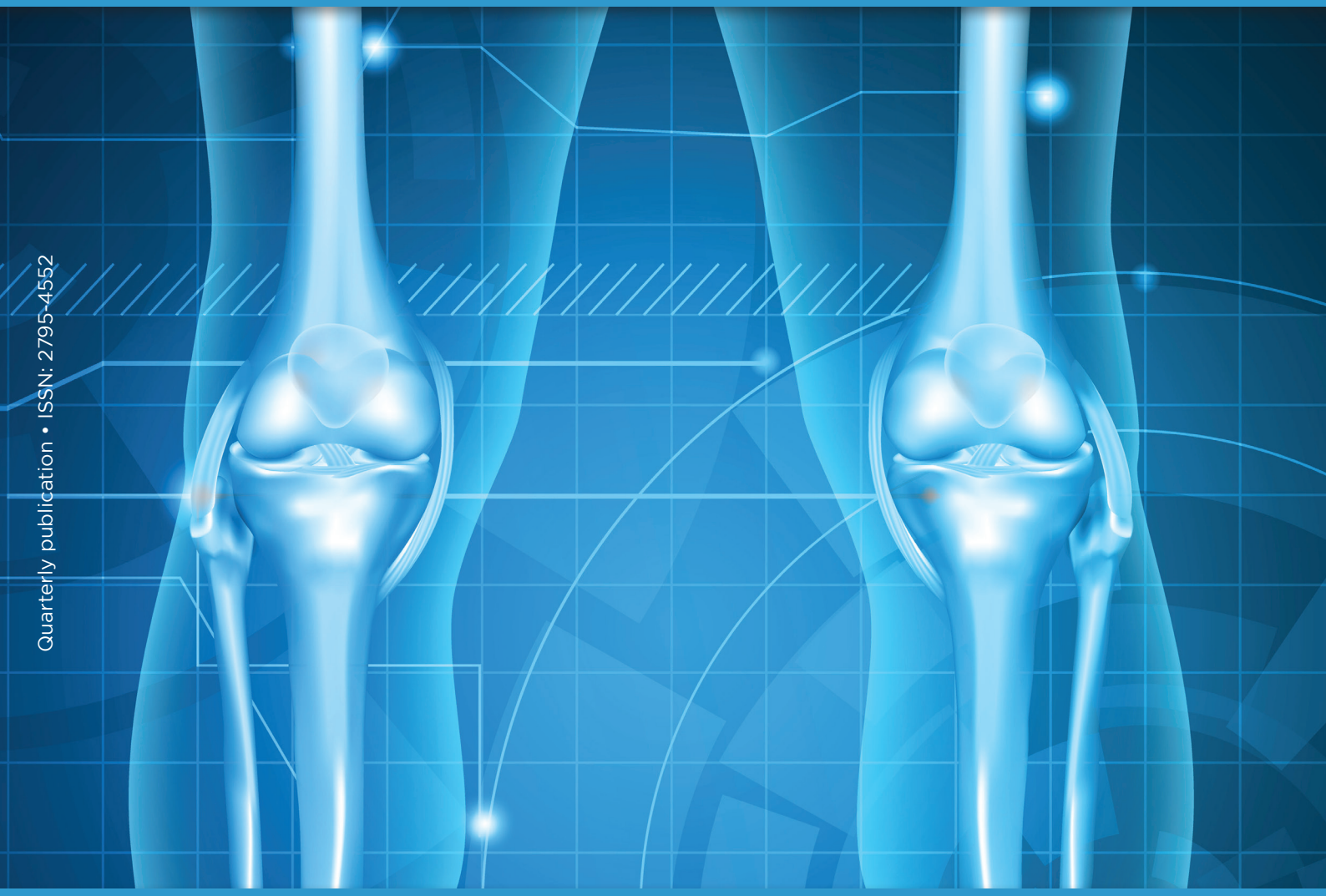
ARP RHEUMATOLOGY

October/December 2022 • Vol 1 • Issue 4 • (XXIV CPR Special Edition)

SPR SOCIEDADE PORTUGUESA DE REUMATOLOGIA

@ARPrheumatology

The official Journal of the Portuguese Society of Rheumatology



Quarterly publication • ISSN: 2795-4552

www.arprheumatology.com

CPR 2022

XXIV CONGRESSO PORTUGUÊS DE REUMATOLOGIA

Sociedade Portuguesa de Reumatologia
Avenida de Berlim, N. 33b
1800-033 Lisboa
secretariado@spreumatologia.pt

Registry
Exempt of registration with the I.C.S. under the terms of paragraph A) of N. 1 of article 12. of Regulatory Decree no. 8/99, of 9 June.

Legal Deposit
86.955/95

Periodicity
Quarterly Publication
ISSN: 2795-4552

Journal referred in Index Medicus, Medline, Pubmed since January 2006.

Journal selected for coverage in Thomson Reuters products and custom information services.
This publication is indexed and abstracted since January 2007 in the following:
- Science Citation Index Expanded (also known as SciSearch®)
- Journal Citation Reports/Science Edition

ARP RHEUMATOLOGY

October/December 2022 • Vol 1 • Issue 4
(XXIV CPR Special Edition)

Sumário

ARP RHEUMATOLOGY
October/December 2022 • Vol 1 • Issue 4
(XXIV CPR Special Edition)
CPR 2022 XXIV CONGRESSO PORTUGUÊS DE REUMATOLOGIA

Sumário

POSTERS – TRABALHOS ORIGINAIS

ARP RHEUMATOLOGY
October/December 2022 • Vol 1 • Issue 4
(XXIV CPR Special Edition)
CPR 2022 XXIV CONGRESSO PORTUGUÊS DE REUMATOLOGIA

Sumário

ARP RHEUMATOLOGY
October/December 2022 • Vol 1 • Issue 4
(XXIV CPR Special Edition)
CPR 2022 XXIV CONGRESSO PORTUGUÊS DE REUMATOLOGIA

Sumário

Sumário

Sumário

CPR 2022
XXIV CONGRESSO PORTUGUÊS DE REUMATOLOGIA

SPR SOCIEDADE PORTUGUESA DE REUMATOLOGIA

Boas-vindas

Caros Colegas,

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

Este é um ano especial, o ano em que se comemoram os 50 anos da Sociedade Portuguesa de Reumatologia!!

A SPR, a Reumatologia portuguesa, os reumatologistas, os profissionais de saúde que trabalham na área da Reumatologia, as Associações de doentes e os doentes reumáticos estão de Parabéns! Nestes 50 anos muito avançámos no diagnóstico, tratamento e reabilitação dos doentes reumáticos, aumentando a sobrevida, a função e a qualidade das suas vidas.

A acessibilidade e os serviços que prestam cuidados aos doentes reumáticos também sofreram uma grande transformação, baseada no aumento do número de especialistas, maior número de serviços, melhor distribuição geográfica, mais organização e maior diferenciação na prestação de cuidados.

O XXIV Congresso Português de Reumatologia é por isso um momento de celebração.

É também um momento de estarmos juntos de novo, enquanto comunidade preocupada e interessada no cuidado aos doentes reumáticos.

Os últimos anos têm sido desafiantes com a pandemia e o isolamento, com a inflação e a guerra. E como bem sabemos, os determinantes sociais influenciam de sobremaneira a saúde das populações em geral e dos doentes reumáticos em particular.

O CPR decorre de 12 a 15 de outubro nos Salgados, Albufeira, Algarve.

Começamos com dois Cursos Pré-Congresso – Vasculites e Ecografia, seguido da Cerimónia de Abertura e atribuição dos Reuméritus. 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, da celebração dos 50 anos, seja um momento de excelência científica, aprendizagem e partilha que todos desejamos.

A atual Direção da SPR cessa funções neste Congresso. É com muita honra que agradecemos a todos os membros da direção, aos membros da assembleia geral, do conselho fiscal, ao presidente eleito, ao secretariado da SPR, equipa do Reuma.pt, equipa da ARP e a todos os que connosco colaboraram nestes dois anos.

Aproveitamos também para desejar as maiores felicidades para a próxima direção.

Parabéns SPR!

Desejamos a todos um Excelente e Muito Participado CPR!

Muito obrigada!!

Helena Canhão
Presidente da Sociedade Portuguesa de Reumatologia

Tiago Meirinhos
Secretário Geral da Sociedade Portuguesa de Reumatologia

CPR 2022
XXIV CONGRESSO PORTUGUÊS DE REUMATOLOGIA

SPR SOCIEDADE PORTUGUESA DE REUMATOLOGIA

COMISSÕES

COMISSÃO ORGANIZADORA

PRESIDENTE: Tiago Meirinhos

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

COMISSÃO CIENTÍFICA

PRESIDENTE: Helena Canhão

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

JÚRIS

JÚRI MELHOR IMAGEM EM REUMATOLOGIA

José Canas da Silva
Teresa Nóvoa
Pedro Gonçalves

JÚRI POSTER CASO CLÍNICO

Viviana Tavares
José Melo Gomes
Cláudia Miguel

JÚRI POSTER TRABALHO ORIGINAL

Carmo Afonso
Cândida Silva
Raquel Marques

JÚRI COMUNICAÇÃO ORAL

Guilherme Figueiredo
Cláudia Vaz
Sandra Falcão

Programa

QUARTA-FEIRA – 12 OUTUBRO

CURSOS PRÉ-CONGRESSO
VASCULITES – COM APOIO VIFOR PHARMA

09:00–10:30 **Vasculites de grandes vasos**
Introdução – Cristina Ponte, Vítor Teixeira, Sofia Barreira
- Como diagnosticar? – Cristina Ponte
- Tratamento: das recomendações à prática clínica – Ana Filipa Águeda

Doença de Behçet
- Manifestações clínicas e tratamento individualizado – Nikita Khmenlinskii

10:30–11:00 *COFFEE BREAK*
11:00–13:00 **Vasculites associadas aos ANCA**
- Diagnóstico e classificação – Carla Macieira
- Atualizações terapêuticas – Vítor Teixeira
- Envolvimento renal: sinais de alerta para o Reumatologista – Estela Nogueira

Conclusão – Vítor Teixeira, Cristina Ponte
13:00–14:30 ALMOÇO

CURSOS PRÉ-CONGRESSO
9º WORKSHOP DE ECOGRAFIA DA ESPER
A ECOGRAFIA NAS DOENÇAS MUSCULOESQUELÉTICAS REUMÁTICAS INFLAMATÓRIAS – COM APOIO ABBVIE

09:00–09:30 **O contributo da US no diagnóstico e monitorização das doenças musculoesqueléticas inflamatórias –** Fernando Saraiva
09:30–10:00 **Quantificação da sinovite por US** – Flávio Costa
10:00–10:30 **Quantificação da entesite por US** – Filipa Teixeira
10:30–10:40 Discussão
10:40–11:00 COFFEE BREAK
11:00–12:30 **Projeção e discussão interativa de imagens US** – Miguel Sousa e Margarida Oliveira
13:00–14:30 ALMOÇO

AUDITÓRIO

14:30–16:00 **Sessão de abertura e entrega Prémio Reuméritus**
- Revisão da evolução da Reumatologia nos últimos 30 anos – Jaime Branco
- Evolução epidemiológica das Doenças Reumáticas em Portugal: para onde caminhamos? – Raquel Lucas

16:00–17:00 **SIMPÓSIO AMGEN**
Moderador: Carlos Vaz
- É fácil implementar e articular uma FLS/Consulta de fraturas de fragilidade? Experiência prática – José Manuel Cancio
- Tratamento sequencial na osteoporose. Uma estratégia a implementar? – Ana Maria Rodrigues

17:00–17:30 COFFEE BREAK
17:30–19:00 **Sessão I – SESSÃO PATROCINADA ASTRAZENECA**
Moderadores: Filipe Barcelos, Luís Inês
- Atualização da abordagem diagnóstica e terapêutica na Síndrome de Sjögren – Vasco Romão
- Atualização terapêutica no Lúpus Eritematoso Sistémico – Filipa Farinha

Comunicações Orais:
- The role of facial, occipital, subclavian and carotid arteries ultrasound in the diagnostic assessment of giant cell arteritis (97) – Joana Martins Martinho
- Predictors of muscle involvement in systemic sclerosis (60) – Eduardo Dourado
- Giant cell arteritis relapse risk – could the extent of vessel involvement on temporal and axillary arteries ultrasound be a prognostic marker? (185) – Diogo Esperança Almeida

SALA MGF

17:30–19:00 **Sessão I MGF: Avaliação diagnóstica e referenciação em Reumatologia**
Coordenador: José António Costa
Moderadores: José António Costa, José António Pereira da Silva
- Grandes Síndromes em Reumatologia – Catarina Dantas Soares, Francisca Guimarães
- Referenciação em Reumatologia – José António Pereira da Silva
- Discussão e closing remarks

QUINTA-FEIRA – 13 OUTUBRO

AUDITÓRIO

09:00–10:30 **Sessão II – SESSÃO PATROCINADA BOEHRINGER-INGELHEIM**
Moderadores: Ana Cordeiro, Carlos Vaz
- Agentes antifibróticos nas Doenças Reumáticas: Quais? Quando e como usar? – Ana Catarina Duarte
- Painéis de anticorpos extraíveis do núcleo: O que há de novo? Aplicações práticas – Maria José Sousa

Comunicações Orais:
- Interstitial lung disease in mixed connective tissue disease: clinical and serological associations (51) – Manuel António
- Nintedanib in Connective tissue disease-interstitial lung disease: experience of a tertiary pneumology center (236) – Francisca Guimarães
- Cardiac involvement in idiopathic inflammatory myopathies: when should we look for it? (95) – Matilde Bandeira

10:30–11:30 COFFEE BREAK | **SESSÃO DE POSTERS 1**

11:30–12:30 **SIMPÓSIO PFIZER**
- Shedding Light upon Challenging Topics in RA Management
  Palestrantes: José António Pereira da Silva, Miguel Bernardes, Vasco Romão

12:30–14:00 ALMOÇO

14:00–15:00 **SIMPÓSIO NOVARTIS Há conversa sobre AP, com ...**
Tiago Meirinhos – Reumatologista
Pedro Mendes Bastos – Dermatologista
Gonçalo Proença – Cardiologista
Gustavo Jesus – Psiquiatra

15:00–16:30 **Sessão III - A importância do Reuma.pt para os médicos, doentes e sociedade**
Moderadores: Ana Maria Rodrigues, Maria José Santos
- Importância do Reuma.pt para os doentes – Elsa Frazão Mateus
- Importância do Reuma.pt para a sociedade – Valter Fonseca
- Importância do Reuma.pt para os médicos – Helena Canhão
- Lançamento da nova área do doente reuma.pt

Comunicações Orais:
- Reuma.pt/myositis – the Portuguese registry of inflammatory myopathies (22) - Eduardo Dourado

- Cycling versus Swapping strategies in Psoriatic Arthritis: results from the Rheumatic Diseases Portuguese Register (150) – Francisca Guimarães
- Adverse events in patients with inflammatory joint diseases: results from the EULAR Coronavirus Vaccine (COVAX) physician-reported registry (183) – Ana Lúcia Fernandes

16:30–17:30 COFFEE BREAK | **SESSÃO DE POSTERS 2**

17:30–18:30 **SIMPÓSIO MSD**

Persisting for the future

- Abertura – Paula Martins de Jesus
- Adiposopatia nas espondilartrites – Paula Freitas
- Conclusões & Encerramento

SALA MGF

09:00–10:30 **Sessão II MGF – Terapêutica da dor**

Coordenadora: Daniela Santos Faria

Moderadores: Daniela Santos Faria, Miguel Martins da Cunha

- Caso clínico "Ombro doloroso" – Hugo Parente
- Anti-inflamatórios e analgésicos – Teresa Martins Rocha
- Infiltrações periarticulares e articulares – Diogo Esperança Almeida
- Discussão / debate: a visão da Reumatologia e da MGF

10:30–11:30 COFFEE BREAK | **SESSÃO DE POSTERS 1**

11:30–12:30 **Sessão III MGF – Imagiologia musculo-esquelética para MGF**

Coordenadores: Augusto Faustino, Tiago Saldanha

- A especificidade das doenças reumáticas e a sua implicação na imagiologia músculo-esquelética – Augusto Faustino
- Exames de Imagem em Reumatologia – Tiago Saldanha

12:30–14:00 ALMOÇO

18:00–19:30 **Assembleia Geral Eleitoral**

SEXTA-FEIRA – 14 OUTUBRO

AUDITÓRIO

09:00–10:30 **Sessão IV** – Moderadores: Patricia Nero, Miguel Bernardes

- Manifestações cutâneas das Doenças Reumáticas – Cristina Sousa
- Manifestações gastrintestinais das Doenças Reumáticas – Fernando Magro

Comunicações Orais:

- Enthesitis of the hand is a dominant lesion in Psoriatic Arthritis and may help distinguishing it from Rheumatoid Arthritis: case-control, single-centre, Ultrasound study (131) – Diogo Esperança Almeida
- Is Inflammation-driven Bone Loss Associated with Two-year Bone Formation at the Same Vertebra in Axial Spondyloarthritis? – a Multilevel MRI and Low Dose CT Analysis from the Sensitive Imaging of Axial Spondyloarthritis (SIAS) Cohort (161) – Mary Lucy Marques
- Direct and indirect effect of TNFi on BASMI components in people with axial spondyloarthritis: a longitudinal study (190) – Ana Sofia Pinto

10:30–11:30 COFFEE BREAK | **SESSÃO DE POSTERS 3**

11:30–12:30 **SIMPÓSIO ABBVIE**

RINVOQ: desafie as suas expectativas na AR

Escolha as perguntas, nós damos a resposta!

Moderadora: Filipa Teixeira

Palestrantes: João Lagoas Gomes, Joaquim Polido Pereira, Pedro Madureira, Vítor Teixeira

12:30–14:00 ALMOÇO

14:00–15:00 **SIMPÓSIO LILLY**

2 Perspetivas, 2 Centros, 2 Doses
Baricitinib na Artrite Reumatóide
Moderadores: João Eurico Fonseca, Augusto Faustino
Palestrantes: Helena Santos, Joaquim Polido Pereira

15:00–16:30 **Sessão V** – Moderadores: Inês Cunha, Sérgio Alcino
- Dieta, exercício físico e aconselhamento na mudança comportamental como pilares da terapêutica dos doentes reumáticos – Conceição Calhau, Margarida Rodrigues, Marta Moreira Marques
- Predictors Of 1-Year Readmission in Patients with Hip Fracture in A Monocentric Cohort (33) – Filipe Pinheiro
- Outcomes of SARS-CoV-2 infection in children and adolescents followed at a Portuguese Pediatric Rheumatology Unit (136) – Ana Teresa Melo
- Determinants of Patient and Physician Global Assessment of Disease Activity in Spondyloarthritis (151) – Catarina Dantas Soares

16:30–17:30 COFFEE BREAK | **SESSÃO DE POSTERS 4**

17:30–18:30 **SIMPÓSIO JANSSEN**
What have we learnt with TREMFYA® in PSA so far?
Palestrantes: Patrícia Nero, Paulo Ferreira

20:00–22:00 **Jantar comemorativo dos 50 anos SPR**

SALA MGF

09:00–10:30 **Sessão IV MGF – Lombalgia para MGF**
Coordenador: Miguel Guerra
- Lombalgia – História Clínica e abordagem diagnóstica – Ana Valido
- Exames auxiliares de imagem na Lombalgia – José Marona
- Casos clínicos – João Lagoas Gomes

10:30–11:30 COFFEE BREAK | **SESSÃO DE POSTERS 3**

11:30–12:30 **Sessão V MGF – Osteoporose para MGF**
Coordenador: Filipe Araújo
- Caso clínico – Filipe Santos
- A prevenção da fratura no hospital e no centro de saúde – Liliana Saraiva
- Farmacoterapia: o velho, o novo e o que está para vir – Carolina Mazeda
- Discussão/debate

12:30–14:00 ALMOÇO

AUDITÓRIO

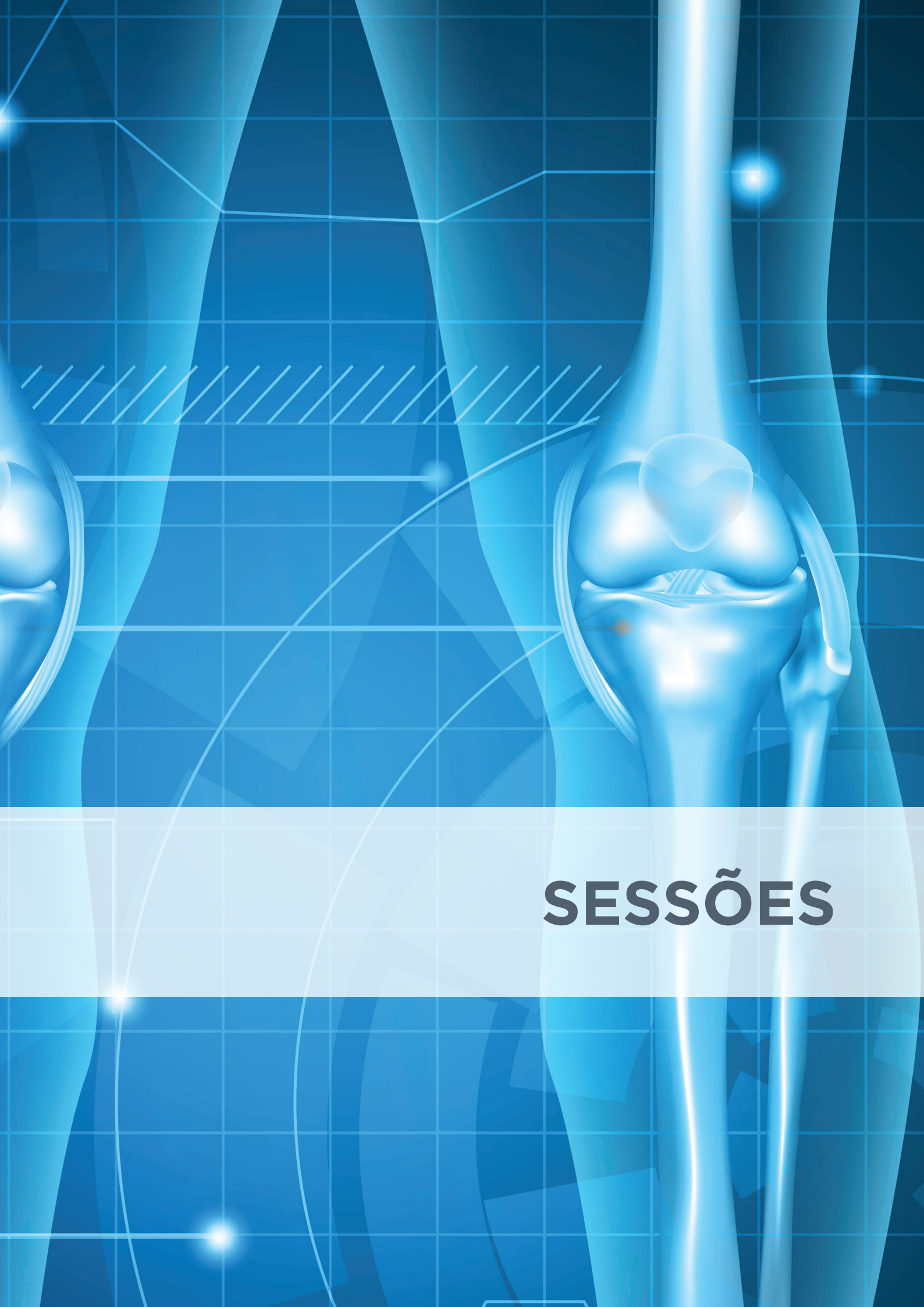
09:00–10:00 **Sessão VI –** Moderadores: Luís Cunha Miranda, José Bravo Pimentão
- Diagnóstico diferencial das Artrites Microcristalinas e atuação – Inês Silva
- Dual-target em Artrite Reumatoide – Aplicação prática – Cátia Duarte

10:00–11:00 **Conferência**
- "Best of" Reumatologia (Últimos 5 anos)
- Investigação e desenvolvimento por Portugueses – Alexandre Sepriano
- Investigação e desenvolvimento no mundo – João Eurico Fonseca

11:00–11:30 COFFEE BREAK

11:30–12:30 **Sessão de Encerramento**
Entrega de Bolsas SPR e Prémios do Congresso

12:30–14:00 ALMOÇO



SESSÕES

ARP Rheumatology 2022;3:13-17 (SUP)

Sessões

SESSÃO I

Atualização da abordagem diagnóstica e terapêutica na Síndrome de Sjögren

Vasco C. Romão[1]
[1]Serviço de Reumatologia, Hospital de Santa Maria, Centro Hospitalar Universitário Lisboa Norte; Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular João Lobo Antunes; Clínica Universitária de Reumatologia, Faculdade de Medicina da Universidade de Lisboa, Centro Académico de Medicina de Lisboa

Os últimos anos têm trazido alguns desenvolvimentos entusiasmantes na área do diagnóstico e tratamento da síndrome de Sjögren. Tratando-se de uma doença clinicamente heterogénea, têm surgido na literatura estudos que identificam a existência de subgrupos de doentes, com características de doença distintas. Aspetos demográficos, o padrão de autoanticorpos (anti-Ro52 ± anti--Ro60 ± anti-La; anti-centrómero, entre outros), o perfil de sintomas gerais (síndrome sicca, fadiga, dor, ansiedade, depressão) ou de manifestações extra-glandulares (envolvimento neurológico, pulmonar, linfadenopático, ...) são alguns dos fatores avançados na direção da estratificação de doentes. Por outro lado, a biópsia e a ecografia das glândulas salivares são centrais para estabelecer o diagnóstico e podem também ajudar na referida individualização de subgrupos de doentes com diferentes prognósticos. A abordagem multidisciplinar é fundamental quer na fase de diagnóstico, quer no tratamento dos doentes com síndrome de Sjögren. Neste sentido, tem havido avanços encorajadores na direção do reforço do arsenal terapêutico desta patologia. Foi publicado recentemente o primeiro ensaio de fase 2 (ianalumab) a atingir o endpoint primário (ESSDAI), bem como outros ensaios clínicos e estudos observacionais *proof-of-concept* de fármacos inovadores (iscalimab, baricitinib) e clássicos (leflunomida) com resultados positivos. Vários ensaios clínicos encontram-se em curso, sugerindo um futuro animador aos doentes com síndrome de Sjögren, que — esperemos — poderão vir a contar em breve com terapêuticas eficazes, seguras e especificamente aprovadas para esta doença.

Atualização terapêutica no Lúpus Eritematoso Sistémico

Filipa Farinha[1]
[1] Hospital Distrital de Santarém

Quando tratamos doentes com Lúpus Eritematoso Sistémico (LES), o nosso objetivo é atingir a remissão ou uma baixa atividade da doença, e prevenir a ocorrência de *flares*, mantendo a mínima dose possível de corticoides. No entanto, com as terapêuticas imunomoduladoras disponíveis, ainda encontramos doentes refratários ou com *flares* frequentes, levando ao acúmulo de dano e redução da qualidade de vida.

Inúmeros novos fármacos têm sido testados em ensaios clínicos ao longo dos últimos anos, mas infelizmente a maioria acaba por ter resultados negativos. Nas últimas décadas, apenas o belimumab tinha sido aprovado para o tratamento do LES, em 2011; no entanto, os doentes com envolvimento ativo e severo do sistema nervoso central ou nefrite lúpica severa haviam sido excluídos dos estudos.

Entretanto, um ensaio clínico na nefrite lúpica acabou por conduzir à aprovação do belimumab também nesta indicação. Dois outros fármacos foram também aprovados recentemente - o anifrolumab, um antagonista do recetor do interferão tipo I, aprovado pela EMA em 2022 no tratamento do LES ativo moderado a severo; e a voclosporina, um novo inibidor da calcineurina, aprovada pela FDA no tratamento da nefrite lúpica.

Serão apresentados sumariamente os estudos que conduziram à aprovação destes fármacos, bem como uma proposta de enquadramento dos mesmos à luz das atuais recomendações para o tratamento do LES.

SESSÃO II

Agentes antifibróticos nas doenças reumáticas: Quais? Quando e como usar?

Ana Catarina Duarte[1]
[1] Serviço de Reumatologia, Hospital Garcia de Orta

O envolvimento pulmonar está descrito na maioria das doenças reumáticas sistémicas (DRS), sendo a doença pulmonar intersticial (DPI) a manifestação mais frequente. O diagnóstico atempado e o tratamento adequado são fundamentais para uma melhoria da morbi--mortalidade associada a esta patologia. Os antifibróticos têm ganho um interesse crescente como terapêutica alternativa/adjuvante à imunosupressão na DPI secundária às DRS.

Presentemente dispomos de dois antifibróticos, o nintedanib e a pirfenidona, ambos aprovados inicialmente para o tratamento da fibrose pulmonar idiopática (FPI). Posteriormente, tendo em conta a fisiopatologia semelhante com a FPI, o seu uso na DPI secundária

às DRS, em particular nos subtipos fibrosantes, tem mostrado resultados promissores em ensaios clínicos e séries de casos.

O nintedanib e a pirfenidona exercem a sua ação anti-fibrótica atenuando a proliferação dos fibroblastos e a produção de proteínas/citocinas associadas à fibrose. Além disso, a pirfenidona parece reduzir a acumulação de células inflamatórias.

Ambos os fármacos são de administração oral, com duas (nintedanib) ou três (pirfenidona) tomas diárias. As queixas gastrointestinais (diarreia, náuseas e vómitos) são as reações adversas mais frequentes, estando também descritas com alguma frequência reações de fotossensibilidade com a pirfenidona. No caso do nintedanib, a inibição do recetor do fator de crescimento endotelial vascular pode estar associada a um risco aumentado de hemorragia. Ambos os fármacos são potencialmente hepatotóxicos, devendo ser feita uma avaliação inicial e monitorização regular dos níveis de transaminases e bilirrubina. O seu uso não está recomendado em doentes com clearance creatinina <30 mL/min ou sob terapêutica de substiuição renal.

Painéis de anticorpos extraíveis do núcleo: o que há de novo? Aplicações práticas

Catarina Castaldo

SESSÃO III

Importância do Reuma.pt para os doentes

Elsa Frazão Mateus

Experiência com o Reuma.pt durante campanha de vacinação contra a COVID-19

Válter Fonseca[1]

[1] Médico; Professor Auxiliar na Faculdade de Medicina de Lisboa; Diretor do Departamento de Qualidade em Saúde da Direção Geral de Daúde, 2018-2022; Coordenador da Comissão Técnica de Vacinação contra a COVID-19, 2020-2022

Nas funções que tive na Comissão Técnica de Vacinação contra a COVID-19 (CTVC) da Direção-Geral da Saúde (DGS) foi necessário, em linha com as recomendações internacionais de boas práticas para decisões de saúde pública, utilizar este Registo Nacional de Doentes Reumáticos (Reuma.pt) que conta já com registos de mais de 30 mil utentes.

