO CLÍNICO

Diogo Guimarães da Fonseca[1], Beatriz Samões[1], Tiago

Beirão[1], Joana Aleixo[1], Romana Vieira[1], Taciana Videira[1], Flávio Campos Costa[1], P Pinto[1]
[1]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Introdução: A dermatomiosite (DM) é uma doença do tecido conjuntivo que habitualmente se caracteriza por manifestações cutâneas clássicas, tais como pápulas/placas de Gottron, rash heliotropo, sinal do xaile, sinal de Holster, telangiectasias periungueais e "mãos de mecânico". Adicionalmente os doentes apresentam miosite, que se caracteriza por défice de força muscular de predomínio proximal e simétrico. Existem diversos auto-anticorpos específicos associados às miopatias inflamatórias, sendo que a sua identificação tem importância prognóstica. No que concerne ao prognóstico do doente, um dos principais fatores a ter em consideração é o risco paraneoplásico, sendo que os doentes com DM têm maior risco de neoplasia associada, que pode atingir os 30% dos casos. Este risco encontra-se particularmente aumentado nos primeiros anos após o diagnóstico, e as neoplasias frequentemente associadas são do pulmão, ovário, mama, colón e linfoma não-Hodgkin.
Caso clínico: Sexo feminino, de 59 anos de idade, encaminhada para consulta de Reumatologia por síndrome constitucional, com perda de 10kg nos últimos 3 meses e mialgias de predomínio proximal. Ao exame objetivo apresentava livedo reticular dos membros inferiores persistente, pápulas de Gottron, rash do decote, sinal do xaile, sinal de Holster bilateral, telangiectasias faciais e "mãos de mecânico". Descrevia um fenómeno de Raynaud e apresentava ainda défice de força muscular de predomínio proximal nos membros superiores e inferiores. Analiticamente apresentava elevação dos reagentes de fase aguda, enzimas musculares normais e anticorpos anti-nucleares 1:160 com padrão homogéneo. A eletromiografia revelou sinais de processo miopático em curso nos músculos proximais com ondas lenta e fibrilações. O estudo imunológico dirigido revelou a presença de anticorpos anti-antigénio muscular específico, mais concretamente anti-MDA5/CADM-140, tendo-se estabelecido o diagnóstico de dermatomiosite. A doente iniciou corticoterapia 1mg/kg/dia de prednisolona com redução subsequente da dose e hidroxicloroquina 400mg/dia. Devido à possível associação a um quadro neoplásico, foi realizado estudo dirigido, tendo sido evidenciado em exame tomográfico lesões nodulares peritoneais, suspeitas de carcinomatose. O estudo de neoplasia primária evidenciou tratar-se de um adenocarcinoma seroso do ovário com metastização hepática, peritoneal e do cólon sigmoide. Foi encaminhada para oncologia e está sob quimioterapia paliativa.
Conclusão: Este caso reporta um diagnóstico de DM paraneoplásica, anti-MDA5/CADM-140 positivo, realçando a importância de exclusão de neoplasia oculta associada ao diagnóstico recente de DM. Os autores destacam ainda a particularidade de a doente não apresentar um autoanticorpo habitualmente associado a um quadro paraneoplásico e a evidência de padrão miopático mas sem elevação das enzimas musculares, descrito na literatura como sendo frequente na DM paraneoplásica ao contrário da DM clássica.

272 - DERMATOMIOSITE AMIOPÁTICA COM CALCINOSE SUBCUTÂNEA EXTENSA - CASO CLÍNICO

Diogo Guimarães da Fonseca[1], Beatriz Samões[1], Tiago Beirão[1], Flávio Campos Costa[1], Joana Aleixo[1], Taciana Videira[1], Romana Vieira[1], P Pinto[1]
[1]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Introdução: As miopatias inflamatórias idiopáticas, são um grupo raro de doenças imuno-mediadas, que se caracterizam por défice de força muscular proximal e simétrico. Apresentam envolvimento extra-muscular característico, e dividem-se em 3 subtipos principais: dermatomiosite (DM), polimiosite (PM) e miosite por corpos de inclusão. A DM amiopática é um subtipo raro, que se caracteriza pela presença das manifestações cutâneas clássicas por mais de 6 meses, na ausência de sinais clínicos ou laboratoriais de miosite. Habitualmente associa-se à presença de anticorpo específico, anti-MDA5/CADM-140. A calcinose subcutânea pode ser encontrada em algumas doenças do tecido conjuntivo, entre as quais a DM, contudo é habitualmente associada à DM de início em idade juvenil.
Caso clínico: Apresenta-se o caso clínico de um indivíduo do sexo masculino, de 36 anos de idade, encaminhado para consulta de Reumatologia por poliartralgias de ritmo inflamatório. Ao exame objetivo apresentava rarefação capilar, pápulas cicatriciais sobre tatuagens existentes, rash heliotropo, rash do decote, sinal de Holster à direita, pápulas de Gottron, telangiectasias periungueais e úlceras digitais. Descrevia um fenómeno de Raynaud e apresentava ainda

poliartrite com envolvimento simétrico das metacarpofalângicas e joelhos. Não apresentava qualquer dor à palpação das massas musculares ou défice de força muscular. Analiticamente apresentava elevação dos reagentes de fase aguda, enzimas musculares normais e anticorpos anti-nucleares <1:160. A eletromiografia não evidenciou qualquer sinal de miopatia e a ressonância magnética das coxas não evidenciou lesões inflamatórias de miosite ou áreas de atrofia adiposa. O estudo imunológico dirigido revelou a presença de anticorpos anti-antigénio muscular específico, mais concretamente anti-MDA5/CADM-140 tendo-se concluído por Dermatomiosite amiopática. Foi iniciada prednisolona 10mg/dia e hidroxicloroquina 400mg/dia, com posterior desenvolvimento de rash eritematoso generalizado, não puriginoso, o que levou à sua suspensão. O doente foi medicado com imunoglobulina endovenosa 2mg/kg mensal durante 6 meses mas dado que mantinha as lesões cutâneas foi decidido alterar para azatioprina titulada até 1.5mg/kg/dia, com resolução das queixas cutâneas e articulares. Cerca de 6 meses após o diagnóstico, o doente desenvolve lesões subcutâneas de consistência pétrea, nodulares da parede abdominal, coxas, cotovelos, antebraço distal e dedos, com tradução radiográfica compatível com calcinose subcutânea. Optou-se por associar pamidronato 60mg endovenoso, inicialmente trimestral e com posterior alargamento para semestral. O doente apresentou uma melhoria clínica, com regressão parcial das lesões de calcinose.

Conclusão: Este caso reporta um diagnóstico raro de DM amiopática, realçando a sua associação com uma apresentação cutânea ampla, típica da DM clássica, mas sem qualquer evidência de atingimento muscular. A identificação dos diversos tipos de auto-anticorpos específicos das miosites inflamatórias, veio igualmente permitir um diagnóstico mais preciso e identificar diferentes subtipos, entre os quais o anticorpo anti-MDA5/CADM-140, associado a esta entidade rara. Destaca-se ainda o surgimento posterior de múltiplos focos de calcinose subcutânea, uma característica ocasionalmente encontrada na DM do adulto, mas muito mais comum na DM juvenil, o que torna este caso ainda mais relevante do ponto de vista clínico.

273 - MYOSITIS OSSIFICANS – AN UNFAMILIAR DIAGNOSIS

Joao Paulo Vilas-Boas[1], Carolina Mazeda[1], Susana P. Silva[1], Barcelos A.[1], Graca Costa[1], Maria Céu Portelada[1]

[1]Rheumatology Department, Centro Hospitalar Entre Douro e Vouga, Aveiro, Portugal

Clinical Case: A 39 years old female patient, without previous relevant past medical history, was referred from the primary care to our rheumatology outpatient center due to sudden intense pain in the right thigh associated with erythema and increased temperature in a well circumscribed area. The symptoms were present in the last two weeks, making the patient unable to walk properly. She denied previous local trauma or unusual physical activity. No other osteoarticular or systemic symptoms such as fever were reported. Physical examination showed a palpable tender hardened mass in the right thigh, with increasing pain associated with internal hip rotation and knee extension. Analgesics and NSAIDs were only moderately successful in reducing the pain intensity. Blood analysis was unremarkable. She maintained analgesic therapy with NSAIDs and in the next 2 weeks after the first evaluation, the pain decreased slowly in intensity allowing the patient to return to her daily activities despite a slight limitation in her left knee extension. A MRI scan was performed (image 1 and 2), showing a significant area of muscle edema in the distal component of the right vastus medialis with a central ovaloid well-defined mass (3x3 cm). The diagnosis of myositis ossificans was made.

Discussion: Myositis Ossificans is a self-limiting pseudo-inflammatory benign ossifying lesion that originates from the skeletal muscle. It is most found in as a solitary lesion in a large skeletal muscle such as the quadriceps. The etiology and potential predisposing factors of non-hereditary myositis ossificans remains unclear. Possible risk factors include history of trauma, burns, coagulation disorders or paraplegy. In most cases no causative factor can be identified. The clinical presentation is variable, but usually presents initially with a 2-4 weeks painful phase with local inflammatory signs and decreased limited range of motion of the adjacent joint. Later, as the lesion matures a circumscribed soft tissue-mass becomes apparent on physical examination with decreased pain and swelling. CT scan and MRI allow for a definitive diagnosis, however it may be difficult in the early stages. Differential diagnosis includes benign lesions such as abscess or malignant conditions like osteosarcomas.

Myositis ossificans is a self-resolving process. A brief period with rest, ice and NSAIDs use is recom-

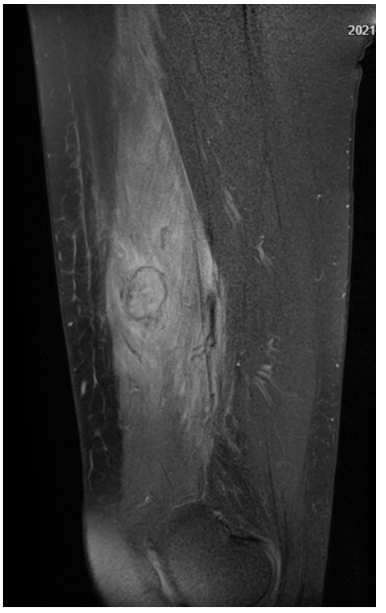


FIGURE 1. LONGITUDIUNAL VIEW (T2 SFAT)

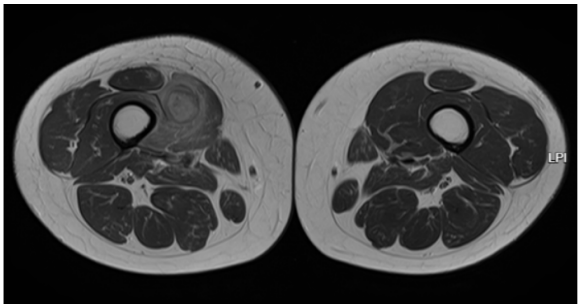


FIGURE 2. TRANSVERSAL VIEW (FSE)

mended. Physical therapy rehabilitation is considered if joint pain and stiffness persists.

274 - ESCLEROSE SISTÉMICA E CIRROSE BILIAR PRIMÁRIA: EVOLUÇÃO NO PERÍODO PÓS-TRANSPLANTE HEPÁTICO

Ana Margarida Correia[1], Emanuel Costa[1], Diogo Esperança Almeida[1], Joana Leite Silva[1], Joana Sousa-Neves[1], Ana Ribeiro[1], Marcos Cerqueira[1]

[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

Introdução: A esclerose sistémica (ES) é uma doença do tecido conjuntivo multi-sitémica, de etiologia não esclarecida, caracterizada pela presença de auto-anticorpos específicos e por desenvolvimento de fibrose e vasculopatia em múltiplos órgãos. A cirrose biliar primária (CBP) é a forma de acometimento hepático mais comum em doentes com ES, correlacionando-se com o subtipo ES cutânea limitada e a presença do anticorpo (Ac) anti-centrómero. A associação ES e CBP é denominada por Síndrome de Reynolds.

Caso clínico: Apresenta-se o caso clínico de uma doente do sexo feminino com 47 anos de idade, seguida em CE de Reumatologia desde 2013 com o diagnóstico de ES forma cutânea limitada em associação com CBP; clinicamente com fenómeno de Raynaud, telangiectasias malares, espessamento cutâneo (face, antebraço e pernas), com hiperpigmentação cutânea marcada dispersa por todo o tegumento, lesões ulceradas nos membros inferiores, artralgias de pequenas articulações das mãos e disfagia; analiticamente com Ac anti-nucleares positivos (1/1280), Ac anti-centrómero (Cenp-B) e Ac anti-mitocondrial também positivose e elevação marcada da fosfatase alcalina e da gama glutamil transferase. A capilaroscopia revelava padrão esclerodérmico com áreas avasculares, presença de megacapilares e hemorragias peri-ungueais. Em termos de terapêutica, foi inicialmente medicada com hidroxicloroquina, que suspendeu por agravamento da hiperpigmentação cutânea; posteriormente, pela patologia hepática, iniciou azatioprina, entretanto suspensa por ineficácia; por último, realizou terapêutica com micofenolato de mofetil (MMF), com resposta significativa ao nível cutâneo, mas com necessidade de suspensão por intolerância gastrointestinal. Ao longo do seguimento, apresentou agravamento progressivo do quadro hepático, com refratariedade terapêutica ao ácido ursodesoxicólico, evoluindo para cirrose hepática com icterícia e prurido marcados, hipertensão portal e varizes esofágicas, com várias complicações associadas, nomeadamente descompensação edemato-ascítica e consequente anasarca, e ainda hemorragia por ruptura de varizes a condicionar anemia com necessidade de suporte transfusional. Por falência a shunt portossitémico intra-hepático transjugular, foi encaminhada para consulta de transplante hepático, ao qual acabou por ser submetida em setembro de 2020, sem intercorrências de relevo, estando atualmente medicada com MMF 1g/dia, tacrolimus 5 mg/dia e prednisolona 5 mg/dia, com boa tolerância. No período pós-transplante, 6 meses após o mesmo, apresentava notória melhoria das alterações cutâneas associadas à ES, nomeadamente completa cicatrização das úlceras que apresentava nos MI's e melhoria significativa da hiperpigmentação e do espessamento cutâneos.

Conclusão: O caso apresentado pretende alertar

para a melhoria clínica significativa do envolvimento cutâneo, numa doente com ES e CBP, observada no período pós-transplante hepático. Tal melhoria pode dever-se ao uso de MMF como terapêutica imunossupressora para prevenção da rejeição, fármaco que tem revelado benefício nas manifestações cutâneas da ES, e ao qual a doente já teria demonstrado resposta previamente ao transplante. Contudo, não se podem excluir outros mecanismos responsáveis pelos efeitos positivos a nível cutâneo inerentes à própria transplantação hepática, sendo que está descrito que se associa a melhoria de algumas manifestações cutâneas associadas à CBP, como também não se pode excluir o possível efeito de outros fármacos como o tacrolimus.

276 - DOENÇA DE LYME DISSEMINADA PRECOCE EM DOENTE COM SÍNDROME DE SJOGREN PRIMÁRIA: RELATO DE CASO

Beatriz Mendes[1], Adriana Carones[1], Ana Rita Prata[1], Marlene Sousa[1], André Saraiva[1], Cátia Duarte[1, 2], Mariana Luis[1]

[1]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [2]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal

Introdução: No diagnóstico diferencial de um quadro clínico caracterizado por poliartralgias de início agudo e febre é essencial considerar, para além de diversas etiologias infecciosas, as doenças do tecido conjuntivo. É, no entanto, reconhecido que quadros infecciosos podem originar positividade transitória para alguns auto-anticorpos. Por outro lado, estão também descritos falsos positivos para serologias infecciosas na presença de doenças auto-imunes. Tais situações podem ocasionar verdadeiros dilemas de diagnóstico diferencial.

Caso Clínico: Doente do sexo feminino, de 51 anos de idade, sem antecedentes pessoais de relevo e com história familiar de artrite reumatóide. Foi internada no Serviço de Reumatologia por quadro de poliartralgias de ritmo inflamatório, aditivas, simétricas, com envolvimento de pequenas e grandes articulações, com um mês de evolução, associado a febre, odinofagia, hiperémia conjuntival indolor e rash cutâneo maculopapular eritematoso, não pruriginoso, no tronco e nos membros, sintomas com quatro dias de evolução.

À revisão de aparelhos e sistemas destacava-se xerostomia e xeroftalmia com alguns anos de evolução, bem como episódio de parotidite no passado.

Analiticamente apresentava linfopenia, reagentes de fase aguda normais e serologias infecciosas, incluindo para *Coxiella burnetii*, HIV 1 e 2 e hepatite B e C negativas, apresentando, unicamente, positividade por imunoensaio por quimioluminescência para IgM de *Borrelia burgdoferi* sensu latu.

Foi então colocada a hipótese de Doença de Lyme forma disseminada precoce, tendo sido introduzida antibioterapia com doxiciclina oral 100 mg de 12/12h, associada a anti-inflamatório, com melhoria significativa das artralgias, bem como resolução das lesões cutâneas e apirexia sustentada. Foi posteriormente conhecida a positividade para anticorpos antinucleares, num título de 1:1280, padrão nuclear mosqueado fino, bem como para anti-Ro/SSA e anti-La/SSB. Foi adicionalmente realizado teste de Schirmer, o qual se revelou positivo bilateralmente, bem como ecografia das glândulas salivares major, evidenciando heterogeneidade marcada e irregularidade dos bordos glandulares compatíveis com Síndrome de Sjögren. Foi assim assumido o diagnóstico de Síndrome de Sjögren primária e equacionada a possibilidade de falsa positividade para IgM de *Borrelia burgdoferi* por eventual reactividade cruzada. Tal hipótese foi, no entanto, afastada, após positividade do teste confirmatório por Western Blot para IgG e IgM de *Borrelia burgdoferi*. Foram, desta forma, assumidos os diagnósticos concomitantes de Doença de Lyme (forma disseminada precoce) e Síndrome de Sjögren primária.

Conclusão: A doença de Lyme, causada pela espiroqueta *Borrelia burgdoferi*, é uma doença multissistémica que envolve frequentemente o sistema músculo-esquelético. Na fase disseminada precoce são frequentes as queixas de poliartralgias migratórias e mialgias. A síndrome de Sjögren pode igualmente cursar com poliartralgias, geralmente dos punhos e pequenas articulações das mãos. Estão descritas reacções cruzadas entre auto-anticorpos e testes serológicos. Realça-se assim a importância da correlação dos achados laboratoriais com a clínica e da sua valorização quando os sinais e sintomas não podem ser inequivocamente atribuídos a uma única patologia.

283 - PANNICULITIS ASSOCIATED WITH DERMATOMYOSITIS: WHEN TREATMENT IS MORE CHALLENGING THAN THE DISEASE

Carolina Mazeda[1], Susana P. Silva[1], Joao Paulo Vilas-Boas[1], Renata Aguiar[1], Manuela Loureiro[2], Catarina

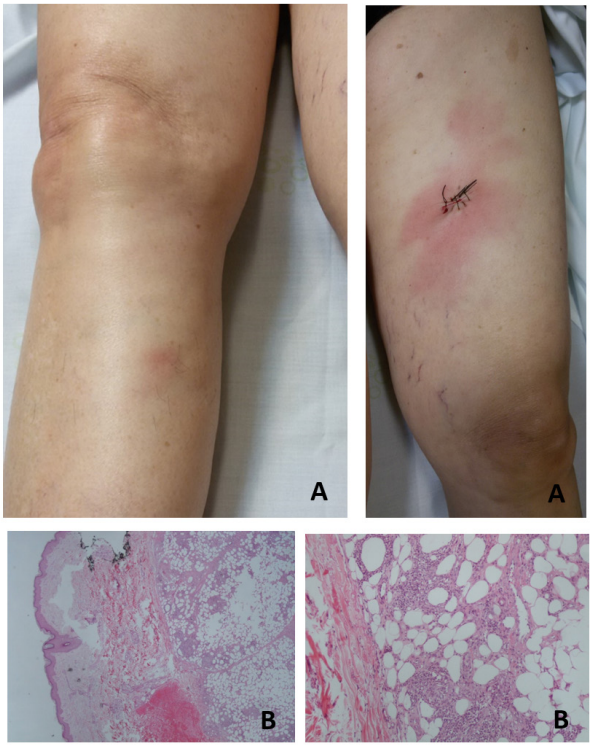
Ambrósio[3], Barcelos A.[1]
[1]Rheumatology Department, Hospital Infante D. Pedro (CHBV), Aveiro, Portugal, [2]Dermatologia, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [3]Instituto de Biomedicina – iBiMED, Universidade de Aveiro, Aveiro, Portugal

Introduction: Panniculitis is a rare disorder characterized by inflammation of the subcutaneous adipose tissue. Clinically characterized by the presence of hard, infiltrative, erythematous, often painful plaques and nodules located mainly in the arms, thighs and buttocks. There are multiple etiologies for MP including surgery, trauma, cancer and less frequently with autoimmune diseases. It was first described in association with dermatomyositis (DM) by Weber and Gray in 1924, but few cases are reported in the literature.

Clinical case: A 61-year-old female patient, retired, autonomous, with a personal history of arterial hypertension and hypothyroidism, being treated with losartan 100 mg and levothyroxine 0.05 µg daily. Initially, the patient had proximal and symmetrical muscle weakness involving upper and lower limbs, shortness of breath, severe asthenia and dysphagia for fluids with XX weeks of evolution. Physical examination was remarkable for violaceous edematous periorbital erythema (heliotrope rash) and presence of violaceous erythematous papules over the dorsal metacarpal and interphalangeal joints of both hands (Gottron's papules). Testing the muscle strength revealed weakness in bilateral shoulder abduction (4/5) and bilateral hip flexion (4/5). Initial investigations revealed elevation in acute phase reactants (VS 68 mm in the 1st hour and PCR 13.43 mg/dL) and creatine kinase at 353 U/L. Immunologic studies showed a positive antinuclear antibody title (1:320, speckled pattern) by immunofluorescence. High-resolution chest computed tomography (HRCT) scan revealed alterations suggestive of cryptogenic organizing pneumonia. Electromyography and muscle biopsy was performed, which showed results consistent with the diagnosis of DM. The workup for malignancy was negative. She was treated with deflazacort oral 1mg/kg/day (given being allergic to prednisolone/prednisone and methylprednisolone), followed by 6 monthly intravenous pulses of cyclophosphamide (1000 mg) with clinical improvement. Deflazacort was tapered and she started azathioprine as steroid-sparing agent. For two and a half years, patient was stable, when new lesions appear with erythematous, hard, infiltrative, and painful plaques and nodules on the anterior surface of both thighs and legs (Figures 1A). Skin biopsy showed "dermo-hypodermis with lobular predominance, without significant vasculitis or necrosis and that is fit for panniculitis of DM "(Figure 1B). Treatment was increased to 30 mg of deflazacort and 150 mg of azathioprine daily. Azathioprine treatment was discontinued three months later secondary to hepatic toxicity. As an alternative steroid-sparing agent, cyclosporin was started after discussion at a multidisciplinary meeting with Dermatology and Pneumology. Patient is currently on cyclosporin A 150 mg/day oral and deflazacort 6 mg/day, with remission of the dermatological lesions.

Conclusion: Panniculitis is an uncommon manifestation of DM and symptoms may occur before, concurrently with or after the diagnosis of DM. There are no defined therapeutic guidelines, the treatment of these cases relies on the off-label use of drugs, given the scarce number of patients. The first-line approach focuses on the use of corticosteroids and immunosuppressants. Beyond diagnosis, a big challenge was also the treatment in an allergic patient to prednisolone/prednisone.

FIGURE 1 - PANNICULITIS IN THE THIGHS (A) AND HISTOLOGY OF CUTANEOUS BIOPSY (B).



289 - TRÊS EXEMPLOS DE IMITADORES DA ESCLEROSE SISTÉMICA.

Emanuel Costa[1], Diogo Esperança Almeida[1], Ana Margarida Correia[1], Marcos Cerqueira[1], Joana Leite Silva[1], Ana Ribeiro[1], Joana Sousa-Neves[1]
[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

Introdução: O espessamento cutâneo e o edema global das mãos são manifestações inespecíficas de diversas patologias nas quais se inclui a esclerose sistémica. O diagnóstico diferencial é essencial para a correta orientação clínica.

Caso clínico 1: Doente do sexo feminino, de 45 anos com antecedentes de dois episódios de taquicardia supraventricular de causa não estabelecida. Apresenta queixas de noção de edema das mãos desde há dois anos com rigidez de curta duração e artralgia esporádica de ritmo mecânico. Negava história de episódios de Raynaud ou úlceras digitais.

Do exame objetivo assinala-se a presença de edema global das mãos com dificuldade na flexão total dos dedos. Dos seus antecedentes familiares, história de doença de Fabry em dois irmãos e filho (todos do sexo masculino).

O resultado da determinação de atividade enzimática de alfa-galactosidase A foi inconclusivo pela existência de deficiência parcial. O estudo prosseguiu com a sequenciação do gene GLA onde foi detetada a presença, em heterozigotia, da variante genética c. 427G>A(p.A143T) cujo papel na patogénese na doença de Fabry não é, ainda, consensual.

Caso clínico 2: Mulher, de 27 anos com queixas com dois meses de evolução, de edema periférico dos membros, simétrico, distal ao joelho e cotovelo de aparecimento súbito e agravamento progressivo, por vezes com pápulas eritematosas pruriginosas. Afirmava, desde o início do quadro, temperatura subfebril (37-38ºC) e mialgias generalizadas. Negava história de episódios de Raynaud ou úlceras digitais.

Ao exame objetivo, apresentava non-pitting edema e endurecimento global dos membros com presença de Groove sign.

A avaliação analítica revelou leucocitose (26 300/µL) com eosinofilia 33% (8 700/µL), trombocitose (650 000/µL), hipergamaglobulinemia com pico monoclonal IgG lambda na imunoelectroforese de proteínas do soro, aumento dos parâmetros inflamatórios com VS 58 mm/h e PCR 18.10 mg/L. Os anticorpos antinucleares foram positivos em título 1/320 sem especificidade ENA.

Na biópsia cutânea profunda, entre o tecido celular subcutâneo e o músculo esquelético, representação de fáscia que se encontrava envolvida por numerosos polimorfonucleares eosinófilos, confirmando o diagnóstico clínico de fasceíte eosinofílica

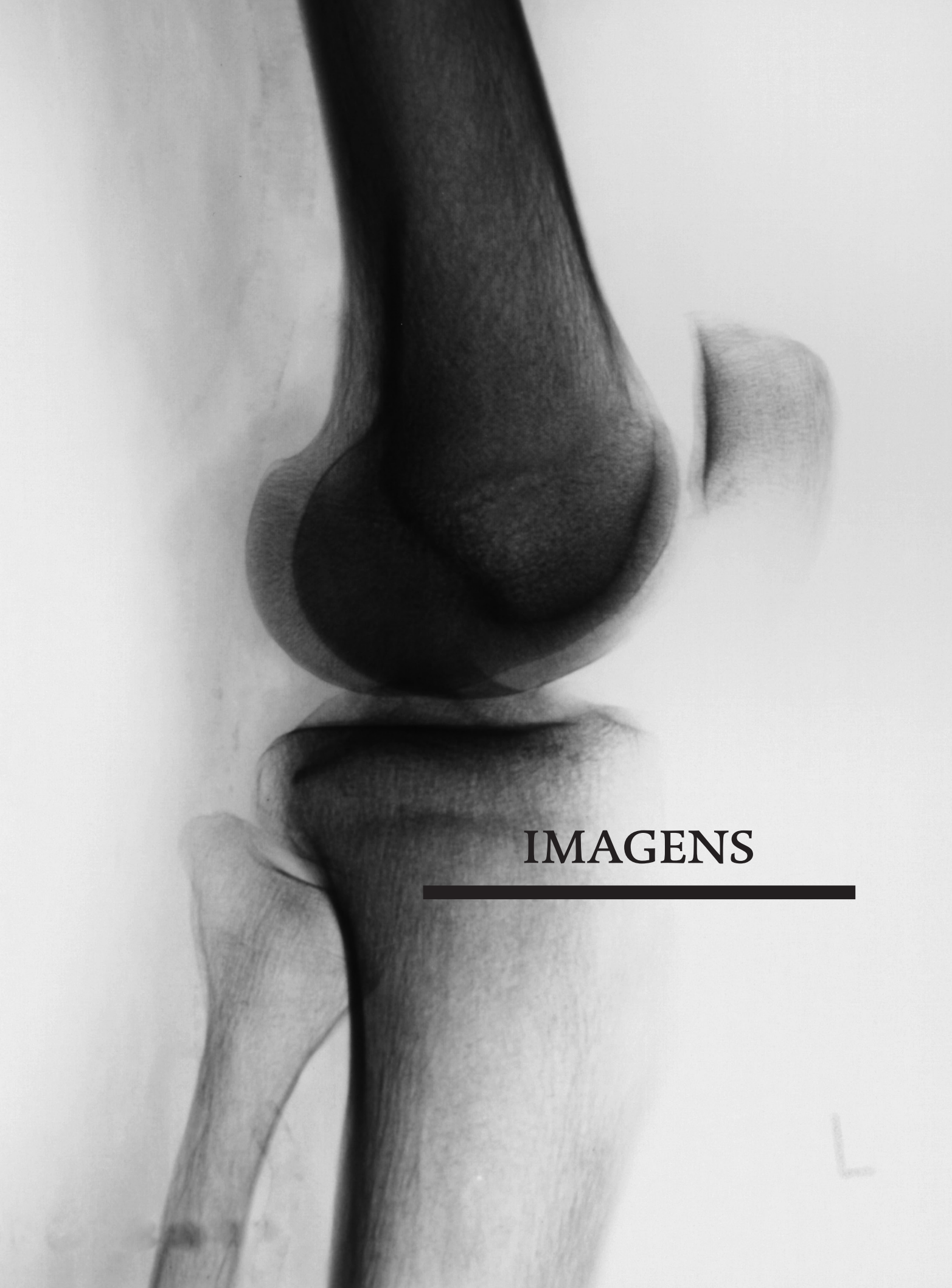
Caso clínico 3: Homem de 68 anos com história de eczema recidivante das mãos com 20 anos de evolução, até então interpretado como de contacto pela atividade profissional e positividade dos testes epicutâneos. Afirmava que desde que se reformou as lesões em ambas as mãos se tornaram mais exuberantes. Afirmava ainda noção de edema global das mãos e, à direita, com extensão ao punho e antebraço.

Ao exame objetivo, com lesões eczematosas, papulares de pequenas dimensões na face dorsal das mãos, com edema acentuado e, à direita, com extensão ao antebraço.

O estudo analítico realizado excluiu a existência de alteração da função tiroideia, paraproteinemia, doença renal ou infeção pelos vírus de hepatite C, B ou vírus da imunodeficiência humana.

O estudo histológico da pele revelou depósitos focais de mucina e ausência de fibroblastos pelo que foi feito o diagnóstico de líquen mixedematoso localizado.

Conclusão: Os três casos representam diferentes diagnósticos cuja apresentação clínica mimetiza a esclerose sistémica e que devem ser tidos em consideração no diagnóstico diferencial. A avaliação dos doentes com sinais e sintomas relacionados com espessamento cutâneo e edema acral global na ausência de fenómeno de Raynaud, negatividade de anticorpos antinucleares e/ou alterações capilaroscópicas deve levantar suspeita para outras patologias.



IMAGENS

Imagens

ACTA REUMATOL PORT. 2021:46:??-?? (SUP)

013 - TENOSSINOVITE DOS EXTENSORES DO PUNHO - UM QUADRO INAUGURAL DE ARTRITE REUMATOIDE

Rafaela Nicolau[1], Nádia Martins[1], Paulo Monteiro[1]
[1]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Doente do sexo feminino de 69 anos, agricultora, sem antecedentes pessoais relevantes e com antecedentes familiares de AR, referenciada à consulta de Reumatologia por quadro de poliatralgias de ritmo inflamatório com noção de tumefação envolvendo mãos, pés, joelhos e tibio-társicas associado a massa nodular da superfície extensora de ambos os punhos com cerca de 7 meses de evolução. Além da tumefação descrita e francamente pior do lado direito, não apresentava outras alterações ao exame objetivo.(Figura 1)

Portadora de ecografia com power doppler da mão direita que revelou a presença de tenossinovite do extensor comum dos dedos.