Na verdade, a importância dos registos a nível nacional e global é fundamental para a tomada de decisões em saúde que devem ser cada vez mais sustentadas em dados e evidências. Estes registos permitem, não só, fazer uma boa análise de situação e adaptar as recomendações de saúde a cada contexto, mas também, numa fase seguinte, monitorizar e avaliar o impacto dessas medidas.

E, por isso, considero fundamental a prática do registo destas e de outras doenças numa perspetiva de políticas de saúde e para cada vez mais tomarmos decisões baseadas em dados e, sobretudo, dados nacionais. Durante as fases iniciais da campanha de vacinação contra a COVID-19 um dos aspetos críticos para fazer face à ainda pouca disponibilidade de vacinas contra a COVID-19 foi necessariamente priorizar as pessoas que, devido a fatores de risco, mais beneficiavam com a vacinação contra a COVID-19.

Naturalmente que as pessoas com doenças autoimunes e reumatológicas foram avaliadas pela CTVC, e isso foi feito através de um excelente trabalho colaborativo com a Sociedade Portuguesa de Reumatologia (SPR) que, através dos dados do Reuma.pt, nos permitiu perceber aquilo que era o panorama Nacional, na altura, destes doentes quando desenvolviam COVID-19. Com isto, conseguimos ter uma estratificação de risco destes doentes, compará-los com outras doenças e, dessa forma, definir grupos elegíveis para a vacinação contra a COVID-19 de uma forma equitativa e priorizando sempre as pessoas que, numa primeira fase, mais beneficiavam da vacinação contra a COVID-19.

Por isso, tenho uma excelente experiência com este processo que suporta as decisões técnicas em saúde pública, nomeadamente a vacinação, aquilo que é informação que já se tem em Portugal. É exemplar a forma como a SPR tem lidado com esta matéria.

O que nos demonstra, com este breve exemplo, numa área tão crítica da nossa história da saúde - a vacinação contra a COVID-19 - as inúmeras potencialidades de registos nacionais de doentes para a tomada de decisão em saúde no futuro.

A minha mensagem final é de apelo à continuidade deste tipo de iniciativas para aumentar o o conhecimento científico e, em última instância, melhorar os cuidados que prestamos aos nossos doentes em Portugal.

Importância do Reuma.pt para os médicos

Helena Canhão

SESSÃO IV

Manifestações cutâneas das Doenças Reumáticas

Cristina Sousa

Manifestações gastrintestinais das Doenças Reumáticas

Fernando Magro

SESSÃO V

Dieta, exercício físico e aconselhamento na mudança comportamental como pilares da terapêutica dos doentes reumáticos

Conceição Calhau, Margarida Rodrigues,
Marta Moreira Marques

Exercício físico

Margarida Rodrigues[1]
[1] Centro de Reabilitação do Norte

A actividade física, seja ela planeada ou não, resulta em múltiplos benefícios a vários níveis: função cardiovascular e respiratória, força muscular, equilíbrio, coordenação, flexibilidade, assumindo um papel fundamental na prevenção primária e secundária de diversas patologias. Sendo isto verdade para a população geral, os doentes reumáticos não são excepção, sobretudo se atentarmos na grande diversidade de patologias. Considerando os défices e sequelas decorrentes quer da afectação multissistémica, quer dos tratamentos a que estes doentes são sujeitos, existem múltiplos alvos terapêuticos para os quais a actividade/exercício físico deve ser entendido com uma modalidade co-adjuvante, nomeadamente nas síndromes de desuso e défice de força muscular, tendinopatias, dor de ritmo mecânico/inflamatório, fadiga e baixa tolerância ao esforço, alteração da densidade mineral óssea, alteração da função respiratória, entre outros.

Nos indivíduos saudáveis, os benefícios ultrapassam largamente os efeitos adversos. Contudo, entre doentes, é necessário ter em conta que para que tal se verifique é obrigatória a sua avaliação prévia, assumindo especial importância as suas comorbilidades, nomeadamente do foro cardiovascular, as quais poderão beneficiar da prática de actividade física ou constituir um factor limitante à sua realização. O mesmo se aplica à presença de patologia músculo-esquelética e alterações do hemograma. A intensidade adequada é um factor essencial para garantir a segurança do exercício.

Outro aspecto relevante é a estabilidade clínica do doente. Numa fase aguda ou de exacerbação da doença de base, a sua recuperação e a resolução da inflamação, em associação com a prevenção de sequelas e controlo da dor, constituem o principal objectivo de uma intervenção que inclui a prescrição de exercício terapêutico de forma especializada por médico Fisiatra, tendo em vista, simultaneamente, a protecção das estruturas articulares/periarticulares, uma vez que estes doentes apresentam particularidades que poderão contraindicar a prática arbitrária de actividade física.

A fase da doença é igualmente pertinente. Veja-se o exemplo da osteoporose, em que a intervenção para a sua prevenção primária pode ser danosa aquando da instalação da diminuição significativa da densidade mineral óssea.

Por ser algo acessível e benéfico, a prática regular de actividade/exercício físico deverá ser aconselhada e encorajada entre os doentes reumáticos, tendo em vista o seu bem-estar e prevenção quer primária, quer secundária de diferentes complicações/sequelas. Contudo, este aconselhamento deverá ser individualizado e criterioso, tendo em conta as comorbilidades do doente, garantindo a segurança desta intervenção.

SESSÃO VI

Diagnóstico diferencial das artrites microcristalinas e atuação

Inês Silva[1]
[1] CHLO – Centro Hospitalar Lisboa Ocidental

As artrites microcristalinas caraterizam-se pelo depósito intra-articular/periarticular de cristais, sinais inflamatórios exuberantes de início súbito e ativação de resposta sistémica mediada pelo inflamassoma *Nod-Like Receptor Protein 3* (NLRP3). O depósito pode preceder anos até ao surgimento das manifestações clínicas. As formas mais comuns são mediadas pela acumulação de cristais de monourato de sódio (gota úrica) e pirofosfato de cálcio (doença por deposição de cristais de pirofosfato de cálcio - DDCPC); outras menos frequentes contam com cristais de fosfato de cálcio básico, oxalato de cálcio, colesterol e lípidos. Em todos os casos a observação do líquido sinovial por microscopia otica de luz polarizada compensada a fresco ou com uso de corantes especiais é o *gold standard* para o diagnóstico. Recomendações internacionais de gota (ACR 2020, EULAR 2018, *British Society* 2017) e DDCPC (EULAR 2011) incluem no diagnóstico também o recurso a imagem (radiografia, ecografia e tomografia computorizada de dupla energia). O ácido úrico é um metabólico ativo em vários processos patológicos da síndrome metabólica. O tratamento da hiperuricemia é fundamental na prevenção desses mecanismos; alopurinol e colchicina veem reconhecidos os seus benefícios de proteção cardiovascular, assim como alguns fármacos com efeito uricosúrico ligeiro a moderado no tratamento das comorbilidades associadas à gota/hiperuricemia assintomática (losartan, fenofibrato, atorvastatina, inibidores SGLT-2). Novas terapias em investigação para a gota parecem ter resultados promissores. Serão abordadas as diferentes manifestações clínicas, microscópicas e imagiológicas das diferentes artrites microcristalinas; opções de tratamento, assim como as inovadoras técnicas de diagnóstico na avaliação do líquido sinovial e de imagem.

Dual-target em artrite reumatóide: aplicação prática

Cátia Duarte[1]
[1]Assistente Graduada Reumatologia, Centro Hospitalar e Universitário de Coimbra

The prognosis of RA patients changed over the last years due several factors including the treat-to-target (T2T) strategy, which recommends that disease activity is regularly assessed and the immunosuppressive treatment is adjusted as needed to achieve and maintain the target of remission.

However, previous research showed that a large proportion of patients fails a remission status only due high Patient Global Assessment (PGA) of disease activity, despite absence of objective inflammation. PGA is not an indicator of disease activity but rather a marker of disease impact and its inclusion in disease activity scores could lead to a high risk of immunosuppressive overtreatment. Despite control of inflammation is crucial, is not exclusive to reduce impact, and other factors can influence the impact of the disease perceived by the patient.

To overcome such limitations, a Dual-T2T (dT2T) strategy was proposed, considering two separate targets: the inflammatory and the impact target, which should be pursued in parallel and in a complementary way. The inflammatory target should be assessed through objective markers of inflammation (tender and joint counts, C-reactive protein). The impact target should be assessed through Patient Reported Outcomes Measures. We purpose the Patient Experienced symptom State (PESS) as a screening tool and in patients who report a state less than good, the RAID.7 should be applied to identify the domain(s) most affected to guide the most adequate adjunctive therapy.

This presentation aims to present the dT2T strategy and its applicability in clinical practice as a strategy to improve the global prognostic of RA patients.

COMUNICAÇÕES ORAIS

Comunicações Orais

022 - REUMA.PT/MYOSITIS – THE PORTUGUESE REGISTRY OF INFLAMMATORY MYOPATHIES

Eduardo Dourado[1, 2], Ana Teresa Melo[1, 2], Patrícia Martins[1, 2], Matilde Bandeira[1, 2], Vanessa Fraga[3], José Luís Ferraro[4], André Saraiva[5], Marlene Sousa[5], Hugo Parente[6], Catarina Soares[6], Ana Margarida Correia[7], Diogo Esperança Almeida[7], Sara Paiva Dinis [8], Ana Sofia Pinto[8], Filipe Oliveira Pinheiro[9], Maria Seabra Rato[9], Tiago Beirão[10], Beatriz Samões[10], Bernardo Santos[11], Carolina Mazeda[11], Ana Teodósio Chícharo[12], Margarida Faria[13], Agna Neto[13], Maria Helena Lourenço[14, 15], Luisa Brites[16], Marília Rodrigues[16], Joana Silva-Dinis[17], João Madruga Dias[18], Filipe Araújo[19], Nádia Martins[20], Couto M[20], Ana Valido[21], Maria José Santos[2, 3], Sofia C Barreira[1, 2], JE Fonseca[1, 2], Raquel Campanilho-Marques[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]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [4]Serviço de Reumatologia, Hospital Beatriz Ângelo, Loures, Portugal, [5]Rheumatology Department, Centro Hospitalar Universitário de 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, Centro Hospitalar Universitário de São João, Porto, Portugal, [10]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [11]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [12]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, [13]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [14]Serviço de Reumatologia, Hospital de Egas Moniz, Centro Hospitalar de Lisboa Ocidental, Lisboa, Portugal, [15]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [16]Serviço de Reumatologia, Centro Hospitalar de Leiria, Leiria, Portugal, [17]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Central, Lisboa, Portugal, [18]Rheumatology Department, Centro Hospitalar Médio Tejo, Torres Novas, Portugal, [19]Serviço de Reumatologia, Hospital CUF Cascais, Cascais, Portugal, [20]Rheumatology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [21]Serviço de Reumatologia, Unidade Local de Saúde do Litoral Alentejano, Santiago do Cacém, Portugal

Background: The idiopathic inflammatory myopathies (IMM) module of the Rheumatic Diseases Portuguese Register (Reuma.pt/Myositis) is a tool used to systematically evaluate IIM patients.

Objective: To clinically characterise the Reuma.pt/Myositis cohort.

Methods: Multicentre open cohort study, including IIM patients registered in Reuma.pt up to January 2022. Data collected included demographic, clinical, and treatment data and patient-reported outcomes. Data were presented as frequencies and median (interquartile range) for categorical and continuous variables, respectively.

Results: 280 patients were included, 71.4% female, 89.4% Caucasian, with a median age at diagnosis and disease duration of 48.9 (33.6-59.3) and 5.3 (3.0-9.8) years, respectively. Patients were classified as having definite (N=57/118, 48.3%; N=35/224, 15.6%), likely (N=23/118, 19.5%; N=50/224, 22.3%), or possible (N=2/118, 1.7%; N=46/224, 20.5%) IIM by 2017 EULAR/ACR and Bohan-Peter criteria, respectively. Disease subtypes included dermatomyositis (DM, N=122/280, 43.6%), polymyositis (N=59/280, 21.1%), myositis in overlap syndromes (N=41/280, 14.6%), clinically amyopathic DM (N=17/280, 6.1%), nonspecific myositis (N=13/280, 4.6%), mixed connective tissue disease (N=12/280, 4.3%), immune-mediated necrotizing myositis (N=9/280, 3.2%), and inclusion bodies myopathy (N=7/280, 2.5%). Over the course of the disease, the most common symptoms were proximal muscle weakness (N=180/215, 83.7%), arthralgia (N=127/249, 52.9%), erythema (N=63/166, 38.0%), fatigue (N=47/127, 37.0%), Raynaud's phenomenon (N=76/234, 32.5%), and dysphagia (N=33/121, 27.3%), and the most common clinical signs were Gottron's sign (N=75/184, 40.8%), heliotrope rash (N=101/252, 40.1%), Gottron's papules (N=93/237, 39.2%), and arthritis (N=38/98, 38.8%). Organ involvement included lung (N=78/230, 33.9%), oesophageal (N=40/221, 18.1%), and heart (N=11/229, 4.8%) involvements. Most patients expressed myositis-specific (MSA, N=158/242, 65.3%) and/or myositis-associated (MAA, 112/242, 46.3%) antibodies. The most frequent antibodies were anti-SSA/SSB (N=70/231, 30.3%), anti-Jo1 (N=56/236, 23.7%), and anti-Mi2

Table I. Autoantibodies in cancer-associated myositis

Cancer	IIM	Autoantibodies	p value
Breast	DM (3)	Mi2, SRP (+ SSA/SSB), Pm/Scl	
Skin (non-melanoma)	Clinically amyopathic DM, PM	Jo1, SAE1 (+SSA/SSB)	<0.001
Colorectal	DM (2)	Mi2 (2)	<0.001
Kidney	DM	-	<0.001
Lung	DM	-	<0.001
Lymphoma	Inclusion bodies myopathy	-	<0.001
Unknown	DM	-	

(N=31/212, 14.6%). Most patients had a myopathic pattern on electromyogram (N=101/138, 73.2%), muscle oedema in magnetic resonance (N=33/62, 53.2%), and high CK (N=154/200, 55.0%) and aldolase levels (N=74/135, 54.8%) at diagnosis, with median highest CK levels of 1308 (518-3172) and aldolase of 42 (12-121) mg/dL. Neoplasia was found in 11/127 patients (8.7%), most commonly breast (N=3/11, 27.3%), non-melanoma skin (N=2/11, 18.2%), and colorectal (N=2/11, 18.2%) cancer (Table 1). Most patients with cancer-associated myositis had DM (N=8/11, 72.7%) and expressed MSA (N=6/11) and/or MAA (N=3/11). The most used drugs over the course of disease were glucocorticoids (N=201/280, 71.8%), methotrexate (N=117/280, 41.8%), hydroxychloroquine (N=87/280, 31.1%), azathioprine (N=85/280, 30.4%), mycophenolate mofetil (N=56/280, 20.0%), intravenous immunoglobulin (N=55/280, 19.6%), and rituximab (N=45/280, 16.1%). At the last follow-up, there was a median MMT8 of 150 (142-150), modified DAS skin of 0 (0-1), global VAS of 10 (0-50) mm, and HAQ of 0.125 (0.000-1.125).

Conclusions: Reuma.pt/Myositis adequately captures the main features of inflammatory myopathies' patients, depicting in this first report a heterogeneous population with frequent muscle, joint, skin and lung involvements. Of interest, most patients reached low disease activity at the last follow-up appointment.

033 - PREDICTORS OF 1-YEAR READMISSION IN PATIENTS WITH HIP FRACTURE IN A MONOCENTRIC COHORT

Filipe Oliveira Pinheiro[1], Pedro Madureira[1], Maria Seabra Rato[1], Lúcia Costa[1]

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

Background: Readmission after hip fractures is associated with further morbidity and mortality in hip fracture patients, and some risk factors may be preventable. The aim of this study is to evaluate possible predictors of hospital readmission in patients with hip fractures.
Materials and Methods: Retrospective study that included patients admitted to our hospital with hip fracture for 3 consecutive months. Data collected were hospitalization in the past 6 months, hemoglobin (Hb), calcium and vitamin D upon admission, length of stay, and surgery within 48 hours. To assess comorbidities, the Charlson Comorbidity Index (CCI) score was used. Anemia was defined as Hb less than 12g/dl in women and 13g/dl in men. Readmission was evaluated at 1 year. Comparison between groups was performed using the chi-square test, t-test and Mann-Whitney U test. Linear regression analysis was performed.
Results: Eighty patients were included, 69 (86.3%) females, aged 81.2±6.5 years; eleven patients (14.9%) had previous hospitalization; mean hemoglobin level was 12.5±1.2 g/dl, with anemia seen in 34 (42.5%) patients, mean serum calcium was 9.8±1.2 mg/dl, and mean vitamin D was 28.5±14.3 ng/ml. The median

Table I. Measured variables at baseline

Female gender – n (%)	69 (86.3)
Age, years – mean (SD)	81.2 (6.5)
Age > 85 years – n (%)	32 (40.0)
Previous fragility fracture – n (%)	21 (28.4)
Hb, g/dl – mean (SD)	12.5 (1.2)
Vitamin D, ng/ml– mean (SD)	28.5 (14.3)
Calcium, mg/dl– mean (SD)	9.8 (1.2)
CCI – mean (SD)	5.0 (1.2)
Surgery within 48 hours – n (%)	54 (67.5)
Readmission at 1-year – n (%)	29 (36.3)
Kawasaki disease	1 (2.9%)
Chronic nonbacterial osteomyelitis	1 (2.9%)
Rheumatic fever	1 (2.9%)
Comorbilities	
Asthma	1 (2.9%)
Allergic rhinitis	1 (2.9%)
Celiac disease	1 (2.9%)

length of hospital stay was 7 (5-9.5) days and surgery was performed within 48 hours in 54 (67.5%) patients. Mean CCI score was 5.0±1.2. Twenty-nine patients (36.3%) were readmitted within 1 year.

Patients who were readmitted were found to have more frequent anemia (62.1% vs 31.4%, p=0.008), age over 85 years (55.2% vs 31.4%, p=0.037) and hospitalization in the past 6 months (26.9% vs 8.3%, p =0.043). Although without statistically significant differences, there were lower levels of Hb (11.9±0.8 vs 12.6±1.3) and vitamin D (26.5±14.3 vs 29.0±14.6), higher CCI score (6.3±2.3 vs 4.6±1.4) and lower percentage of surgery performed in the first 48h (65.5% vs 68.7%) in patients who were readmitted.

When adjusted for comorbidities, age, gender, length of stay and timing of surgery, anemia (OR 6.805, 95% CI 1.769-26.175, p=0.005) and hospitalization in the past 6 months (OR 6.321, 95% CI 1.177-33.940, p=0.032) were independent predictors of readmission.
Conclusion: Anemia and previous hospitalization within 6 months were associated with 1-year readmission after hip fracture, while other factors such as comorbidities, length of stay and timing of surgery have not been shown to be predictors of readmission in this study.

051 - INTERSTITIAL LUNG DISEASE IN MIXED CONNECTIVE TISSUE DISEASE: CLINICAL AND SEROLOGICAL ASSOCIATIONS

Manuel Silvério-António[1, 2], Martins-Martinho J [1, 2], Ana Teresa Melo[1, 2], Francisca Guimarães[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[3], Daniela Peixoto[3], Eduardo Dourado[1, 2], Nikita Khmelinskii[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]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [4]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [5]Faculdade de Medicina da Universidade do Porto, Departamento de Medicina e CINTESIS, Porto, Portugal, [6]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal, [7]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [8]Serviço de Reumatologia do Hospital Egas Moniz - Centro Hospitalar Lisboa Ocidental (CHLO- E.P.E.), Lisboa, Portugal, [9]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal, [10]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal, [11]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, [12]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [13]Serviço de Reumatologia, Centro Hospitalar do Baixo Vouga, E.P.E., Aveiro, Portugal, [14]Rheumatology Department, Centro Hospitalar Trás-os-Montes e Alto Douro, Vila Real, Portugal, [15]Rheumatology Department, Centro Hospitalar Vila Nova de Gaia/Espinho, Gaia, Portugal, 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) is a rare systemic rheumatic disease characterized by the expression of autoantibodies targeting the U1-ribonucleoprotein and overlapping clinical features of systemic sclerosis, systemic lupus erythematosus, and inflammatory myopathies. Interstitial lung disease (ILD) is present in 47% to 78% of patients and has been associated with higher mortality rates. Associations of ILD with Raynaud's phenomenon, dysphagia, anti-Ro52 antibodies, and a scleroderma pattern on nailfold capillaroscopy have been reported in MCTD patients.
Objective: This study aims to identify clinical and serological associations and independent predictors of ILD for patients with MCTD.
Methods: Multicenter retrospective study using data collected from clinical records. Adult patients who underwent lung computed tomography (CT) and met at least one of four MCTD diagnostic criteria (Sharp, Alarcón-Segovia, Kasukawa, or Kahn criteria) were included. Univariate analysis was performed using Chi-Square, Fischer's Exact, and Mann-Whitney tests, as appropriate. Multivariate analysis was performed using binary logistic regression modelling. The linearity of the continuous variables concerning the logit of the dependent variable was assessed via the Box-Tidwell procedure. Cases with missing information and outliers were excluded from the multivariate analysis to fulfil all assumptions necessary to assure the validity of the regression.
Results: Fifty-seven patients, of whom 37 were Caucasian (64.9%) and 48 were females (84.2%), with a mean age of 39.4±14.0 years, were included. Twenty-seven patients had ILD (47.4%), of whom 22 had nonspecific interstitial pneumonia (81.5%), 4 had usu-

al interstitial pneumonia (14.8%), and 1 had lymphoid interstitial pneumonia (3.7%) pattern on CT. Among patients with ILD, 13 were asymptomatic (48.1%), while 14 had respiratory symptoms (51.9%), including dyspnea (N=13, 48.1%), cough (N=7, 25.9%), and pleuritic chest pain (N=1, 3.7%). Pulmonary function tests were performed in 22 patients (81.5%), 20 of whom had a restrictive pattern (90.9%).
In the univariate analysis, lymphadenopathy at disease onset (22.2% vs 3.3%, p=0.045) and esophageal involvement at any time point (40.7% vs 16.7%, p=0.043), were associated with ILD.

The binary logistic regression model predicting ILD included 56 patients, and the model explained 36.5% (Nagelkerke R2) of the variance in ILD and correctly classified 75% of all cases. Older age at diagnosis (OR 1.10/year, 95%CI: 1.00-1.12, p=0.046) and lymphadenopathy at disease onset (OR 19.65, 95%CI: 1.91-201.75, p=0.012) were identified as predictors of ILD in MCTD patients, irrespective of sex and esophageal involvement.
Conclusions: Older age at diagnosis and lymphadenopathy at disease onset were independent predictors of ILD in MCTD. Therefore, these factors should be considered when evaluating MCTD patients, especially at the time of diagnosis. To the best of our knowledge, this is the largest study ever describing predictors of ILD for MCTD patients.

060 - PREDICTORS OF MUSCLE INVOLVEMENT IN SYSTEMIC SCLEROSIS

Eduardo Dourado[1, 2], Raquel Freitas[3], Patrícia Martins[1, 2], Ana Teresa Melo[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], Inês Genrinho[10], Couto M[10], Marília Rodrigues[9], Maria João Salvador[4, 5], Ana Catarina Duarte[3], Ana Cordeiro[3], Maria José Santos[2, 3], JE Fonseca[1, 2], Inês Cordeiro[1, 2], Catarina Resende[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]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [4]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [5]Faculdade de Medicina, Universidade de 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, Centro Hospitalar de Leiria, Leiria, Portugal, [10]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Introduction: Only one study has ever reported independent predictors of myositis in systemic sclerosis (SSc). Lung and heart involvements were reported as positive predictors, and anti-centromere antibodies (ACA) positivity has been described as a negative predictor. Several studies have associated different autoantibodies and clinical manifestations with the occurrence of myositis in SSc, but these associations were not validated through multivariate analysis and can, therefore, result from the interference of confounders.
Methods: Multicentre open cohort study including adult SSc patients registered in the Rheumatic Diseases Portuguese Register (Reuma.pt) and with information regarding the occurrence of myositis up to February 2021. Univariate analysis was performed using Chi-Square, Fischer's Exact Test and Mann-Whitney Test. The Bonferroni correction for multiple comparisons was applied to get $\alpha \leq 0.05$. Definite associations were defined by $p \leq 0.002$, and likely associations by $p \leq 0.05$. Multivariate analysis was performed using binary logistic regression modelling. Cases with missing information and outliers were excluded from the multivariate analysis to fulfil all assumptions necessary to assure the validity of the regression.
Results: 984 patients were included, with a mean age and disease duration of 60.0±14.7 and 12.5±10.1 years, respectively. Most were females (87.5%) and had European ancestry (EA, 93.0%). The most common disease subtypes were limited cutaneous (lcSSc, 58.0%) and diffuse cutaneous (dcSSc, 13.6%). Most patients expressed antinuclear antibodies (93.7%), and the most frequent were ACA (54.3%), anti-topoisomerase I (21.5%), and anti-Pm/Scl (4.9%) antibodies. Myositis was reported in 6.3% of the patients.

Male sex, African ancestry, younger age at diagnosis, higher mRSS, flexion contractures, oesophageal involvement, interstitial lung disease, and the presence of anti-PmScl or anti-U1-RNP antibodies were positively associated with myositis. Conversely, ACA were negatively associated with myositis.

The multivariate analysis included 359 patients (624 patients had missing information, and one patient was an outlier). The logistic regression model was statistically significant, $\chi 2(7)=62.13$, $p<0.0005$. The model explained 62.6% (Nagelkerke R2) of the variance in myositis and correctly predicted 96.9% of all cases. Male sex [odds ratio (OR) 43.0, 95% confidence interval (95%CI): 5.0-369.5, p=0.001], oesophageal involvement (OR 19.9, 95%CI: 2.7-145.3, p=0.003), and positivity for anti-Pm/Scl (OR 12.3, 95%CI: 1.4-106.6,

Table I. Binary logistic regression predicting the likelihood of having muscle involvement based on sex, race, age at diagnosis, oesophageal involvement and positivity for anti-centromere, anti-PmScl and anti-U1RNP antibodies

	p-value	Odds ratio	95% C.I. for the odds ratio	
			Inferior	Superior
Male sex	0,001	42,970	4,997	369,521
African ancestry	0,274	4,077	0,328	50,661
Age at diagnosis	0,164	0,957	0,899	1,018
Oesophageal involvement	0,003	19,879	2,720	145,287
Anti-centromere antibodies	0,994	0,000	0,000	Not calculable
Anti-PmScl antibodies	0,022	12,328	1,425	106,627
Anti-U1RNP antibodies	0,002	40,506	3,762	436,110

p=0.022) or anti-U1RNP (OR 40.5, 95%CI: 3.8-436.1, p=0.002) antibodies were identified as independent predictors of myositis in SSc, irrespective of age at diagnosis, ancestry or the presence of ACA.

Conclusion: In our SSc cohort, male sex, oesophageal involvement, and positivity for anti-Pm/Scl or anti-U1RNP antibodies were independent predictors of myositis. Clinicians should be particularly alert to the possible occurrence of myositis in SSc patients with these risk factors.

095 - CARDIAC INVOLVEMENT IN IDIOPATHIC INFLAMMATORY MYOPATHIES: WHEN SHOULD WE LOOK FOR IT?

Matilde Bandeira[1, 2], Eduardo Dourado[1, 2], Ana Teresa Melo[1, 2], Patrícia Martins[1, 2], Vanessa Fraga[3], José Luís Ferraro[4], André Saraiva[5], Marlene Sousa[5], Hugo Parente[6], Catarina Soares[6], Ana Margarida Correia[7], Diogo Esperança Almeida[7], Sara Paiva Dinis [8], Ana Sofia Pinto[8], Filipe Oliveira Pinheiro[9], Maria Seabra Rato[9], Tiago Beirão[10], Beatriz Samões[10], Bernardo Santos[11], Carolina Mazeda[11], Ana Teodósio Chícharo[12], Margarida Faria[13], Agna Neto[13], Maria Helena Lourenço[14, 15], Luisa Brites[16], Marília Rodrigues[16], Joana Silva-Dinis[17], João Madruga Dias[18], Filipe Araújo[19], Nádia Martins[20], Couto M[20], Ana Valido[21], Maria José Santos[1, 3], Sofia C Barreira[1, 2], JE Fonseca[1, 2], Raquel Campanilho-Marques[1, 2]

[1]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [4]Serviço de Reumatologia, Hospital Beatriz Ângelo, Loures, Portugal, [5]Serviço de Reumatologia, Centro Hospitalar e Universitário de 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]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [10]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [11]Serviço de Reumatologia, Centro Hospitalar do Baixo Vouga, E.P.E., Aveiro, Portugal, [12]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal, [13]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [14]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [15]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [16]Serviço de Reumatologia, Centro Hospitalar de Leiria, Leiria, Portugal, [17]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Central, Lisboa, Portugal, [18]Rheumatology Department, Centro Hospitalar Médio Tejo, Torres Novas, Portugal, [19]Serviço de Reumatologia, Hospital CUF Cascais, Cascais, Portugal, [20]Rheumatology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [21]Serviço de Reumatologia, Unidade Local de Saúde do Litoral Alentejano, Santiago do Cacém, Portugal

Background: Idiopathic inflammatory myopathies (IIM) are a group of rare disorders that can affect the heart. It is unclear which patients are at higher risk. Anti-signal recognition particle (anti-SRP) antibody has been inconsistently suggested as a risk factor. This work aims to find predictors of heart involvement in IIM.

Methods: Multicenter open cohort study, including patients registered in the IIM module of the Rheumatic Diseases Portuguese Register (Reuma.pt/Myositis) until January 2022. Patients without heart involvement information were excluded. Myo(peri)carditis, dilated cardiomyopathy, conduction abnormalities and/or premature coronary artery disease were considered. Data from continuous variables are presented as mean ± standard deviation. Univariate analysis was performed using chi-square, Fisher's exact, Mann-Whitney or t-test, as appropriate. Independent predictors of cardiac involvement, adjusted for sex and age at diagnosis, were identified through binomial logistic regression.