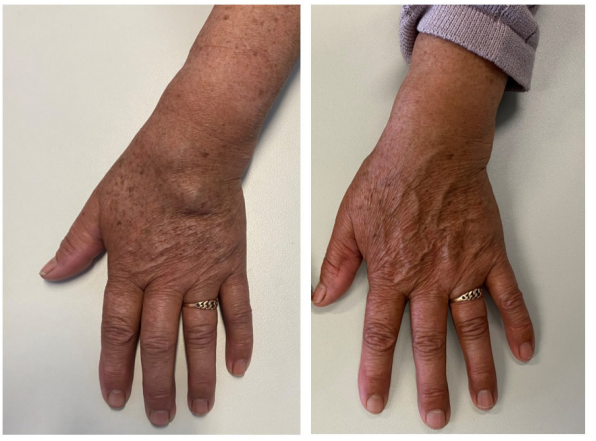
À exceção de um aumento da velocidade de sedimentação (48 mm/h), não apresentava outras alterações analíticas (hemograma e proteína C reativa normal, anticorpos antinucleares, fator reumatoide, anticorpo anti-peptídeo cíclico citrulinado negativo, enzima de conversão da angiotensina e uricemia normais, IGRA negativo, serologias para VIH, VHB, VHC, VDRL, Brucelose e doença de Lyme não reativas).

Foi efetuada infiltração com 40 mg de acetato de metilprednisolona da tenossinovite do extensor comum dos dedos da mão direita com resolução do quadro (Figura 1)

Durante o seguimento desta doente foi objetivado quadro de poliartrite envolvendo as metacarpofalângicas e interfalângicas proximais de ambas as mãos, cerca de 12 meses após o quadro inaugural.

Foi admitido o diagnóstico de AR seronegativa e iniciada terapêutica com metotrexato na dose de 10 mg/semana, folicil 10 mg/semana e metilprednisolona na dose inicial de 6 mg id.

FIGURA. SUPERFÍCIE EXTENSORA DA MÃO DIREITA ANTES E APÓS INFILTRAÇÃO COM METILPREDNISOLONA



018 - JACCOUD'S ARTHROPATHY

Eduardo Dourado[1, 2], Sofia C Barreira[1, 2]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

FIGURE. JACCOUDS ARTHROPATHY (PANELS A, B AND C)

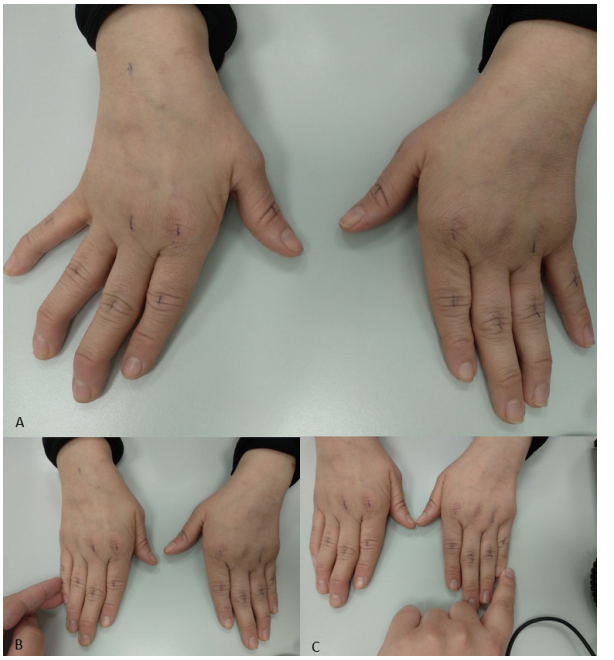


Figure caption: Jaccoud's arthropathy is a deforming non-erosive arthropathy characterised by ulnar deviation of the second to fifth fingers with metacarpophalangeal joint subluxation that is correctable or reducible with physical manipulation. Jaccoud's arthropathy can be associated with systemic lupus erythematosus, rheumatic fever and other rheumatic and non-rheumatic conditions. It occurs due to ligament and/or joint capsule laxity rather than primary articular involvement. This patient presented to the Rheumatology clinic at the age of 34 with fully developed Jaccoud's arthropathy. Although she had arthritis since the age of 13 and hand deformities for several years, she never sought medical assistance. The patient was finally diagnosed with polyarticular juvenile idiopathic arthritis and treated with prednisolone and hydroxychloroquine due to planned pregnancy, with good clinical response. Figure 1-A shows typical Jaccoud's arthropathy deformities, corrected with side pressure on the right hand (Figure 1-B) and left hand (Figure 1-C). The marks on the patient's hands signal the swollen joints.

027 - AXIAL OCHRONOTIC ARTHROPATHY

Eduardo Dourado[1, 2], Ana Sofia Pinto[3], Anabela Oliveira[4], Nikita Khmelinskii[1, 2]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, [4]Serviço de Medicina I, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

FIGURE: CONVENTIONAL RADIOGRAPHS OF THE LUMBAR SPINE AND SACROILIAC JOINTS OF A PATIENT WITH ALKAPTONURIA

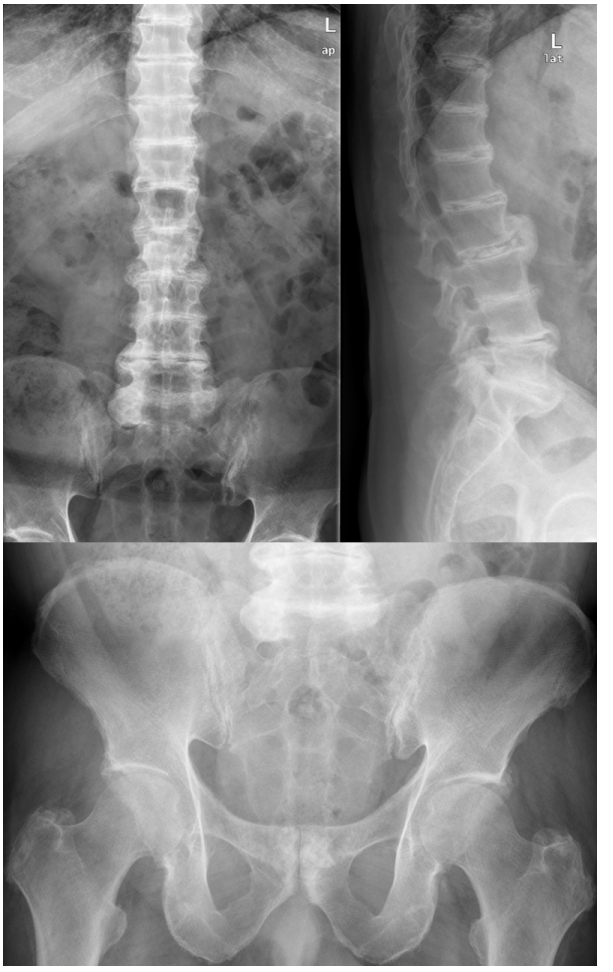


Figure caption: Conventional radiographs of the lumbar spine and sacroiliac joints of a patient with alkaptonuria. The diagnosis was based on the identification of black tendons during orthopaedic surgery after a spontaneous Achilles tendon rupture, grey deposits in the sclerae, ears and nose, and the patient's history, including the passage of dark urine since childhood. The high urine homogentisic acid concentration confirmed the diagnosis at the age of 61 years. The patient presented to the Rheumatology clinic a few weeks after the diagnosis, complaining of low back pain for more than ten years. A few years before, the pain was elicited by movement but was constant by the time of the first Rheumatology appointment. He had a morning stiffness of over 2 hours. Similarly to axial spondyloarthritis, axial ochronotic arthropathy typically presents as low back pain and stiffness and evolves to ankylosis of the spine. However, as shown in these radiographs, there is extensive spine involvement with relative sparing of the sacroiliac joints. The multilevel calcification (including the nucleus pulposus) and collapse of the intervertebral discs are also typical and can be seen at different levels of the lumbar and dorsal spine of this patient. Pubic symphysitis, another typical finding, can also be seen in the posteroanterior pelvic radiograph.

035 - TOPHACEOUS GOUT: THE NATURAL EVOLUTION OF A COMMON DISEASE

Ana Margarida Duarte-Monteiro[1, 2], Eduardo Dourado[1, 2], JE Fonseca[1, 2]

[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas,

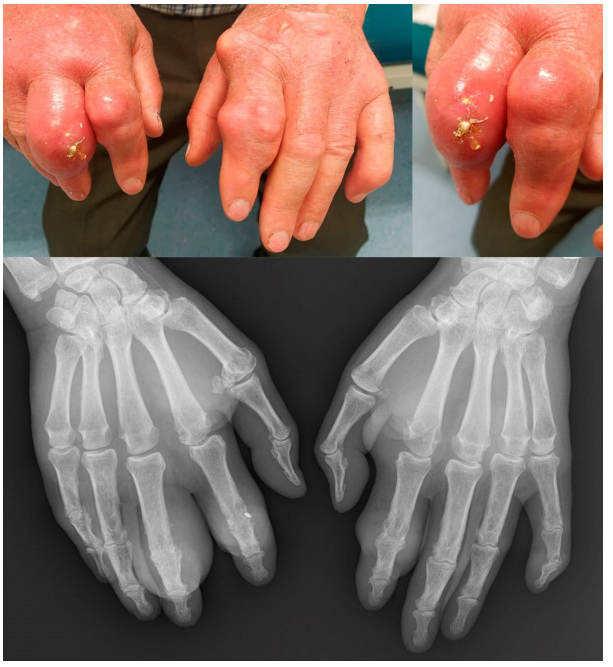
Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introduction: Gout is a common inflammatory arthritis in which monosodium urate crystals deposit in the synovial membrane, articular cartilage and periarticular tissues. Chronic tophaceous gout usually develops after several years of active disease, with recurrent flares and uncontrolled hyperuricemia. There are effective treatments to control uricemia, prevent and control gout flares and tophi formation. However, patients are often non-compliant, and the condition can become disabling.

Clinical vignette: A 76-year-old man presented to the emergency department complaining of oedema on the dorsal aspect of his right hand and pain and swelling of the third right proximal interphalangeal joint (PIP) with white discharge, but no fever. The symptoms began seven days before, and the patient took colchicine 2mg/day (for three days) and acemetacin 60mg/day, with no improvement.

There was a history of gout since the age of 45 and treatment with allopurinol for a brief period. Flares were common (5 to 6/year) and self-managed.

FIGURE. ADVANCED CHRONIC TOPHACEOUS GOUT



An exuberant tenderness and swelling adjacent to the third right PIP due to an exuberant gouty tophus with a white discharge was evident on physical examination. There were no other painful or swollen joints, but several tophi on the metacarpophalangeal and interphalangeal joints of both hands, knees and elbows could be observed.

The blood workup revealed leucocytosis (13.300/µL), elevated C-reactive protein (4.35 mg/dL) and hyperuricaemia (10.6 mg/dL). Radiographic evaluation of the hands revealed rat-bite erosions, and soft tissue swelling was also documented. drained the tophus, which contained chalk-like material, were surgically drained. The patient started prednisolone 15 mg/day for seven days and was re-evaluated within a week, with significant improvement of pain and swelling. Febuxostat 40mg/day was started with a good but incomplete hyperuricaemia response after two months of therapy. Dose escalation ensued.

Conclusions: We present a case of untreated gouty arthritis, reflecting the natural history of the disease evolving into its chronic tophaceous form. Early and effective management of the condition is critical for preventing flares and avoiding structural damage and tophi formation, which can be disabling and prone to severe complications, such as infection.

049 - PROSPECTIVE CLINICAL AND ULTRASONOGRAPHIC ASSESSMENT OF THE TEMPORAL ARTERY IN A PATIENT WITH GIANT CELL ARTERITIS

Rita Cunha[1], Ana Margarida Duarte-Monteiro[2,3], Catarina Tenazinha[2,3], Nikita Khmelinskii[2, 3], Cristina Ponte[2, 3]

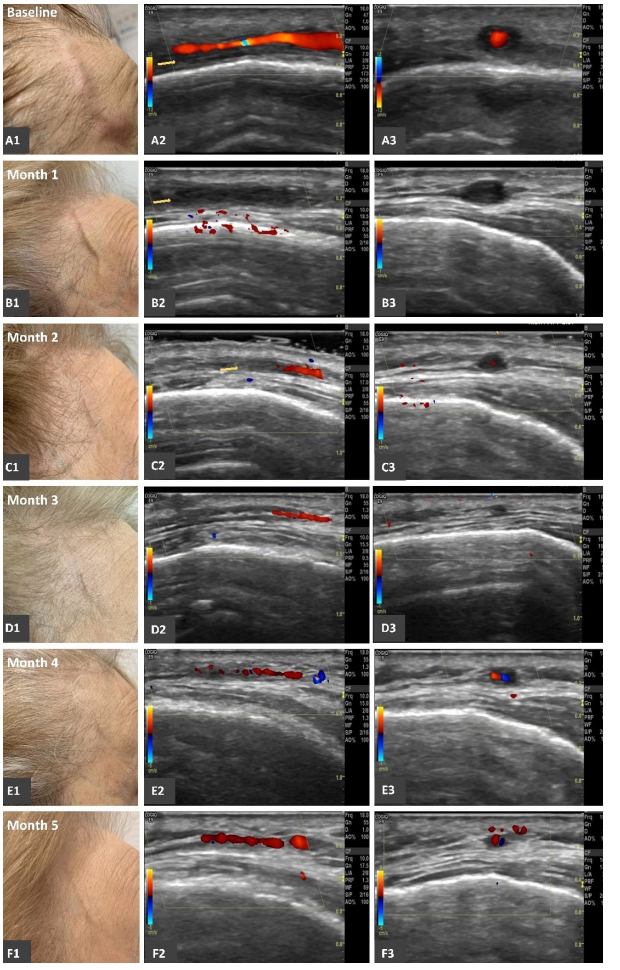
[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [2]Rheumatology and Bone Metabolic Diseases Department, Centro Hospitalar Lisboa Norte, EPE - Hospital de Santa Maria, Lisbon Academic Medical Centre, Lisboa, Portugal, [3]Rheumatology Research Unit, Instituto de Medicina Molecular, Faculdade de Medicina da Universidade de Lisboa, Lisboa, Portugal

A 72-year-old female presented for medical attention with a 2-month history of bilateral temporal headache, jaw claudication and visual disturbances. She had a background history of arterial hypertension and dyslipidaemia and had been diagnosed with polymyalgia rheumatica six months ago, treated with a maximum prednisolone dose of 15mg/day. On vascular examination, a thickened, tender and hard 'cord-like'

frontal branch of the right temporal artery (TA), with reduced pulse, was observed (Fig. A1). Ophthalmological examination was unremarkable. Laboratory tests showed elevated acute phase reactants (erythrocyte sedimentation rate of 120mm/hr, C-reactive protein of 6.5mg/dL). Ultrasound of the TAs revealed the presence of "halo sign" in all TA branches, including the thickened frontal branch (Fig. A2-3). The patient was diagnosed with giant cell arteritis (GCA) and was started on 60mg/day of prednisolone with progressive tapering [1]. Her symptoms and laboratory findings resolved after three weeks and her TA thickening slowly returned to normal after five months of treatment (Fig. F). She underwent monthly ultrasonographic assessments depicting the TA abnormalities found on physical examination (Figs. B-F). This case demonstrates the good correlation between clinical and ultrasonographic findings in GCA and highlights the potential role of the ultrasound in monitoring this disease [2].

(A1) Right temporal artery (frontal branch) at baseline showing arterial thickening; corresponding colour Doppler ultrasonographic assessment in longitudinal (A2) and transverse view (A3) revealing the presence of hypoechoic wall thickening, i.e. "halo sign", and stenosis (arrow).(B1) Reduction of the temporal artery thickening after 1 month of treatment; corresponding ultrasonographic assessment in longitudinal (B2) and transverse view (B3) revealing progression to arterial occlusion due to wall thickening (arrow). (C1) Continuous improvement of the temporal artery thickening at 2 months; corresponding ultrasonographic assessment in longitudinal (C2) and transverse view (C3) showing a less pronounced occlusion and diminished "halo sign" (arrow), with reduction of the arterial wall hypoechogenicity. (D1) Minor temporal artery thickening after 3 months of treatment; corresponding ultrasonographic assessment in longitudinal (D2) and transverse view (D3) revealing a minor wall thickening with persistent distal occlusion. (E1) Normal aspect of the temporal artery after 4 months of treatment; corresponding ultrasonographic assessment in longitudinal (E2) and transverse view (E3) showing a minor wall thickening with improved arterial flow of the previously occluded arterial segment. (F1) Normal aspect of the temporal artery after 5 months of treatment; corresponding ultrasonographic assessment in longitudinal (F2) and transverse view (F3) without the presence of wall thickening and revealing complete resolution of the previously occluded arterial segment.

FIGURE. ASSOCIATION BETWEEN CLINICAL AND ULTRASONOGRAPHIC FINDINGS IN A PATIENT WITH GCA



REFERENCES:

1. Hellmich B, Agueda A, Monti S, Buttgereit F, de Boysson H, Brouwer E, et al. 2018 Update of the EULAR recommendations for the management of large vessel vasculitis. Annals of the Rheumatic Diseases 2020; 79:19-30.
2. Dejaco C, Ramiro S, Duftner C, et al. EULAR recommendations for the use of imaging in large vessel vasculitis in clinical practice. Annals of the Rheumatic Diseases 2018; 77:636-643

074 - BIPARTITE ULNAR NERVE: A RARE ANATOMICAL VARIATION

Marta Henriques Oliveira[1], Jorge Garcia[2], João Madruga Dias[2]

[1]USF Cardilium, ACES Médio Tejo, Torres Novas, Portugal, [2]Rheumatology Department, Centro Hospitalar Médio Tejo, Torres Novas, Portugal

Introduction: The diagnosis of peripheral neuropa-

FIGURE. RIGHT ELBOW ULTRASOUND TRANSVERSE CUT (A-C) AND MRI T1 WEIGHTED AXIAL CUT (D-F)

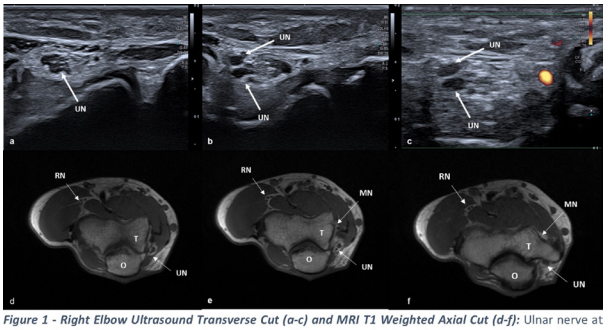


Figure 1 - Right Elbow Ultrasound Transverse Cut (a-c) and MRI T1 Weighted Axial Cut (d-f): Ulnar nerve at different levels of its course going distal to proximal (1a to 1c and 1d to 1f). It starts with its usual unique configuration (1a, 1d) and then starts to split (1b, 1c, 1e), returning to its unique configuration (1f). Through Fig. 1c it is possible to distinguish the ulnar nerve (without Power Doppler signal) from blood vessels (with Power Doppler Signal signal). **Abbreviations:** MN - *Median Nerve*, O - *Olecranon process*, RN - *Radial Nerve*, T - *Trochlea*, UN - *Ulnar Nerve.*

thies and motor deficits requires a deep knowledge of the peripheral nerves anatomy. We present an image of an elbow ultrasound that revealed a rare anatomical variation of the ulnar nerve (UN) in a patient with symptoms compatible to an ulnar entrapment syndrome.

Case Report: A 45-year-old woman, housekeeper, was sent to the Rheumatology appointment for pain, functional limitation in extension and flexion movements of the fingers, accompanied numbness in the 4th and 5th finger of the right hand. This clinical depiction had 2 to 3 years of evolution, with progressive worsening. The physical exam showed pain at palpation of UN in the ulnar fossa of the right elbow. Blood tests and electromyography studies did not show any significant changes. Ultrasonography of the elbow revealed a rare anatomical variation that was then confirmed by MRI as illustrated in the image that we present. Since the remaining complementary means of diagnosis were normal, the bipartite UN described seems to be the probable cause of the patient's complaints.

Discussion: Anatomical variations of the UN can occur in all its course however, the ones occurring along UN proximal course are rarely described in the literature. Although there are descriptions of peripheral neuropathies associated with bipartite median nerve our research did not find a similar description of bipartite UN at a proximal level.

Conclusion: This case demonstrates the importance of deep anatomy knowledge in the evaluation of peripheral neuropathies and motor deficits, that overlooked can result in delays of diagnosis and consequently treatment of these pathologies

098 - AN UNUSUAL CAUSE OF SHOULDER PAIN IN A YOUNG WOMAN WITH SYSTEMIC SCLEROSIS: ATYPICAL INTRA-ARTICULAR CALCINOSIS

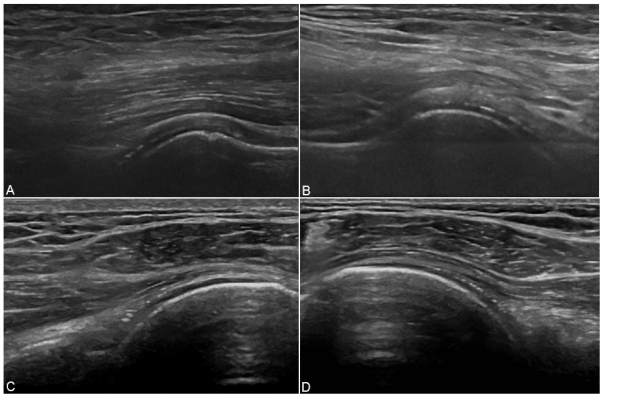
Tomás Fontes[1], Manuel Silvério-António[2, 3], Carla Macieira[2, 3], Joaquim Polido Pereira[2, 3]

[1]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal, [2]Rheumatology Department, Hospital Santa Maria, CHLN, Lisbon Academic Medical Center, Lisboa, Portugal, [3]Rheumatology Research Unit, Instituto de Medicina Molecular, Faculdade de Medicina da Universidade de Lisboa, Lisboa, Portugal

Calcinosis is a frequent manifestation of systemic sclerosis and is caused by the deposition of insoluble calcium in soft tissues, especially in the skin and the subcutaneous tissue.1 It can also occur in periarticular structures, such as muscles, tendons and ligaments,2 but intra-articular calcinosis (IAC) is not frequently documented in systemic sclerosis,3 with no reports of IAC solely to date. Calcium crystal arthropathies are mainly caused by calcium pyrophosphate and basic calcium deposition, the first being extremely rare in the young, and the second almost only occurring associated with osteoarthritis, when deposits are intra-articular.4

We report the case of a 25-year-old woman with a recent diagnosis of systemic sclerosis, associated with the anti-topoisomerase-1 antibodies, with rapidly progressive scleroderma and severe esophageal dysmotility, serositis and Raynaud's phenomenon, but no signs of skin calcinosis. Bilateral shoulder pain and limited mobilization prompted an ultrasound scan. No synovitis or signs of tenopathy

FIGURE. AXILLARY (A–LEFT; B–RIGHT) AND POSTERIOR RECESSES (C–LEFT; D–RIGHT) OF THE GLENOHUMERAL JOINT. CALCIUM DEPOSITS.



were found. However, there was evidence of intra-articular calcium deposits adhered to the surface of the hyaline cartilage of both humerus heads (which moved with it on dynamic exploration) (Figure 1). Deposits on the glenoid labrum were also documented. Ultrasound exploration of the wrists, hands and knees, as well as peripheral joint radiographs, showed no similar signs in these locations. Secondary causes of calcium deposition disease were excluded. The cause of this arthralgia probably relates to the intra-articular calcium compounds, its etiology being either an unusual calcium crystal disease or an atypical and isolated calcinosis phenomenon in the setting of systemic sclerosis.

REFERENCES:

1. Fallet GH, Vischer TL, Micheli A. La chondrocalcinose [Chondrocalcinosis]. Schweiz Med Wochenschr. 1982 Jun 19;112(25):888-97. French. PMID: 7112058.
2. Valenzuela A, Song P, Chung L. Calcinosis in scleroderma. 2018 Nov;30(6):554-561. doi: 10.1097/BOR.0000000000000539. PMID: 30124603.
3. Resnick D, Scavulli JF, Goergen TG, Genant HK, Niwayama G. Intra-articular calcification in scleroderma. Radiology. 1977 Sep;124(3):685-8. doi: 10.1148/124.3.685. PMID: 887759.
4. McCarthy GM, Dunne A. Calcium crystal deposition diseases - beyond gout. Nat Rev Rheumatol. 2018 Oct;14(10):592-602. doi: 10.1038/s41584-018-0078-5. PMID: 30190520.

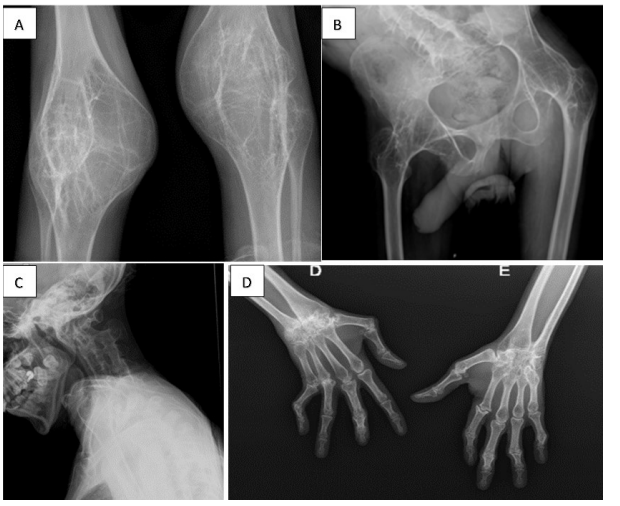
111 - A SEVERE, DEFORMING CASE OF JUVENILE PSORIATIC ARTHRITIS

Carolina Ochôa Matos[1,2], Joana Antunes[3,4], Carlos Miranda Rosa[1], João Eurico Fonseca[1,2], Elsa Vieira-Sousa[1,2]

[1] Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [2] Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular João Lobo Antunes, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [3] Serviço de Dermatologia, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal; [4] Clínica Universitária de Dermatologia, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introduction: Juvenile psoriatic arthritis (JPsA) is a subtype of juvenile idiopathic arthritis (JIA) that can be associated with severe joint and bone deformities, as well as growth impairment, due to persistence of inflammation. Epiphyseal vascular congestion can lead to premature closure of growth plates, causing alterations in the shaping of bone (such as balloon-shaped

FIGURE. RADIOGRAPHIC FEATURES OF A SEVERE AND DEFORMING JPSA DISEASE IN A MALE PATIENT.



hypertrophy) and a tendency to joint ankylosis. There is typical involvement of the cervical spine, often with ankylosis of the facet joints. Radiographic changes also include periarticular osteoporosis, which can be generalized in advanced forms, and periosteal new bone formation. Peripheral joints erosions, when severe, can shape into a "pencil-in-cup" appearance.

Case Report: We herein present the case of a JPsA male patient, initially diagnosed with JIA. The disease first manifested at the age of 13, with enthesitis-related arthritis features (involvement of the hips, knees, tibiotarsal joints and the cervical spine) and positivity for human leucocyte antigen-B27. The disease was characterized by periods of remission and exacerbation but showed a rapid progression in the first 2 years, with asymmetric, addictive and deforming polyarthritis, as well as further axial involvement, with radiographic sacroiliitis, and severe enthesitis. Methotrexate was initiated after nonsteroidal anti-inflammatory drugs failure. The first biologic disease-modifying antirheumatic drug, infliximab, was started 10 years after the diagnosis. At the age of 28, a few years after initiating treatment with this drug, he was diagnosed with psoriasis after the appearance of typical erythematous, scaly plaques. In this context, he switched to etanercept, and later to adalimumab due to psoriasis worsening. After this cutaneous manifestation, the diagnosis was changed to JPsA. Additionally, he was diagnosed with severe, diffuse osteoporosis at 15 years old, for which he started treatment with zoledronate. From an early age, the disease caused significant in-

capacity due to multiple joint ankylosis and muscular atrophy, with little recovery even after effective treatment and motor rehabilitation. Currently, the patient needs substantial support in the daily activities, requiring the use of a wheelchair.

Discussion: With this case, we want to highlight the radiologic features of JPsA and its importance in the diagnosis and follow-up of JIA patients. This case is also notable for its rarely seen images of severe joint ankylosis.

Fig.1. Radiographic features of a severe and deforming JPsA disease in a male patient. Knee ankylosis with balloon-shaped hypertrophy (A). Bilateral grade IV sacroiliitis, fusion of pubic symphysis and complete joint ankylosis of the hips (B). Bamboo spine with facet joint ankylosis from C2–C5 and loss of cervical lordosis (C). Severe joint space narrowing at radiocarpal and carpal joints, erosions and "pencil-in-cup" deformity well defined at the left 2nd metacarpophalangeal joint (D).

130 - KNUCKLE PADS - A RARE FINDING

Ana Teodósio Chícharo[1], Margarida Lucas Rocha[1], Diana Silva[2], Graça Sequeira[1]

[1]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal, [2]Unidade de Cuidados de Saúde Primários de Quarteira, Quarteira, Portugal

A 27-year-old male, electrician, presented in the rheumatology appointment due to local swelling of the proximal interphalangeal joints (PIP) of all fingers of the right hand and second finger of the left hand for the past 3 years. He reported pain with movement, restricted mobility and morning stiffness for 30 minutes. He had no pain at rest. He denied any major trauma or prior injury. He did not report fever, weight loss, rash or any other complaints.

He had history of asthma since childhood and smoking habits of 20 smoking-pack-years, with no relevant alcohol consumption.

His family history was positive for Dupuytren disease in his father and grandfather.

Physical examination showed soft subcutaneous nodules located at the dorsal surface of the PIP joints. There was no local tenderness, and the overlying skin showed a normal temperature. There was also thickening of the palmar fascia, and contracture of the third finger of the right hand.

Laboratory workup was normal, with no elevation of erythrocyte sedimentation rate or C-reactive protein. Immunological studies, including rheumatoid factor, cyclic citrullinated peptide antibody, antinuclear antibodies and anti-double stranded DNA showed no alterations.

The soft tissue ultrasound revealed hypoechogenic lesions with irregular borders at the subcutaneous aspect of PIP joints on all fingers of the right hand and second finger of the left hand, of maximum dimension of 10x3mm, without internal flow signals at color doppler. The ultrasound also showed hypoechogenic flat lesions in the superficial fascia of the palm, adhering to the flexor tendons.

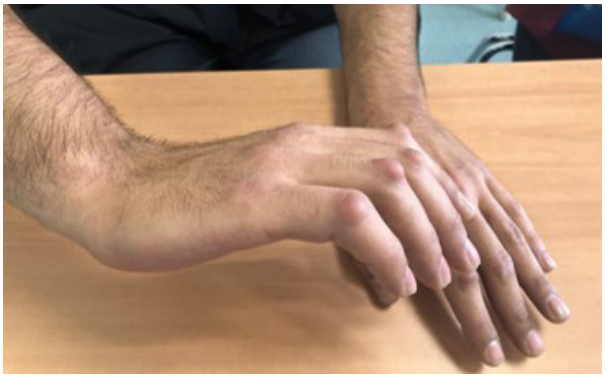
Therefore, the clinical suspicion of Knuckle pads and Dupuytren disease was confirmed by ultrasound.

Due to the complaints of limited function, that interfered with his job, the patient was referred to orthopedic surgery and the PIP nodules were removed. He referred resolution of the symptoms on the first months after the procedure.

Now, two years after the surgery, the nodules reappeared in the same fingers, and are also currently on other fingers previously not affected. He has again limited range in the involved joints, due to local discomfort.

This image intends to show a rare finding of Knuckle pads in a young male, who was submitted to surgery with reoccurrence of the nodules.

FIGURE. KNUCKLE PADS IN A YOUNG MALE.