Table I. Clinical and serological features of patients with and without heart involvement

	Patients with heart	Patients without heart involvement	Univariate analysis
Age at disease onset, median ± SD (N)	57.6 ± 11.9 (11)	46.6 ± 19.2 (193)	p=0.035
Age at diagnosis, median ± SD (N)	58.3 ± 12.2 (11)	48.0 ± 18.9 (189)	p=0.049
Disease duration (in years), median ± SD (N)	3.9 ± 4.1 (11)	7.2 ± 6.7 (193)	p=0.019
Female, n/N (%)	7/11 (63.6)	156/218 (71.6)	p=0.734
Deceased patients, n/N (%)	1/11 (9.1)	9/218 (4.1)	p=0.395
Clinical data			
Musculoskeletal involvement			
Muscle involvement			
Proximal muscle weakness, n/N (%)	7/11 (63.6)	145/176 (82.4)	p=0.128
Myositis, n/N (%)	9/11 (81.8)	175/213 (82.2)	p=1.000
Worse MMT8, median ± SD (N)	109.0 ± 36.4 (5)	135.0 ± 23.9 (126)	p=0.024
Joint involvement			
Arthralgia (without arthritis), n/N (%)	0/1 (0.0)	2/56 (3.6)	p=1.000
Arthritis, n/N (%)	3/5 (60.0)	30/87 (34.5)	p=0. 346
Skin involvement			
Gottron' sign, n/N (%)	1/10 (10.0)	68/153 (44.4)	p=0.045
Heliotrope rash, n/N (%)	4/11 (36.4)	92/216 (42.6)	p=0.764
Gottron's papules, n/N (%)	1/11 (9.1)	84/214 (39.3)	p=0.056
Periungual changes, n/N (%)	2/11 (18.2)	51/206 (24.8)	p=1.000
Malar rash, n/N (%)	2/7 (28.6)	28/122 (23.0)	p=0.664
Oedema, n/N (%)	5/11 (45.5)	41/211 (19.4)	p=0.053
Shawl sign, n/N (%)	1/7 (14.3)	25/121 (20.7)	p=1.000
Mechanic's hands, n/N (%)	3/7 (42.9)	20/121 (16.5)	p=0.109
Calcinosis, n/N (%)	0/10 (0.0)	21/216 (9.7)	p=0.604
Worse DAS skin, median ± SD (N)	0.6 ± 1.1 (8)	1.4 ± 1.8 (123)	p=0.348
Vascular involvement			
Raynaud's phenomenon, n/N (%)	6/10 (60.0)	64/214 (29.9)	p=0.075
Digital ulcers, n/N (%)	0/4 (0.0)	1/97 (1.0)	p=1.000
Internal organ involvement			
Lung involvement, n/N (%)	9/11 (81.8)	68/217 (31.3)	p=0.001
Gastrointestinal involvement, n/N (%)			
Dysphagia, n/N (%)	4/7 (57.1)	29/112 (25.9)	p=0.092
Dysphonia, n/N (%)	2/7 (28.6)	10/111 (9.0)	p=0.149
Esophageal involvement, n/N (%)	5/10 (50.0)	34/208 (16.3)	p=0.018
Gastric involvement, n/N (%)	0/10 (0.0)	2/207 (1.0)	p=1.000
Intestinal involvement, n/N (%)	1/10 (10.0)	2/209 (1.0)	p=0.131
Systemic involvement			
Fatigue, n/N (%)	3/7 (42.9)	42/118 (35.6)	p=0.702
Weight loss, n/N (%)	3/7 (42.9)	17/118 (14.4)	p=0.081
Fever, n/N (%)	0/7 (0.0)	5/119 (4.2)	p=1.000
Neoplasia, n/N (%)	0/7 (0.0)	11/118 (9.3)	p=1.000

continues on the next page

Table I. continuation

Complementary diagnostic exams	Patients with heart	Patients without heart involvement	Univariate analysis
Muscle enzymes			
High CK levels, n/N (%)	10/10 (100.0)	132/178 (74.2)	p=0.122
Maximum CK, median ± SD (N)	3680.7 ± 4235.5 (9)	2544.0 ± 3242.8 (79)	p=0.577
High aldolase levels, n/N (%)	1/3 (33.3)	69/126 (54.8)	p=0.592
Electromyogram			
Myopathic pattern	7/9 (77.8)	89/123 (72.4)	p=1.000
Muscle magnetic resonance			
Muscle oedema (STIR)	0/1 (0.0)	31/59 (52.5)	p=0.483
Antibodies			
SSA/SSB, n/N (%)	4/11 (36.4)	59/197 (29.9)	p=0.738
Anti-RNP, n/N (%)	0/10 (0.0)	11/197 (5.6)	p=1.000
Anti-Mi2, n/N (%)	1/10 (10.0)	26/185 (14.1)	p=1.000
Tif1gamma, n/N (%)	1/9 (11.1)	5/173 (2.9)	p=0.266
Anti-MDA5, n/N (%)	0/9 (0.0)	8/173 (4.6)	p=1.000
Anti-NXP2, n/N (%)	1/9 (11.1)	5/171 (2.9)	p=0.268
Anti-SAE1, n/N (%)	0/9 (0.0)	6/171 (3.5)	p=1.000
Anti-SRP, n/N (%)	3/9 (33.3)	10/175 (5.7)	p=0.018
Anti-Jo1, n/N (%)	4/10 (40.0)	47/202 (23.3)	p=0.257
Anti-PL7, n/N (%)	1/9 (11.1)	7/181 (3.9)	p=0.327
Anti-PL12, n/N (%)	1/9 (11.1)	6/181 (3.3)	p=0.292
Anti-EJ, n/N (%)	1/9 (11.1)	3/175 (1.7)	p=0.183
Anti-OJ, n/N (%)	1/9 (11.1)	2/174 (1.1)	p=0.141
Anti-Pm/Scl, n/N (%)	0/10 (0.0)	16/187 (8.6)	p=1.000
Anti-Ku, n/N (%)	0/10 (0.0)	9/179 (5.0)	p=1.000

Abbreviations: ACR – American College of Rheumatology, ALT – alanine transaminase, AST – aspartate transaminase, CK – creatine kinase, LDH – lactate dehydrogenase, n – number of patients positive for the variable of interest, N – number of patients without missing information regarding the variable of interest, STIR – short tau inversion recovery; MMT8 – manual muscle testing; DAS – disease activity score

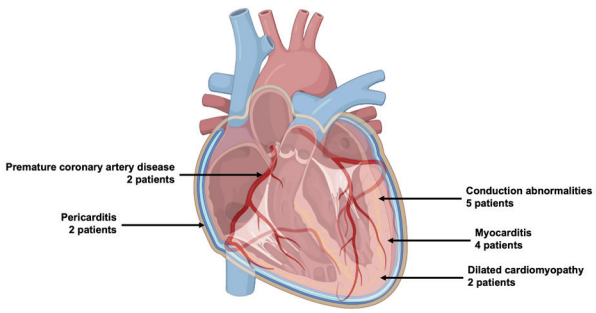


Figure 1. Types of heart involvement in our cohort. Some patients had more than one manifestation

The linearity of the continuous variables was assessed via the Box-Tidwell procedure. Correlated variables, cases with missing information and outliers were excluded from the multivariate analysis to assure the validity of the regression.

Results: 229 patients were included, 163 (71.2%) females. Patients were classified as having definite [56/115 (48.7%)], likely [23/115 (20.0%)], or possible [2/115 (1.7%)] IIM by 2017 EULAR/ACR classification criteria. Cardiac involvement was present in 11 (4.8%) patients (Figure 1), of whom 42.9% were classified as likely and 57.1% as definite IIM. The mean age at disease onset was 47.1 ± 19.0 years, the mean age at diagnosis was 48.6 ± 18.7 years, and the mean disease duration at the last follow-up was 7.0 ± 6.6 years. Compared to other IIM patients (Table 1), patients with cardiac involvement were older at disease onset (57.6

± 11.9 vs 46.6 ± 19.2 years, p=0.035) and diagnosis (58.3 ± 12.2 vs 48.0 ± 18.9 years, p=0.049), and had a shorter disease duration at the last follow-up (3.9 ± 4.1 vs 7.2 ± 6.7 years, p=0.019). Clinically, patients with cardiac involvement had a lower manual muscle testing score (MMT), comparing the lowest value throughout follow-up, and more frequently had esophageal [5/10 (50.0%) vs 34/208 (16.3%), p=0.018] and lung involvement [9/11 (81.8%) vs 68/217 (31.3%), p=0.001]. Conversely, this group less frequently had Gottron's sign [1/10 (10.0%) vs 68/153 (44.4%), p=0.045]. Anti-SRP antibodies were more commonly identified in patients with cardiac involvement [3/9 (33.3%) vs 10/175 (5.7%), p=0.018]. No differences were found between the two groups for other demographical or clinical data or serum biomarkers. In the multivariate analysis, lung involvement (OR 7.064, 95%CI: 1.246-40.057, p=0.027) and positivity of anti-SRP antibodies (OR 7.886, 95%CI: 1.333-46.666, p=0.023) were confirmed as independent predictors of heart involvement in IIM patients, regardless of sex and age at diagnosis.
Conclusion: Lung involvement and anti-SRP antibodies were independent predictors of heart involvement in our cohort of IIM patients. We suggest considering a closer screening for heart involvement in these patients.

097 - THE ROLE OF FACIAL, OCCIPITAL, SUBCLAVIAN AND CAROTID ARTERIES ULTRASOUND IN THE DIAGNOSTIC ASSESSMENT OF GIANT CELL ARTERITIS

Martins-Martinho J [1, 2], Matilde Bandeira[1, 2], Tomás Fontes[1, 2, 3], Sofia C Barreira[1, 2], Nikita Khmelinskii[1, 2], Cristina Ponte[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]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal

Background: Giant cell arteritis (GCA) is the most common form of primary systemic vasculitis in elderly patients. It predominantly affects the cranial arteries; however, extra-cranial disease involving the aorta and its major branches can also be present. Currently, ultrasound of the temporal (TA) and axillary (AX) arteries is the first imaging modality recommended in patients with suspected predominantly cranial GCA. Nevertheless, other arteries such as facial (FA), occipital (OC), subclavian (SC), and common carotid (CC) arteries can also present with vasculitic changes on ultrasound. However, there are still conflicting data to support the inclusion of these arteries in the routine ultrasound assessment of patients with suspected GCA.
Objectives: To assess the value of adding the evaluation of the FAs, OCs, SCs and CCs in the ultrasonographic diagnosis of patients with GCA.
Methods: Single-center retrospective study, using data from patients diagnosed with GCA registered at the Rheumatic Diseases Portuguese Registry (Reuma.pt). All patients underwent ultrasound of the TAs and AXs ± FAs, OCs, SCs or CCs at the time of diagnosis. The halo sign was considered a positive ultrasonographic finding for GCA. Only patients with the presence of halo sign in at least one of the arterial segments evaluated were included. Binary logistic regression modelling was performed to explore associations between the presence of halo sign in different arterial segments.
Results: We included 84 patients, 57 (67.9%) females, with a mean ± standard deviation age at diagnosis of 75.6 ± 8.8 years. Halo sign was found on the TAs of 66/84 (78.6%) patients, AXs of 40/84 (47.6%) patients, FAs of 37/74 (50.0%) patients, OCs of 15/61 (24.6%) patients, SCs of 30/49 (61.2%) patients and CCs of 13/60 (21.7%) patients. Of the 18/84 patients with GCA without the presence of TA halo, 17/18 (94.4%) showed halo on the AXs, 1/18 (5.6%) on the FAs, 0/18 (0%) on the OCs, 15/17 (88.2%) on the SCs and 6/16 (37.5%) on the CCs. Of the 44/84 patients with GCA without the presence of AX halo, 43/44 (97.7%) showed halo on the TAs, 24/39 (61.5%) on the FAs, 12/32 (37.5%) on the OCs, 4/18 (22.2%) on the SCs and 3/33 (9.1%) on the CCs arteries. A total of 83/84 (98.8%) patients had halo sign on the ultrasound of TA or AX arteries. The patient with normal TA and AX ultrasound had the presence of halo sign in the SCs. Table 1 shows the proportion of patients with positive TA and AX ultrasounds according to the presence of halo on the FA, OCs, SCs or CCs arteries. Patients with involvement of the cranial arteries were more likely to have a TA halo (FA: OR 30.6, 95%CI:3.8-247.3; OC: OR not applicable) and less likely to have an AX halo (FA: OR 0.37, 95%CI:0.14-0.95; OC: OR 0.19, 95%CI: 0.05-0.77). As opposed to patients with involvement of the extra-cranial arteries in whom the halo sign was more frequently found in the AXs (SC: OR 18.2 95%CI 4.2-78.9; CC: OR 5.9 95%CI 1.4-24.4) but not in the TAs (SC: OR 0.12 95%CI 0.02-0.60; CC: OR 0.315 95%CI 0.086-1.151).
Conclusions: Our results support the need to assess both TAs and AXs in patients with suspected GCA. Only by adding the evaluation of the SCs to the already recommended TAs and AXs increased the diagnostic sensitivity of ultrasound from 99% to 100%. All patients with a positive FA, OC or CC ultrasound for GCA also showed a halo sign on either the TAs or AXs. Hence,

Table I. Differences in the presence of halo sign in the temporal and axillary arteries according to the arterial segment affected

Arterial segment with halo	Temporal arteries with halo	Axillary arteries with halo
Facial arteries (n=37)	36/37 (97.3%)	13/37 (35.1%)
Occipital arteries (n=15)	15/15 (100%)	3/15 (20.0%)
Subclavian arteries (n=30)	15/30 (50.0%)	26/30 (86.7%)
Common carotid arteries (n=13)	7/13 (53.8%)	10/13 (76.9%)

the additional assessment of these arteries did not improve the diagnostic yield of ultrasound and, therefore, should not be recommended in routine practice.

131 - ENTHESITIS OF THE HAND IS A DOMINANT LESION IN PSORIATIC ARTHRITIS AND MAY HELP DISTINGUISHING IT FROM RHEUMATOID ARTHRITIS: CASE-CONTROL, SINGLE-CENTRE, ULTRASOUND STUDY

Diogo Esperança Almeida[1], Emanuel Costa[1], Ana Margarida Correia[1], Joana Sousa-Neves[1], Joana Leite Silva[1], Ana Ribeiro[1], Marcos Cerqueira[1]

[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

Background: Enthesitis is the hallmark of psoriatic arthritis (PsA) and may assist in distinguishing PsA from other forms of arthritis. Its clinical evaluation is hampered by lack of specificity of physical examination. Ultrasound (US) may provide information about its presence and response to treatment. Although some previous works have shown that enthesitis of the hand is important in PsA, most US studies and scores focused on greater size entheses.

Objectives: To explore the prevalence of enthesitis of the hand in PsA patients as evaluated by US, and compare it with other inflammatory arthritides, namely rheumatoid arthritis (RA).

Methods: Cross-sectional study in which consecutive patients with PsA and RA were recruited for an US protocol evaluating 4 entheses of the hand including: 1. measurement of the extensor digitorum tendon central slip at its insertion at the middle phalanx of the 2nd and 3rd finger bilaterally; 2. search for the presence of power-Doppler (PD) sign; 3. identification of structural lesions.

Linear regression models were built to test if diagnosis (PsA vs RA) explained part of the variance of the thickness of tendons insertion while controlling possible influences of age, type of work and body surface area. A ROC curve was built to find a mean thickness cut-off allowing distinction between PsA and RA. The prevalence of PD sign and structural lesions of the entheses was compared between groups.

Results: Fifty-eight patients were recruited (29 PsA and 29 RA) and a total of 232 entheses were evaluated.

Mean thickness of the interest entheses was superior in PsA patients compared to RA patients (2nd finger – 0.96 ± 0.16mm vs. 0.74 ± 0.09mm; 3rd finger – 0.96 ± 0.20mm vs. 0.76 ± 0.11mm).

Linear regression models including diagnosis and potential confounders significantly explained mean thickness of both entheses (2nd finger – R2=0.56, p<.001; 3rd finger – R2=0.41, p<.001), with the diagnosis group being the most important predictor (Table I). ROC curve (AUC 0.897, p<.001) showed a cut-off value of 0.925mm for the mean of the 4 entheses had a specificity of 93.1% for the identification of PsA patients.

In our sample, 8 (3.5%) entheses had a measured thickness above mean + 2 SD, all belonging to PsA patients; 6 (75%) had signs of ongoing inflammatory process as proved by the presence of PD sign (figure 1). Regarding structural lesions, enthesophytes or bone irregularities/erosions were found in 13.8% of PsA entheses, which compared to 1.7% of RA entheses.

Conclusions: This work reinforces enthesitis as a key lesion in PsA. It also shows enthesitis occurs significantly in small entheses, like the ones of the hand and that, in some instances, it may be the dominant lesion in a swollen joint. US may be valuable for establishing a diagnosis in the setting of inflammatory arthritis of unknown etiology.

136 - OUTCOMES OF SARS-COV-2 INFECTION IN CHILDREN AND ADOLESCENTS FOLLOWED AT A PORTUGUESE PEDIATRIC RHEUMATOLOGY UNIT

Ana Teresa Melo[1, 2, 3], Miguel Bernardo[1, 4], Bianca Paulo Correia[1, 2, 3], Raquel Campanilho-Marques[1, 2, 3], Patrícia Costa Reis[1, 3, 5], Filipa Oliveira Ramos [1, 2, 3]

[1]Unidade de Reumatologia Pediátrica, Hospital de Santa Maria, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]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, [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 Pediatria, Hospital São Pedro, Centro Hospitalar de Trás-os-Montes e Alto Douro, Vila Real, Portugal, [5]Departamento de Pediatria, Hospital de Santa Maria, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introduction: The infection with SARS-CoV-2 is generally mild in children. However, the outcomes and predictors of severe disease in children and adolescents with rheumatic and musculoskeletal diseases (RMD) are still being evaluated.
Objectives: To assess the epidemiological features and clinical outcomes of children and adolescents followed at a Pediatric Rheumatology Unit, who were infected with SARS-CoV-2 and evaluate possible predictors of severe disease.
Methods: A prospective study was performed. Data on demographic variables and clinical features were collected, using the Rheumatic Diseases Portuguese Register (Reuma.pt) and complemented with data from the hospital clinical records between March 2020 and April 2022.

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 performed using binary logistic regression modelling.
Results: Ninety-four patients infected with SARS-CoV2 were identified, 65% were female (n=61), with a median age of 13.5 [8-17] years. Sixty-four patients (68%) had a confirmed inflammatory disease. The most frequent diagnosis was juvenile idiopathic arthritis (n=31, 33%), followed by systemic lupus erythematosus (SLE) (n=11, 12%).

No deaths were registered. Thirty-four patients (36%) had an asymptomatic infection of which half had a confirmed inflammatory disease (n=17). Most patients had milder symptoms (61%, n=57), the majority of whom with an inflammatory disease previously diagnosed (n=44). The most frequent symptoms were cough (n=27), fever (n=19), headache (n=15) and rhinorrhea (n=14). Three patients were hospitalized (3%). Only one of those patients was previously vaccinated. He had a diagnosis of SLE and had thoracalgia without troponin elevation. He was treated with nonsteroidal anti-inflammatory drugs. Other patient was healthy before the SARS-CoV2 infection and developed a severe case of hemolytic anaemia (lower hemoglobin 3.7 mg/dL) with hemodynamic instability requiring pediatric intensive care. SLE, triggered by SARS-CoV2, was later diagnosed. Other patient had an undefined autoinflammatory disease. She developed fever, abdominal pain and diarrhea 7 weeks after the COVID-19 diagnosis. She had elevated liver transaminases. A Multisystem inflammatory syndrome in children (MIS-C) was diagnosed.

Twenty-eight patients (30%) changed the medication during the infection (temporary withheld or dose reduction), while 9 patients (10%) had an adjustment done in medication after the infection.

SLE diagnosis was positively associated with hospitalization (18% vs 1%, p=0.035) and no other associations have been found.

The multivariate analysis predicting hospitalization explained 38.4% (Nagelkerke R2) of the variance and correctly classified 96.8% of all cases. SLE diagnosis was identified as an independent predictor of hospitalization (OR 55.8 95%CI: 1.7-1822), regardless of sex, age, ethnicity and vaccination status.
Conclusions: SLE diagnosis was an independent predictor of hospitalization. Although a milder illness was predominant, we suggest that patients with SLE and COVID-19 infection should maintain a tighter follow-up.

150 - CYCLING VERSUS SWAPPING STRATEGIES IN PSORIATIC ARTHRITIS: RESULTS FROM THE RHEUMATIC DISEASES PORTUGUESE REGISTER

Francisca Guimarães[1], Maria Pontes Ferreira[1], Catarina Soares[1], Hugo Parente[1], Carolina Ochôa Matos[2], Roberto Costa[2], Daniela Santos Oliveira[3], Catarina Abreu[4], Rafaela Nicolau[5], Sofia Ferreira Azevedo[6], João Madruga Dias[7], Filipe Araújo[8], Carla Campinho Ferreira[9], Daniela Santos-Faria[1], José Tavares-Costa[1]
[1]Rheumatology Department, Hospital Conde de Bertiandos (ULSAM), Ponte de Lima, Portugal, [2]Rheumatology Department, Centro Hospitalar Universitário de Lisboa Norte, Lisboa, Portugal, [3]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [4]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [5]Rheumatology Unit, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [6]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [7]Rheumatology Department, Centro Hospitalar Médio Tejo, Torres Novas, Portugal, [8]Rheumatology and Osteopororis Unit, Hospital de Sant Ana, Lisboa, Portugal, [9]Rheumatology Department, Hospital de Braga, Braga, Portugal

Background: In Psoriatic arthritis (PsA) management, after an inadequate response to a first tumour necrosis factor alfa inhibitor (TNFi), patients may receive a second TNFi – "cycling" or a drug with a different mode

Table I. Patients' baseline characteristics

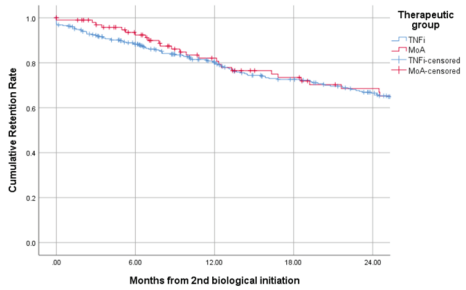
	All patients N=454	TNFi N=352	MoA N=102	p-value
Sociodemographic characteristics				
Age (years)	50±12	50±11	49±12	NS
Gender (Female)	261 (57.5%)	203/352 (57.7%)	58/102 (56.9%)	NS
Race (White European origin)	322/336 (95.8%)	251/352 (71.3%)	71/102 (69.6%)	NS
Smoking status (Never smoked)	192/311 (61.7%)	121/191 (63.4%)	23/38 (60.5%)	NS
Alcohol consumption (occasional/never consumed)	234/293 (79.9%)	145/182 (79.7%)	30/39 (76.9%)	NS
BMI (Kg/m2)	28.3±5.4	28.26±5.37	27.23±4.98	NS
Disease characteristics				
Age at diagnosis (years)	41±12	41±12	41±12	NS
Disease duration until 1st biologic (years)	6.9±13.3	6.2±9.2	6.0±13.8	NS
Axial disease	18/378 (4.8%)	9/211 (4.3%)	5/58 (8.6%)	NS
Peripheral disease	245/378 (64.8%)	136/211 (64.5%)	38/58 (65.5%)	NS
Axial and peripheral disease	115/378 (83.3%)	66/211 (31.3%)	15/58 (25.9%)	NS
Enthesitis (yes)	128/388 (33.0%)	70/220 (31.8%)	15/58 (25.9%)	NS
Psoriasis (yes)	267/291 (91.8%)	159/171 (93%)	40/42 (95.2%)	NS
Nail psoriasis (yes)	77/223 (34.5%)	46/129 (35.7%)	12/34 (35.3%)	NS
Dactylitis (yes)	84/227 (37.0%)	47/130 (36.2%)	16/37 (43.2%)	NS
Uveitis (yes)	22/209 (10.5%)	19/122 (15.6%)	1/30 (3.3%)	NS
HLAB27 (positive)	51/213 (23.9%)	33/122 (27%)	5/25 (20%)	NS
Extra-articular manifestations (yes)	281/454 (61.9%)	168/253 (66.4%)	40/69 (58.0%)	NS
Comorbidities				
Hypertension (yes)	108/376 (28.7%)	64/216 (29.6%)	10750 (20%)	NS
Dyslipidemia (yes)	4/376 (1.1%)	47/212 (1.9%)	0/50 (0%)	NS
Diabetes mellitus (yes)	38/376 (10.1%)	22/216 (10.2%)	5/50 (0%)	NS
DMARDs therapy				
Discontinuation of the 1st TNFi due to ineffectiveness (yes)	314/451 (69.6)	245/352 (69.6%)	69/33 (67.6%)	NS
Disease duration until 2nd biologic (years)	9.00±9.30	9.9±12.0	7.05±5.6	0.004*
Methotrexate association (yes)	222/448 (49.6%)	140/250 (56.0%)	17/68 (25%)	<0.001*
Leflunomide association (yes)	24/448 (5.4%)	15/250 (6.0%)	4/68 (5.9%)	NS
Glucocorticoid (yes)	160/447 (35.8%)	134/346 (38.7%)	26/101 (25.7)	0.018*
NSAIDs (yes)	136/446 (30.5%)	110/346 (31.8%)	26/100 (26%)	NS
Baseline disease activity				
Tender joints 68	7.63±8.19	7.11±7.92	7.32±8.88	NS
Swollen joints 68	3.86±4.66	3.61±4.27	3.98±5.39	NS
ESR (mm/1st hour)	27.64±24	28.23±24.72	20.20±15.07	0.007*
CRP (mg/dL)	1.48±2.33	1.67±2.60	0.89±1.73	0.017*
Patients VAS	59.72±25.96	57.65±27.47	54.47±24.00	NS
Pain VAS	59.34±25.55	56.50±26.66	56.61±25.07	NS
Physician VAS	44.29±23.14	41.83±24.17	41.08±21.39	NS
DAS 28 4V CRP	4.08±1.30	4.00±1.34	3.72±1.38	NS
DASPSA	25.73±13.41	24.31±13.54	25.59±15.41	NS
HAQ	1.10±0.75	1.04±0.73	1.00±0.73	NS

continues on the next page

Table I. continuation

	All patients N=454	TNFi N=352	MoA N=102	p-value
BASDAI	5.68±2.46	5.28±2.69	5.20±2.35	NS
ASDAS-CRP	3.30±1.18	3.24±1.26	2.94±1.14	NS
BASFI	5.35±2.77	5.07±2.74	5.19±3.11	NS
MASES	1.39±2.67	1.47±2.81	1.16±2.10	NS
Number of dactylitis (median-IQR)	0 (0)	0 (0)	0 (0)	NS
Psoriasis VAS (median-IQR)	1 (2)	0 (1)	1 (2)	NS

Abbreviations: ACR – American College of Rheumatology, ALT – alanine transaminase, AST – aspartate transaminase, CK – creatine kinase, LDH – lactate dehydrogenase, n – number of patients positive for the variable of interest, N – number of patients without missing information regarding the variable of interest, STIR – short tau inversion recovery; MMT8 – manual muscle testing; DAS – disease activity score



Months	0	6	12	24
TNFi	351	285	230	168
MoA	100	79	59	41

Figure. Drug retention by months for each group; n of patients at risk by months.
TNFi: tumour necrosis factor inhibitor; MoA: drug with different mode of action

of action (MoA) – "swapping". Yet, data about the comparative effectiveness of different switching strategies (cycling VS swapping) in daily practice are scarce. This study aimed to compare the effectiveness and safety of switching to secukinumab (SEK) or ustekinumab (UST) versus a second TNFi measured by retention rates during a 2-year period of follow-up, in PsA patients with previous inadequate response to a first TNFi.

Methods: Retrospective longitudinal cohort study with a 2-year period of follow-up using real-world anonymous patient-level data from the Reuma.pt Portuguese nationwide database. Patients with a diagnosis of PsA, fulfilling the CASPAR classification criteria and previous treatment failure to a first-line TNFi that started a second biotechnological drug (TNFi or SEK/UST) were included. Sociodemographic data, disease characteristics, disease activity scores and physical function at baseline and after 6,12 and 24 months were recorded. Persistency of TNFi ("cycling group") and SEK/UST ("swapping group") was estimated using Kaplan-Meier analysis. Cox regression was used to obtain a predictor model of discontinuation. Crude and LUNDEX adjusted response rates were evaluated at 6, 12 and 24 months, and reasons for discontinuation were compared between groups. SPSS v25 was used for statistical analysis and significance level was defined as 2-sided $p< 0.05$.

Results: In total, 454 patients were included, 57.5% were female, with a mean age of 50±12 years old. Baseline characteristics are described in Table 1. Of those, 77.5% initiated a 2nd TNFi and 22.5% a drug with a different MoA (64 SEK and 36 UST). Most patients discontinued the 1st TNFi due to inefficacy (69.2%), mainly secondary (56.6%).

The retention rates at 6, 12 and 24 months of follow-up in the cycling group and the swapping group were 80%, 72% and 24%; and 82%, 74% and 27%, respectively. There were no significant differences in drug retention (Figure 1), when considering all sample (p=0.711), but also after sub analysis regarding both axial, peripheral, and extra-articular involvement (p=0.824). Older age at diagnosis (HR=1.052, p=0.03), higher DAPSA at baseline (HR=1.059, p=0.013) and extra-articular manifestations (HR=4.854, p=0.015) were independent predictors of suspension of the 2nd biologic.

Regarding peripheral involvement, the proportion of patients in remission or low disease activity according to DAPSA at 6, 12 and 24 months was, respectively, 60.0%/58.2%/75.3% for TNFi, and 50%/63.2%/50% for SEK/UST. After LUNDEX adjustment, response rates were respectively, 48.6%/43.1%/17.3% for 2nd TNFi, and 42.5%/46.8%/12.0% for SEK/UST.

Concerning axial involvement, the proportion of patients in remission or low disease activity according to ASDAS-PCR at 6, 12 and 24 months was, respectively, 36.8%/50.0%/62.5% for TNFi, and 0.0%/50%/20%

for SEK/UST. After LUNDEX adjustment, response rates were 29.8%/37.5%/15.6% for 2nd TNFi, and 0%/33.0%/3.6% for SEK/UST, respectively. The main reason for discontinuation of the 2nd biologic was inefficacy for both groups.

Conclusions: After a 1st TNF inhibitor, "cycling" and "swapping" strategies are both acceptable due to similar retention rates. Further studies with greater samples are needed in order to perform survival subgroup analysis regarding different extra-articular manifestations and PsA subtypes.

151 - DETERMINANTS OF PATIENT AND PHYSICIAN GLOBAL ASSESSMENT OF DISEASE ACTIVITY IN SPONDYLOARTHRITIS

Catarina Dantas Soares[1], Soraia Azevedo[1], Francisca Guimarães[1], Hugo Parente[1], Maria Pontes Ferreira[1], Daniela Peixoto[1], Filipa Teixeira[1], José Tavares-Costa[1], Carmo Afonso[1], Daniela Santos-Faria[1]

[1]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal

Background: Patient's and Physician's Global Assessment of Disease Activity (PtGA and PhGA) are important measures in Spondyloarthritis (SpA), but often provide discordant results. Some data show that patient-physician discordance can result in patient poor adherence to treatment and healthcare costs in SpA. Identifying the factors associated with PtGA and PhGA may facilitate shared decision making and optimal treatment management.

Objective: We intended to assess the principal determinants of both PtGA and PhGA in SpA patients under biologic treatment.