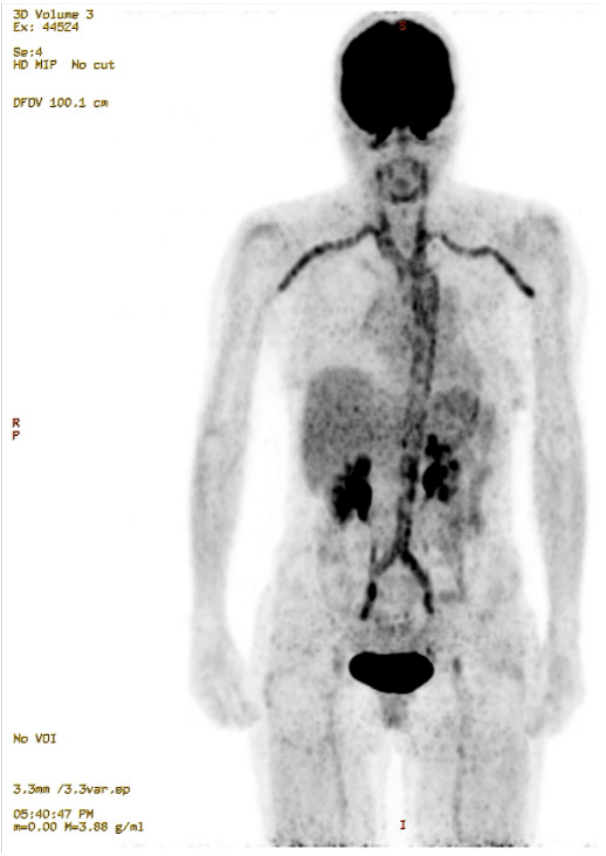
151 - THE GHOST

Liliana Saraiva[1], Luisa Brites[1], André Saraiva[1], Marlene Sousa[1], Ana Isabel Maduro[1], Adriana Carones[1], Beatriz Mendes[1], Helena Assunção[1], Ana Rita Prata[1], Cátia Duarte[1, 2]

[1]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [2]Coimbra Institute for Clinical and Biomedical Research (iCBR) - Faculty of Medicine, University of Coimbra, Coimbra, Portugal

A 62-year-old woman with rheumatic polymyalgia in remission and under treatment with 2.5mg/day of prednisolone, presented a new increase in acute

FIGURE. VASCULITIS OF LARGE VESSELS, INVOLVING SEVERAL ARTERIAL VASCULAR TERRITORIES



phase markers (Erythrocyte sedimentation rate was 64 mm/1st hour and C-reactive protein 3.8 mg/dL). She had no complaints suggesting a relapse of rheumatic polymyalgia, a new onset of giant cell arteritis (GCA), or other conditions such as tumors or infections. A Positron emission tomography (PET) scan was performed to exclude tumors and vasculitis and showed an extensive involvement of different vascular territories compatible with large vessel vasculitis. We established the diagnosis of extracranial GCA and the patient initiated treatment with prednisolone 1 mg/kg/day.

Extracranial GCA are lesser symptomatic that cranial ACG, and patients usually required higher doses of corticosteroids, have higher rates of relapsing, more risk to develop aortic aneurysms and higher mortality rate. In this context, and because our patient had an increased risk to osteoporosis we proposed tocilizumab 162mg weekly subcutaneous.

In this type of vasculitis with such a silent presentation, the high suspicious of this diagnosis is crucial and PET has an increased value to stablish the diagnosis.

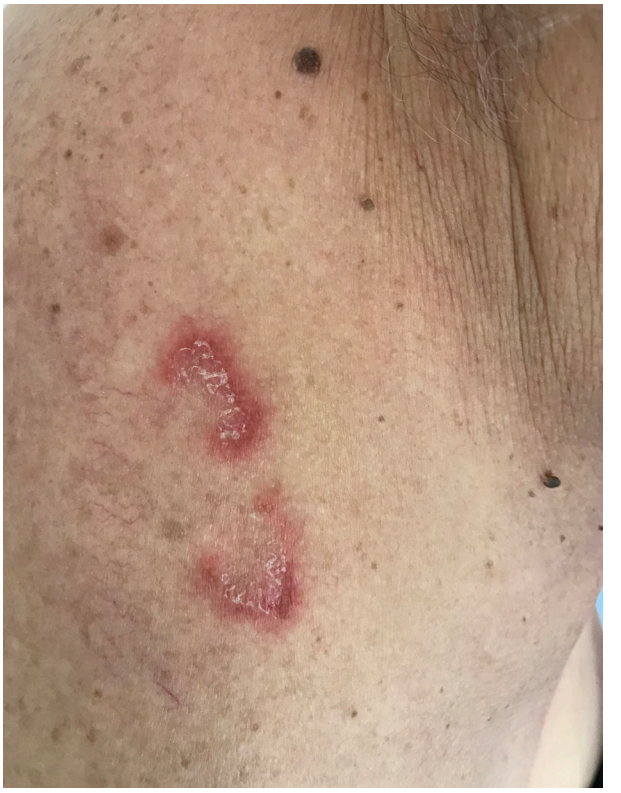
155 - CUTANEOUS SUBACUTE LUPUS ERYTHEMATOSUS IN A PATIENT WITH PRIMARY SJOGREN SYNDROME

Augusto Silva[1]

[1]Rheumatology and Bone Metabolic Diseases Department, Centro Hospitalar Lisboa Norte, EPE - Hospital de Santa Maria, Lisbon Academic Medical Centre, Lisboa, Portugal

74-year-old female with primary Sjogren's Syndrome under 400mg Hydroclichloroquine. She developed 2 pruriginous skin lesions, at the level of the shoulders, in an arcuate shape with an erythematous desquamative plaque. Skin biopsy revealed dense lymphocytic infiltrate and perivascular and V-shaped perianexial sleeve in the dermis, with negative PAS staining for microorganisms. These findings are consistent with subacute lupus erythematosus.

FIGURE: PRURIGINOUS SKIN LESION, WITH ARCUATE SHAPE AND ERYTHEMATOUS DESQUAMATIVE PLAQUE. SKIN BIOPSY WAS COMPATIBLE WITH SUBACUTE LUPUS

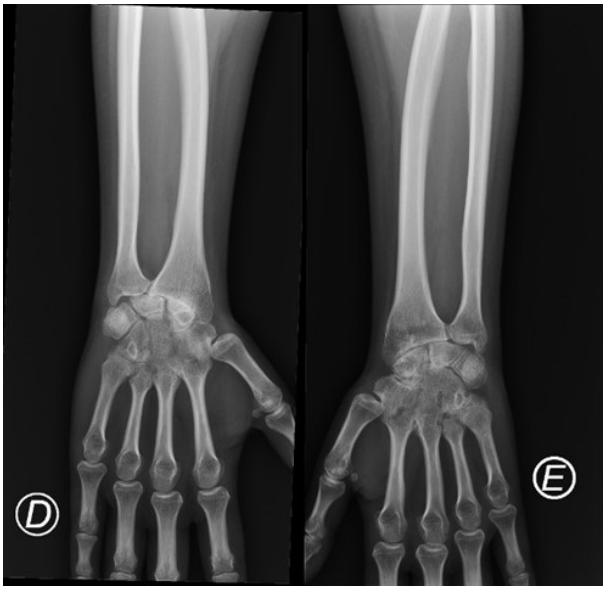


160 - IDADE APARENTE SUPERIOR À IDADE REAL

Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Tiago Beirão[1], Flávio Campos Costa[1], Romana Vieira[1], Taciana Videira[1], Joana Aleixo[1], P Pinto[1]
[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Mulher de 18 anos portadora de Artrite Idiopática Juvenil Forma Poliarticular com positividade do fator reumatoide, com 7 anos de evolução. Medicada com metotrexato 20 mg via subcutânea desde o diagnóstico e com tocilizumab desde há 6 meses, tendo tido falência prévia ao adalimumab introduzido há 2 anos. Atualmente, mantém atividade da doença, particularmente nos punhos, que apresentam dor e limitação à mobilidade passiva, com necessidade de infiltrações intra-articulares adjuvantes. Na radiografia dos punhos verifica-se uma discreta dissociação escafo-lunar à direita e marcada redução da interlinha radiocárpica, intercárpica e carpo-metacárpica bilateralmente, com esclerose subcondral, geodes e erosões, em especial à esquerda, sugestivo de uma artrose secundária à doença articular inflamatória subjacente (Figura 1). Este caso destaca-se pelo dano radiográfico extenso numa idade tão jovem, apesar de tratamento adequado e precoce, traduzindo uma doença agressiva e com prognóstico reservado.

FIGURA: ARTROSE DO PUNHO SECUNDÁRIA A AIJ

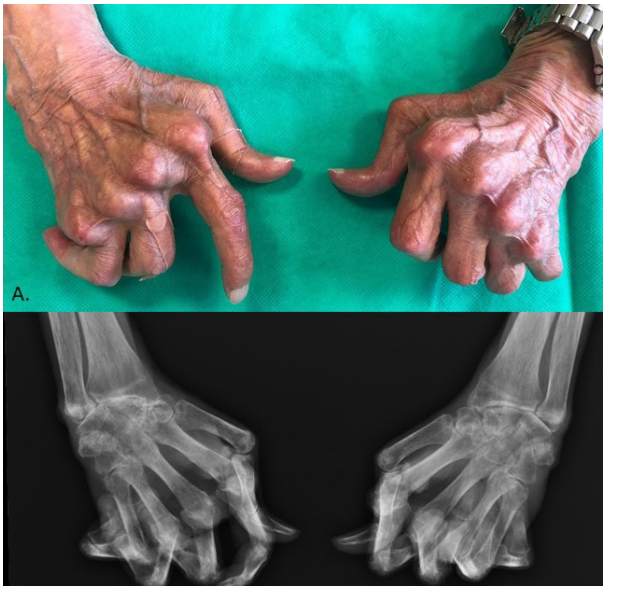


161 - UMA ARTRITE REUMATÓIDE DE LIVRO

Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Tiago Beirão[1], Romana Vieira[1], Taciana Videira[1], Joana Aleixo[1], Flávio Campos Costa[1], P Pinto[1]
[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Homem de 77 anos, com antecedentes de doença pulmonar obstrutiva crónica com enfisema pulmonar, foi enviado à consulta de Reumatologia com o diagnóstico de Artrite Reumatóide desde os 28 anos. Estava medicado com metotrexato 20 mg/semana, celecoxib 200 mg id e deflazacort 6 mg/dia. Ao exame objetivo apresentava deformidades exuberantes típicas de Artrite Reumatóide, nomeadamente alargamento dos punhos, polegar em Z, desvio cubital dos dedos, subluxação das articulações metacarpo-falângicas com limitação da extensão dos dedos, dedos em botoeira e atrofia dos interósseos (fig. 1-A). O estudo radiográfico das mãos, para além das deformidades atrás descritas, permitiu identificar múltiplas erosões marginais associadas a redução da interlinha articular das radiocárpicas, intercárpicas, carpo-metacárpicas e metacarpo-falângicas, bilaterais e simétricas, bem como erosão praticamente completa da apófise estilóide cubital (fig. 1-B). Pretende-se alertar para o facto de, apesar da evolução terapêutica que tem decorrido nos últimos anos e da maior consciencialização por parte da comunidade médica para a doença reumática inflamatória, casos como o descrito continuam a chegar à consulta de Reumatologia, com deformidades irreversíveis e limitativas nas AVDs. Realça-se, assim, a importância da referenciação atempada e do diag-

FIGURA: UM CASO DE ARTRITE REUMATÓIDE AVANÇADA



nóstico e tratamento precoces da Artrite Reumatóide para que, futuramente, casos como este passem a ser encontrados apenas em livros.

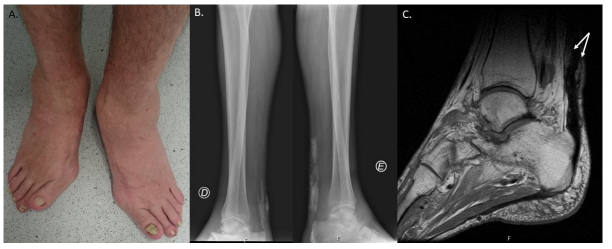
162 - UM CASO RARO DE OSSIFICAÇÃO HETEROTÓPICA DO TENDÃO DE AQUILES

Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Tiago Beirão[1], Flávio Campos Costa[1], Romana Vieira[1], Taciana Videira[1], Joana Aleixo[1], P Pinto[1]
[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Homem de 47 anos, com antecedentes de múltiplas cirurgias de correção de malformação mal-esclarecida dos tornozelos e pés durante a infância e de nefrectomia total direita após acidente de viação aos 13 anos. Enviado à consulta de Reumatologia por quadro compatível com artrite gotosa. Concomitantemente, referia artralgias mecânicas crónicas dos tornozelos com franca limitação na marcha e necessidade do uso de canadiana. Ao exame objetivo, apresentava deformidade em valgus dos tornozelos e hálux, tumefação pétrea dos tornozelos, limitação da mobilidade passiva das articulações tibio-társica e subtalar e nódulos duros no trajeto do tendão de Aquiles, bilateralmente (fig. 1-A). A radiografia de perfil em carga dos tornozelos mostrou extensas calcificações do tendão de Aquiles e artrose da tibiotársica e subtalar, bilateralmente (fig. 1-B). A ressonância magnética dos tornozelos confirmou a presença de ossificações intratendinosas do Aquiles bilateralmente (Figura 1-C). Foi instituída terapêutica hipouricemiante com alopurinol juntamente com profilaxia de crises de gota e otimizada analgesia. A Ortopedia propôs realização de artrodese do tornozelo, que o doente recusou, e foram realizados bloqueios dos nervos peroneal superficial, sural e tibial posterior pela Fisiatria de Intervenção com alívio parcial da dor.

A ossificação heterotópica do tendão de Aquiles é uma patologia incomum que acomete predominantemente o sexo masculino. A sua etiologia é desconhecida embora seja mais observada em contexto de cirurgia prévia, rotura do Aquiles, microtrauma repetitivo, existindo também alguma associação com a presença de gota, como neste caso[1,2].

FIGURA: OSSIFICAÇÃO HETEROTÓPICA DO TENDÃO DE AQUILES



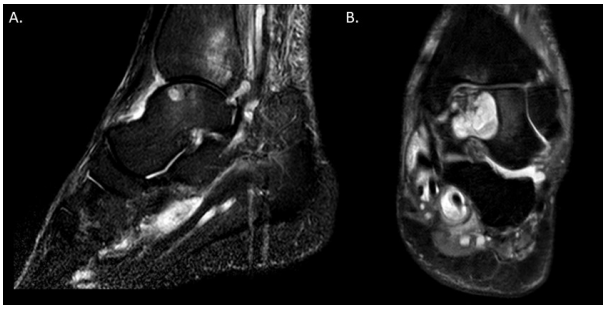
REFERÊNCIAS:
1. Francesco Manfreda *et al*. A silent massive ossification of Achilles tendon as a suspected rare late effect of surgery for club foot. SAGE Open Medical Case Reports 2018; 6:1–5
2. Raju Vaishya *et al*. Heterotopic Ossification of Tendo Achilles: An Uncommon Clinical Entity. J Orthop Case Rep. 2019; 9(2): 45–47.

163 - UM ESTRANHO CASO DE OSTEONECROSE BILATERAL E NÃO TRAUMÁTICA DO ASTRÁGALO

Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Tiago Beirão[1], Flávio Campos Costa[1], Romana Vieira[1], Taciana Videira[1], Joana Aleixo[1], P Pinto[1]
[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Homem de 68 anos, com múltiplos fatores de risco cardiovasculares e antecedentes de cardiomiopatia de stress e de doença de Peyronie, enviado à consulta de Reumatologia por artralgias dos tornozelos de ritmo mecânico mas com tumefação recorrente, com dois anos de evolução, e alívio com toma de anti-inflamatório. Objetivamente sem dor ou tumefação à palpação da interlinha nem limitação da mobilidade. A radiografia em carga dos tornozelos mostrou alterações degenerativas ao nível da tibiotársica bilateralmente, com evidência de imagens líticas no astrágalo. A ressonância magnética mostrou extensas lesões csticas na cúpula do astrágalo bilateralmente e na superfície articular da tíbia à direita, envoltas em edema da medula óssea, e à esquerda com comunicação com a superfície articular, evidenciando algum grau de colapso (figura 1 - sequências STIR corte sagital do tornozelo direito (A) e corte coronal do tornozelo esquerdo (B)). Apresentava também alterações degenerativas difusas da articulação tibiotársica com moderado a volumoso derrame articular bilateralmente. Foi assumido o diagnóstico de osteonecrose do astrágalo com artrose secundária da tibiotársica. O doente foi orientado para a consulta de Ortopedia onde foi proposta intervenção cirúrgica (osteotomia e preenchimento com enxerto).

FIGURA: OSTEONECROSE BILATERAL E NÃO TRAUMÁTICA DO ASTRÁGALO



A osteonecrose bilateral não traumática do astrágalo é muito rara, havendo poucos casos descritos na literatura[1].

REFERÊNCIAS:

1. VRM Reddy, G Selzer, M Faisal, MJ Aldridge. Bilateral idiopathic avascular necrosis of talus: a case report. Foot and Ankle Surgery 2003;9(1):35-39

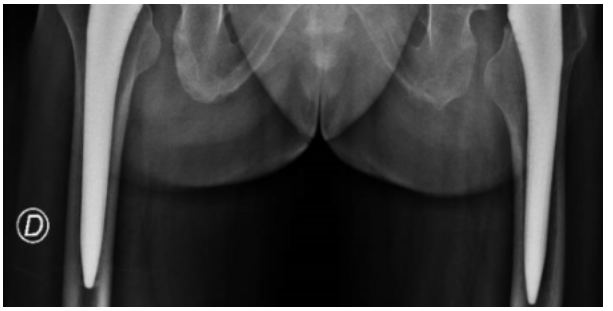
200 - NEM TUDO O QUE RELUZ É OURO

Patrícia Pires[1], Ricardo Pinto Rocha[1], Nuno Mendonça[1]

[1]Serviço de Ortopedia B, Hospital Prof. Doutor Fernando Fonseca, Amadora, Portugal

Vinheta clínica: Homem de 48 anos, submetido a artroplastia total da anca bilateral.
Comentário: A dor na anca em jovens deve despertar diagnósticos diferenciais para além da coxartrose isolada. Manifestações de sacroileíte bilateral radiográfica com erosões, esclerose, redução do espaço articular ou anquilose, são um sinal clássico de espondiloartropatia axial (ex. Espondilite Aquilosante) que reflectem um dano estrutural.

FIGURA.



201 - SPIDER HAND

Patrícia Pires[1], Nuno Mendonça[1], Ricardo Pinto Rocha[1]

FIGURE.

[1]Serviço de Ortopedia B, Hospital Prof. Doutor Fernando Fonseca, Amadora, Portugal

56-year-old man with boutonnière fingers in both hands. The boutonnière deformity is defined as the lesion of the zone III of the extensor tendon, manifesting by the flexion of the proximal interphalangeal (PIP) joint and extension of the distal interphalangeal (DIP) joint. The diagnosis is clinical, with a positive Elson´s test.

The treament of acute lesions involves splinting of the PIP. On the other hand, surgical treatment is an option in chronic symptomatic lesions. This deformity is common in rheumatoid arthritis, since there is capsular distension leading to rupture of the central slip; more than half of these patients develop the deformity in at least one finger.

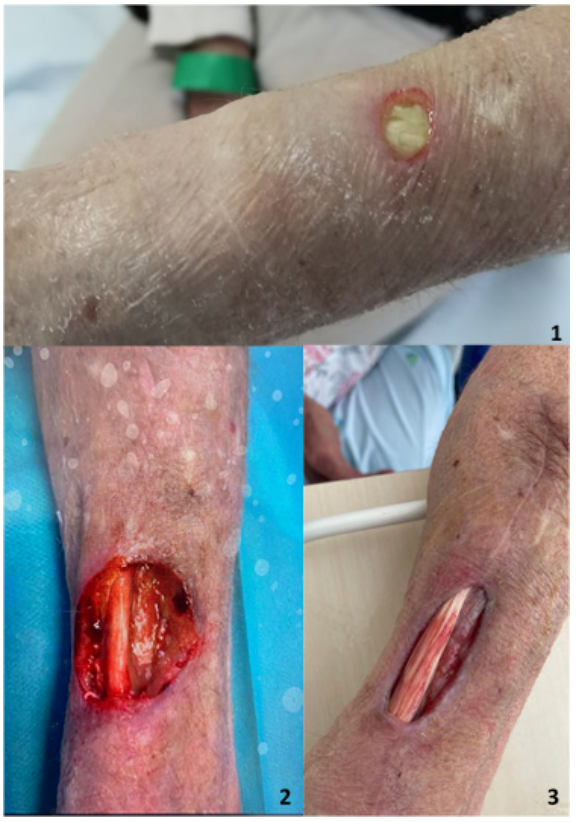
204 - CUTANEOUS TUBERCULOSIS IN A PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS

Nuno Pina Gonçalves[1, 2, 3], Maria Helena Lourenço[2, 3], Sara Pocinho[4], João Borralho[4], Maria Manuela Costa[3], Sandra Falcao[3], Kamal Mansinho[4, 5], Jaime C. Branco[2, 3]

[1]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [2]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [3]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [4]Infectious Diseases and Tropical Medicine, Centro Hospitalar de Lisboa Ocidental, Lisboa, Portugal, [5]Institute of Hygiene and Tropical Medicine - NOVA University of Lisbon, Lisboa, Portugal

Female, 67 years old, with a history of systemic lupus erythematosus with mucocutaneous, articular, renal

FIGURE: LEFT FOREARM CUTANEOUS TUBERCULOSIS IN A PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS



and hematologic involvement, medicated with micophenolic acid 500mg/day and prednisolone 5mg/day, with good disease control, presents to a rheumatology apointment with a white circular lesion in the left forearm, with one year of progression (suggestive of caseum) (1). Several courses of antibiotics were previously made, with only minor improvement. At 4 months with no improvement, surgical debridement was performed and a deep tissue sample was obtained (2). Mycobacterium tuberculosis was isolated (resistant to streptomycin) and the patient was admitted to the infectious diseases ward. Quadruple therapy was started with isoniazid, rifampicin 600mg, ethambutol and pyrazinamide. The patients is now at follow-up, slowly recovering (3) with an apparent good response to anti-bacillary therapy that should ensue for at least 9 months. In the setting of an immunocompromised patient with a long lasting infection with no response to initial therapy, a high index of suspicion to indolent agents (such as Mycobacterium tuberculosis) should be present.

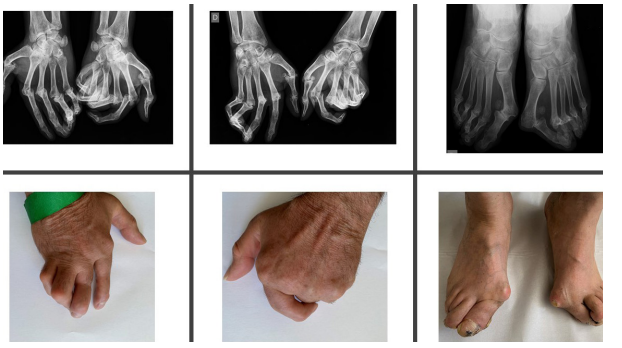
208 - ARTRITE PSORIÁTICA MUTILANTE

Laura Gago[1], Ana Filipa Mourão[1, 2], Jaime C. Branco[1, 2], Maria Manuela Costa[1], Maria João Gonçalves[1]
[1]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [2]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

A forma mutilante de artrite psoriática ocorre em 1-5% dos doentes com esta patologia.

Homem, 47 anos, com psoríase desde os 18 anos de idade, enviado à consulta de reumatologia em 2009 por artralgias das mãos com meses de evolução. À observação, apresentava desvio das mãos em rajada de vento, dedos em botoeira e em colo de cisne e deformação óssea de ambos os pés (pés em tripé) e analiticamente fator reumatoide e parâmetros inflamatórios negativos. Abandonou a consulta, até 2016 e regressou por agravamento das deformações ósseas e da psoríase. Iniciou metotrexato 20mg/semana, suspenso por intolerância gastrointestinal. Fez switch para sulfassalazina, porém perdeu follow-up novamente. Regressou em 2019 por artralgias do punho direito e cotovelo esquerdo, agravamento das deformações ósseas, objectivando-se tumefação do punho direito, mobilidade axial limitada e lesões de psoríase generalizadas – DAS28 4v 4.011 e PASI 19.6. Estava medicado com corticóide tópico e diclofenac 75mg em SOS. A radiografia das mãos apresentava luxação das MCF2-3 bilateral e MCF4,5 da mão esquerda, anquilose das IFD de 2-5 da mão direita, erosões ósseas justa-articulares da ulna direita e todos os metacarpos e primeiras falanges, com lesões pencil in cup nas MCF4,5 direitas. A radiografia dos pés revelava osteopenia periarticular, exuberante hallux valgus bi-

FIGURA: MÃOS, PÉS E RADIOGRAFIAS DE DOENTE COM ARTRITE PSORIÁTICA MUTILANTE



lateral, subluxações das MTFs direitas com pencil in cup das MTFs 2-5 e luxação das MTF2-4 esquerdas.

Por intolerância a dois cDMARDs foi proposto para terapêutica biológica. Dada tuberculose latente, foi prescrita isoniazida, sem adesão do doente (confirmado com a farmácia hospitalar).

Pela tuberculose latente, má adesão terapêutica e predomínio do envolvimento articular periférico e cutâneo, optou-se pelo início de ustecinumab 45mg em ambiente hospitalar.

Apresentou excelente resposta cutânea e articular, com ΔDAS284v -3.021 e resposta PASI 90 ao 7º mês de tratamento, com franca melhoria na adesão do doente aos cuidados de saúde.

210 - TREATMENT OF CALCINOSIS CUTIS WITH COMBINATION OF MINOCYCLINE AND DILTIAZEM

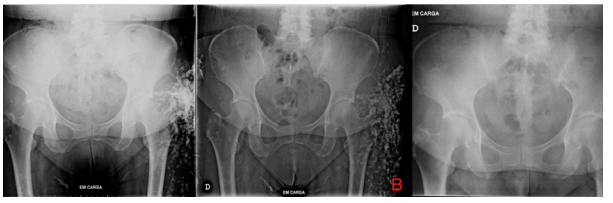
Margarida Lucas Rocha[1], Ana Teodósio Chícharo[1], Vítor Teixeira[1], Graça Sequeira[1]

[1]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal

A currently 61 years-old woman was diagnosed with anti-MDA5 positive amyopathic dermatomyositis characterized by several cumulative manifestations since the age of 51 including gottron sign, calcinosis, Raynaud's phenomenon, polyarthralgias and mild interstitial lung disease. Over the years deposits of calcinosis have increased and appeared in other places, such as the elbows and fingers. Lesions in the left hip were particularly large and painful. Treatment of disease activity with methotrexate and hydroxychloroquine associated with corticosteroids was not successful in reducing calcinosis, leading to progressive functional limitation and chronic pain.

Combination treatment with minocycline 50 mg bid and diltiazem 90 mg/day were performed by the age of 59. The figure shows fast improvement on hip calcinosis after only 3 months of treatment (B) and sustainable improvement after one year of treatment (C).

FIGURE: X-RAY OF THE PATIENTS HIP AT BASELINE, 3 MONTHS (B) AND 1 YEAR (C) AFTER STARTING TREATMENT



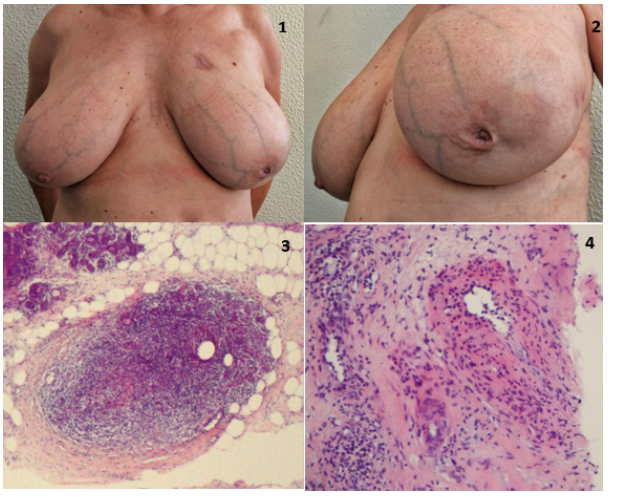
224 - AN IGG4 RELATED DISEASE MAMMARY MANIFESTATION MIMICKING BREAST CANCER

Rita Pinheiro Torres [1, 2], Sandra Falcao[1, 2, 3], Eduardo Cruz[4], Fernando Pimentel-Santos[1, 2, 3], Jaime C. Branco[1, 2, 3]

[1]Serviço de Reumatologia, Centro Hospitalar Lisboa Ocidental | Hospital Egas Moniz, Lisboa, Portugal, [2]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [3]NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [4]Anatomia Patológica, Hospital Egas Moniz, Centro Hospitalar Lisboa Ocidental, Lisboa, Portugal

A fifty-eight-year-old female, with history of IgG4 related vasculitis, under rituximab (375 mg/m2, weekly for four weeks, every six months), methotrexate (25mg subcutaneous, weekly) and prednisolone (5mg daily) therapy, with low disease activity, presented in the Rheumatology appointment with left breast pain, with a two-week evolution. She denied fever, galactorrhea and other symptoms. On the physical examination, it was obvious a left nipple inversion, the presence of orange peel like skin surrounding the aureole, a hard painful swelling with no defined limits located within the upper outer quadrant of the left breast and skin redness of the same area (Figures 1 and 2). She was empirically medicated with an antibiotic, because there was a suspicion of mastitis; no clinical improvement was verified. Therefore, an ultrasound and mammography were requested. The ultrasound raised the hypothesis of breast cancer, reinforced by the findings in the mammography. Simultaneously, the patient underwent a breast biopsy with six frag-

FIGURE. BREAST VASCULITIS



ments, that revealed an intense mixed inflammatory infiltrate, with neutrophil granulocytes and vasculitis lesion, with no malignant neoplasm tissue were identified (Figures 3 and 4). The patient is now repeating the rituximab cycle. The last cycle was not completed due to infectious intercurrence.

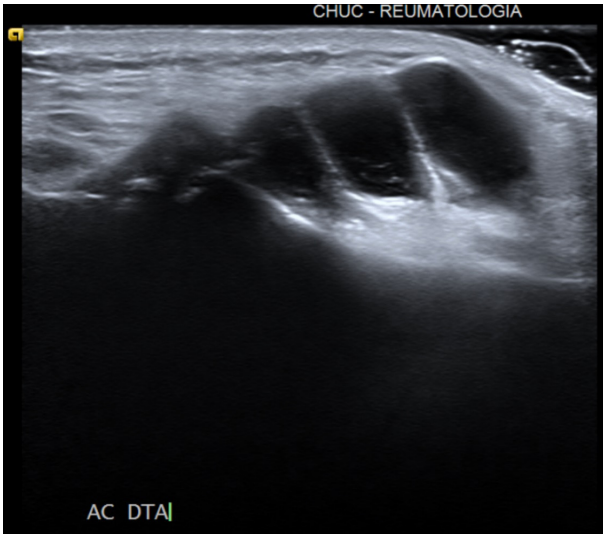
The breast involvement by IgG4 related disease has been described since 2005, under different denominations. However, this appears to be a rare manifestation of a very rare disease. Thus, because of its characteristics, the possibility of breast cancer should always be ruled out.

227 - SINAL DE GEYSER: UM ACHADO ECOGRÁFICO A RECONHECER!

André Saraiva[1], Ana Isabel Maduro[1], Ana Rita Prata[1], Helena Assunção[1], Margarida Coutinho[1]
[1]Serviço de Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

Doente do sexo feminino de 82 anos, com antecedentes de artrite psoriática poliarticular (medicada com salazopirina 1500mg/dia e prednisolona 5mg/dia) e rutura completa do tendão do supra-espinhoso direito. Em consulta de seguimento refere omalgia direita, com noção de tumefação, instalada ao longo de uma semana, sem rubor ou história de traumatismo, febre ou infeção precedente. O exame objetivo revelou marcada tumefação na região da articulação acrómio-clavicular (AC) de consistência duro-elástica e indolor à palpação. O estudo analítico foi normal, incluindo reagentes de fase aguda. Foi realizada ecografia do ombro na qual foi confirmada a rutura total do tendão supra-espinhoso e identificada acentuada distensão da articulação AC por derrame articular apresentando continuidade com estrutura hipoecoica de limites bem definidos de 2.7cm de maior eixo compatível com formação quística (sinal de geyser). (figura 1)

FIGURA: SINAL DE GEYSER- QUISTO DA ARTICULAÇÃO AC



Este sinal ecográfico é caracterizado pelo movimento de fluido desde o local da rutura da coifa dos rotadores, através da bursa subacromial, com descompressão superior através da articulação AC, servindo esta como válvula unidirecional. A identificação deste sinal durante a avaliação ecográfica de uma tumefação/nódulo do ombro indica um processo benigno. Os quistos da articulação AC são, geralmente, indolores e apresentam-se como uma massa firme imediatamente acima da articulação. São mais comuns no sexo masculino e em faixas etárias mais avançadas.

A RMN é o gold standard para o diagnóstico, embora a ecografia também o possa estabelecer

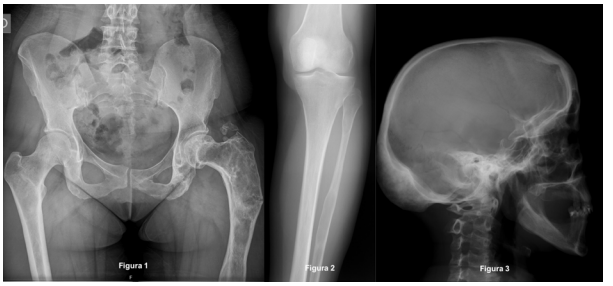
O tratamento poderá ser conservador ou cirúrgico, dependendo da idade, grau de limitação, estado geral de saúde e comorbilidades do doente. Em doentes com patologia reumática ou imunocomprometidos poderá ser prudente optar por terapêutica conservadora, tal como se procedeu neste caso.