Methods: We performed a cross-sectional study, including patients with SpA under biologic treatment registered in the Rheumatic Diseases Portuguese Register (Reuma.pt), consecutively evaluated in a tertiary hospital center. Sociodemographic (age, gender, marital status, educational level, employment status) and clinical data [comorbidities, daily medication, inflammatory parameters, Health Assessment Questionnaire (HAQ), Functional Assessment of Chronic Illness Therapy (FACIT), Short Form (36) Health Survey (SF-36), EuroQol-5 dimension (EQ5D), Hospital Anxiety and Depression scales (HADS), Bath Ankylosing Spondylitis Activity Index (BASDAI) and Bath Ankylosing Spondylitis Functional Index (BASFI)], were collected.

PtGA and PhGA were measured on a Visual Analogue Scale of 0-100, with 0 indicating best status and 100 indicating worst status.

To identify determinants of PtGA and PhGA, we performed firstly a univariate analysis with the independent variables and subsequently a multiple linear regression. SPSS® v.24 was used for statistical analysis and significance level was defined as 2-sided $p< 0.05$.

Table I. Clinical and laboratory characteristics of patients with spondyloarthritis

Age (years), mean ± SD	52.2 ± 12.9
Gender – male, %(n/N)	53.2% (99/186)
Years from diagnosis, mean ± SD	11.1 ± 7.9
Biologic DMARD position, %(n/N)	1st: 74.2% (138/186) 2nd: 17.7% (33/186) Others: 8.1% (15/186)
Patient Global VAS, mean ± SD	34.4 ± 27.2
Patient pain VAS, mean ± SD	31.9 ± 26.3
Patient back pain VAS, mean ± SD	25.0 ± 26.9
Patient nocturnal back pain VAS, mean ± SD	27.4 ± 26.1
Physician Global VAS, mean ± SD	7.4 ± 12.7
Patient-physician discordance mean ± SD***	27.8 ± 24.0
Tender joints count (n), median (IQR)	0.0 (1.0)
Swollen joints count (n), median (IQR)	0.0 (0.0)
CRP (mg/dL), median (IQR)	0.29 (0.6)
ESR (mm/hr), mean ± SD	11.0 ± 17.7
HAQ, median (IQR)	0.8 (1.1)
BASDAI, median (IQR)	2.5 (3.7)
BASFI, median (IQR)	2.4 (4.4)
Short Form (36) Health Survey (SF-36), mean ± SD	455.7 ± 167.2
FACIT, mean ± SD	35.6 ± 11.4
HADS, median (IQR)	Anxiety: 6 (8.0) Depression: 5 (8.0)
EQ5D, median (IQR)	0.4824 ± 0.4058
Daily medication, median (IQR	3 (3)
Prednisolone, %(n/N)	17.7% (33/186)
Comorbidities, median (IQR)	1 (2)
Osteoarthrosis, %(n/N)	15.6% (29/186)
Fibromyalgia, %(n/N)	6.5% (12/186)

BASDAI: Ankylosing Spondylitis Disease Activity Index; BASFI: Bath Ankylosing Spondylitis Functional Index; CRP: C reactive protein; EQ5D: EuroQol-5 dimension; ESR: erythrocyte sedimentation rate; FACIT: Functional Assessment of Chronic Illness Therapy; HADS: Hospital Anxiety and Depression scales; HAQ: Health Assessment Questionnaire; SF-36: Short Form (36) Health Survey; VAS: Visual Analogue Scale

Results: We evaluated 186 patients with SpA according to ASAS criteria, under biologic treatment. Most patients were male (53.20%) with a mean age of 52.15 (SD=12.9) years-old at the time of last medical appointment. PtGA and PhGA were significantly different. Clinical and laboratory characteristics of patients are shown in Table I.

There was a positive correlation between higher PtGA and older age, unemployment, number of tender joints, HAQ, HADS, BASDAI, number of comorbidities

and daily medication. There was also an association with the concomitant presence of osteoarthrosis, fibromyalgia, C reactive protein (CRP), erythrocyte sedimentation rate (ESR) and daily prednisolone intake. On the other side, we found a negative correlation with SF-36, FACIT and EQ5D. The multiple linear regression shows that the SF-36 (p=0.001), BASDAI (p<0.001) and being unemployed (p=0.042) were the most preponderant determinants in PtGA explaining 85% of the variability noted in PtGA (R2 = 0.846; R2a= 0.828).

Regarding PhGA we found a positive correlation between the number of tender and swollen joints, CRP and daily prednisolone intake. In multivariable analyses the main determinants of PhGA were the number of swollen joints and higher CRP (R2 = 0.867; R2a= 0.829).

Conclusions: This study reinforces, in a real-life setting, the notion of variability in disease activity measured by PtGA and PhGA. We have demonstrated that comorbidities, employment status, and other factors not directly related to the disease are also determinants in PtGA. On the other hand, more objective data such as swollen joints and increased CRP were predominant in PhGA construct.

161 - IS INFLAMMATION-DRIVEN BONE LOSS ASSOCIATED WITH TWO-YEAR BONE FORMATION AT THE SAME VERTEBRA IN AXIAL SPONDYLOARTHRITIS? – A MULTILEVEL MRI AND LOW DOSE CT ANALYSIS FROM THE SENSITIVE IMAGING OF AXIAL SPONDYLOARTHRITIS (SIAS) COHORT

Mary Lucy Marques[1, 2], Nuno Pereira da Silva[3], Désirée van der Heijde[1], Rosalinde Stal[1], Xenofon Baraliakos[4], Juergen Braun[4], Monique Reijnierse[5], Caroline Bastiaenen[6], F vanGaalen[1], Sofia Ramiro[1, 7]

[1]Rheumatology, Leiden University Medical Center, Leiden, Netherlands, [2]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [3]Serviço de Radiologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [4]Department of Rheumatology, Rheumazentrum Ruhrgebiet, Herne, and Ruhr-University Bochum, Bochum, Germany, [5]Radiology, Leiden University Medical Center, Leiden, Netherlands, [6]Epidemiology Department, Maastricht University, Maastricht, Netherlands, [7]Rheumatology Department, Zuyderland Medical Center, Heerlen, Netherlands

Background: In radiographic axial Spondyloarthritis (r-axSpA), inflammation-driven vertebral bone loss has been hypothesised to trigger pathological bone formation (i.e., to maintain spinal stability, trabecular bone loss is compensated by syndesmophyte formation) [1]. While this has been possible to study in animal models, given the limitations of DXA scans, radiography, and conventional Computed Tomography (CT) it has been difficult to study in patients.

Aims: Using sensitive imaging techniques, namely low dose CT (ldCT) and magnetic resonance imaging (MRI), we aimed to investigate whether inflammation is associated with lower bone density (surrogate of bone loss) and subsequently, if lower bone density is associated with a higher likelihood of 2-year bone formation at the same vertebra in r-axSpA, from C3 to L5.

Methods: Data from the multicentre 2-year Sensitive Imaging in Ankylosing Spondylitis (SIAS) cohort was used. Baseline vertebral bone density Hounsfield Units (HU) were assessed on ldCT scans by two readers (Figure 1). Baseline magnetic resonance imaging bone marrow edema (MRI-BME) status-scores, and 2-year ldCT syndesmophyte formation and/or growth change-scores were assessed by three and two readers respectively. Inter-reader reliability for imaging scorings was assessed by vertebra. Average of readers´ continuous scores or readers´ agreement in binary scores were used at the same vertebra (1-present in ≥1 quadrant/0-absent in all quadrants). Multilevel generalised estimating equations models were used, the unit of analysis being the vertebra.

Results: We analysed 1,100 vertebrae in 50 patients with r-axSpA. Intraclass correlation coefficients for HU measurements:0.89-0.97, Fleiss-Kappa (MRI-BME status-scores):0.41-0.78, and Cohen´s kappa (syndesmophyte formation/growth change-scores):0.36-0.74. Bone density HU decreased from cranial to caudal vertebrae. Baseline MRI-BME was present in 300/985 (30%) and syndesmophytes in 588/910 (65%) vertebrae, both most prevalent at the thoracolumbar region. Syndesmophyte formation or growth was observed in 18% of at-risk vertebrae (124/691). A cross-sectional significant confounder-adjusted association was found between inflammation and lower bone density (regression coefficient=-51;95% CI:-63 to -39) (Table 1A). Bone density was not associated with 2-year syndesmophyte formation or growth (adjOR=1.00;95% CI:0.99 to 1.00) (Table 1B).

Conclusion: While in r-axSpA vertebral inflammation is associated with low vertebral bone density, lower vertebral bone density itself did not increase the risk for subsequent bone formation at the same vertebra. These data highlight inflammation as a major factor in r-axSpA bone disease.

REFERENCES

1. Lories RJ. Best Pract Res Clin Rheumatol. 2018 Jun;32(3):331–41.

SIAS study was funded by the Dutch Rheumatism Association ("ReumaNederland"). MLM is supported by the Fundação para a Ciência e Tecnologia (FCT) grant SFRH/BD/143744/2019.

Table I. Relationships between baseline MRI detected spinal inflammation and bone density (A), and between baseline bone density and ldCT bone formation after two years (B), at the same vertebra.

A.	**Independent variables**	**Bone density (ldCT Hounsfield Units)**	
		Univariable analysis Reg coeff. (95% CI) N = 910 to 985	Multivariable analysis Adj Reg coeff. (95% CI) N = 985
	MRI-BME (presence)	**-51 (-63 to -39)**	**-51 (-63 to -39)**
	Age (years)	-1 (-2 to 1)	-1 (-2 to 1)
	Gender (male)	21 (-20 to 63)	16 (-24 to 57)
	TNFi treatment (yes)	26 (-7 to 59)	27 (-6 to 61)
	Baseline syndesmophytes (presence)*	**-42 (-54 to -30)**	-
B.	**Independent variables**	**Syndesmophyte formation or growth§**	
		Univariable analysis OR (95% CI) N = 672 to 691	Multivariable analysis AdjOR (95% CI) N = 672
	Bone density (ldCT Hounsfield Units)	1.00 (0.99 to 1.00)	1.00 (0.99 to 1.00)
	Age (years)	1.02 (0.99 to 1.06)	1.02 (0.98 to 1.05)
	Gender (male)	0.44 (0.13 to 1.52)	0.56 (0.15 to 2.06)
	Smoking (current)	0.89 (0.40 to 1.97)	1.02 (0.42 to 2.44)
	Treatment with TNFi (yes)	1.34 (0.56 to 3.21)	1.30 (0.43 to 3.90)
	MRI-BME (presence)	**2.03 (1.23 to 3.71)**	**1.73 (1.06 to 3.34)**
	Baseline syndesmophytes (presence)*	**2.84 (1.83 to 4.41)**	-

*Multicollinearity with MRI-BME. § Absolute agreement of readers. **adjOR,** adjusted odds ratio; **CI,** confidence interval; **BME,** bone marrow edema, **HU,** Hounsfield units; **ldCT,** low dose computed tomography; **MRI,** magnetic resonance imaging; **TNFi,** Tumour necrosis factor inhibitors. Statistical significance highlighted in bold.

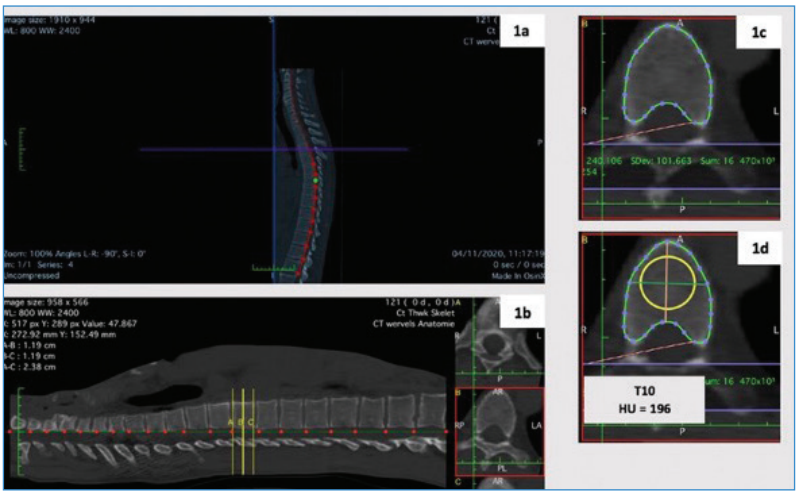


Figure 1. Methodology of low dose Computed Tomography Hounsfield Units (HU) measurement. 1a: Using a three-dimensional curved-multiplanar reconstruction, the curve of the spine adjacent to the spinal canal was delimit. 1b: On the obtained sagittal image, each vertebra (from C3 to L5) was identifiable. At each vertebra, two lines of reference were 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 was automatically positioned by the software at the center of the vertebral body. 1c: In the reconstructed cross-sectional slice, the vertebral body was manually delimited. 1d: A region of interest was manually selected, having a diameter equal to 75, of the average of anteroposterior and transverse diameters. The density of the vertebra was displayed by the software as the average image intensity within the sample region, reported in HU.

183 - ADVERSE EVENTS IN PATIENTS WITH INFLAMMATORY JOINT DISEASES: RESULTS FROM THE EULAR CORONAVIRUS VACCINE (COVAX) PHYSICIAN-REPORTED REGISTRY

Ana Lúcia Fernandes[1], José A Gómez-Puerta[2], Juan Camilo Sarmiento-Monroy[2], Saskia Lawson-Tovey[3], Kimme Hyrich[3], Laure Gossec[4], Loreto Carmona[5], Anja Strangfeld[6], Elsa Mateus[7, 8], Rodrigues AM[9, 10], Eric Hachulla[11], Marta Mosca[12], Patrick Durez[13], Bernd Raffeiner[14], Nicolas Roux[15], Eric Veillard[16], Olivier Brocq[17], Julija Zepa[18], Inita Bulina[18], Eva Strakova[19], Vanda Mlynarikova[20], Emoke Stenova[21], Martin Soubrier[22], Xavier Mariette[23], Pedro M. Machado[24, 25]
[1]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal, [2]Reumatologia, Hospital Clinic, Barcelona, Spain, [3]Centre for Genetics and Genomics Versus Arthritis, Centre for Musculoskeletal Research, University of Manchester, Manchester, United Kingdom, Manchester, United Kingdom, [4]Department of rheumatology, AP-HP, Pitié Salpêtrière, Paris, France, [5]Instituto de Salud Musculoquelética, Madrid, Spain, [6]Epidemiology Unit, German Rheumatism Research Center (DRFZ Berlin), Berlin, Germany, [7]Liga Portuguesa Contra as Doenças Reumáticas, Lisboa, Portugal, [8]European League Against Rheumatism (EULAR) Standing Committee of People with Arthritis/Rheumatism in Europe (PARE), Kilchberg,, Zurich, Switzerland, [9]Sociedade Portuguesa de Reumatologia, Lisboa, Portugal, [10]CEDOC, NOVA Medical School, Nova University, Lisboa, Portugal, [11]Service de Médecine Interne et Immunologie Clinique, Centre de référence des maladies autoimmunes systémiques rares du Nord et Nord-Ouest de France (CeRAINO),, Lille, France, [12]University of Pisa and Azienda Ospedaliero Universitaria Pisana, Pisa, Italy, Roma, Italy, [13]SINT LUC, Brussels, Belgium, Brussels, Belgium, [14]Department of Rheumatology, Central Hospital of Bolzano, Bolzano, Italy, Roma, Italy, [15]Service de Rhumatologie, Hôpital Robert Schuman, Metz, France, [16]Cabinet de Rhumatologie des "Marines de Chasles", Saint Malo, Paris, France, [17]Department of Rheumatology, Princess Grace Hospital, Monaco, Monaco, [18]Pauls Stradins Clinical University Hospital, Riga, Latvia, [19]Department of Internal Medicine, Faculty Hospital Prešov, Prešov, Bratislava, Slovakia, [20]National Institute of Rheumatic Diseases, Pieštany, Bratislava, Slovakia, [21]1st Department of Internal Medicine, Faculty of Medicine, Comenius University and University Hospital, Bratislava, Slovakia, [22]Department of Rheumatology, CHU Clermont-Ferrand, Hopital Gabriel Montpied, Clermont-Ferrand, France, [23]Department of Rheumatology, Université Paris-Saclay, Assistance Publique-Hôpitaux de Paris, Paris, France, [24]Centre for Rheumatology Research & MRC Centre for Neuromuscular Diseases, University College London, London, United Kingdom, [25]Rheumatology, Northwick Park Hospital, London North West University Healthcare NHS Trust, London, United Kingdom

Introduction: Patients with inflammatory/autoimmune rheumatic and musculoskeletal diseases (I-RMDs) were excluded from SARS-CoV-2 vaccination development programs. Therefore, concerns regarding the safety and effectiveness of SARS-CoV-2 vaccines in this population arose. Previous reports capturing a wide range of I-RMDs have been reassuring [1], but more granular data on specific conditions is desirable.
Aims: To describe adverse events (AEs) in the most common inflammatory joint diseases (IJD), namely rheumatoid arthritis (RA), axial spondyloarthritis (axSpA), psoriatic arthritis (PsA), other peripheral spondyloarthritis (pSpA), and gout/other crystal arthritis (CA), in comparison with a group of patients with non-inflammatory rheumatic and musculoskeletal diseases (NI-RMDs).
Methods: Physician-reported registry of RMDs patients vaccinated against SARS-CoV-2. From 5 February 2021 to 3 March 2022, data were collected on demographics, vaccination, RMD diagnosis, immunomodulatory/immunosuppressive treatments and both early AEs and AEs of special interest. Data were analyzed descriptively.
Results: A total of 7625 patients from 31 different countries were included: 6870 with IJD (63.9% female, mean age 58.8 years), namely 3639 with RA, 1680 with axSpA, 1205 with PsA, 220 with pSpA and 126 with CA, and 755 with NI-RMDs (83.2% female, mean age 68.5 years). Main results are presented on Table 1. Most patients received a full scheme of vaccination (IJD: n=5964, 86.8%; NI-RMDs: n=612, 81.1%), and the most commonly administered vaccine was Pfizer/BioNTech (first dose: IJD n=4385, 63.8%; NI-RMDs n=534, 70.7%). AEs were observed less frequently in IJD than in NI-RMDs, including early AEs (vaccine reaction) (IJDs: n=3743, 54.5%; NI-RMDs: n=543, 71.9%) and AEs of special interest (IJDs: n=129, 1.9%; NI-RMDs: n=57, 7.5%). The pSpA group was an exception, presenting a higher rate of early AEs (n=185, 84.1%) and AEs of special interest (n=13, 5.9%). The overall rate of serious AEs was very low (IJD: n=22, 0.3%; NI-RMDs: n=19, 2.5%), and similar across IJDs. The serious AE included events of arrythmia, coronary heart disease, syncopes, arterial hypertension, telogen effluvium, eczema/rash, erythema nodosum, gengivitis, abdominal pain, lymphadenopathy, dyspnoea, pharyngitis exacerbation of asthma, thoracic pain, pulmonary embolism, herpes zoster and shingles. The registry being mainly dedicated to inflammatory RMDs, there was probably a bias favoring registration pf patients with mechanical RMDs having had AE. No deaths were reported and most patients recovered from the AE without sequelae.

Table I. Adverse events in patients with most common inflammatory joint diseases and non-inflammatory rheumatic and musculoskeletal diseases

Adverse events	NI-RMDs (n=755)		RA (n=3639)		axSpA (n=1680)		PsA (n=1205)		pSpA (n=220)		CA (n=126)		Total IJDs (n=6870)	
	n	%	n	%	n	%	n	%	n	%	n	%	n	%
Total AEs (early and special interest adverse events)	600	79.5	1973	54.2	907	56.1	735	61	198	90	59	46.8	3872	56.4
Early AEs	543	71.9	1910	52.5	882	54.5	710	58.9	185	84.1	56	44.4	3743	54.5
AEs of special interest	57	7.5	63	1.7	25	1.5	25	2.1	13	5.9	3	2.4	129	1.9
AE seriousness	-	-	-	-	-	-	-	-	-	-	-	-	-	-
Non-serious	38	5	45	1.2	24	1.4	20	1.7	10	4.5	2	1.6	101	1.5
Serious (total)	19	2.5	15	0.4	1	0.1	3	0.2	2	0.9	1	0.8	22	0.3
Serious – important medical event	16	2.1	8	0.2	1	0.1	2	0.2	2	0.9	0	0	13	0.2
Serious - hospitalisation (or prolongation of existing hospitalisation)	3	0.4	5	0.1	0	0	1	0.1	0	0	1	0.8	7	0.1
Serious – life threatening	0	0	2	0.1	0	0	0	0	0	0	0	0	2	0.03
Unknown/missing	0	0	3	0.1	0	0	2	0.2	1	0.5	0	0	6	0.1
Total	57	7.5	63	1.7	25	1.5	25	2.1	13	5.9	3	2.4	129	1.9
AE outcome	-	-	-	-	-	-	-	-	-	-	-	-	-	-
Ongoing/continuing	13	1.7	7	0.2	3	0.2	7	0.6	1	0.5	0	0	18	0.3
Recovered/resolved without sequelae	41	5.4	43	1.2	22	1.3	16	1.3	10	4.5	3	2.4	94	1.4
Recovered/resolved with sequelae	2	0.3	5	0.1	0	0	0	0	1	0.5	0	0	6	0.1
Unknown/missing	1	0.1	8	0.2	0	0	2	0.2	1	0.5	0	0	11	0.2
Total	57	-	63	-	25	-	25	-	13	-	3	-	129	-

Conclusion: Serious AEs and breakthrough infections were infrequently reported in patients with RA, PsA, axSpA, pSpA and CA. The safety profile of SARS-CoV-2 vaccines in patients with IJDs is reassuring.

REFERENCES:

1. Machado PM, et al. Ann Rheum Dis 2022;81:695–709.

184 - GIANT CELL ARTERITIS RELAPSE RISK – COULD THE EXTENT OF VESSEL INVOLVEMENT ON TEMPORAL AND AXILLARY ARTERIES ULTRASOUND BE A PROGNOSTIC MARKER?

Diogo Esperança Almeida[1, 2], Kate Smith[3], Borsha Sarker[3], Andrew Barr[2, 4], Richard Wakefield[2, 3], Sarah Mackie[2, 3]

[1]Rheumatology Department, Hospital de Braga, Braga, Portugal, [2]University of Leeds, Leeds Institute of Rheumatic and Musculoskeletal Medicine, Leeds, United Kingdom, [3]NIHR Leeds Musculoskeletal Biomedical Research Unit, Leeds Teaching Hospitals Trust, Leeds, United Kingdom, [4]Rheumatology Department, Chapel Allerton Hospital, Leeds, United Kingdom

Background: Giant cell arteritis (GCA) is the most common form of vasculitis after the age of 50. It is considered an emergency in Rheumatology context as prompt treatment mitigates the chance of permanent blindness. Because of the need for a timely diagnosis, ultrasound (US) of temporal and axillary arteries constitutes is a valuable element of the diagnostic work-up in GCA pathways. Although its role in diagnosis of GCA is now established, its role in defining the prognosis of GCA remains less clear.

Objective: To determine if disease extent on ultrasound of temporal and axillary arteries predicts a relapsing course of giant cell arteritis.

Methods: We conducted a single-centre retrospective study in which consecutive patients diagnosed with GCA between January 2019 (beginning of GCA fast-track US evaluations in the department) and May 2021 records were reviewed. Using Cox proportional hazards regression, we evaluated if disease extent, defined as the number of vessels showing non-compressible halo – 'halo count' – on baseline US scan performed as part of the diagnostic work-up is able to predict the risk of relapse in GCA.

Results: A total of 72 patients with a clinical diagnosis of GCA in which an US scan was performed in the diagnostic work-up were included. Thirty-seven (51.4%) experienced a relapse of GCA (median follow-up of 20.9 months; median time-to-first relapse of 6.3 months), ultimately needing treatment escalation. In a multivariable Cox regression model (n=72; -2 log likelihood = 275.63; $\chi 2 = 8.03$; df = 3; P = 0.045), halo count was found to be a significant predictor of time-to-relapse in GCA (HR = 1.19; CI 95% 1.04, 1.35; P = 0.012) after adjustment for patient sex and presence of ischaemic symptoms.

Conclusion: GCA disease extent as defined by US halo count at the time of diagnosis may predict relapsing disease and may help clinicians stratify care for these patients. Larger prospective studies are needed to confirm our findings.

190 - DIRECT AND INDIRECT EFFECT OF TNFI ON BASMI COMPONENTS IN PEOPLE WITH AXIAL SPONDYLOARTHRITIS: A LONGITUDINAL STUDY

Ana Sofia Pinto[1], Claire Harris[2], Rhys Hayward[2], Andrew Keat[2], Pedro M Machado[3]

[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]Rheumatology, Northwick Park Hospital, London North West University Healthcare NHS Trust, London, United Kingdom, [3]Centre for Rheumatology & MRC Centre for Neuromuscular Diseases, University College London, London, UK., London, United Kingdom

Background: The Bath Ankylosing Spondylitis Metrology Index (BASMI) is an index of spinal mobility for people with axial spondyloarthritis (axSpA). BASMI is one of the few objective measures of spinal disease progression that does not involve imaging. The assessment of mobility and physical function are essential components of the management of axSpA patients and, with the use of early therapeutics that are effective on improving/preserving function, mobility and, potentially structural damage, understanding the interplay between treatments and the various health outcomes affected by axSpA will contribute to better patient management.

Objective: To describe the long-term effect of TNF inhibitors (TNFi) on spinal mobility (each component of BASMI) in patients with axSpA, and to determine whether the use of TNFi treatment influences spinal mobility and if this is due to a direct or indirect effect (mediated by disease activity).

Methods: We performed a longitudinal study, using data routinely collected from patients with a clinical diagnosis of axSpA treated with TNFi at a specialist tertiary care center. Patients with at least two BASMI measurements (one before and one after TNFi initiation) were included. The relationship between TNFi treatment and BASMI over time was investigated using binomial generalized estimating equations (GEE). GEE is a technique that makes use of all available longitudinal data, allows unequal numbers of repeated measurements, corrects for within-subject correlation, and has some robustness against deviation from normality. We built 5 multivariable models; in the first three we investigated the isolated effect of TNFi (model 1), ASDAS (model 2) and BASDAI+CRP (model 3) on spinal mobility (each BASMI component); then we built two additional models, adjusting simultaneously for TNFi treatment and disease activity - one adjusted for TNFi and ASDAS (model 4) and one adjusted for TNFi and BASDAI+CRP (model 5). Other demographic and clinical variables were included as covariates in all multivariable models, including the time period between starting TNFi and the spinal mobility assessment.

Results: Data from 188 patients and 1326 visits were

Table I.

	bDMARD	ASDAS	BASDAI
Tragus-wall distance	β 0.874 (-0.104, 1.851); p=0.080	β 0.782 (0.261, 1.304); p=0.003	
	β 0.592 (-0.109,1.293); p=0.098		B 0.240 (0.056, 0.424); p=0.011
mSchober	β 0.178 (-0.016, 0.371); p=0.072	β -0.174 (-0.243, -0.105); p<0.001	
	β 0.184 (0.006, 0.362); p=0.043		β -0.086 (-0.119, -0.053); p<0.001
Mean lateral flexion	β 0.530 (0.062, 0.998); p=0.027	β -0.372 (-0.572, -0.172); p<0.001	
	β 0.457 (-0.002, 0.915); p=0.051		β -0.230 (-0.338, -0.123); p<0.001
Mean cervical rotation	β 2.945 (1.063, 4.828); p=0.002	β -1.785 (-2.462, -1.108); p<0.001	
	β 2.618 (0.785, 4.451); p=0.005		β -1.103 (-1.477, -0.729); p<0.001
Intermalleolar distance	β 2.532 (0.466, 4.598); p=0.016	β -1.392 (-2.454, -0.330); p=0.010	
	β 2.242 (0.266, 4.218); p=0.026		β -0.908 (-1.377, -0.440); p<0.001

analysed. Mean age was 45.6 (SD 11.6) years, mean disease duration was 15.8 (SD 9.64) years, 152 (80.9%) were male, 120 (73.6%) had radiographic axSpA, and 83 (74.8%) were HLA-B27 positive. Mean follow-up time was 8.0 (SD 4.4) years, ranging from 0.8 to 18.2 years. In the first three models, we observed a significant effect of TNFi in all the BASMI components, as we did for ASDAS and BASDAI (the only exception being for BASDAI, in the tragus-wall distance model). In the models combining TNFi treatment and disease activity, we found that TNFi treatment was significantly associated with improvement in the majority of BASMI components, even after controlling for disease activity (Table I).

Conclusion: TNFi has a long-term beneficial effect on all BASMI components, which seems to be both due to an indirect effect (mediated by disease activity) and a direct effect of TNFi treatment.

236 - NINTEDANIB IN CONNECTIVE TISSUE DISEASE-INTERSTITIAL LUNG DISEASE: EXPERIENCE OF A TERTIARY PNEUMOLOGY CENTER

Francisca Guimarães[1], Mariana Serino[2], Maria Pontes Ferreira[1], Hugo Parente[1], André Terras Alexandre[2], Natália Melo[2], Patrícia Mota[2, 3, 4], Hélder Novais Bastos[2, 3, 4], António Morais[2, 3, 4]

[1]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [2]Interstitial Lung Diseases Unit, Pneumology department, Centro Hospitalar Universitário do São João, Porto, Portugal, [3]Faculdade de Medicina da Universidade do Porto, Porto, Portugal, [4]Instituto de Investigação e Inovação em Saúde, Universidade do Porto, Porto, Portugal

Background: Progressive fibrosing interstitial lung disease (PF-ILD) is an important cause of morbidity and mortality in connective tissue diseases (CTD). Recently, two large randomized clinical trials (RCTs) - SENSCIS and INBUILD- allowed the approval of nintedanib, an inhibitor of multiple tyrosine kinases with antifibrotic properties, in Systemic Sclerosis (SSc)-ILD and PF-ILD (which includes other CTDs), respectively. Gastrointestinal toxicity was the main adverse event reported on RCTs with nintedanib. Real word data are needed to better understand safety and effectiveness of nintedanib in PF-ILD. With this study we aimed to characterize our cohort of patients with CTD-ILD that started on nintedanib.

Methods: Retrospective and longitudinal study including patients diagnosed with CTD-ILD followed between January 2019 until February 2022, in a tertiary pneumology centre. Sociodemographic data, CTD-ILD characterization and treatment information were collected. Baseline pulmonary functional tests (PFT) and changes on high-resolution computer tomography (HRCT), and at 3, 6 and 12 months after the beginning of nintedanib therapy were recorded. Data on adverse events and need to suspension of antifibrotic were collected. Descriptive analysis was used to characterize the cohort and adverse events. Paired-sample T-test was performed to compare baseline Functional Vital Capacity (FVC) and Diffusing Capacity of Lung for Carbon Monoxide (DLCO) with FVC and DLCO at 3, 6 and 12 months after therapy. SPSS v25 was used for statistical analysis and significance level was defined as 2-sided $p< 0.05$.