240 - DISPLASIA FIBROSA POLIOSTÓTICA - DIAGNÓSTICO EM IDADE ADULTA

Maria Seabra Rato[1], Filipe Oliveira Pinheiro[1], Alexandra Bernardo[1], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal

A displasia fibrosa (DF) é uma doença rara, causada pela mutação no gene GNAS1, que pode conduzir a deformidades ósseas e ao aumento do risco de fratura. A forma poliostótica, que ocorre em 30% dos casos, é diagnosticada habitualmente durante a infância. Descreve-se o caso de uma doente do sexo feminino, de 36 anos, avaliada em consulta por dores ósseas generalizadas com um ano de evolução e história de fraturas do fémur esquerdo aos 8 e 10 anos de idade e fratura do úmero esquerdo aos 13 anos, em contexto de quedas da própria altura. Ao exame objetivo apresentava coxa esquerda vara e dismetria dos membros inferiores. O estudo analítico revelou défice de 25-OH-vitamina D (9ng/mL), com restante metabolismo fosfo-cálcio normal. Realizou radiografias osteoarticulares que evidenciaram múltiplas lesões quísticas,

FIGURA 1. DEFORMIDADE EM CAJADO DO FÉMUR
FIGURA 2: LESÕES QUÍSTICAS E EXPANSIVAS NO PERÓNIO
FIGURA 3: LESÕES ESCLERÓTICAS OCCIPITAIS



com aspeto em fundo de garrafa, expansivas e com adelgaçamento cortical em ambos os fémures, ossos ilíacos, perónio esquerdo, úmeros e cúbito direito. A radiografia da bacia evidenciou também deformidade em cajado da diáfise proximal do fémur esquerdo e a radiografia do crânio mostrou lesões escleróticas occipitais. Realizou também cintigrafia óssea de corpo inteiro que mostrou hiperfixação do radiofármaco em múltiplas localizações compatíveis com DF poliostótica. Iniciou suplementação com vitamina D e pamidronato endovenoso (90mg em dois dias consecutivos de 6-6 meses) com melhoria das dores ósseas. A DF poliostótica é uma doença de prognóstico favorável, mas que pode estar associada a incapacidade significativa. Nesta doente, apesar das manifestações sugestivas da doença em idade pediátrica, o diagnóstico apenas foi estabelecido na idade adulta.

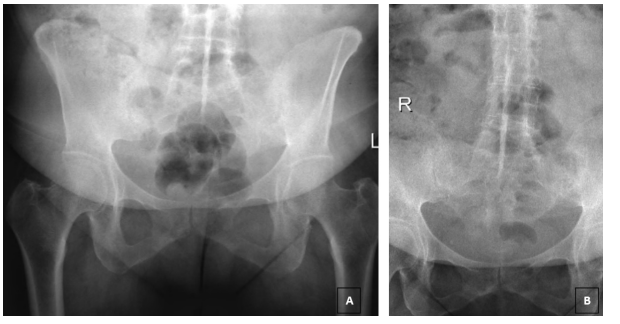
246 - AN INCREASINGLY RARE RADIOGRAPHIC SIGN – THE DAGGER SIGN

Margarida Faria[1], Ricardo Figueira[1], Lídia Teixeira[1], Agna Neto[1], Daniel Carvalho[1], Mário Rodrigues[1]
[1]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal

Introduction. Dagger sign is a characteristic radiographic feature of advanced ankylosis spondylitis (AS) that corresponds to a central radiodense line in the anteroposterior (AP) radiograph of the spine and/or pelvis. It results from the ossification of supraspinous and interspinous ligaments, secondary to the enthesitis. Currently, with timely diagnosis and the new treatments, these signs are less seen in our patients, and due to that we decided to present this case, in which a pelvic and a lumbar AP radiographs of an AS patient presented a dagger sign.

Clinical case. A 57 years-old woman, presented at our Rheumatology center with a prior diagnosis of AS and IgA nephropathy since 18 years old, treated with adalimumab 40 mg, subcutaneous, every 15 days, since the age of 46. At physical examination, she presented a cervical kyphosis and loss of lumbar lordosis along with high limitation in all planes of axial mobility, and a BASDAI score of 4. On pelvic AP radiograph, the patient presented bilateral narrowing of the joint space in both sacroiliac joints along with subcondral sclerosis and erosions (grade 3 sacroiliitis according to the New York criteria) and also a central radiodense line corresponding to the dagger sign (figure 1A). The dagger sign is also visible in the lumbar spine (figure 1B).
Conclusions. The radiographic examination of pelvic and lumbar radiographies of a patient with AS, the radiographic phase of axial spondyloarthritis (AxSp), revealed signs of bilateral sacroiliitis along with signs of enthesopathy – ossification of supraspinous and interspinous ligaments (that results in a radiodense central line that resembles a dagger). AxSp is an inflammatory systemic disease affecting mostly the sacroiliac joints, spine and the enthesis. Nowadays, it is less common to see these characteristic radiographic findings, secondary to chronic enthesitis, but in cases of longstanding disease, these abnormalities can appear and help a diagnosis.

FIGURE. PELVIC AP RADIOGRAPH SHOWING BILATERAL GRADE 3 SACROILIITIS AND A DAGGER SIGN (A), ALSO VISIBLE IN THE LUMBAR AP RADIOGRAPH (B).



REFERENCES:
1. Rajiv Ranjan Kumar, Saket Jha, Aman Sharma, Dagger sign of ankylosing spondylitis, Oxford Medical Case Reports, Volume 2019, Issue 12, December 2019, Pages 502–503,

255 - CLINICAL IMAGES OF ENT INVOLVEMENT IN ANCA ASSOCIATED VASCULITIS

Raquel Freitas[1], Henrique Teixeira[2], Jorge Pestana Lopes[1], Ricardo São Pedro[2], Jorge Dentinho[2], Pedro

Gonçalves[1], Ana Cordeiro[1], Maria José Santos[1, 3]
[1]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [2]Otorhinolaryngology Department, Hospital Garcia de Orta, Almada, Portugal, [3]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Anti-neutrophil cytoplasmic autoantibody (ANCA) associated vasculitis (AAV) are rare necrotizing vasculitides. Ears, nose and throat (ENT) involvement is common in AAV, particularly in Granulomatosis with Polyangiitis (GPA) and Eosinophilic Granulomatosis with Polyangiitis (EGPA). ENT features account for the initial presenting symptoms in the majority of AAV patients with GPA and EGPA.

We report 4 clinical images of 4 patients with different types of ENT involvement in AAV. The first patient is a 47-year old woman with GPA that presented with pulmonary, renal and musculoskeletal involvement associated with epistaxis, rhinorrhea and saddle nose deformity (Fig1.A).

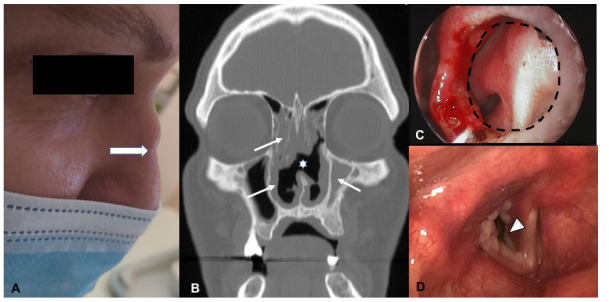
The second patient is a 45-year old man with long standing GPA with chronic sinonasal inflammation and a progressive midline destructive lesion (Fig1.B). He also has interstitial lung disease.

The third patient is a 46-year old woman with limited GPA, with only ENT involvement. She presented with nasal septum perforation and sinonasal inflammation (Fig1.C).

The fourth patient is a 58-year old woman with multi-systemic involvement, including otitis media with effusion, nasal crusting and subglottic stenosis (Fig1.D).

FIGURE 1. DIFFERENT TYPES OF ENT INVOLVEMENT IN ANCA ASSOCIATED VASCULITIS

A – Saddle nose deformity; B – Nasal septum perforation (asterisk) and sinonasal inflammation (arrows); C – Nasal septum perforation on nasal endoscopy. D – Subglottic stenosis (arrow head)

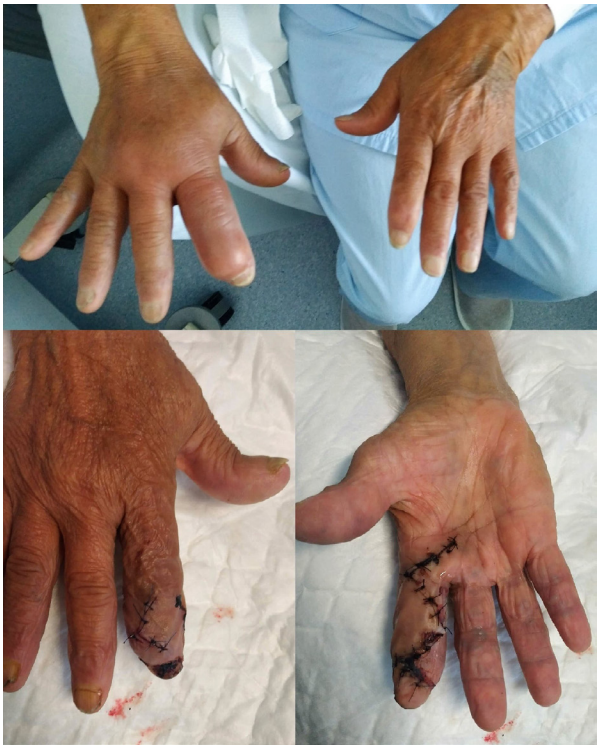


257 - DACTILITE - DIAGNÓSTICO DIFERENCIAL COM O AUXÍLIO DA HISTOLOGIA

Filipe Oliveira Pinheiro[1], Pedro Madureira[1], Ana Martins[1], Daniela Santos Oliveira[1, 2], Maria Seabra Rato[1], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Center for Health Technology and Services Research (CINTESIS),, Faculdade de Medicina da Universidade do Porto, Porto, Portugal, Porto, Portugal

Doente do sexo feminino, 81 anos, recorreu ao SU por dor e sinais inflamatórios exuberantes do 2º dedo da mão direita com 2 semanas de evolução, sem resposta a 2 cursos de antibioterapia oral. Fez ecografia que mostrou marcada densificação dos tecidos moles envolventes do dedo indicador da mão direita, sobretudo na articulação interfalângica distal. Perante a suspeita de etiologia infeciosa foi submetida a desbridamento cirúrgico de necrose dos tecidos profundos com atingimento da bainha, polia e do tendão flexor superficial do dedo, com isolamento de S. epidermidis multissensível e identificação de tofo gotoso em histologia. Melhoria dos sinais inflamatórios após o procedimento e posteriormente com a instituição de prednisolona e antibioterapia dirigida.

FIGURA.



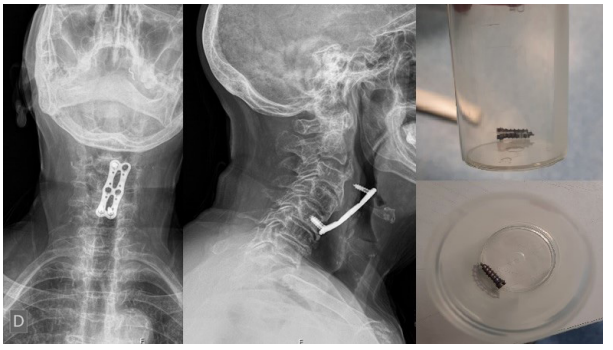
261 - UMA CAUSA EXCECIONALMENTE RARA DE CERVICALGIA E DISFAGIA

Filipe Oliveira Pinheiro[1], Pedro Madureira[1], Flávia Moreira[2], Joana Almeida[2], Maria Seabra Rato[1], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Serviço de Ortopedia e Traumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal

Doente do sexo feminino, 82 anos, com história de cirurgia cervical prévia em 2013. Nos últimos 3 anos vinha apresentando quadro de cervicalgia de intensidade progressiva, e associadamente quadro de diafagia progressiva com 5 meses de evolução. Recorreu ao SU por ter apresentando, aquando da alimentação, episódio de disfagia e regurgitação, com conteúdo metálico, que a doente assegurava ser um parafuso colocado aquando da cirurgia cervical - trouxe o mesmo ao SU. As radiografias no SU demonstraram deslocação da parte superior da placa com ausência de um dos parafusos. A doente foi internada tendo-se evidenciado fístula com o esófago, com necessidade de extração do material e encerramento da fístula.

FIGURE.



270 - OSTEOCONDROMATOSE: UM ACHADO RADIOLÓGICO

Susana P. Silva[1], Carolina Mazeda[1], Joao Paulo Vilas-Boas[1], Gisela Eugénio[1], Anabela Barcelos[1]

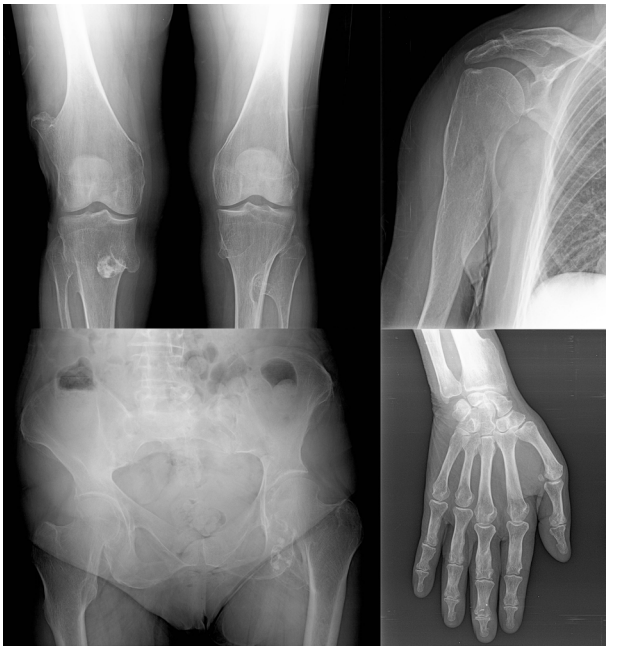
[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal

Introdução: A osteocondromatose múltipla hereditária, previamente denominada exostose múltipla hereditária, carateriza-se pelo crescimento de tumores ósseos benignos, solitários ou múltiplos, os osteocondromas. A maioria são assintomáticos, e achados incidentais, sendo a deformação a maior queixa associada.

Caso Clínico: Mulher de 61 anos, sem antecedentes pessoais de relevo, enviada à consulta de reumatologia, por deformação óssea associada a dor de ritmo inespecífico no joelho direito. Ao exame objetivo, apresentava 145cm de altura e 55kg, mobilização dos ombros e das coxofemorais limitada e tumefações ósseas do joelho direito na face lateral e posterior. Tinha 3 filhos, dos quais o mais novo também apresentava deformações num joelho e braço. Além disso, referia também que todos os seus irmãos apresentavam uma baixa estatura e deformidades ósseas associadas. Foram realizadas radiografias do esqueleto que revelaram alargamento das metáfises do rádio, do úmero, do fémur e da tíbia, bilateralmente, e osteocondromas (figura 1). Analgesia e fisioterapia contribuíram para a melhoria dos sintomas, sem necessidade de intervenção cirúrgica.

Discussão: Os osteocondromas podem surgir em qualquer região do esqueleto e podem levar à diminuição do crescimento do esqueleto e a baixa estatura, a deformidade óssea, a limitação na mobilidade, a osteoartrose precoce e a conflitos com compressão de estruturas neuro-vasculares. Estima-se que a maior parte dos indivíduos sejam diagnosticados na infância. O risco de transformação maligna, embora aumente com a idade, não ultrapassa os 5%. A sua gestão passa pela vigilância e abordagem cirúrgica nos casos de estética, limitação ou conflito.

FIGURA: RAIO-X DOS JOELHOS, ÚMERO DIREITO, BACIA, PUNHO E MÃO DIREITA COM MÚLTIPLOS OSTEOCONDROMAS.



275 - PARTO OU ARTROPLASTIA? DILEMA DE UMA OSTEOPOROSE TRANSITÓRIA DA ANCA

Diogo Guimarães da Fonseca[1], Beatriz Samões[1], Tiago Beirão[1], Romana Vieira[1], P Pinto[1]
[1]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Doente do sexo feminino, 30 anos, com fratura de fragilidade do colo do fémur, após queda da própria altura, às 38 semanas da gravidez (Figura 1). Foi realizado parto por cesariana (Figura 2) e passado 1 semana foi submetida a artroplastia total da anca direita (figura 3). No seguimento do estudo desta doente, foi evidenciado um quadro de osteoporose transitória da anca, uma patologia rara, mas que pode acometer as mulheres principalmente no terceiro trimestre da gravidez e nas situações mais graves pode inclusivamente condicionar uma fratura de fragilidade do colo do fémur como se verificou neste caso.