Results: In total, 135 patients with CTD-ILD were included, 68.9% were female, with a mean age at diagnosis of 60.05±12.77. Of these, 21 patients were medicated with nintedanib, after a mean follow up of 4.14±3.00 years. Most of these patients were female (66.7%), with a mean age of diagnosis of 61.12±9.36 years old. The most common CTD was Rheumatoid Arthritis and SSc (38.1 and 33.3%, respectively) and Usual Interstitial Pneumonia was the most common radiological pattern (52.4%). Most patients (76.2%) presented extensive disease with more than 20% of the lung affected. Most of these patients were also medicated with immunosuppressive drugs, mainly rituximab (57.1%) and mycophenolate mofetil (28.6%).

After 10±3.71 months on nintedanib, 63.2% of the patients had at least one adverse event, namely diarrhoea (75.0%), that leaded to temporary discontinuation in 31.6% and definitive drug suspension in 10.5% of the patients. FVC and DLCO were stable after 3, 6 and 12 months of therapy. Six patients performed a 12-month HRCT after the start of nintedanib, where most presented (66.7%) a stable disease.

Conclusions: Adverse events are common, early after the beginning of antifibrotic. Gastrointestinal manifestations were the most common, especially diarrhoea, as reported in literature. Nevertheless, most adverse events didn't require definitive suspension of the drug. Immunosuppressive and antifibrotic association was common, and even thought INBUILD trial excluded patients on rituximab, in our cohort, concomitant use of rituximab was common with no red alert regarding safety.

Yet, our follow up time after the beginning of the antifibrotic therapy was short and longer follow up times and larger sample are needed to better assess safety and efficacy of nintedanib in CTD-ILD.

Table I. Patient and disease characteristics at baseline

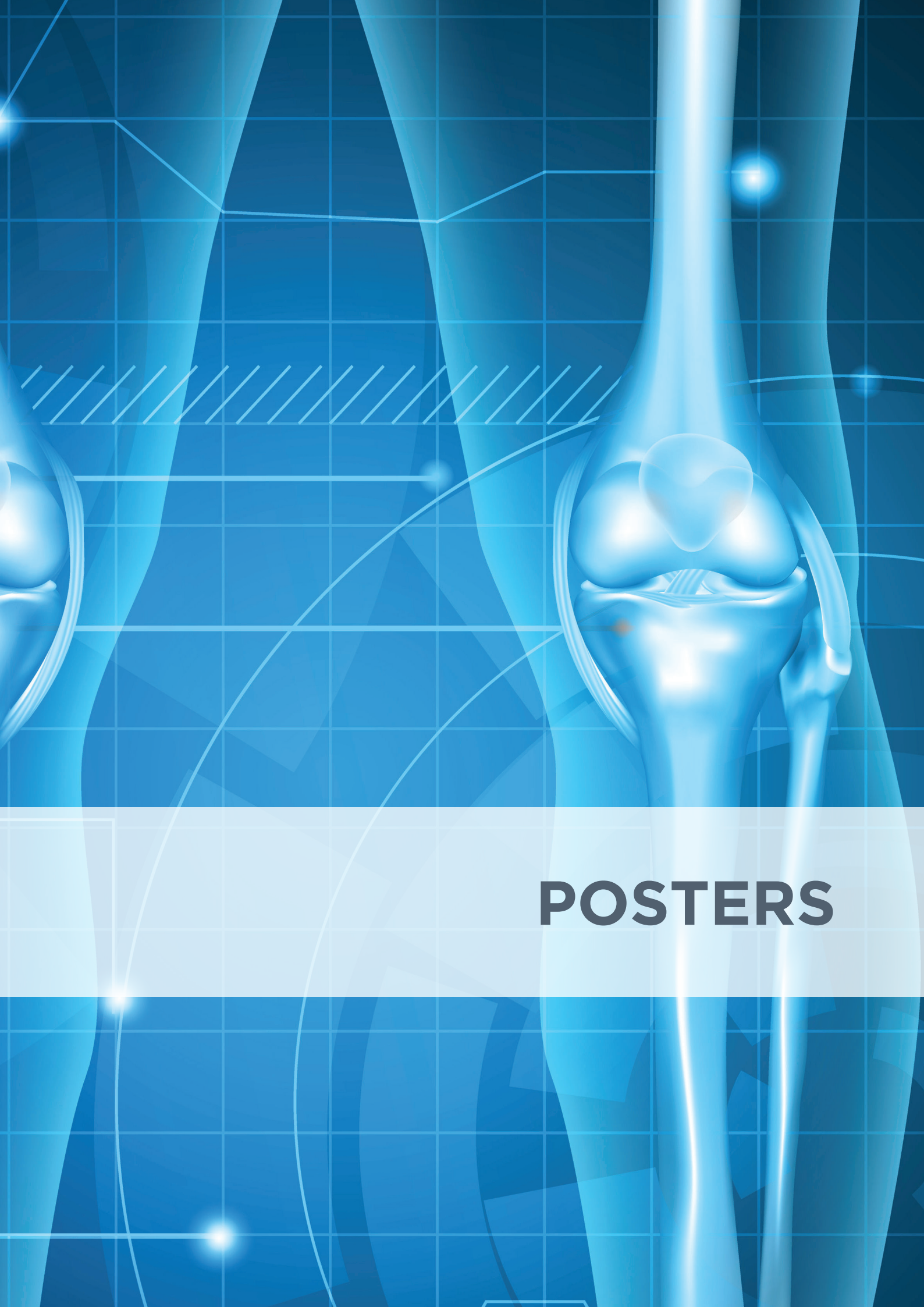
	All patients
Sociodemographic characteristics	
Age (years)	66.60±8.90
Gender (Female)	14/21 (66.7%)
Race (White European origin)	20/21 (95.2%)
Smoking status (Never smoked)	11/20 (55.0%)
CTD- Disease characteristics	
Diagnosis	
Rheumatoid arthritis	8/21 (38.1%)
Systemic sclerosis	7/21 (33.3%)
Inflammatory immune mediated myopathy	0/21 (0.0%)
Sjögren Syndrome	2/21 (9.5%)
Systemic lupus erythematosus	1/21 (4.8%)
Overlap syndrome Rheumatoid arthritis/Systemic sclerosis	1/21 (4.8%)
Mixed connective tissue disease	1/21 (4.8%)
Undifferentiated Connective Tissue disease	1/21 (4.8%)
ILD-disease characteristics	
Age at diagnosis (years)	61.12±9.36
Previous CTD diagnosis (yes)	18/21 (85.7%)
RFT pattern	
Normal	0/21 (0.0%)
Restrictive	13/21 (61.9%)
Obstructive	1/21 (4.8%)
Isolated decreased DLCO	7/21 (33.3%)
ILD classification	
Non-specific interstitial pneumonia	9/21 (42.9%)
Usual interstitial pneumonia	11/21 (52.4%)
Organizing pneumonia	1/21 (4.8%)
Baseline PFT of antifibrotic	
FVC, %	80.26±19.54
DLCO, %	33.97±15.10
Baseline changes in HCRT	
Reticulations (yes)	21/21 (100%)
Ground Glass (yes)	9/21 (42.9%)
Traction bronchiectasis (yes)	21/21 (100%)
Honeycombing (yes)	12/21 (57.1%)
Fibrosis (yes)	21/21 (100 %)
Lung extension > 20% (yes)	16/21 (76.2%)

continues on the next page

Table I. continuation

	All patients
Treatment	
Glucocorticoid (yes)	12/21 (57.1%)
Hydroxychloroquine (yes)	5/21 (23.8%)
Methotrexate association (yes)	4/21 (22.2%)
Mycophenolate mofetil association (yes)	6/21 (28.6%)
Cyclophosphamide (yes)	0/21 (0.0%)
Rituximab (yes)	12/21 (57.1%)
Antifibrotic therapy	
Adverse events (yes)	12/19 (63.2%)
Diarrhea (yes)	9/12 (75.0%)
Nausea/vomits (yes)	3/12 (25.0%)
Weight Loss (yes)	1/12 (8.3%)
Hepatitis/cholestasis (yes)	2/12 (16.6%)
Suspension	
Temporary	6/19 (31.6%)
Definitive	2/19 (10.5%)
Follow-up on antifibrotic (months)	10±3.71

CTD: connective tissue disease; DLCO: diffusion lung carbon oxide; FVC: functional vital capacity; HRCT: high resolution computerized tomography; ILD: Interstitial lung disease; PFT: Pulmonary functional tests.
Continuous variables are presented as mean ± standard deviation and categorical variables as number/total population available (percentage).



POSTERS

Trabalho Original

001 - IMMUNE RESPONSES TO mRNA VACCINES AGAINST SARS-CoV-2 IN PATIENTS WITH IMMUNE-MEDIATED INFLAMMATORY RHEUMATIC DISEASES

Cristiana Sieiro Santos[1], Ruben Rego Salgueiro[2], Clara Moriano[1], Carolina Álvarez[1], Sara Calleja[3], Jose Garcia Ruiz de Morales[3], Elvira Díez[1]
[1]Reumatología, Complejo Asistencial Universitario de León, León, Spain, [2]Medicina Interna, ULS Guarda, Guarda, Portugal, [3]Immunology, Complejo Asistencial Universitario de León, León, Spain

Background: Patients with immune-mediated rheumatic diseases (IMRDs) are commonly treated with immunosuppressors and prone to infections. Recently introduced mRNA SARS-CoV-2 vaccines have demonstrated extraordinary efficacy across all ages. Immunosuppressed patients were excluded from phase III trials with SARS-CoV-2 mRNA vaccines.
Objectives: To fully characterise B-cell and T-cell immune responses elicited by mRNA SARS-CoV-2 vaccines in patients with rheumatic diseases under immunotherapies, and to identify which drugs reduce vaccine's immunogenicity.
Methods: Humoral, CD4 and CD8 immune responses were investigated in 100 naïve patients with SARS-CoV-2 with selected rheumatic diseases under immunosuppression after a two-dose regimen of SARS-CoV-2 mRNA vaccine. Responses were compared with age, gender and disease-matched patients with IMRD not receiving immunosuppressors and with healthy controls.
Results: Patients with IMRD showed decreased seroconversion rates (80% vs 100%, p=0.03) and cellular immune responses (75% vs 100%, p=0.02). Patients on methotrexate achieved seroconversion in 62% of cases and cellular responses in 80% of cases. Abatacept decreased humoral and cellular responses. Rituximab (31% responders) and belimumab (50% responders) showed impaired humoral responses, but cellular responses were often preserved. Antibody titres were reduced with mycophenolate and azathioprine but preserved with leflunomide and anticytokines.
Conclusions: Patients with IMRD exhibit impaired SARS-CoV-2 vaccine immunogenicity, variably reduced with immunosuppressors. Among commonly used therapies, abatacept and B-cell depleting therapies show deleterious effects, while anticytokines preserved immunogenicity. The effects of cumulative methotrexate and glucocorticoid doses on immunogenicity should be considered. Humoral and cellular responses are weakly correlated, but CD4 and CD8 tightly correlate. Seroconversion alone might not reflect the vaccine's immunogenicity.

002 - FACTORS ASSOCIATED WITH ADVERSE OUTCOMES IN UVEITIS RELATED TO SPONDYLARTHRITIS (SpA-U)- DEVELOPMENT OF A PROGNOSTIC OUTCOME SCORE IN PATIENTS WITH SpA-U

Cristiana Sieiro Santos[1], Isabel Sendino Tenorio[2], Carolina Álvarez[1], Miguel Cordero[2], Clara Moriano[1], Elvira Díez[1]
[1]Reumatología, Complejo Asistencial Universitario de León, León, Spain, [2]Ophtalmology, Complejo Asistencial Universitario de León, León, Spain

Background: Uveitis is the most frequent extra-articular manifestation of spondylarthritis (SpA), characterized by a sudden onset, often unilateral, anterior and recurrent and may be the first clinical manifestation of the disease. The lack of standardized and validated outcome measures in uveitis makes it difficult to evaluate the efficacy and refractoriness to treatment and determine factors associated with adverse outcomes.
Objectives: To develop a prognostic outcome score for patients with uveitis associated with spondylarthritis (SpA-U) and determine factors associated with adverse outcomes in uveitis associated to SpA-U in patients under systemic treatment.
Methods: Clinical records of patients with SpA-U from 1990 to 2020 were retrospectively reviewed, including sociodemographic features, factors related to articular involvement, therapeutic choices and data related to uveitis outbreaks. The prognostic outcome score was defined by visual acuity, inflammation in anterior chamber (anterior chamber cells, hypopyon, presence of fibrin, active posterior keratic precipitates), presence of synechia, pupilar membrane, epiretinal membrane or any complications (macular oedema, vitritis, panuveitis, peripheral ulcerative keratitis), and refractoriness to 2 or more d/csDMARDs (conventional synthetical disease-modifying anti-rheumatic drug) or 1 or more bDMARD (biological disease-modifying anti-rheumatic drug) treatment. The prognostic outcome score ranked from 0 (good) to 5 (bad). Factors associated with adverse outcomes in uveitis were studied using linear regression. For categorical factors, marginal averages and their standard errors are displayed together with linear regression coefficients with 95% confidence intervals (CI). For continuous factors, averages and standard

deviations are reported in addition to linear regression coefficients with 95% CI. For each variable, two regression coefficients are reported: unadjusted and adjusted for age at diagnosis and sex.

Results: 42 patients were included, 59.5% were male, with a mean age at diagnosis of 36.6±11.9 years and with a total of 190 uveitis outbreaks. Time since diagnosis was 12.5±7.9 years. 64.4% of patients had uveitis as disease onset. 52.4% were overweight (BMI≥30 kg/m2), 16.7% were former/active smokers. 28.6% of patients had a family history of SpA. 14.3% had 1-2 uveitis outbreaks, 47.6% had 3-5 uveitis outbreaks and 38.1% had 6-11 uveitis outbreaks. 102 (53.7%) uveitis outbreaks fulfilled 1 criterion, 38 (20%) uveitis outbreaks fulfilled 2 criteria, 19 (10%) uveitis outbreaks fulfilled 3 criteria and 5 (2.6%) uveitis outbreaks fulfilled 4 or more. The results of the linear regression model revealed that the uveitis was more severe in patients with smoking history (β=0.34), axial and peripheral involvement (β=0.43), a BASDAI (Bath Ankylosing Spondylitis Disease Activity Index)>4 (β=0.32), positive HLA-B27 (β=0.29), female sex (β=0.19), patients with CRP (C-reactive protein) elevation (β=0.002) and a history of bilateral ocular involvement (β=0.32) while shorter disease evolution (β=-0.02) and normal vitamin D levels (β= -0.03) were associated with a better outcome.

Conclusion: We identified factors associated with adverse outcomes in SpA-U by developing a prognostic outcome score that integrates ocular inflammatory activity, ocular complications and refractoriness to treatment.

003 - FACTORS ASSOCIATED WITH ADVERSE PREGNANCY OUTCOMES IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS (SLE)

Cristiana Sieiro Santos[1], Ruben Rego Salgueiro[2], Clara Moriano[1], Carolina Álvarez[1], Elvira Díez[1]

[1]Reumatología, Complejo Asistencial Universitario de León, León, Spain, [2]Medicina Interna, ULS Guarda, Guarda, Portugal

Background: Pregnancies in systemic lupus erythematosus (SLE) are considered high risk and associated with maternal and obstetric complications.

Objectives: Our goal with this study was to determine the most important predictors for each of the main adverse pregnancy outcomes in SLE patients.

Methods: We conducted a retrospective case-controls study by including multiparous women diagnosed with SLE from 1980 to 2020 followed in our unit and compared the clinical profile of patients with adverse pregnancy outcomes to control SLE patients. We excluded elective terminations of pregnancy and cases lost to follow-up. Qualitative data were analyzed by Chi-square test and Fisher's exact test and continuous variables were analyzed by using Student's t test. Multiple logistic regression models were performed to determine the predictive factors for adverse pregnancy outcomes with adjustment of confounding factors. In all tests, P values less than 0.05 were considered to be statistically significant.

Table I. Multiple logistoc analysis

	Abortion	Stillbirth	Pre-eclampsia	PROM	Ectopic pregnancy	Neonatal Lupus
Anti-DNAds	β =0,71, p=0.04	β =0.26, p=0.04	β =0.12, p=0.38	β =0.15, p =0.24	β = 0.10, p =0.64	β = 0.16, p=0.24
APA	β = 0.2, p = 0.03	β = 0.22, p=0.03	β = 0.11, p = 0.85	β = 0.26, p = 0.04	β = 0.16, p = 0.21	β = 0.83, p = 0.08
Renal involvement	β=0,28, p 0.03	β = 0.26, p = 0.38	β = 0.33, p = 0.83	β = 0.07, p = 0.53	β = 0.17, p = 0.20	β = 0.58, p = 0.07
Serositis	β = 0.85, p = 0.95	β = 0.11, p = 0.41	β=0.31, p=0.02	β = 0.06, p = 0.46	β = 0.13, p = 0.35	β = 0.08, p = 0.46
Direct Coombs positivity	β =0.11, p = 0.41	β = 0.03, p = 0.81	β = 0.42, p=0.01	β=0.03, p =0.83	β= 0.14, p = 0.81	β=0.03, p=0.83
Anti-Ro/SSA	β=0.19, p = 0.13	β=0.03, p = 0.83	β=0.07, p =0.62	β=0.11, p=0.39	β=0.09, p=0.52	β= 0.16, p 0.02
Anti-RNP	β =0.5, p=0.69	β = 0.09, p=0.49	β = 0.16, p= 0.23	β = 0.09, p= 0.81	β=0.03, p=0.81	β=0.16, p=0.03

Results: 135 multiparous women were included (43% with adverse pregnancy outcomes). A total of 57 pregnancies (42%) were linked to adverse outcomes. The occurrence of abortion was correlated with anti-DNAds (β=0.71. p=0.04), renal involvement (β=0.28, p 0.03), antiphospholipid antibodies (β=0.29, p 0.03), ESR elevation (β =0.81, p=0.02) and CPR elevation (β =0.91, p=0.01). Stillbirth was also correlated with renal involvement (β= 0.26, p =0.04), antiphospholipid antibodies (β=0.22, p=0.03) and ESR elevation (β =0.53, p=0.02). Preeclampsia was correlated with direct Coombs positivity (β=0.42, p=0.01), serositis (β=0.31, p=0.02), ESR elevation (β =0.52, p=0.03) and CPR elevation (β =0.32, p=0.04). Neonatal Lupus was correlated with anti-RNP (β=0.16, p=0.03) and anti-Ro/SSA (β=0.16, p 0.02).

Conclusion: The most unfavorable pregnancy outcome in women with SLE was spontaneous abortion. Renal involvement, anti-DNAds positivity, antiphospholipid antibody positivity, anti-Ro/SSA, elevated ESR and a younger age at disease onset increased the risk of pregnancy complications.

004 - PREDICTORS OF INTERSTITIAL LUNG INVOLVEMENT AND TIMING OF ONSET IN SYSTEMIC SCLEROSIS

Cristiana Sieiro Santos[1], Ruben Rego Salgueiro[2], Clara Moriano[1], Elvira Díez[1]

[1]Reumatología, Complejo Asistencial Universitario de León, León, Spain, [2]Medicina Interna, ULS Guarda, Guarda, Portugal

Background: Interstitial lung disease (SSc-ILD) and pulmonary hypertension are the leading causes of

death in patients with systemic sclerosis (SSc). Identifying SSc-ILD development and initiating treatment is essential to optimize therapeutic benefit.
Objectives: We aimed to identify predictors of SSc-ILD and compared early (<5 years from diagnosis) versus late (>5 years from diagnosis) onset.
Methods: We conducted a retrospective cohort study by including patients diagnosed with SSc from 1980 to 2020 followed in our unit and compared the clinical profile of patients with SSc-ILD to control SSc-non-ILD patients. Demographic features, clinical and immunological characteristics, baseline pulmonary function and capillaroscopy data were retrieved. Logistic regression modelling was run to identify factors associated with SSc-ILD development. Factors associated with ILD were then determined as factors associated with early or late onset using multivariate analysis. Bonferroni correction was used to limit Type I errors.
Results: We have included 103 patients from our patient registry from 1980 to 2021 (42% with SSc-ILD). Logistic regression identified risk factors associated with increased or decreased odds ratio for developing ILD is summarized in table 1. Smoking history, male sex, the presence of myositis, anti-Scl70 and anti-Ro52 positivity, baseline pulmonary function including FVC and DLCO, mMRC (Modified Medical Research Council) dyspnea scale>2, mMSS (Modified Rodnan Skin Score), and late pattern in capillaroscopy were identified as SSc-ILD predictors. Older age at SSc diagnosis, the presence of telangiectasias and smoking status were correlated with of SSc-ILD onset before 5 years, while male gender, the presence of myositis and antiphospholid antibodies were correlated with late-onset SSc-ILD.
Conclusion: We identified 10 factors significantly associated with risk of developing SSc-ILD: smoking, male sex, diffuse cutaneous involvement, the presence of myositis, shorter Raynaud duration to SSc diagnosis, anti-Scl70 and anti-Ro52 positivity and baseline pulmonary function (lower baseline DLCO and FVC increasing risk) and late capillaroscopy pattern and identified predictors for early and late-onset SSc-ILD.

005 - CANCER IN SYSTEMIC SCLEROSIS: ASSOCIATION BETWEEN ANTIBODIES AND MALIGNANCY

Cristiana Sieiro Santos[1], Ruben Rego Salgueiro[2], Clara Moriano[1], Elvira Díez[1]
[1]Reumatología, Complejo Asistencial Universitario de León, León, Spain, [2]Medicina Interna, ULS Guarda, Guarda, Portugal

Background: Systemic sclerosis (SSc) is associated with increased risk of malignancy. Risk factors predisposing a SSc patient for development of malignancy are not well defined, and the pathogenic basis of the association is yet to be explained. Some autoantibodies have been associated with a close temporal relationship with cancer. The absence of malignancy screening guidelines tailored for SSc patients raise the importance of the need for more studies on the association of SSc and cancer.
Objectives: To study the prevalence of cancer in SSc and the association between SSc-specific and SSc-associated autoantibodies and cancer in a third-level center.
Methods: We conducted a retrospective cohort study by including patients diagnosed with SSc followed from 1980 to 2020 fulfilling the 2013 ACR/EULAR SSc criteria. Demographic features, clinical and immunological characteristics were retrieved. The primary outcome was cancer-associated SSc, defined as cancer occurring within 2, 3 and 5 years of first non-Raynaud SSc manifestation. The exposure was defined by the presence of SSc-specific/associated autoantibodies, including anti-centromere (ACA), topoisomerase I (Scl70), RNA polymerase III, fibrillarin, Th/To, PM-Scl, Ku, TIF1g, Ro52. Descriptive analysis was used to compare clinical characteristics of subjects with cancer to those without cancer. Univariate logistic regression was used to compare the odds of cancer-associated SSc between the autoantibody subgroups.
Results: Out of 103 SSc subjects, 27 (26%) had a history of cancer following SSc diagnosis. Mean age was 61.9 (57-69) years, 70% were female and 88% had a smoking history. Median time between cancer and disease onset was 6.33 (3-9) years. Among patients with cancer, 12 (44%), 8 (29%) and 7 (26%) were diagnosed within 2, 5 and 10 years of SSc onset. The most frequent types were breast cancer (n=9), gastrointestinal cancer (n=5), prostatic cancer (n=4), hematological (n=3) cancers, cervical/uterine cancers (n=2), non-melanoma skin (n=2), lung cancer (n=2). Patients with cancer were more likely to be Sc70+(OR 2.55, 95% CI1.03-6.3, p 0.04), anti-TIF1g (OR 19.5, 95% CI 5.6 – 68.3, p 0.001) and RNA pol III (OR 10.9 CI 95% 1.08-109.3, p 0.04), have a history of smoking (OR 7.24, 95% CI 2.6-197, p 0.001), myositis (OR 5.2 IC 95% 2.06-13.2, p 0.005) and older age at SSc onset (61.9 vs 57, p 0.04). Breast cancer was more frequent in anti-TIF1g (OR 3.75 IC 95% 1.8-17.5) and RNA pol-III (OR 7.14 IC 95% 1.56-90.8) subgroups. The risk of cancer-associated SSc was significantly increased among anti-TIF1g-positive subjects at 5 years after SSc onset (OR 2.1 CI 95% (1.45-9.94), p 0.04) and among RNA-pol III-positive subjects at 2 years after SSc onset (OR 3.5 95% CI (1.2-51.4), p 0.02) (table 1).
Conclusion: Anti-Scl70, anti-TIF1g and RNA pol III were predictive of cancer-associated SSc for cancers.

Breast cancer was the most frequent. Subjects with cancer were more likely to have a history of smoking, myositis and an older age at SSc onset. Autoantibodies should be taken into account in cancer screening. Larger studies are needed to define the risk of cancer-associated SSc in different autoantibody subgroups.

006 - CLINICAL PHENOTYPE IN SCLERODERMA PATIENTS WITH LIMITED AND DIFFUSE CUTANEOUS DISEASE BASED ON AUTOIMMUNITY

Cristiana Sieiro Santos[1], Ruben Rego Salgueiro[2], Clara Moriano[1], Elvira Díez[1]

[1]Reumatología, Complejo Asistencial Universitario de León, León, Spain, [2]Medicina Interna, ULS Guarda, Guarda, Portugal

Background: Classically, anti-centromere antibodies (ACA) are associated with limited cutaneous involvement (lcSSc) and pulmonary hypertension, whereas anti-topoisomerase I (Sc70) are associated with diffuse skin involvement (dcSSc) and pulmonary fibrosis (ILD). Patients with lcSSc and Sc70 antibodies draw particular attention, which is why characterization of clinical phenotypes can help distinguish patient subgroups and assessing the prognosis of the disease.

Objectives: We aimed to characterize the clinical phenotype of patients with SSc based on autoantibodies.

Methods: We included patients with SSc, fulfilling the 2013 ACR/EULAR criteria, with disease duration ≤10 years. We have compared different subgroups of patients with lcSSc: Scl70-lcSSc (group1), ACA-lcSSc (group 2) and ANA-lcSSc (group 3), (table 1). Next, we compared patients with Scl70-lcSSc (group 1) to Scl70-dcSSc (group 4) and ANA-lcSSC (group 3) to ANA-dcSSC (group 5). In the ANA subgroup we included patients with negative Scl70 and ACA antibodies. We have assessed the risk of ILD, myositis, scleroderma renal crisis, cardiac and gastrointestinal involvement, myositis, pulmonary hypertension (systolic pulmonary arterial pressure sPAP>45 mmHg at transthoracic echocardiography or sPAP>25 mmHg at right heart catheterization), cancer and all-cause-mortality.

Results: 103 SSc patients were included: 72 (69%) females, 82 (79%) lcSSc and 21 (20%) dcSSc. Among lcSSc patients, 43 (52%) had ACA, 16 (19%) Scl70 and 23 (28%) ANA. Among dcSSC patients, 9 (43%) had Scl70 and 12 (57%) had ANA. Scl70-lcSSc patients had significantly shorter time from Raynaud's phenomenon (RP) to SSc diagnosis (p=0.02), younger age at SSc onset (p=0.04), higher mRSS (p=0.03), higher rate of myositis (p=0.04) and renal scleroderma crisis (p=0.04) than ACA-lcSSc patients. The risk of ILD in Scl70-lcSSC compared to ACA-lcSSC is 3.8 higher (95% IC 1.2-14.5) and 1.5 higher (95% IC 1.05-5.61) than ANA-lcSSC. All-cause mortality was higher in Scl70-lcSSC (p=0.04) compared to ACA-lcSSC. Scl70-dcSSC patients had a shorter time from RP to SSc diagnosis (p=0.02), higher CRP (p=0.04), mRSS (p=0.001), higher rate of myositis (p<0.05) and ILD (p=0.04) and all-cause mortality (p=0.04) than Scl70-lcSSc patients, while renal and cardiac involvement was similar. ANA-dcSSC patients also had a shorter time from RP to SSc diagnosis (p=0.03), higher mRSS (p=0.02) and higher rate of ILD (p=0.02).

Conclusion: Scl70-lcSSc patients show the major organ involvement, followed by ANA-lcSSc and ACA-lcSSc. Scl70-dcSSc and ANA-dcSSC patients exhibit more cutaneous involvement and ILD than Scl70-lcSSc and ANA-lcSSc. These results may provide new ways to help in early diagnosis, management and assessing the prognosis of the disease.

Table I. Clinical characteristics of patients with lcSSc

	Scl70-lcSSc	P value	ACA-lcSSc	P value	ANA-lcSSc
Female	9 (56.3%)	**0.03**	36 (83.7%)	0.80	12 (52%)
Time from RP to SSc (years)	1.28 (0.25-3.5)	**0.02**	6.23 (3-9)	**0.002**	4.91 (2-5)
Age at SSc onset	56.5 (48-66)	**0.04**	62.5 (54-73)	0.76	59.2 (44-66)
CRP elevation	30 (22-39)	**0.02**	22 (16-29)	0.21	10 (6-19)
mRSS	4 (2-4.2)	**0.03**	3.2 (2.1-4.0)	0.92	3.9 (2.5-4.2)
Joint synovitis	6 (37.5%)	0.47	12 (28%)	0.71	10 (43%)
Tendon friction rubs	2 (12.5%)	0.50	3 (7%)	0.19	0
Myositis	4 (25%)	**0.04**	2 (5%)	0.81	5 (21.7%)
Gastrointestinal involvement	6 (37.5%)	0.19	27 (63%)	**0.04**	16 (69%)
Renal crisis	4 (25%)	**0.04**	2 (5%)	0.18	2 (9%)
ILD	10 (63%)	**0.001**	7 (16%)	**0.04**	7 (30%)
Pulmonary hypertension	4 (25%)	0.47	7 (16%)		3 (13%)
Arrythmia	5 (21.7%)	0.08	6 (14%)	0.35	7 (30%)
Conduction defects	1 (6.3%)	0.59	5 (11.6%)	0.79	1 (4%)
Diastolic disfunction	7 (43.8%)	0.36	17 (39.5%)	0.77	9 (39%)
Immunosuppressants	10 (63%)	**0.01**	13 (30.2%)	0.52	12 (52%)
Steroids	9 (56%)	**0.04**	12 (28%)	0.98	13 (56.6%)
Cancer	8 (50%)	**0.04**	10 (23.2%)	0.22	7 (30%)
Mortality	3 (18.8%)	**0.04**	6 (14%)	0.82	5 (21.7%)

Table I. Logistic regression analysis for the risk of cancer within 2,5 and 10 years of SSc onset according to antibody positivity

	OR 95% CI for cancer diagnosis within 2 years of SSc onset	OR 95% CI for cancer diagnosis within 5 years of SSc onset	OR 95% CI for cancer diagnosis within 10 years of SSc onset
Anti-Scl70	1.56 (0.51-44.5)	1.41 (0.27-12.7)	2.1 (0.55-13.4)
Anti-centromere	1.72 (0.62-52.1)	1.52 (0.32-32.1)	1.69 (0.14-21.3)
Anti-TIF1g	3.9 (0.35-43.4)	**2.1 (1.45-9.94)**	0.42 (0.04-5.32)
ARN Pol III	**3.5 (1.2-51.4)**	1.9 (0.72-62.1)	-
Anti-PM Scl75/100	-	-	0.87 (0.04-1.98)
Anti-Ro52	2.5 (0.66-66.8)	2.0 (0.45-50.2)	0.63 (0.05-7.74)

010 - JAKS AND STATS: WHICH GAME DO THEY PLAY IN CHRONIC ARTHRITIS?

Rui Lourenço Teixeira[1, 2, 3], Rita A. Moura[2], Alcobia[3], 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]Instituto de Histologia e Biologia do Desenvolvimento, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal

Rheumatoid arthritis (RA) is a chronic systemic immune-mediated inflammatory disease that mainly affects the joints. Albeit targeted treatments have a significative impact on disease control, no more than one third of patients with chronic arthritis achieve remission. Instead, they suffer from a difficult to treat, disabling disease. Upadacitinib (UPA) is a novel JAK1 antagonist approved for refractory RA. Although UPA is already in use by rheumatologists, most of the mechanisms that underlie its efficacy are yet to be understood. Our group has been studying JAK-STAT signaling pathway in early drug-naïve arthritis patients. We are now focusing on the effects of the clinical administration of UPA in established RA. Our aim is to characterize an RA cohort of patients when clinically exposed to UPA, having clinical remission as primary endpoint. We aim to analyze the effect of UPA on JAK-STAT signaling pathway activation, namely on STAT phosphorylation of peripheral blood mononuclear cells (PBMC) and synovial inflammatory infiltration.

For this purpose, we have designed an investigator-initiated clinical and translational study, approved and funded by the pharmaceutical company Abbvie. A pilot study is currently being held at our center. The clinical study has been submitted and obtained approval by both national and local ethics committees and has been registered on the EUDRA-CT. Patients are invited to perform a blood collection and a synovial biopsy before starting UPA and after 6 months of therapy. Patients receive UPA under close clinical supervision. STAT activation levels are accessed in both timepoints by PBMC flow cytometry with a focus at T cells, B cells, monocytes and dendritic cells. Synovial inflammation is characterized by applying Krenn's score and immunohistochemistry for lymphocytes and macrophages.

Under the light of its clinical efficacy, we expect to find a great proportion of patients achieving remission with a significant STAT activation reduction.

013 - START FROM SCRATCH: ANALYSING THE IMPACT OF A RHEUMATOLOGY CENTRE BORN IN THE REUMA.PT ERA

Eduardo Dourado[1, 2], Ana Valido[3]

[1]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Serviço de Reumatologia, Unidade Local de Saúde do Litoral Alentejano, Santiago do Cacém, Portugal

Background: According to the Portuguese Epidemiologic Study of Rheumatic Diseases, the Alentejo region has the highest estimated prevalence of rheumatoid arthritis (RA, 1.8%) and the second-highest estimated prevalence of axial spondyloarthritis (axSpA, 2.3%) in Portugal. However, until 2017, National Health Service hospitals in Alentejo had no rheumatologists. From 2017 to 2020, Centro Hospitalar Universitário Lisboa Norte developed a monthly Rheumatology appointment in Unidade Local de Saúde do Litoral Alentejano (ULSLA), which serves a population greater than 100.000 people. In November 2020, ULSLA hired a rheumatologist, and the first Rheumatology centre in Alentejo was finally founded.

Objective: To describe the impact of a new Rheumatology centre in a Portuguese secondary care hospital.

Methods: Retrospective study comparing analogous periods before (November 2018 to December 2019) and after (November 2020 to December 2021) the implementation of the new Rheumatology centre. Accessibility and productivity data were obtained from the hospital's Planning and Management Office. Clinical data were retrieved from the Rheumatic Diseases Portuguese Register (Reuma.pt). Disease activity data refers to the score registered at the last appointment.

Results: The number of Rheumatology appointments performed in ULSLA increased from 210 to 1754, and the number of patients observed increased from 191 to 708 compared to the analogous period. The percentage of appointment requests resulting in an appointment raised from 70% (N=58/83) to 84% (218/260), and the percentage of appointment requests not resulting in an appointment due to reasons imputable to scheduling delays decreased from 10% (N=8/83) to 3% (N=7/260). The median referral-to-appointment time decreased from 264 days in 2019 to 84 days in 2021, and the percentage of appointments performed within the legally defined maximum response time increased from 0% to 71%. In addition, the median primary care referral-to-Rheumatology appointment time was reduced irrespective of the triage priority: from 287 to 110 days for normal priority, 285 to 76 days for urgent priority, and 348 to 87 days for very urgent priority.

About a quarter (N=163/708, 23.0%) of the patients evaluated in the period of interest had a diagnosis of either inflammatory arthropathy or connective tissue disease and were included in the centre's Reuma.pt da-

tabase. Most of these patients were female (N=117/163, 71.7%) and Caucasian (N=144/150, 89.3%), with a mean age at the last appointment of 58.3±14.2 years. The most common diagnoses were RA (N=68, mean DAS28 of 3.28±1.57 [n=64]), axSpA (N=31, mean ASDAS of 1.98±0.86 [n=28]; mean SPARCC of 0.3±0.9 [n=19]), psoriatic arthritis (N=15), Sjögren's syndrome (N=11), systemic lupus erythematosus (N=10, mean SLEDAI of 2.7±4.6 [n=7]), and systemic sclerosis (N=9, mean mRSS of 5.0±9.0 [n=5]). The most used disease-modifying anti-rheumatic drugs (DMARD) were methotrexate (N=72), sulfasalazine (N=31), and hydroxychloroquine (N=30), but also included biological and targeted synthetic DMARD such as adalimumab (N=16), etanercept (N=7), secukinumab (N=3), and tofacitinib (N=3). The most common comorbidities were arterial hypertension (N=29), dyslipidaemia (N=21), and psoriasis (N=16), and nine patients had neoplasms.
Discussion: Hiring a rheumatologist significantly improved the accessibility to a Rheumatology appointment for patients in the ULSLA referral zone. Besides, the ULSLA quality indicators, such as the accomplishment of the legally defined maximum response time, dramatically improved.

017 - IMPACT OF SARS-COV-2 INFECTION ON THE DISEASE ACTIVITY OF PATIENTS WITH PSORIATIC ARTHRITIS UNDER BDMARDS: REAL LIFE DATA

Rafaela Nicolau[1], Ana Martins[2], Daniela Santos Oliveira[2, 3], Beatriz Samões[4], Frederico Rajão Martins[5], Maria Seabra Rato[2], Filipe Oliveira Pinheiro[6], Miguel Bernardes[2], Lúcia Costa[2]

[1]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar de São João, Porto, Portugal, [3]Faculdade de Medicina da Universidade do Porto, Departamento de Medicina e CINTESIS, Porto, Portugal, [4]Rheumatology Department, Centro Hospitalar Vila Nova de Gaia/Espinho, Gaia, Portugal, Vila Nova de Gaia, Portugal, [5]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [6]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal

Background: SARS-CoV-2 infection can lead to severe inflammation and has been suggested to induce Psoriatic Arthritis (PsA) flares.1 However, the impact on disease activity and response to biological disease modifying anti-rheumatic drugs DMARDs (bDMARDs) remains unknown.
Objectives: To evaluate the effect of SARS-CoV-2 infection on disease activity and bDMARDs responses in patients with PsA.
Methods: We performed a retrospective analysis including all the patients with PsA, meeting the CASPAR criteria and under biologic therapy, followed in the Rheumatology department of a tertiary university hospital. Demographic and clinical data, including occurrence of SARS-CoV-2 infection, were collected from our national database (reuma.pt). Disease activity (CDAI, SDAI, DAS28 4v, BASDAI, ASDAS) and bDMARDs responses (EULAR, ASDAS, ASAS, ACR and PsARC responses) were evaluated before and after SARS-Cov-2 infection. Statistical analysis was performed with SPSS. Continuous variables were compared through paired samples t-test.
Results: A total of 102 patients with PsA were included. Fifty-two were females (51%).The mean age was 53 ± 11.09 years and the median disease duration was 15 years [min 2, max 47]. Overall, 54 (53%) patients had predominant axial involvement, 26 (26%) peripheric and 36 (37%) enthesopathic. The most used bDMARD was etanercept (n=28, 27.5%) followed by adalimumab (n=22, 21.6%) and secukinumab (n=18, 17.6%).
The prevalence of SARS-CoV-2 infection was 15.7% (n=16). Sixty-three per cent received the BNT162b2 (Pfizer/BioNtech) vaccine, 31% received mRNA-1273 (Moderna), 13% received AZD1222 (AstraZeneca) and 13% received AD26.COV2.S (Janssen/Johnson & Johnson). Sixty-three percent were infected before any vaccination, 13% after the first dose and 25% after the second. The most common symptoms were anosmia (65%), dysgeusia (56%) and cough (56%). All patients fully recovered from the infection, with no need for hospitalization.
Regardless of the score used, the difference between the mean disease activity after SARS-CoV-2 infection and that at baseline did not reach statistical significance. At baseline and after infection, mean (SD) disease activity parameters were, respectively: CDAI 8.6±5.7 vs 8.6±5.7, p=0.997; SDAI 9.3±6.6 vs 9.2±6.1, p=0,928; DAS 28 4v 2.9±1.2 vs 2.9 ±1.2, p= 0.818; BASDAI 3.6 ±2.6 vs 3.2±2.7, p=0.506; ASDAS 2.2±1.2 vs 2.2±1, p=0.721.
The number of patients unresponsive to bDMARDs (according EULAR, ASDAS, ASAS, ACR and PsARC) before the infection wasn't different from post-infection.
Conclusions: Our study suggests that SARS-CoV2 infection has no negative impact on PsA disease activity and bDMARD responses. However, more studies are still needed to better understand the long-term effects of SARS-CoV2 infection.

REFERENCES

Zhou Q et al. SARS-CoV-2 Infection Induces Psoriatic Arthritis Flares and Enthesis Resident Plasmacytoid Dendritic Cell Type-1 Interferon Inhibition by JAK Antagonism Offer Novel Spondyloarthritis Pathogenesis Insights. Front Immunol. 2021 Apr 15; 12:635018.

020 - CLINICAL MANIFESTATIONS PREDICTED BY ANTISYNTHETASE ANTIBODIES IN PATIENTS WITH INFLAMMATORY MYOPATHIES: WHAT SHOULD WE LOOK FOR?

Matilde Bandeira[1, 2], Eduardo Dourado[1, 2], Ana Teresa Melo[1, 2], Patrícia Martins[1, 2], Vanessa Fraga[3], José Luís Ferraro[4], André Saraiva[5], Marlene Sousa[5], Hugo Parente[6], Catarina Soares[6], Ana Margarida Correia[7], Diogo Esperança Almeida[7], Sara Paiva Dinis [8], Ana Sofia Pinto[8], Filipe Oliveira Pinheiro[9], Maria Seabra Rato[9], Tiago Beirão[10], Beatriz Samões[10], Bernardo Santos[11], Carolina Mazeda[11], Ana Teodósio Chícharo[12], Margarida Faria[13], Agna Neto[13], Maria Helena Lourenço[14, 15], Luisa Brites[16], Marília Rodrigues[16], Joana Silva-Dinis[17], João Madruga Dias[18], Filipe Araújo[19], Nádia Martins[20], Couto M[20], Ana Valido[21], Maria José Santos[2, 3], Sofia C Barreira[1, 2], JE Fonseca[1, 2], Raquel Campanilho-Marques[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]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [4]Rheumatology Department, Hospital Beatriz Ângelo, Lisboa, Portugal, [5]Serviço de Reumatologia, Centro Hospitalar e Universitário de 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, Centro Hospitalar Universitário de São João, Porto, Portugal, [10]Rheumatology Department, Centro Hospitalar Vila Nova de Gaia/Espinho, Gaia, Portugal, Vila Nova de Gaia, Portugal, [11]Serviço de Reumatologia, Centro Hospitalar do Baixo Vouga, E.P.E., Aveiro, Portugal, [12]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal, [13]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [14]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [15]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [16]Serviço de Reumatologia, Hospital Distrital de Leiria, Leiria, Portugal, [17]Rheumatology Department, centro Hospitalar de Lisboa Central, Lisboa, Portugal, [18]Rheumatology Department, Centro Hospitalar Médio Tejo, Torres Novas, Portugal, [19]Serviço de Reumatologia, Hospital CUF Cascais, Cascais, Portugal, [20]Rheumatology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [21]Serviço de Reumatologia, Unidade Local de Saúde do Litoral Alentejano, Santiago do Cacém, Portugal

Background: Idiopathic inflammatory myopathies (IIM) are a group of rare multisystemic disorders. Autoantibody expression significantly impacts the clinical features and prognosis of IIM, influencing the follow-up plan and treatment choices. Antisynthetase antibodies are positive in 11-39% of patients with IIM and are usually associated with antisynthetase syndrome classically characterized by the triad of arthritis, myositis and interstitial lung disease (ILD). This work aims to define the clinical characteristics of IIM patients positive for antisynthetase antibodies, compared to IIM patients negative for these autoantibodies, regardless of IIM subtype.

Methods: Multicentre open cohort study, including patients registered in the IIM module of the Rheumatic Diseases Portuguese Register (Reuma.pt/Myositis) until January 2022. Only patients who had information regarding the results of the myositis-specific (MSA) and associated antibodies (MAA) testing were included. Univariate analysis was performed using chi-square, Fisher's exact, Mann-Whitney or t-test, as appropriate. Independent predictors of different clinical manifestations, adjusted for sex and age at diagnosis, were identified through binomial logistic regression modelling. The linearity of the continuous variables with respect to the logit of the dependent variable was assessed via the Box-Tidwell procedure. Correlated variables, cases with missing information and outliers were excluded from the multivariate analysis in order to fulfil all assumptions necessary to assure the validity of the regression.

Results: From the 280 patients registered at Reuma.pt/Myositis, 237 were included, 71.7% females, with a mean age at diagnosis of 49.27±18.40, out of which 72 (25.7%) were positive for either anti-Jo1 (n=56), anti-PL7 (n=10), anti-PL12 (n=8), anti-EJ (n=4) or anti-OJ (n=4). These patients' most common clinical manifestations were myositis (n=54/70, 77.1%), arthritis (n=20/29, 69.0%), and lung involvement (40/65, 61.5%). Compared to other IIM patients, patients positive for antisynthetase antibodies more frequently had arthritis (20/29 vs 15/61, p<0.001), lung involvement (40/65 vs 36/149, p<0.001), mechanic's hands (12/36 vs 11/88, p=0.010), and heart involvement (7/65 vs 3/148, p=0.010). On the contrary, this group less frequently had Gottron's sign (13/51 vs 55/119, p=0.016) and papules (18/69 vs 66/152, p=0.017), heliotrope rash (16/71 vs 76/170, p<0.001), malar rash (2/36 vs 27/89, p=0.002), erythema (11/49 vs 49/105, p=0.005), photosensitivity (3/36 vs 24/89, p=0.029) and the shawl sign (3/36 vs 22/88, p=0.047). In the multivariate analysis, antisynthetase antibodies (OR 19.34, 95%CI: 2.99-125.04, p=0.002) and mechanic's

Table I.

	Antisynthetase antibodies positive (n=72)	Antisynthetase antibodies negative (n=165)	Univariate analysis
Age at disease onset, median ± SD (N)	50.51±17.20 (55)	46.88±19.04 (146)	p=0.199
Age at diagnosis, median ± SD (N)	52.51±16.41 (56)	47.98±19.04 (140)	p=0.098
Disease duration (in years), median ± SD (N)	7.81±6.51 (55)	6.50±6.69 (146)	p=0.212
Deceased patients, n/N (%)	5/72 (6.9)	5/165 (3.0)	p=0.177
Clinical data			
Musculoskeletal involvement			
Muscle involvement			
Proximal muscle weakness, n/N (%)	43/55 (78.2)	118/138 (85.5)	p=0.283
Myositis, n/N (%)	54/70 (77.1)	131/153 (85.6)	p=0.128
Muscle weakness (not predominantly proximal), n/N (%)	6/34 (17.6)	10/84 (11.9)	p=0.392
Joint involvement			
Arthralgia (without arthritis), n/N (%)	1/9 (11.1)	1/44 (2.3)	p=0.313
Arthritis, n/N (%)	20/29 (69.0)	15/61 (24.6)	**p<0.001***
Skin involvement			
Gottron' sign, n/N (%)	13/51 (25.5)	55/119 (46.2)	**p=0.016***
Heliotrope rash, n/N (%)	16/71 (22.5)	76/160 (47.5)	**p<0.001***
Gottron's papules, n/N (%)	18/69 (26.1)	66/152 (43.4)	**p=0.017***
Erythema, n/N (%)	11/49 (22.4)	49/105 (46.7)	**p=0.005***
Periungual changes, n/N (%)	13/61 (21.3)	39/146 (26.7)	p=0.484
Malar rash, n/N (%)	2/36 (5.6)	27/89 (30.3)	**p=0.002***
Oedema			
Periorbital oedema, n/N (%)	2/34 (5.9)	13/85 (15.3)	p=0.227
Generalized subcutaneous oedema, n/N (%)	1/35 (2.9)	5/85 (5.9)	p=0.670
Photosensitivity, n/N (%)	3/36 (8.3)	24/88 (27.3)	**p=0.029***
Shawl sign, n/N (%)	3/36 (8.3)	22/88 (25.0)	**p=0.047***
Mechanic's hands, n/N (%)	12/36 (33.3)	11/88 (12.5)	**p=0.010***
Cutaneous vasculitis, n/N (%)	6/54 (11.1)	17/115 (14.8)	p=0.634
Calcinosis, n/N (%)	4/65 (6.2)	16/150 (10.7)	p=0.443
Alopecia, n/N (%)	1/36 (2.8)	11/88 (12.5)	p=0.177
Skin ulceration, n/N (%)	5/65 (7.7)	11/149 (7.4)	p=1.000
Panniculitis, n/N (%)	0/35 (0.0)	6/87 (6.9)	p=0.181
Livedo reticularis, n/N (%)	0/35 (0.0)	6/87 (6.9)	p=0.181
Vascular involvement			
Raynaud's phenomenon, n/N (%)	27/66 (40.9)	43/150 (28.7)	p=0.084
Periungual capillary changes, n/N (%)	5/33 (15.2)	17/76 (22.4)	p=0.447
Digital ulcers, n/N (%)	0/29 (0.0)	1/73 (1.4)	p=1.000
Internal organ involvement			
Heart involvement, n/N (%)	7/65 (10.8)	3/148 (2.0)	**p=0.010***
Lung involvement, n/N (%)	40/65 (61.5)	36/149 (24.2)	**p<0.001***
Gastrointestinal involvement, n/N (%)	12/36 (33.3)	40/95 (42.1)	p=0.426
Dysphagia, n/N (%)	7/33 (21.2)	25/82 (30.5)	p=0.365
Dysphonia, n/N (%)	4/32 (12.5)	8/82 (9.8)	p=0.737
Abdominal pain, n/N (%)	2/31 (6.5)	3/81 (3.7)	p=0.616
Oesophageal involvement, n/N (%)	8/64 (12.5)	31/143 (21.7)	p=0.129

hands (OR 9.86, 95%CI: 1.15-84.28, p=0.037) were identified as predictors of ILD in IIM patients independently of sex, age at diagnosis, arthritis, and neoplasia. Additionally, antisynthetase antibodies (OR 12.31, 95%CI: 2.41-62.81, p=0.003) and mechanic's hands (OR 21.41, 95%CI: 3.10-147.74, p=0.002) were also predictors of arthritis in IIM patients independently of sex, age at diagnosis, ILD, and neoplasia. On the other hand, no independent predictors of heart involvement were identified.

Conclusions: Patients positive for antisynthetase antibodies are more likely to have arthritis, mechanic's hands, and heart and lung involvements than other IIM patients. Both antisynthetase antibodies and mechanic's hands were identified as independent predictors of ILD and arthritis in IIM patients.

021 - SYSTEMIC INVOLVEMENT IN SJÖGREN SYNDROME: CAN WE PREDICT IT?

Matilde Bandeira[1, 2], Manuel Silvério-António[1, 2], Nikita Khmelinskii[1, 2], JE Fonseca[1, 2], Vasco C Romão[1, 2]

[1]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Background: Sjögren's Syndrome (SS) is a systemic autoimmune disease targeting mainly exocrine glands. Its hallmark is ocular and oral dryness. Systemic extra-glandular involvement including neurological, renal, vascular or pulmonary manifestations are present in one third of patients. Stratification of SS patients is a growing need and predictors for systemic disease are a useful tool for differentiating patients with exclusive glandular involvement from patients with systemic involvement, which may occur years after disease onset.

Methods: Retrospective cohort study based on Reuma.pt, including SS patients followed in our center until March 2022. Only patients who fulfilled the 2016 ACR/EULAR classification criteria for SS were included. Data from univariate analysis was performed using chi-square, Kruskal-Wallis or ANOVA, as appropriate. Independent predictors of systemic involvement were identified through binomial logistic regression modelling.

Results: 216 patients were included; 96.8% females, with a mean age at diagnosis of 51.32±14.85. The most common clinical manifestations were oral dryness (n=203/216, 94.0%) and ocular dryness (n=198/216, 91.7%). Patients were divided into three groups based on both ESSDAI at diagnosis and ESSDAI during follow-up: group A with no systemic involvement (ESSDAI=0 at diagnosis and throughout follow-up), group B with no systemic involvement at diagnosis that developed throughout follow-up (ESSDAI at diagnosis=0 and ESSDAI throughout the patient's follow-up≥1) and group C with systemic involvement from the beginning (ESSDAI at diagnosis≥1). Disease duration was significantly lower in patients with ESSDAI=0 at diagnosis and throughout follow-up (5.90±5.78 years, n=31), intermediate in the group who had systemic involvement at diagnosis (9.25±7.71 years, n=162), and higher in the group with no systemic involvement at diagnosis that developed throughout follow-up (10.96±9.39 years, n=23). Age at diagnosis was significantly higher in patients with no systemic involvement (58.16±13.90 years, n=31), intermediate in patients who developed systemic involvement during follow-up (52.09±11.51 years, n=23) and lower in patients with systemic involvement at diagnosis (49.90±13.90 years, n=162). Both rheumatoid factor (RF) and salivary gland biopsy were more frequently positive in patients with systemic involvement.

In the multivariate analysis, age at diagnosis (OR 0.956, 95%CI: 0.918-0.996, p=0.032), positive biopsy (OR

Table I. Differences between patients with and without systemic involvement

	Patients with no systemic involvement (ESSDAI=0 at diagnosis and throughout follow-up)	Patients with no systemic involvement at diagnosis that developed throughout follow-up (ESSDAI at diagnosis=0 and ESSDAI throughout the patient's follow-up≥1)	Patients with systemic involvement from the beginning (ESSDAI at diagnosis≥1)	Univariate analysis
Age at diagnosis, median ± SD (N)	**58.16±13.90 (31)**	**52.09±11.51 (23)**	**49.90±13.90 (162)**	**p=0.017**
Disease duration (in years), median ± SD (N)	**5.90±5.78 (31)**	**10.96±9.39 (23)**	**9.25±7.71 (162)**	**p=0.036**
Females, n/N (%)	30/31 (96.8)	22/23 (95.7)	157/162 (96.9)	p=0.950
Deceased patients, n/N (%)	2/31 (6.5)	4/162 (2.5)	2/23 (8.7)	p=0.228
Ocular dryness, n/N (%)	29/31 (93.5)	22/23 (95.7)	147/162 (90.7)	p=0.669
Oral dryness, n/N (%)	30/31 (96.8)	22/23 (95.7)	151/162 (93.2)	p=0.701
Schirmer's test < 5mm in 5 minutes, n/N (%)	19/26 (73.1)	13/19 (68.4)	97/141 (68.8)	p=0.906
Unstimulated salivary flow < 1.5mL in 15 minutes, n/N (%)	5/17 (29.4)	4/12 (33.3)	43/92 (46.7)	p=0.323
Positive salivary biopsy, n/N (%)	**12/27 (44.4)**	**16/20 (80.0)**	**94/132 (71.2)**	**p=0.012**
Positive anti-SSA/Ro, n/N (%)	27/31 (87.1)	21/23 (91.3)	150/162 (92.6)	p=0.597
Positive anti-SSB/La, n/N (%)	16/31 (51.6)	13/22 (59.1)	88/162 (54.3)	p=0.864
Positive antinuclear antibodies, n/N (%)	27/31 (87.1)	20/23 (87.0)	154/162 (95.1)	p=0.133
Positive rheumatoid factor, n/N (%)	**7/28 (25.0)**	**92/160 (57.5)**	**11/21 (52.4)**	**p=0.006**
Hematologic neoplasia, n/N (%)	0/31 (0.0)	10/162 (6.2)	1/23 (4.3)	p=0.353

Abbreviations: ESSDAI – EULAR Sjögren's syndrome disease activity index

3.800, 95%CI: 1.330-10.861, p=0.013) and RF (OR 3.853, 95%CI: 1.336-11.114, p=0.013) were identified as predictors of systemic involvement (ESSDAI≥1 at some point during follow-up) in SS patients independently of sex, disease duration, presence of anti-SSA or anti-SSB and sicca symptoms.

Conclusions: Patients with systemic involvement are more likely to have a lower age at diagnosis, positive salivary gland biopsy and positive RF compared to patients with no systemic disease during the disease course. Age at diagnosis, positive salivary gland biopsy and RF were identified as independent predictors of systemic involvement in SS. Its presence may require a closer follow-up.

023 - GENDER DIFFERENCES IN SPONDYLOARTHRITIS – DO WOMEN START AT A DISADVANTAGE?

Filipe Oliveira Pinheiro[1], Pedro Madureira[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], Beatriz Samões[2], Miguel Bernardes[1, 5], Lúcia Costa[1]
[1]Rheumatology Department, 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]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]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal

Background: Unlike many rheumatic diseases, which show a marked female predominance, spondyloarthritis are diagnosed more frequently in men, in a 3:1 ratio. This fact may help to explain the lesser recognition of this pathology in females and the greater lack of knowledge regarding the evolution and prognosis of the disease, when compared to males. The aim of this study is to determine whether gender influences diagnosis, treatment, and disease activity measures in spondyloarthritis.

Materials and Methods: Retrospective study that evaluated patients diagnosed with spondyloarthritis, registered on the Portuguese national database (Reuma.pt) platform, evaluated at the start of their first bDMARD, and at 12 and 24 months. Demographic, clinical and laboratory data were evaluated. Comparison between groups (male vs female) were performed using chi-square (categorical variables), t-test (continuous variables with normal distribution) and Mann-Whitney (continuous variables with non-normal distribution). Differences between assessment measurements taken at 12 and 24 months relative to baseline were represented as delta.

Results: A total of 273 patients were included in this

Table I.

Variables at 24 months	Female	Male	p
TJC – mean (SD)	2,4 (4,5)	0,4 (0,7)	<0,001
SJC – mean (SD)	0,4 (1,1)	0,0 (0,2)	<0,001
ESR – mean (SD)	23,7 (15,1)	12,3 (22,2)	0,002
CRP – mean (SD)	0,70 (0,68)	0,42 (0,76)	0,485
Patient VAS – mean (SD)	40,6 (29,9)	33,8 (26,1)	0,037
Physician VAS – mean (SD)	17,9 (18,3)	13,2 (11,2)	0,272
Pain VAS – mean (SD)	38,6 (26,7)	27,2 (25,9)	0,009
BASDAI – mean (SD)	4,1 (2,3)	2,6 (1,6)	0,001
ΔBASDAI – mean (SD)	-3,2 (2,3)	-3,4 (2,1)	0,358
BASDAI50 – n (%)	32 (64,0)	55 (74,3)	0,218
BASMI – mean (SD)	3,9 (1,6)	3,5 (1,9)	0,785
BASFI – mean (SD)	4,4 (2,7)	3,0 (2,4)	0,021
ASDAS CRP – mean (SD)	2,3 (0,9)	1,9 (0,7)	0,002
ΔASDAS CRP – mean (SD)	-1,7 (1,1)	-2,2 (1,1)	0,023
MASES – median (IQR)	1 (4)	0 (0)	<0,001
ΔMASES – median (IQR)	0 (3)	0 (2)	0,664

study, 123 of which (45.1%) were female. Females were diagnosed, on average, 4.2 years later than males (35.7±11.6 vs 31.5±11.5, p=0.003), although no statistically significant difference was found between age at the start of first bDMARD (females 43.3±12.4 versus males 40.9 ±12.2 years, p=0.112), the time to first bDMARD (females 15.2±12.2 versus males 15.5 ±10.3 years, p=0.809), and the choice of bDMARD. There was a higher proportion of HLA-B27 in male patients (84.5% vs 62.8%, p=0.001), and males had higher C-reactive protein levels (CRP, 3.18±3.45 vs 1.56±1.67, p=0,001) and BASMI (4.8±1.5 vs 3.9±1.6, p=0,001); however, female patients had higher scores in pain visual analog scale (67.2±22.7 vs 59.3±25.2, p=0.005), BASDAI (7.1±1.4 vs 5.9±1.6, p=0,001), BASFI (7.0±2.0 vs 6.0±2.2, p=0,001) and MASES (4±6 vs 1±3, p<0,001) at the start of first bDMARD. No differences were observed in erythrocyte sedimentation rate (ESR), physician and patient visual analog scale, and ASDAS. Twelve months after the start of bDMARD therapy, females have higher ESR (22.2±15.0 vs 8.3±9.4, p=0,001), physician (20.3±20.9 vs 13.7±14.3, p=0,005) and pain visual analog scale (45.5±27.0 vs 33.5±22.8, p=0,020), BASDAI (4.3±2.2 vs 2.8±1.8, p=<0,001), ASDAS-CRP (2.6±1.0 vs 2.0±0.9, p=0,001), MASES (1±4 vs 0±0, p=<0,001), lower proportion of BASDAI50 responses (55.4 vs 71.1, p=0,033) and lower variations in ASDAS-CRP (1.4±1.1 vs 2.1±1.3, p=0,001). Twenty-four months after starting bDMARD, females still have higher ESR (23.7±15.1 vs 12.3±22.2, p=0,002), patient (40.6±29.9 vs 33.8±26.1, p=0,037) and pain visual analog scale (38.6±26.7 vs 27.2±25.9, p=0,009), BASDAI (4.1±2.3 vs 2.6±1.6, p=0,001), BASFI (4.4±2.7 vs 3.0±2.4, p=0.021), ASDAS-CRP (2.3±0.9 vs 1.9±0.7, p=0,002), MASES (1±4 vs 0±0, p=<0,001), and lower variations in ASDAS-CRP (1.7±1.1 vs 2.2±1.1, p=0,023).