FIGURA 1 - 38 SEMANAS DE GRAVIDEZ.
FIGURA 2 - APÓS PARTO POR CESARIANA.
FIGURA 3 - APÓS ARTROPLASTIA TOTAL DA ANCA

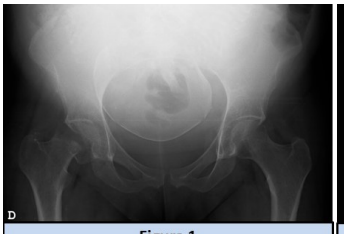


Figura 1

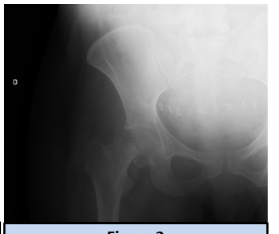


Figura 2

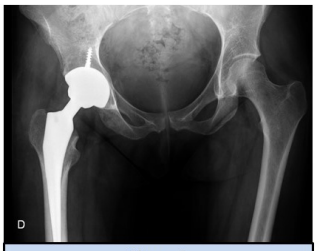


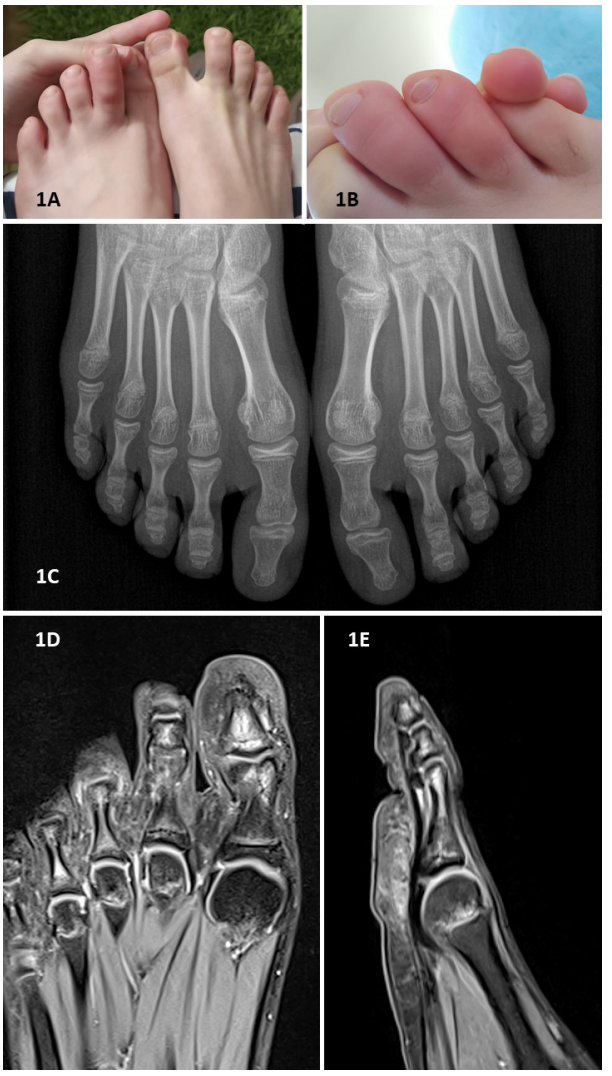
Figura 3

280 - HIPERSINAL MEDULAR ÓSSEO EM RESSONÂNCIA MAGNÉTICA DE LESÕES DE PÉRNIO: COVID OU NÃO? EIS A QUESTÃO.

Francisca Guimarães[1], Diogo Roriz[2], Teresa Fernandes[3, 4], Daniela Peixoto[1]
[1]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [2]Radiologia, Unidade Local de Saúde do Alto Minho, Viana do Castelo, Portugal, [3]Serviço de Radiologia, Hospital Privado de Viana do Castelo, Viana do Castelo, Portugal, [4]Radiologia, JCC diagnóstico por imagem, Viana do Castelo, Portugal

Introdução: As lesões de pérnio resultam de uma resposta inflamatória vascular superficial, que surge tipicamente em resposta à exposição ao frio e que ocorre tipicamente em crianças e mulheres de meia-idade. Durante a pandemia COVID-19, têm sido descritos a nível mundial inúmeros casos de lesões pérnio-like afetando predominantemente os dedos dos pés, mas também mãos, calcanhares, nariz e orelhas, que se pensa que poderão ter relação com a infeção pelo COVID-19, motivo pelo qual têm sido designadas de COVID-toes.
Caso Clínico e descrição de imagem: Adolescente de 12 anos, previamente saudável, foi observada em consulta de Reumatologia por dor e tumefação global de vários dedos de ambos os pés, com cianose e rubor associado, bem como prurido, com 1 mês de evolução. Negava história pessoal ou familiar de psoríase ou doença inflamatória intestinal, bem como sintomatologia evocadora de conectivite. Negava contactos conhecidos com doentes COVID positivos, mas referia período de cefaleias e náuseas no mês prévio, contudo, sem outros sintomas associados e que resolveu espontaneamente. Ao exame objectivo apresentava tumefação global de 3 dedos dos pés, bilateralmente, com períodos de rubor ou cianose associados (Figura 1A/1B). Foi solicitada radiografia onde se constatava apenas tumefação de partes moles, mas sem alterações na estrutura trabecular óssea ou erosões (Figura 1C). Foi solicitada ecografia articular que excluiu artrite, tenossinovite ou entesite, mas demonstrava irregularidades da cortical óssea da falange média. Para melhor caracterização destas alterações foi solicitada ressonância magnética que demonstrou "edema medular ósseo a afectar a epífise da cabeça dos metatarsos e falanges, com hipossinal nas sequências de supressão de gordura, a afectar a falange intermédia do 3º e 4º dedos" (Figura 1D/1E). Analiticamente o hemograma não evidenciava citopenias, os parâmetros inflamatórios eram negativos, a função tiroidea normal e os anticorpos-antinucleares negativos. Colheu serologias IgG e IgM para a infeção COVID que se encontram em curso.
Discussão: Com o número crescente de casos reportado de lesões pérnio-like durante pandemia COVID-19, tem havido uma crescente discussão sobre se estas lesões representam uma manifestação associada a esta infeção ou se apenas um epifenómeno. Esta discussão prende-se pelo facto de vários estudos reportarem que, numa percentagem significativa de doentes, o teste PCR e o estudo serológico é negativo, o que pode ser justificado pelo facto de se tratar

FIGURA 1



maioritariamente de uma manifestação tardia (após o clearance viral), e ocorrer em doentes assintomáticos ou com doença ligeira, com pouca resposta imunitária e reduzida produção de anticorpos. Adicionalmente alguns autores defendem que não existe relação causal com a infeção pelo SARS-COV, mas poder resultar da mudança de estilos de vida verificados durante o confinamento, como maior sedentarismo, deambular descalço dentro da habitação, maior consumo de café, tabaco, etc.

Por outro lado, apesar de existir um crescente interesse no estudo das alterações imagiológicas das manifestações musculoesqueléticas do COVID-19, até à data, nenhum estudo descreve os achados imagiológicos dos COVID-toes. Porém, um estudo recente descreve que, tal como noutras condições microvasculares, poderão surgir alterações de edema medular ósseo distal na ressonância magnética, como verificado no caso descrito.

282 - POLICONDRITE RECIDIVANTE – A IMPORTÂNCIA DA VISÃO INTEGRADA DO REUMATOLOGISTA

Diogo Guimarães da Fonseca[1], Maria Seabra Rato[2], Filipe Oliveira Pinheiro[2], Lúcia Costa[2]
[1]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal

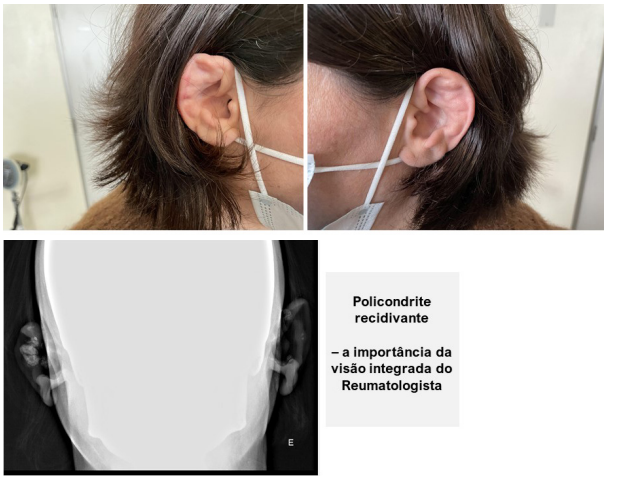
Doente do sexo feminino, 49 anos de idade, encaminhada a consulta de Reumatologia por poliartralgias de ritmo inflamatório a nível dos punhos e pequenas articulações das mãos.

Ao exame objetivo apresentava poliartrite simétrica dos punhos e metacarpofalângicas. À inspeção do pavilhão auricular observou-se deformação da cartilagem auricular bilateralmente, de aspeto lobulado.

Revisitando a anamnese, a doente referiu há cerca de três anos, episódio de tumefação exuberante da cartilagem auricular bilateralmente, tendo sido avaliada nos Cuidados de Saúde Primários e medicada com antibioterapia e posteriormente corticoterapia. Desde então com deformação da cartilagem auricular mas que nunca valorizou.

A radiografia convencional revelou depósitos de

FIGURA: CARTILAGEM AURICULAR EM COUVE-FLOR BILATERALMENTE E CALCIFICAÇÕES RADIOGRÁFICAS AURICULARES



cálcio na cartilagem auricular bilateralmente, em maior número à direita. De destacar ainda que as radiografias das mãos e punhos não apresentavam qualquer lesão erosiva e que os anticorpos - fator reumatóide e anticorpo anti-peptídeo citrulinado cíclico - eram negativos.

Trata-se, portanto, de um caso de condrite auricular bilateral e poliartrite periférica seronegativa não erosiva, em provável contexto de policondrite recidivante, para já sem evidência de atingimento nasal, respiratório, ocular ou vascular.

288 - MANIFESTAÇÕES MUSCULOESQUELÉTICAS DA ACROMEGALIA COMO FORMA DE MANIFESTAÇÃO INICIAL.

Emanuel Costa[1], Diogo Esperança Almeida[1], Ana Margarida Correia[1], Marcos Cerqueira[1], Joana Leite Silva[1], Ana Ribeiro[1], Joana Sousa-Neves[1]

[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

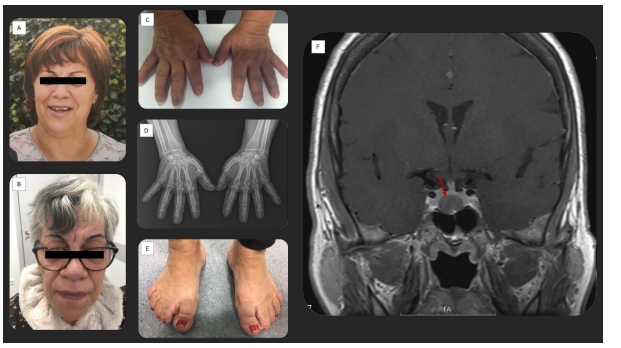
Uma doente do sexo feminino de 55 anos foi referenciada à consulta de reumatologia por artralgias de ritmo mecânico envolvendo as pequenas articulações das mãos e noção de tumefação global dos dedos. À avaliação, a doente referia início das queixas com evolução de dois anos, rigidez de curta duração e parestesias no território do nervo mediano bilateralmente. Não existia história sugestiva de ocorrência de artrite ou fenómeno de Raynaud. Ao exame físico, apresentava fácies grosseira com sulcos marcados, prognatismo, base do nariz alargada e diastemas dentários (A e B), mãos e pés quadrados com alargamento global de tecidos moles (C e E). O teste de Phallen era positivo bilateralmente.

Os achados motivaram o inquérito por outra sintomatologia da qual se destaca a presença de roncopatia de início recente, noção de desfiguração progressiva e necessidade de aumento de tamanho do calçado (37-41).

O estudo analítico mostrou aumento de IGF1 673 ng/mL (N 45-210). A Ressonância Magnética do crânio revelou lesão isointensa em T1 e hipointensa em T2 com 13*15 milimetros de dimensão colocando-se como hipótese de diagnostico principal a de macroadenoma (F).

A doente foi submetida a exérese cirúrgica transesfenoidal e o exame histológico confirmou o diagnóstico clínico de adenoma hipofisário secretor de hormona de crescimento.

FIGURA 1: CARACTERÍSTICAS CLINICAS E IMAGIOLÓGICAS DA ACROMEGALIA.



A acromegalia é uma doença caracterizada pelo aumento da libertação de hormona de crescimento e, consequentemente, fator de crescimento semelhante à insulina 1 (IGF1), mais frequentemente por um adenoma hipofisário. A exposição prolongada ao excesso hormonal conduz a uma desfiguração somática progressiva e a uma ampla variedade de manifestações sistémicas que estão associadas ao aumento da mortalidade[1].

Entre as manifestações clínicas da acromegalia, a artropatia é frequente e pode ser grave e incapacitante. Em alguns casos, a artropatia pode ser a forma de apresentação da doença[2].

Figura 1: A- Fotografia cedida pela doente do ano de 2015; B- Fotografia da doente no momento da observação em 2020; C- Mãos quadradas com aumento de tecidos moles; D- Radiografia das mãos com redução do espaço articular e hipertrofia das falanges distais com aparência de pá; E- Pés quadrados com aumento de tecidos moles; F- Corte coronal em ponderação T1 de RMN do crânio mostrando adenoma hipofisário (seta).

REFERÊNCIAS:

1. Colao A, Grasso L, Giustina A, Melmed S, Chanson P, Pereira A et al. Acromegaly. Nature Reviews Disease Primers. 2019;5(1).
2. Claessen K, Mazziotti G, Biermasz N, Giustina A. Bone and Joint Disorders in Acromegaly. Neuroendocrinology. 2015;103(1):86-95.

290 - MAIS VALE PREVENIR...

Patrícia Pires[1], Nuno Mendonça[1], Ricardo Pinto Rocha[1]

[1]Serviço de Ortopedia B, Hospital Prof. Doutor Fernando Fonseca, Amadora, Portugal

A espondilite anquilosante, patologia reumática, crónica e sistémica, é um protótipo das espondiloar-

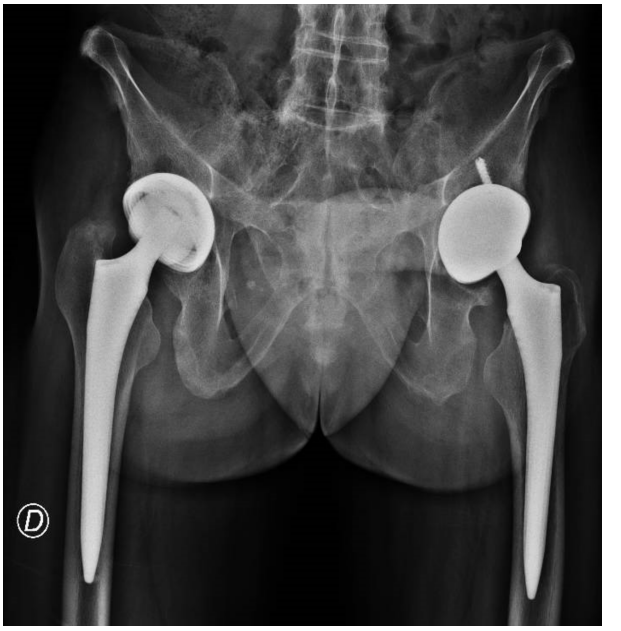
tropatias seronegativas, sendo caracterizada por inflamação e dor em várias articulações, especialmente ao nível da coluna vertebral. Está associada a diminuição da actividade física, a fadiga, a distúrbios do sono, a ansiedade, a depressão e stress, podendo desta forma restringir as actividades da vida diária e a qualidade de vida do indivíduo.

Assim, a espondilite anquilosante é uma patologia que atinge preferencialmente o esqueleto axial, mas pode associar-se a grandes articulações como o ombro e a anca.

Para além da patologia degenerativa da anca pode associar-se à contractura em flexão da anca, o que agrava a deformidade cifótica da coluna.

O tratamento passa por artroplastia bilateral, sendo que o acetábulo antevertido e vertical que aumenta o risco de luxação anterior.

FIGURA: ESPONDILITE



SPR

SOCIEDADE
PORTUGUESA DE
REUMATOLOGIA