Conclusion: In this study, the authors report higher disease activity measures and indices in female patients with spondyloarthritis; in fact, female patients are diagnosed at a later stage and show higher disease activity since the beginning of the disease and treatment, which extended to the next 2 years. Whether or not this worsening is related only to diagnostic delay is not known. The authors reinforce the need for an early diagnosis in women with spondylarthritis, since delayed diagnosis can have devastating consequences.

025 - THE BATTLE OF THE SEXES IN RHEUMATOLOGY – A REAL-WORLD STUDY COMPARING GENDER DIFFERENCES IN PSORIATIC ARTHRITIS

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

[1]Rheumatology Department, Centro Hospitalar Universitário 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 Vila Nova de Gaia/Espinho, Gaia, Portugal, Vila Nova de Gaia, Portugal, [5]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal

Background: Psoriatic arthritis is an inflammatory disease that affects 20 to 30% of patients with psoriasis. It occurs equally in women and men, unlike axial spondyloarthritis, which shows a clear male predominance. It has been reported that females show an overall worse prognosis, with higher rates of treatment failure, and more fatigue and disability than men. The aim of this study is to determine whether gender influences diagnosis, treatment, and disease activity measures in psoriatic arthritis.

Materials and Methods: Retrospective study that evaluated patients diagnosed with psoriatic arthritis, registered on the Portuguese national database (Reuma.pt) platform, evaluated at the start of their first bDMARD, and 12 months after. Demographic, clinical and laboratory data were evaluated. Comparison between groups (male vs female) were performed using chi-square (categorical variables), t-test (continuous variables with normal distribution) and Mann-Whitney U (continuous variables with non-normal distribution). Differences between assessment measurements taken at 12 months relative to baseline were represented as delta.

Results: A total of 81 patients were included in this study, 41 of which (50.6%) were female. There were no differences regarding age of diagnosis, age at the start of first bDMARD, years of disease at the start of first bDMARD, and exposure to glucocorticoids and glucocorticoid dosage at the start of first bDMARD between genders. At baseline, females showed a higher disease activity assessed by different measures, such as CDAI (22.5±11.3 vs 19.0±8.9, p=0.022), SDAI (24.7±11.3 vs 19.9±9.1, p=0.022), DAS4V-CRP (4.4±1.2 vs 3.6±1.2, p=0.004), BASDAI (7.4±1.5 vs 6.7±1.9, p=0.002), MASES (1±5 vs 0±1, p=0.027), and DAPSA (37.3±14.6 vs 29.5±15.2, p=0.046), with a borderline tendency to higher ASDAS (3.9±11.2 vs 3.5±0.7, p=0.051). No differences were observed regarding erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), physician/pain/patient visual analog scale, HAQ, BASMI and BASFI scores. Twelve months after starting bDMARD, female patients had a higher CDAI (8.6±8.1 vs 8.2±6.6, p=0.010), SDAI (9.1±8.0 vs 8.4±6.5, p=0.006), DAS4V-

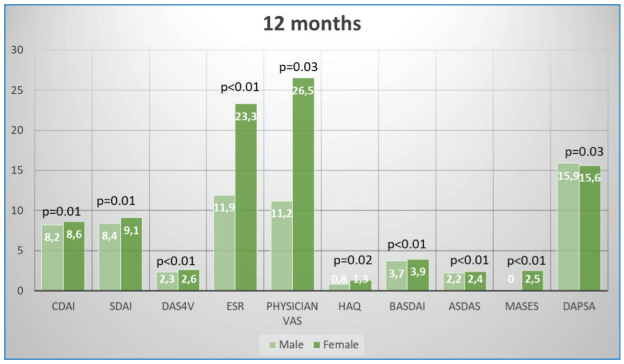


Figure 1.

CRP (2.6±1.1 vs 2.3±1.0, p=0.004), ESR (23.3±22.4 vs 11.9±11.2, p<0.001), physician visual analog scale (26.5±28.2 vs 11.2 vs 9.6, p=0.034), HAQ (1.25±0.58 vs 0.79±0.93, p=0.023), BASDAI (3.9±2.5 vs 3.7±2.3, p=0.002), ASDAS (2.4±1.0 vs 2.2±0.7, p=0.002), MASES (2.5±5 vs 0±1, p=0.<001); interestingly, males showed higher DAPSA scores (15.9±11.1 vs 15.6±12.6, p=0.027). No differences were observed regarding BASDAI50, ASAS20/40/70, ACR20/50/70 and PsARC responses. Females had a higher rate of treatment switch, with a median switch 0.5±1 versus 0±0 observed in males (p=0.017).

Conclusion: In this cohort, there is a marked gender difference in psoriatic arthritis, with women exhibiting higher disease activity, disability, and treatment failure when compared to men. This study highlights the potential role of gender in several outcomes in psoriatic arthritis.

026 - DOSAGE AND EFFICACY OF METHOTREXATE IN PATIENTS WITH RHEUMATOID ARTHRITIS ON bDMARD

Filipe Oliveira Pinheiro[1], Pedro Madureira[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], Beatriz Samões[2], Miguel Bernardes[1, 5], Lúcia Costa[1]
[1]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Rheumatology Department, Centro Hospitalar Vila Nova de Gaia/Espinho, Gaia, Portugal, Vila Nova de Gaia, 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]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal

Background: In rheumatoid arthritis, methotrexate is considered the anchor drug, both in monotherapy and in combination therapy. Several studies have shown an association between an effective response and the dose of methotrexate used, especially when used as monotherapy. The aim of this study was to assess whether the methotrexate dose influences the therapeutic response in patients with rheumatoid arthritis under bDMARD.

Methods: Retrospective study that evaluated patients diagnosed with rheumatoid arthritis, registered in the national database (Reuma.pt), who started bDMARD therapy after 2015 and with at least one year of follow-up. Demographic, clinical and laboratory data were collected. Correlation analyzes were performed (continuous variables and methotrexate dose) and comparison between groups (methotrexate dose ≤15mg/week vs >15mg/week) using chi-square (categorical variables), t-test (continuous variables with normal distribution) and Mann-Whitney U (continuous variables with non-normal distribution). Differences between assessments at 12 months from baseline were represented as delta (Δ).

Results: A hundred and seventy six patients were included, of which 149 (84.7%) were female, aged 53.3±7.6 years. Forty-nine (27.8%) started therapy with etanercept, 41 (23.3%) rituximab, 31 (17.6%) tocilizumab, 26 (14.8%) adalimumab, 11 (14.8%) golimumab, 8 (4.5%) infliximab, 6 (3.4%), abatacept, 3 (1.7%) certolizumab and 1 (0.6%) anakinra; one hundred and thirty-nine (79.0%) were under corticosteroid therapy, 86 (48.9%) were under methotrexate (MTX), with a mean dose of 15.0±4.5 mg/week, and 42 of the patients under methotrexate were taking doses greater than 15mg/week (48.3%). Upon initiation of bDMARD, there was no statistically significant difference between the MTX ≤15mg/week and MTX>15mg/week groups in the variables age, corticosteroid dose, and erythrocyte sedimentation rate, but the MTX>15 group contained elements with higher C-reactive protein (1.4±1.3 vs 0.6±0.9, p=0.040), DAS28 4V-CRP (4.2±1.6 vs 3.0±1.4, p<0.01), CDAI (21.0±14.2 vs 11.5±11.3, p<0.01), and SDAI (24.0±19.4 vs 12.1±11.7, p<0.01). At 6 months, there were no statistically significant differences in these variables between the 2 groups. When evaluated at 12 months, a significantly higher ΔDAS was observed in the MTX>15mg/week group (1.7±1.5 vs 0.7±1.6, p<0.01), with no differences in DAS, CDAI, SDAI, C-reactive protein, erythrocyte sedimentation rate, and ACR20, ACR50 and ACR70 responses. A statistically significant correlation was identified with ΔDAS and methotrexate dose at 6 and 12 months (ρ 0.321, p=0.01; ρ 0.445,p<0.01).

Conclusion: In this work, the authors point out that higher doses of methotrexate were associated with greater DAS variation in the first 6 and 12 months after starting treatment with bDMARD; however, differenc-

Table I.

Results – 12 months	MTX ≤15	MTX >15	p
DAS284V – mean (SD)	2,7 (0,9)	2,7 (0,8)	0,917
ΔDAS284V – mean (SD)	0,7 (1,6)	1,7 (1,5)	0,007
CDAI – mean (SD)	8,4 (6,5)	7,7 (4,7)	0,580
SDAI – mean (SD)	9,1 (7,1)	8,3 (4,9)	0,506
ESR, mm/1ªh – mean (SD)	18,6 (17,5)	18,5 (14,4)	0,508
CRP, mg/L – mean (SD)	0,7 (1,4)	0,6 (0,7)	0,519
DAS ≤3,2 – n (%)	29 (65,9)	30 (71,4)	0,223

es in activity scores and response to therapy were not observed with the use of higher doses of methotrexate. Although safety was not evaluated in this work, the authors emphasize these findings, as they demonstrate that lower doses of methotrexate can be equally effective in patients starting bDMARD therapy, which needs to be confirmed in prospective studies using different methodologies.

027 - SMOKING AND TREATMENT RESPONSE IN PATIENTS WITH RHEUMATOID ARTHRITIS ON BDMARD THERAPY

Filipe Oliveira Pinheiro[1], Pedro Madureira[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], Beatriz Samões[2], Miguel Bernardes[1, 5], Lúcia Costa[1]
[1]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Rheumatology Department, Centro Hospitalar Vila Nova de Gaia/Espinho, Gaia, Portugal, Vila Nova de Gaia, 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]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal

Background: Cigarette smoking is associated with the development of rheumatoid arthritis and ACPA-positive rheumatoid arthritis, higher disease activity, and reduced treatment efficacy. Although smoking cessation reduces the risk of developing rheumatoid arthritis, its relationship to the effectiveness of treatment is more controversial. The aim of this study was to evaluate the response to treatment in active and former smokers, comparing it to patients with rheumatoid arthritis who never smoked.
Methods: Retrospective study that evaluated patients diagnosed with rheumatoid arthritis, followed in our department, and registered in the Portuguese national database (Reuma.pt), who started bDMARD therapy after 2015. Demographic, clinical and laboratory data were collected. Patients were divided into 3 groups: non-smokers, former smokers, and active smokers. The comparison between the 3 groups was performed using chi-square (categorical variables), ANOVA test and Krukal-Wallis test. Differences between assessments at 12 months from baseline were represented as delta (**Δ**).
Results: A total of 176 patients were included, of which 149 (84.7%) were female, aged 53.3±7.6 years. One hundred and twenty (68.2%) had never smoked, 28 (15.9%) were former smokers and 28 (15.9%) were active smokers. Upon initiation of bDMARD, there were no statistically significant differences regarding age, positivity for rheumatoid factor and ACPA, DAS, CDAI, SDAI, erythrocyte sedimentation rate and C-re-

Table I.

Variables – 12 months	Non-smokers	Former smokers	Active smokers	p
DAS284V – mean (SD)	2,8 (1,0)	2,7 (0,9)	2,6 (0,8)	0,486
ΔDAS284V – mean (SD)	1,1 (1,5)	1,4 (1,6)	1,8 (1,4)	0,064
CDAI – median (IQR)	7,9 (6,5)	7,5 (5,6)	7,2 (5,2)	0,343
SDAI – median (IQR)	8,6 (7,1)	8,1 (8,1)	7,3 (7,6)	0,321
ESR, mm/1ªh – mean (SD)	16,0 (19)	8 (34)	13,5 (18)	0,202
CRP, mg/L – mean (SD)	0,3 (0,6)	0,2 (0,8)	0,2 (0,5)	0,703
DAS remission – n (%)	57 (47,5)	13 (46,4)	13 (46,4)	0,753
DAS LDA – n (%)	82 (68,3)	19 (67,9)	21 (75,0)	0,729
ACR20 – n (%)	102 (79,7)	28 (100)	28 (100)	0,305
ACR50 – n (%)	60 (50,0)	14 (50,0)	12 (42,9)	0,339
ACR70 – n (%)	18 (15,0)	7 (25,0)	4 (14,3)	0,671

active protein. One year after starting bDMARD, there was a trend towards lower disease activity in the groups of active smokers and former smokers, compared to non-smokers, assessed using DAS (active smokers 2.6±0.8 vs former smokers 2.7± 0.9 vs non-smokers 2.8±1.0, p=0.486) and ΔDAS (active smokers 1.8±1.4 vs former smokers 1.4±1.6 vs non-smokers 1.1±1.5, p=0.064). There were no differences in ACR responses and inflammatory markers between the 3 groups.

Conclusion: In this study, active smokers and former smokers did not show higher disease activity compared to non-smokers after starting bDMARD; interestingly, there was a trend towards a better response to therapy in patients already exposed to tobacco. Despite the multiple benefits of smoking cessation on the individual's overall health, these findings demonstrate that tobacco exposure may not be as detrimental to the therapeutic response to bDMARD as other publications reported. These results will need to be confirmed in prospective and randomized studies.

REFERENCES

Deane KD, Demoruelle MK, Kelmenson LB, Kuhn KA, Norris JM, Holers VM. Genetic and environmental risk factors for rheumatoid arthritis. Best Pract Res Clin Rheumatol. 2017 Feb;31(1):3-18. doi: 10.1016/j.berh.2017.08.003. Epub 2017 Sep 18. PMID: 29221595; PMCID: PMC5726551.

028 - ASSESSMENT OF THE SWOLLEN TO TENDER JOINT COUNT RATIO AS A PREDICTOR OF RESPONSE IN RHEUMATOID ARTHRITIS PATIENTS: A COHORT STUDY

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], Beatriz Samões[2], Rafaela Nicolau[5], Miguel Bernardes[1, 6], Lúcia Costa[1]

[1]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Rheumatology Department, Centro Hospitalar Vila Nova de Gaia/Espinho, Gaia, Portugal, Vila Nova de Gaia, 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]Rheumatology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [6]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal

Background: Several response predictors have already been studied in rheumatoid arthritis, especially since the introduction of biological therapies. The swollen to tender joint count ratio (STR) has been proposed as a predictor of response in patients receiving anti-TNF therapy, but its usefulness in other therapies is not yet well established.

Methods: Longitudinal and retrospective study that included patients diagnosed with rheumatoid arthritis followed in our department under therapy with b/tsDMARD. Demographic, laboratory, and clinical data were collected and evaluated at 0, 6 and 12 months after starting the first b/tsDMARD therapy performed after 2015. The variation in each parameter compared to baseline was calculated at 6 and 12 months and represented as a delta. A cutoff of 1 was defined for comparison between STR groups. The correlations between the continuous variables were assessed by Pearson's test and comparison between groups of ratios using t test (continuous variables) and chi-square test (categorical variables). Multiple linear regression and multivariate logistic regression were performed to determine response predictors.

Results: A total of 287 patients were included, 238 (82.9%) females, aged 55.7±10.8 years and diagnosed with rheumatoid arthritis for 11.2±8.1 years. Two hundred and sixty-nine (93.7%) were on csDMARD; with regard to b/tsDMARD therapy, 66 started etanercept (23.0%), 62 tocilizumab (21.6%), 58 rituximab (20.2%), 44 adalimumab (15.3%), 17 golimumab (5.9%), 14 abatacept (4.9%), 7 certolizumab (2.4%), 5 upadacitinib and baricitinib (1.7%), 4 infliximab (1.4%), 3 tofacitinib (1.0%), and 2 anakinra (0.7%). At the start of therapy with b/tsDMARD, the mean DAS28 4V was 4.7±1.5, CDAI 20.4±12.2, SDAI 22.8±16.4, erythrocyte sedimentation rate 31.8±24.4, C-reactive protein (CRP) 1.5±1.7, patient VAS 62.6±1.5, physician VAS 41.4±29.7, pain VAS 62.5±24.8, and HAQ 1.5±0.6; median tender joint count was 4 (interquartile range - IQR - 6), swollen joint count was 3 (IQR 6), and STR joint count was 0.9 (IQR 0.5). When the STR < 1 and STR ≥1 groups were compared, it was found that there were no differences in these variables when starting b/tsDMARD, nor in the therapies they performed. At 6 months, the STR ≥1 group showed a higher proportion of patients in CDAI remission (CDAI ≤2.8 – 15.3% vs 6.9%, p=0.033) and in DAS28 4V remission or low disease activity according to DAS28 4V (DAS28 4V ≤3.2 – 36.5% vs 22.4%, p=0.008). At 12 months, the STR ≥1 group exhibited less disease activity (mean DAS28 4V 3.2±1.2 vs 3.6±1.3, p=0.028) and a higher proportion of patients in DAS28 4V remission (25.2% vs 19.7%, p=0.047). In the correlation studies, a weak correlation was identified between STR and CRP at 12 months (r=0,28, p<0,001). In multiple linear regression studies, when adjusted for sex, age, prednisolone, csDMARD, inflammatory parameters, b/tsDMARD, STR was not shown to be a predictor of DAS28 4V disease activity at 1 year; in multivariate logistic regression

Table I.

Results	
Female gender – n (%)	238 (82.9)
Age – years, mean (SD)	55.7 (10.8)
Disease duration – years, mean (SD)	11.2 (8.1)
csDMARD – n (%)	269 (93.7)
bDMARD – n (%)	
Etanercept	66 (23.0)
Tocilizumab	62 (21.6)
Rituximab	58 (20.2)
Adalimumab	44 (15.3)
Golimumab	17 (5.9)
Abatacept	14 (4.9)
Certolizumab	7 (2.4)
Upadacitinib	5 (1.7)
Baricitinib	5 (1.7)
Infliximab	4 (1.4)
Tofacitinib	3 (1.0)
Anakinra	2 (0.7)

studies, when adjusted for the same variables previously described, STR was not shown to be a predictor of remission or remission or low disease activity according to DAS28 4V at 1 year.

Conclusion: The STR is a practical, easy-to-use index that can be used as an adjunct in clinical practice in the evaluation of patients with rheumatoid arthritis, as it is associated with a better response to b/tsDMARD therapy, regardless of patient status and therapy.

029 - SWITCHING FROM BIO-ORIGINATOR TO BIOSIMILAR DMARDS IN RHEUMATOID ARTHRITIS - THE EXPERIENCE OF ONE CENTER

Filipe Oliveira Pinheiro[1], Pedro Madureira[1], Maria Seabra Rato[1], Diogo Guimarães da Fonseca[2], Ana Martins[1], Daniela Santos Oliveira[1, 3], Beatriz Samões[2], Frederico Rajão Martins[4], Rafaela Nicolau[5], Miguel Bernardes[1, 6], Lúcia Costa[1]

[1]Rheumatology Department, 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]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]Rheumatology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [6]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal

Background: Biological DMARDs are an essential class of drugs in the treatment of inflammatory rheumatic diseases, such as rheumatoid arthritis. The more recent development of biosimilars (bsDMARD), agents highly comparable to bio-originators (boDMARDs), has made access to these therapies more widespread, with no apparent compromise in efficacy. However, there are some conflicting data regarding disease activity and response to therapy after switching from a boDMARD to its bsDMARD. The aim of this study is to assess whether switching from boDMARD to its bsDMARD affects disease activity and response to therapy in rheumatoid arthritis in a monocentric cohort.

Methods: Longitudinal and retrospective study that included patients diagnosed with rheumatoid arthritis followed in the Rheumatology Department of a tertiary hospital, who underwent therapy change from boDMARD to bsDMARD.

Demographic, laboratory, and clinical data were collected, including Visual Analog Scale (VAS), DAS28 4V, and HAQ. All patients were evaluated at 0 (baseline) and 12 months after switching to bsDMARD. The variation in each parameter compared to baseline was calculated at 12 months and represented as a delta. The differences between 0 and 12 months were assessed by paired-samples t-test (continuous variables) and chi-square test (categorical variables). Kruskal-Wallis test was used to evaluate the differences between deltas in the bsDMARD groups.

Results: A total of 80 patients were included, 58 (72.5%) females, aged 59.4±10.6 years and diagnosed with rheumatoid arthritis for 25.0±8.5 years. Regarding bDMARD therapy switch, 32 were on rituximab (40.0%), 28 on etanercept (35.0%), 13 on adalimumab (16.3%) and 7 infliximab (8.8%). At the baseline switch from boDMARD to bsDMARD, the mean DAS28 4V was 2.9±0.8, erythrocyte sedimentation rate (ESR) 21.5±17.0, C-reactive protein (CRP) 0.5±0.6, patient VAS 36.4±22.5, physician VAS 9.7±16.9, pain VAS 41.4±42.8, and HAQ 1.1±0.6; 30 patients (37.5%) were on DAS28 4V remission and 55 (68.8%) on remission or low disease activity.

At 12 months after switch, there was a statistically significant difference relative to baseline in patient VAS (42.7±23.3, p=0.024), physician VAS (13.4±15.8, p=0.009), DAS28 4V remission (33.8%, p=0.004) and remission or low disease activity (61.3%, p<0.001). Etanercept bsDMARD at 12 months was associated

with higher CRP (0.9±1.7 vs 0.2±0.2, p=0.044), patient VAS (38.8±19.1 vs 31.5±18.6, p=0.034), physician VAS (15.2±16.8 vs 5.8±5.6, p=0.007), and lower proportion of DAS28 4V remission or low disease activity (60.7% vs 75.0%, p=0.007), when comparing to baseline. Rituximab bsDMARD at 12 months was associated with higher physician VAS (16.8±17.2 vs 8.6±15.0, p=0.008), and lower proportion of DAS28 4V remission or low disease activity (56.3% vs 59.4%, p=0.002), when comparing to baseline. Adalimumab bsDMARD at 12 months was associated with higher patient VAS (39.9±34.2 vs 28.3±26.1, p=0.045) relative to baseline. No differences were observed with infliximab. There were no differences between deltas (DAS28 4V, ESR, CRP, VAS, HAQ) in the various bsDMARD groups.

Conclusion: In this cohort, we found that after switching from boDMARD to its bsDMARD, there was an overall worsening in some, but not all, variables of disease activity, with a lower proportion of patients achieving remission or low disease activity, especially in patients receiving rituximab and etanercept. These results must be confirmed in populations with a larger sample size.

Table. MMF vs AZA at 24 months

	MMF	AZA	p
ESR, mm/1st hour – median (IQR)	13.0 (18)	27.0 (36)	0.048
CRP, mg/L – median (IQR)	2.30 (4.3)	3.20 (5.4)	0.507
Creatinine, mg/dl – mean, SD	0.80 (0.25)	0.84 (0.27)	0.259
C3, mg/dl – median (IQR)	107.0 (35.3)	112.0 (35.0)	0.774
C4, mg/dl – median (IQR)	21 (15.5)	21 (16)	0.250
dsDNA, IU/ml – median (IQR)	74.1 (98.7)	56.6 (165.3)	0.302
Proteinuria, mg/24h – median (IQR)	180.0 (448.5)	135.7 (113.7)	0.886
Hematuria – n (%)	5 (13.5)	1 (5.9)	0.652
Leukocyturia – n (%)	9 (24.3)	4 (23.5)	0.617

030 - MAINTENANCE THERAPY IN LUPUS NEPHRITIS - IS THERE A DIFFERENCE BETWEEN MYCOPHENOLATE MOFETIL AND AZATHIOPRINE

Filipe Oliveira Pinheiro[1], Pedro Madureira[1], Maria Seabra Rato[1], Lúcia Costa[1]

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

Background: Lupus nephritis is one of the most relevant causes of morbidity and mortality in patients with systemic lupus erythematosus. It is divided into distinct histological classes, and in classes III-V immunosuppressive therapy is indicated. After achieving partial or complete responses with induction therapy, patients are switched to maintenance therapy, to prevent relapses. The aim of this work is to evaluate the efficacy of azathioprine (AZA) and mycophenolate mofetil (MMF) regimens in the treatment of lupus nephritis (LN) at our center.

Methods: Retrospective longitudinal study that included patients diagnosed with LN classes III-V followed in a tertiary center. Demographic, laboratory, and clinical data were collected and evaluated at 0, 24 and 48 months after switch from induction to maintenance therapy. Comparison between groups (AZA vs MMF) was performed using the chi-square test (categorical variables), t-test (continuous variables with normal distribution) and Mann-Whitney U test (continuous variables with non-normal distribution).

Results: Fifty-four patients were included, 44 (81.5%) females, aged 31.0±11.6 years at the time of diagnosis of LN; histologically, 39 patients (72.2%) had class IV, 8 (14.9%) class V and 7 (13.0%) class III. Twenty-six patients (48.1%) underwent induction treatment with cyclophosphamide and 28 (51.9%) with MMF; thirty-seven (68.5%) started maintenance with MMF and 17 (31.5%) with AZA. Patients had a median ESR of 20.0 mm/1st hour (IQR 25.0), median CRP was 2.2 mg/L (IQR 4.7), mean creatinine was 0.97±0.22 mg/dl, median C3 100.0 mg/dl (IQR 30.0), median C4 21.0 mg/dl (IQR 11), median dsDNA 30.2 IU/ml (IQR 137.4) and median proteinuria 269.7 mg/24h (IQR 564.9). Fourteen patients (25.9%) had erythrocyturia and 13 (24.1%) had leukocyturia.

At baseline evaluation, there were no differences between both MMF and AZA groups regarding the studied variables: ESR MMF 17.5 (IQR 18) vs AZA 24 (IQR 30), p=0.255; CRP MMF 1.75 (IQR 5.2) vs AZA 2.60 (IQR 4.6), p=0.616; plasma creatinine MMF 0.83±0.23 vs AZA 0.84±0.21, p=0.302; C3 MMF 98.5 (IQR 23.5) vs AZA 118.0 (IQR 34.3), p=0.064; C4 MMF 21 (IQR 13) vs 20 (IQR 4), p=0.678; dsDNA MMF 39.8 (IQR 144.8) vs AZA 27.7 (IQR 132.3), p=0.421; proteinuria MMF 288.7 (IQR 708.4) vs AZA 169.5 (IQR 410.4), p=0.423; hematuria MMF 24.3% vs AZA 29.4%, p=0.745; leukocyturia MMF 29.7% vs AZA 11.8%, p=0.189.

At 24 months, patients on AZA therapy had a higher ESR – 27 (IQR 30) vs 13 (IQR 18), p= 0.048 –, with no other statistically significant differences observed between the two groups.

At 48 months, no statistically significant differences

were seen: ESR MMF 12 (IQR 16) vs AZA 14 (IQR 16), p=0.356; CRP MMF 1.50 (IQR 3.6) vs AZA 1.30 (IQR 5.7), p=0.915; plasma creatinine MMF 0.84±0.29 vs AZA 0.79±0.22, p=0.737; C3 MMF 100.0 (IQR 36.3) vs AZA 100.0 (IQR 34.3), p=0.762; C4 MMF 22 (IQR 13) vs 20 (IQR 12), p=0.423; dsDNA MMF 52.0 (IQR 108.9) vs AZA 26.4 (IQR 342.9), p=0.468; proteinuria MMF 156.7 (IQR 189.3) vs AZA 83.7 (IQR 117.5), p=0.306. Three patients on MMF had relapse, while there were no patients on AZA with relapse. There were no differences between the percentage of patients with proteinuria below 500mg (81.1% with MMF vs 94.1% AZA) and below 1000mg (97.3% MMF vs 100% AZA) at 48 months.

Conclusion: In this real-life study, limited by the small sample size, namely in the number of patients on AZA, it was found that there were no statistically significant differences in the two maintenance therapeutic strategies used in LN.

031 - MYCOPHENOLATE MOFETIL VERSUS CYCLOPHOSPHAMIDE IN LUPUS NEPHRITIS - A COMPARISON STUDY WITH REAL-WORLD DATA

Filipe Oliveira Pinheiro[1], Pedro Madureira[1], Maria Seabra Rato[1], Lúcia Costa[1]

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

Background: Lupus nephritis is one of the most relevant causes of morbidity and mortality in patients with systemic lupus erythematosus. It is divided into distinct histological classes, and in classes III-V immunosuppressive therapy is indicated, usually with cyclophosphamide (CYC) and mycophenolate mofetil (MMF). The aim of this work is to evaluate the efficacy of CYC and MMF regimens in the treatment of lupus nephritis at our center.

Methods: Retrospective longitudinal study that included patients diagnosed with lupus nephritis classes III-V followed in a tertiary center. Demographic, laboratory, and clinical data were collected, including erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), C3, C4, proteinuria, erythrocyturia and leukocyturia; these variables were evaluated at 0, 6 and 12 months after the diagnosis of lupus nephritis. Comparison between groups (CYC vs MMF) was performed using the chi-square test (categorical variables), t-test (continuous variables with normal distribution) and Mann-Whitney U (continuous variables with non-normal distribution).

Results: Sixty patients were included, 49 (82.9%) females, aged 33.8±12.7 years at the time of diagnosis of lupus nephritis, 44 (73.3%) with class IV, 8 (13.3%) class V and 8 (13.3%) class III. Thirty patients underwent induction therapy with CYC and 30 with MMF. As for laboratory data, patients had a mean ESR of 59.2±32.0 mm/1st hour, median CRP was 10.1 mg/L (IQR 17.2), median creatinine was 0.97 mg/dl (IQR 0.51), median C3 69.4 mg/L dl (IQR 50.3), median C4 14.5 mg/dl (IQR 13), median dsDNA 246.1 IU/ml (IQR 576.9) and median proteinuria 3157.3 mg/24h (IQR 1890.4). Thirty-two patients (53.3%) had erythrocyturia and 37 (61.7%) had leukocyturia.

Patients who started CYC had higher mean ESR (62.0±34.6 vs 39.1±25.6, p=0.005), higher creatinine - 0.92 (IQR 0.48) vs 0.74 (IQR 0.36), p=0.030 -, lower C3 - 63.7 (IQR 47.5) vs 79.5 (IQR 52.7), p=0.021 -, and higher proportion of hematuria (66.7% vs 40.0%, p=0.038) and leukocyturia (76.7% vs 46.7%, p=0.017), with no other statistically significant differences.

At 6 months, patients on CYC therapy maintained a higher mean ESR (36.8±27.7 vs 20.1±12.4, p=0.021). No other differences found at baseline were identified at 6 months.

At 12 months, patients on CYC therapy still exhibited a higher mean ESR (33.1±28.1 vs 17.2±13.8, p=0.026). Patients under CYC tended to have higher levels of C3 - 102.0 (IQR 33.0) vs 93.0 (IQR 24.0) - but with an increase in levels from baseline of 58.4% vs 14.5% with MMF, a statistically significant difference (p= 0.012); there was a trend towards higher proteinuria with CYC patients - 348.4 (IQR 828.6) vs 288.7 (IQR 494.4) -, and the percentage of patients with proteinuria below 500mg (66.7% with CYC vs 70% MMF) and below 1000mg (80% CYC vs 86.7% MMF), as well as the percentage reduction in proteinuria from baseline (90.7% CYC vs 88.6% MMF), was not significantly different between the groups.

Conclusion: In this study, the 12-month prognosis of lupus nephritis was quite favorable, with both induction treatments showing comparable efficacy. CYC was chosen in patients with higher ESR, creatinine, active urinary sediment, and with lower levels of C3, but the absolute results and the percentage of change in laboratory parameters were not significantly different from that observed with MMF, except for C3.

032 - THE RELATIONSHIP BETWEEN DYSLIPIDEMIA AND LUPUS NEPHRITIS - RESULTS:FROM A MONOCENTRIC COHORT

Filipe Oliveira Pinheiro[1], Pedro Madureira[1], Maria Seabra Rato[1], Lúcia Costa[1]

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

Background: Cardiovascular risk factors are important predictors of morbidity and mortality in patients with

systemic lupus erythematosus (SLE). Multiple studies have shown that the prevalence of dyslipidemia is higher in patients with SLE, but its impact on kidney disease is not as well established as other risk factors, such as hypertension and diabetes mellitus.

Methods: Retrospective longitudinal study that included patients diagnosed with lupus nephritis followed in a tertiary center. Demographic, laboratory, and clinical data were collected, including presence of dyslipidemia, lupus nephritis' class and therapies performed, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), C3, C4, proteinuria, erythrocyturia and leukocyturia; these variables were evaluated at 0, 12, and 60 months after the diagnosis of lupus nephritis. Comparison between groups (dyslipidemia vs non-dyslipidemia patients) was performed using the chi-square test (categorical variables), t-test (continuous variables with normal distribution) and Mann-Whitney U (continuous variables with non-normal distribution). Linear and multivariate logistic regression analysis was performed to identify predictors of proteinuria.

Results: Sixty patients were included, 49 (82.9%) females, aged 33.8±12.7 years at the time of diagnosis of lupus nephritis, 44 (73.3%) with class IV, 8 (13.3%) class V, and 8 (13.3%) class III. Twenty-six (43.3%) had dyslipidemia. As for laboratory data, patients had a mean ESR of 59.2±32.0 mm/1st hour, median CRP was 10.1 mg/L (IQR 17.2), median creatinine was 0.97 mg/dl (IQR 0.51), median C3 69.4 mg/L dl (IQR 50.3), median C4 14.5 mg/dl (IQR 13), median dsDNA 246.1 IU/ml (IQR 576.9) and median proteinuria 3157.3 mg/24h (IQR 1890.4). Thirty-two patients (53.3%) had erythrocyturia and 37 (61.7%) had leukocyturia.

At baseline, patients with dyslipidemia were found to have lower ESR values - 41.6 (IQR 24.5) vs 58.5 (IQR 36.0), p=0.046 - and lower percentage of leukocyturia - 30.8% vs 85.3%, p<0.001, with no differences regarding the remaining laboratory and clinical variables.

In the evaluation at 12 months, there were no differences between the groups. In the 60-month evaluation, patients with dyslipidemia were found to have significantly more proteinuria than patients without dyslipidemia - 178.1 (IQR 206.3) vs 82.6 (IQR 116.6), p=0.016, with a tendency towards higher ESR values, CRP, and creatinine.

In regression analyses, when adjusted for hypertension, nephritis class, creatinine, treatment and age, dyslipidemia was not a significant predictor of proteinuria.

Conclusion: In this study, dyslipidemia was associated with higher proteinuria at 5 years in patients diagnosed with lupus nephritis, with a tendency towards higher values of inflammatory markers and creatinine. Although the effect of dyslipidemia on proteinuria may be associated with other changes, it is important to treat dyslipidemia in these patients with lupus nephritis, to improve long-term prognosis.

034 - HYPOCALCEMIA LINKED TO 1-YEAR MORTALITY IN PATIENTS WITH HIP FRACTURE - A MONOCENTRIC COHORT STUDY

Filipe Oliveira Pinheiro[1], Pedro Madureira[1], Maria Seabra Rato[1], Lúcia Costa[1]
[1]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal

Background: Hypocalcemia is associated with acute complications in patients hospitalized for hip fracture, but its role in 1-year mortality is not well established. The aim of this study is to evaluate the association between hypocalcemia and mortality.

Materials and Methods: Retrospective study that included patients admitted to our hospital with hip fracture for 3 consecutive months. Data collected were cerebrovascular disease, heart failure (HF), dementia, chronic obstructive pulmonary disease (COPD), diabetes mellitus (DM), chronic kidney disease (CKD), neoplasm, previous fragility fracture, calcium, and vitamin D. These variables were evaluated upon admission and mortality was seen at 1-year. Calcium was assessed as a continuous variable and hypocalcemia was defined as a categorical variable if calcium was less than 8.6mg/dl. Comparison between groups was performed using the chi-square test and t-test. Linear regression analysis was performed.

Results: Eighty-two patients were included, 71 (86.6%) females, aged 81.5±6.4 years. Five (6.1%) patients had acute myocardial infarction, 14 (17.1%) stroke, 11 (13.4%) HF, 15 (18.3%) dementia, 5 (6.1%) COPD, 8 (9.8%) CKD, 14 (17.1%) neoplasm, and 22 (28.9%) had previous fragility fracture. Mean calcium was 9.6 (0.4) mg/dl, and mean vitamin D was 30.1 (13.5) ng/ml. Nineteen patients (23.2%) died within 1 year of fracture.

Patients who died were found to have more frequent hypocalcemia (60.0% vs 13.2%, p=0.01), age over 85 years (63.2% vs 33.3%, p=0.02) and dementia (36.8% vs 12.7%, p =0.04). There were no statistically significant differences in the remaining variables studied, although there was a tendency for the deceased patients to have a higher percentage of previous fragility fracture, previous acute myocardial infarction, HF and CKD, and lower values of vitamin D.

In linear regression analyses, when adjusted for comorbidities, age and gender, hypocalcemia was an independent predictor of 1-year mortality (OR 9.37, 95%CI 1.73-50.68, p=0.01).

Conclusion: Hypocalcemia was associated with 1-year mortality after hip fracture, regardless of age, gender,

Table I. Measured variables at baseline

Female gender – n (%)	71 (86.6)
Age, years – mean (SD)	81.5 (6.4)
Age > 85 years – n (%)	33 (40.2)
Previous fragility fracture – n (%)	22 (28.9)
Vitamin D, ng/ml– mean (SD)	30.1 (13.5)
Calcium, mg/dl– mean (SD)	9.6 (0.4)
Comorbidities – n (%)	
•Acute myocardial infarction	5 (6.1)
•Stroke	14 (17.1)
•HF	11 (13.4)
•Dementia	15 (18.3)
•COPD	5 (6.1)
•CKD	8 (9.8)
•Neoplasm	14 (17.1)
Mortality at 1-year – n (%)	19 (23.2)

Table I. Measured variables at baseline

Female gender – n (%)	103 (81.7)
Age, years – mean (SD)	82.2 (7.2)
Previous fragility fracture – n (%)	45 (35.7)
- Distal radius	- 19 (42.2)
- Hip	- 13 (28.9)
- Proximal humerus	- 6 (13.3)
- Pelvis	- 3 (6.7)
- Malleolar	- 2 (4.4)
- Spine, clavicle	- 1 (2.2)
Anti-osteoporotic therapy – n (%)	9 (20.0)
Readmission at 1-year – n (%)	29 (36.3)

or comorbidities. The identification and treatment of hypocalcemia may play an important role in the prognosis of these patients, and further studies are needed to better assess this association.

035 - PREVIOUS FRAGILITY FRACTURES IN HIP FRACTURE PATIENTS - AN IGNORED WARNING

Filipe Oliveira Pinheiro[1], Pedro Madureira[1], Maria Seabra Rato[1], Lúcia Costa[1]
[1]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal

Background: Many hip fracture patients have had fragility fractures, which, if treated, can prevent future complications from occurring. The aim of this study is to assess the type and timing of the occurrence of previous fragility fractures in patients with hip fractures.
Materials and Methods: Retrospective study that included patients admitted to our hospital with hip fracture for 6 consecutive months. Data were collected regarding the existence of previous fragility fractures, their location and timing of occurrence, as well as previous anti-osteoporotic therapy. Comparison between groups of patients was made using chi-square and Mann-Whitney U test.
Results: One hundred and twenty-six patients were included, 103 (81.7%) females, aged 82.2±7.2 years. Forty-five (35.7%) patients had had a previous fragility fracture, occurring a median of 3 (1-8) years before; nineteen (42.2%) fractures of the distal radius, 13 (28.9%) hip, 6 (13.3%) proximal humerus, 3 (6.7%) pelvis, 2 (4.4%) malleolar and 1 (2.2%) of the spine and clavicle. Only 9 (20.0%) of the patients with a previous fragility fracture underwent anti-osteoporotic therapy. There were no statistically significant differences between the different fracture sites and timing of occurrence; there was no fracture site significantly more associated with the prescription of anti-osteoporotic therapy, and it should be noted that, of the most prevalent fractures, only 2 (15.4%) patients with hip fracture underwent anti-osteoporotic therapy, 4 (21.1%) patients with fracture of the distal radius and 1 (16.7%) patient with fracture of the proximal humerus.
Conclusion: In this series, it was demonstrated that a relevant percentage of patients with hip fractures already have a history of previous fragility fracture; the identification, evaluation and prescription of adequate and timely anti-osteoporotic therapy in this context can change the functional and vital prognosis of patients with severe osteoporosis.

044 - ASSOCIATION BETWEEN ABNORMAL BODY COMPOSITION PARAMETERS AND DISEASE CHARACTERISTICS IN RHEUMATOID ARTHRITIS PATIENTS: A CROSS-SECTIONAL STUDY USING BIOIMPEDANCE ANALYSIS

João Lagoas Gomes[1], José Adriano Oliveira Marona[2], Pedro Carvalho[1], Ana Raposo[1], Tiago Meirinhos[1]
[1]Rheumatology Unit, Centro Hospitalar do Tâmega e Sousa, Penafiel, Portugal, [2]Rheumatology Department, Centro Hospitalar e Universitário Cova da Beira, Covilhã, Portugal

Introduction: Rheumatoid arthritis is a chronic systemic inflammatory disease with negative effects that go beyond the musculoskeletal systems. Although a sig-

nificant amount of data has been published assessing abnormal body composition phenotypes in Rheumatoid Arthritis (RA) patients, this analysis was not been performed yet in the Portuguese RA population and certain patients reported outcome measures (PROMs) were not fully analyzed.

Objective: To describe body composition parameters in RA patients using bioimpedance analysis (BIA) and evaluate the association between changes in muscle mass and visceral fat with disease activity, pain, disability, quality of life and fatigue.

Methods: In this cross-sectional observational cohort study, RA patients were recruited consecutively during their routine visit to the RA clinic at a secondary care hospital in northern Portugal between January and April 2022. Exclusion criteria were patients having implantable devices or hydroelectrical disturbances that could disturb body composition assessment, alcohol consumption or vigorous exercise in the previous 24 hours and eating or drinking in the preceding 3 hours. To assess body fat, body mass and visceral fat, a multi-frequency BIA instrument was used (OMRON BF511 Body composition monitor; Omron Healthcare Co., Ltd, Kyoto, Japan). Measurements were taken using a fixed protocol to avoid measurement bias. At study enrolment, demographic characteristics were recorded, as well as disease outcomes, including: pain using the visual analogue scale (PVAS), disease activity using the DAS28 (4 variables CRP), disability using the Health Assessment Questionnaire (HAQ), quality of life using the EuroQol 5 dimensions (EQ5D) and fatigue using the Functional Assessment of Chronic Illness Therapy (FACIT) Fatigue Scale (Version 4).

Patients were then stratified into 2 group: low muscle mass (LMM) and normal/high muscle mass (N/HMM) by adjusting for age, gender and BMI. Demographic and disease characteristics were compared between groups using independent-samples t-test for continuous variables and Chi2 for categorical variables. Two linear regression models were created to assess the effect of muscle mass and visceral fat on the studied clinical outcomes.

Results: Overall, 73 patients were included in this study, 28 (38.3%) having LMM and 45 (61.7%) N/HMM. Mean age was 61 in the LMM group and 57 in the N/HMM group. A similar proportion of females was observed between groups (68% vs 64%) and seropositivity rates were also similar (78.6% vs 77.8%). As expected, patients presenting low muscle mass had higher total body and visceral fat percentages. Concerning clinical outcomes, low muscle mass patients presented higher DAS 28 (2.46±1 vs 2.29±1), higher HAQ scores (0.63±0.6 vs 0.53±1) and slightly higher FACIT (44.7±4.0 vs 44±6.7), but none of these differences achieved statistical significance ($p>0.05$). When performing the univariable analysis, neither muscle mass nor visceral fat demonstrate an effect on the studied clinical variables.

Conclusions: The prevalence of low muscle mass in this RA cohort was high and patients with low muscle mass presented a tendency towards unfavorable RA outcomes, but these findings were not statistically significant.

050 - ENTHESITIS, FEMALE GENDER AND VITALITY PERCEPTION AS FATIGUE DETERMINANTS IN SPONDYLOARTHRITIS PATIENTS UNDER BDMARD

Frederico Rajão Martins[1], Ana Martins[2], Beatriz Samões[3], Daniela Santos Oliveira[2, 4], Filipe Oliveira Pinheiro[2], Maria Seabra Rato[2], Miguel Bernardes[2, 5], Lúcia Costa[2]

[1]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, [2]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [3]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, 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: Fatigue is an important domain in quality of life of spondyloarthritis patients, not always directly associated with disease course. The explanatory factors of fatigue in these patients are still not clearly understood.

Objectives: To assess the determinants of fatigue in patients with SpA under biologic disease modifying antirheumatic drugs (bDMARDs).

Methods: A retrospective observational study was performed using registry data of patients with SpA under bDMARD therapy followed at a tertiary level hospital. Data regarding disease activity, response criteria measures, analytic markers, function, metrology, pain, general health and fatigue (using FACIT score) was gathered at baseline, 6 months (t6) and 12 months (t12) after introduction of bDMARD. Statistical analysis (significance at $p < 0.05$) was performed using paired T-test, Wilcoxon test and McNemar tests for paired samples, Mann Whitney-U, Kruskall-Walis and One Way ANOVA for independent samples. Linear and logistic regression models were performed to assess direction and strength of association.

Results: A total of 46 patients were analysed, most male (24, 52.2%) with a predominantly axial involvement (31, 68.9%) and 74.4% of HLA-B27 positive. Most patients had high school or lower education (29,

69.1%), never smoked (26, 61.9%), never drank (34, 79.1%) and had a full-time job (38, 88.4%). TNF inhibitors were employed in all patients, mostly adalimumab (23, 50%). Patient, physician and night pain VAS were significantly lower (p< 0.001) at t6 and t12, but spine VAS only varied significantly between t0 - t6 (p = 0.021) and not t0 - t12 (p=0.405). FACIT didn't vary significantly after bDMARD initiation, and only SF36 vitality score varied significantly (p<0.05). No significant changes were observed in EQ5D, HADs anxiety or depression scores.
At baseline, there was a strong negative correlation between FACIT score, and pain VAS (R = 0.9, p=0.037). Several positive correlations with fatigue at t6 were observed, the strongest with anxiety and depression HADs (p < 0.001), BASFI (p < 0.001) and ASDAS-CRP (p< 0.001). Other positive associations were seen with 66 TJC (p <0.01), patient VAS (p<0.001, pain VAS (p = 0.001), nocturnal pain VAS (p <0.001), BASDAI (p <0.001) and MASES (p <0.001). Fatigue at t6 had a negative correlation with increased ASAS response measures mean value (p = 0.014), EQ5D (p<0.001) and all domains of SF36 (p < 0.001), specially the general health domain (R=-0.811). At t6 there was more fatigue in the female group (p=0.01) and in patient with higher ASDAS disease activity score (p < 0.05), with no differences according to work status, alcohol consumption or tobacco use, educational level, TNF inhibitor exposure or ASAS response. Prediction analysis showed univariable association between several baseline variables and fatigue (lower FACIT scoring) at t6: age at bDMARD introduction (B = - 0,405, p = 0.02), age at diagnosis (B = - 0.43, p = 0.02), physician VAS (B = 0.149, p < 0.05), MASES (B = -1.732, p <0.001), SPARCC (p < 0.05) and female gender (B = -7.95, p = 0.01). Multiple linear regression analysis allowed for creation of a predictive model for FACIT scoring at t6 (R2 = 0.900, p = 0.019): (-3.426) x MASES t0 + (-24.074) x female gender + 0.949 x SF36 vitality score.
Conclusion: Enthesitis, female gender and subjective assessment of vitality seem to be determinants of fatigue in SpA patients under bDMARD. Fatigue in this population is associated with diverse factors that should be optimized in a holistic approach to the patient.

052 - PREDICTORS OF MUSCLE INVOLVEMENT IN PORTUGUESE PATIENTS WITH MIXED CONNECTIVE TISSUE DISEASE

Ana Teresa Melo[1, 2], Manuel Silvério-António[1, 2], Martins-Martinho J [1, 2], Eduardo Dourado[1, 2], Francisca Guimarães[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], Carmo Afonso[3], Daniela Peixoto[3], Nikita Khmelinskii[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]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [4]Serviço de Reumatologia, Centro Hospitalar de São João, Porto, Portugal, [5]Center for Health Technology and Services Research (CINTESIS),, Faculdade de Medicina da Universidade do Porto, Porto, Portugal, Porto, Portugal, [6]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal, [7]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [8]Serviço de Reumatologia, Centro Hospitalar Lisboa Ocidental | Hospital Egas Moniz, Lisboa, Portugal, [9]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal, [10]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal, [11]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, [12]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [13]Serviço de Reumatologia, Centro Hospitalar do Baixo Vouga, E.P.E., Aveiro, Portugal, [14]Serviço de Reumatologia, Centro Hospitalar Trás-os-Montes e Alto Douro, Vila Real, Portugal, [15]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/ Espinho, Gaia, Portugal, [16]Serviço de Reumatologia, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal, [17]Serviço de Reumatologia, Unidade Local de Saúde de Castelo Branco, Castelo Branco, Portugal

Background: Mixed connective tissue disease (MCTD) is a rare heterogeneous disease, characterized by overlapping features of classic connective tissue diseases. Myositis may be present in up to two-thirds of patients with MCTD and it is included in all diagnostic criteria available. Although some possible associations have been reported, to the best of our knowledge, no independent predictors of MCTD-related myositis have been described. We aimed to identify clinical and laboratorial predictors for muscular involvement in a cohort of Portuguese patients with MCTD.
Methods: Multicentre retrospective cohort study including adult-onset patients with a clinical diagnosis of MCTD and fulfilling at least one of the following diagnostic criteria: Sharp, Kasukawa, Alarcón-Segovia or Kahn criteria. Myositis was defined as proximal muscle weakness, creatine kinase elevation, electromyography (EMG) suggestive changes or a positive muscular biopsy. Univariate analysis was performed using Chi-

Square, Fischer's Exact Test and Mann-Whitney Test, as appropriate. Multivariate analysis was performed using binary logistic regression modelling. The linearity of the continuous variables concerning the logit of the dependent variable was assessed via the Box-Tidwell procedure. Cases with missing information and outliers were excluded from the multivariate analysis to fulfil all assumptions necessary to assure the validity of the regression.

Results: A total of 98 patients were included, 43 (44.3%) of whom had muscular involvement at any time of the disease course. Concerning patients with MCTD-related myositis, the mean age at diagnosis was 34.8±12.5 years and the mean disease duration of 4.1±4.9 years. The majority of patients were female (90.7%) and of European ancestry (66.7%).

EMG was performed in 24 patients, of whom 10 (41.7%) had a myopathic pattern. Seventeen patients were submitted to a muscular biopsy, of whom 8 (47.1%) had histological myositis features. Capillaroscopy was performed in 24 patients and 12 (50%) had a scleroderma pattern.

African ancestry and leukopenia were positively associated with myositis at disease onset. Furthermore, fever at the onset of disease, younger age at diagnosis and shorter disease duration were positively associated with the occurrence of myositis at any phase of the disease.

The multivariate analyses predicting myositis at diagnosis included 54 patients and at any time of the disease included 90 patients. These models explained 37.8% and 26.9% (Nagelkerke R2) of the variance in myositis and correctly classified 79.6% and 73.3% of all cases, respectively.

African ancestry (OR 8.39, 95%CI: 1.43-49.37, p=0.019), leukopenia (OR 6.24, 95%CI: 1.32-29.48, p=0.021) and younger age at diagnosis (OR 1.07/year, 95%CI: 1.01-1.14, p=0.035) were identified as independent predictors of myositis at diagnosis. Fever (OR 6.51, 95%CI: 1.23-34.37, p=0.027) was an independent predictor of muscular involvement at any time of the disease in MCDT patients.

Conclusions: African ancestry, leukopenia and younger age at diagnosis are independent predictors of myositis at presentation in MCTD patients, while fever is an independent predictor of myositis at any time of the disease. While evaluating patients with MCTD, these predictive factors should be considered.

054 - RECOMMENDATIONS FOR THE PERFORMANCE AND REPORTING OF SKIN ULTRASOUND IN SYSTEMIC SCLEROSIS: AN INTERNATIONAL COLLABORATION UNDER THE WSF SKIN ULTRASOUND GROUP

Tânia Santiago[1, 2], Eduardo Santos[3], Barbara Ruaro[4], Gemma Lepri[5], L Green[6], M Wildt[7], S Watanabe[8], Alain Lescoat[9], R Hesselstrand[7], Francesco Del Galdo[6], J Pauling[10], L Reeve[11], MA Agostino[12], Marco Matucci[5], Annamaria Iagnocco[13], JAP da Silva[1, 14]

[1]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal, [2]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [3]Escola Superior de Tecnologia e Gestão, Instituto Politécnico de Viseu, Viseu, Portugal, [4]Department of Pulmonology, University Hospital of Cattinara, University of Trieste, Italy., Trieste, Italy, [5]Department of Experimental and Clinical Medicine, University of Florence, and Division of Rheumatology AOUC, Firenze, Italy, [6]Leeds Institute of Rheumatic and Musculoskeletal Medicine, University of Leeds, Leeds, United Kingdom, [7]Department of Rheumatology, Skåne University Hospital, Lund University, Lund, Stockholm, Sweden, [8]Department of Allergy and Rheumatology, Nippon Medical School Graduate School of Medicine, Tokyo, Japan., Tokyo, Japan, [9]Univ Rennes, CHU Rennes, Inserm, EHESP, Irset (Institut de Recherche en Santé, Environnement et Travail) - UMR_S 1085, Rennes, France, Rennes, France, [10]Royal National Hospital for Rheumatic Diseases (at Royal United Hospitals NHS FT), Bath, UK., Bath & North East Somerset, United Kingdom, [11]Occupational Therapist, Pain Management Centre., London, United Kingdom, [12]Rheumatology Department, Ambroise Paré Hospital, AP-HP, Boulogne-Billancourt, Île-de-France, France, Paris, France, [13]Academic Rheumatology Centre, Department of Clinical and Biological Science, University of Turin, Turin, Italy, Torino, Italy, [14]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

Objective: Ultrasound is a promising tool to foster much-needed improvement of skin assessment in systemic sclerosis (SSc). Our aim was to develop evidence and expert-opinion based recommendations to promote the standardization and harmonization of technical execution and reporting of skin ultrasound studies in SSc.

Methods: A multidisciplinary taskforce of 16 members (including 12 rheumatologists, 2 health professionals, 1 methodologist, and, 1 patient research partner) from five European countries and Japan was convened under the auspices of World Scleroderma Foundation. First, a systematic literature review was performed. (1) Then, each member proposed and formulated items to the overarching principles, recommendations, and research agenda. Two rounds of mails exchange for consensus as well as an on-line meeting were performed to debate and refine the proposals. Two Delphi rounds of voting resulted in the final recommendations. Levels of evidence and strengths of recommendations were assigned, and taskforce members voted anonymously on the level of agreement with each of the items.

Table I. WSF recommendations for the execution and reporting of skin ultrasound in systemic sclerosis

	Overarching principles	LoE	GoR	LoA†
A.	B-mode ultrasound and elastography are promising tools to assess skin involvement, but their role in the management of patients with SSc has yet to be defined.	n.a.	n.a.	9.1 (2.3) 87.5%
B.	Report of ultrasound studies in rheumatic and musculoskeletal diseases, including systemic sclerosis, should consider the recommendation checklist developed by EULAR.	n.a.	n.a.	9.0 (1.7) 81.3%
C.	Standardization of the technical aspects for skin ultrasound, in particular image acquisition and analysis, is essential to foster progress in this field.	n.a.	n.a.	8.8 (2.4) 87.5%
D.	The level of training of the examiner and use of appropriate ultrasound equipment and settings, are critical in the assessment of the skin in SSc.	n.a.	n.a.	9.9 (2.0) 87.5%
E.	These recommendations are designed to promote the full validation of skin ultrasound in SSc through optimized objectivity, reliability, and sensitivity of evaluations.	n.a.	n.a.	9.2 (2.2) 93.8%
	Recommendations for the execution of skin ultrasound in SSc			
1.	The examination of the skin in patients with SSc should, whenever possible, include B-mode ultrasound, to measure thickness and echogenicity; and elastography, to measure stiffness.	3b	B	9.3 (1.1) 87.5%
2.	Skin ultrasound should be performed at the standardized areas used in the modified Rodnan skin score.	3b	B	8.7 (1.2) 81.3%
3.	Skin ultrasound should be performed with a high-frequency linear probe (≥18MHz), and with the probe perpendicular to skin surface. Operators should use a generous amount of gel and minimal pressure to avoid tissue compression.	3b	B	9.7 (0.6) 100.0%
4.	Stands-offs should not be used in skin ultrasound in SSc.	5	D	8.6 (2.6) 86.6%
5.	Skin ultrasound should only be performed by well-trained examiners.	3b	B	9.3 (1.4) 87.5%
	Recommendations for the reporting of skin ultrasound in SSc			
6.	Regarding image acquisition, always specify			
	a) the quality criteria for acceptance of an ultrasound image			8.6 (1.9) 87.5%
	b) the skin layers evaluated (epidermis, dermis, hypodermis, subcutaneous layers, others)			9.6 (0.8) 93.8%
	c) the exact location of the skin site/area assessed			9.6 (0.7) 100.0%
	d) the number of images acquired per skin site			8.6 (1.8) 81.3%
7.	Regarding image analysis always specify:			
	a) the number of measurements per skin image/scan and their location within the image			8.9 (1.5) 81.3%
	b) with shear-wave elastography, the size and shape of the region of interest.			9.1 (1.8) 93.8%
	c) how individual measures were processed to calculate the site value			9.2 (1.2) 93.8%

These recommendations should be interpreted in the light of the clarifications provided in the body of the text and by the supporting SLR. †Numbers in column 'LoA' indicate the mean and SD (in parentheses) of the LoA, as well as the percentage of task force members with an agreement ≥8. LoE, level of evidence; GoR, Grade of recommendation; LoA, level of agreement; n.a., not applicable. SSc, systemic sclerosis; EULAR, European Alliance of Associations for Rheumatology.

Results: Five overarching principles and 7 recommendations were developed, based on a SLR and expert opinion, through consensus procedures (Table 1). The overarching principles highlight the promising role of skin ultrasound in SSc assessment, the need for standardization of technical aspects, sufficient training, and adequate equipment. The recommendations provide standards for the execution and reporting of skin ul-

Table II. Research agenda

I.	Validity
a.	Does ultrasound echogenicity have convergent validity against mRSS and/or skin histological findings?
b.	Does ultrasound stiffness have convergent validity against mRSS and/or skin histological findings and/or durometer?
c.	What is the correlation between skin ultrasound domains and different clinical scorings (mRSS)?
d.	What is the correlation between skin ultrasound domains and skin histological findings in different disease clinical phases?
e.	What is the correlation between skin ultrasound domains and patient reported outcome measures, such as Scleroderma Skin Patient-Reported Outcome (SSPRO)?
f.	Is there an association between skin ultrasound domains and disease activity?
g.	Is there a relation between skin ultrasound domains and hand function?
h.	What is the best core of parameters/settings for image acquisition and analysis?
II.	Reliability
a.	What is the test-retest reliability of skin ultrasound in the different SSc subsets?
b.	What is the intra and inter-reader reliability of skin ultrasound in the different SSc subsets?
III.	Discriminatory capacity
a.	Does skin ultrasound domains discriminate between:
1.	early phases of the disease and normal controls
2.	disease subsets, i.e., VEDOSS vs early inflammatory and dcSSc vs lcSSc vs sine?
3.	phase of cutaneous involvement, i.e., edematous vs fibrotic vs atrophic?
IV.	Responsiveness to change
a.	What is the sensitivity to change over time/treatment of skin ultrasound in SSc, in different disease subsets, in observational studies and randomized clinical trials?
b.	What is the correlation between changes in skin ultrasound measurements and in mRSS/skin histology over time/treatment?
c.	Can skin ultrasound separate between the effects of normal ageing and that of the disease and treatments upon the skin?
d.	How frequently should skin ultrasound be repeated in SSc patients?
V.	Threshold of meaning
a.	What is the smallest detectable change and minimal clinically important difference for the diverse skin ultrasound domains (with stratification based on disease subsets)?
VI.	Feasibility
a.	What is the feasibility of skin ultrasound, in particular: cost of equipment and software, time taken for image acquisition and analysis?
b.	What is the best trade-off of validity and feasibility regarding of the minimum number (and sites) of skin regions examined by ultrasound in SSc?
c.	Is there an advantage in performing skin ultrasound examination in symmetrical Rodnan skin sites?
d.	Is skin ultrasound useful in a combined multi-organ ultrasound approach (e.g., digital ulcers, lung, vascular, joints)?
e.	What is the availability and current practice of skin ultrasound in SSc worldwide?
VII.	Contextual factors
a.	What is the impact of patient factors (age, gender, BMI, smoking, sun exposure, …) on skin ultrasound domains?
b.	What is the impact of ambient contextual factors (hour of the day, room temperature, time of acclimatization, patient position,...) on skin ultrasound domains?
VIII.	Educational agenda
a.	Ultrasound courses and process of certification of competencies, specifically focused on skin ultrasound in SSc

trasound in SSc. The research agenda (table 2) includes the need for more research into unmet needs according to OMERACT Algorithm requirements.

Conclusion: These are the first recommendations providing guidance on the execution and reporting of skin ultrasound in SSc patients, aiming at improving the

interpretability, reliability, and generalisability of skin ultrasound, thus consolidating its role in research and practice.

REFERENCES

1. Santiago T, Santos E, Ruaro B, et al. Ultrasound and elastography in the assessment of skin involvement in systemic sclerosis: A systematic literature review focusing on validation and standardization. Semin Arthritis Rheum. Jan 10 2022:151954. doi:10.1016/j.semarthrit.2022.151954

057 - COVID19 IN SPONDYLOARTHRITIS PATIENTS UNDER BDMARD – A COMPREHENSIVE ASSESSMENT

Frederico Rajão Martins[1], Ana Martins[2], Daniela Santos Oliveira[2, 3], Beatriz Samões[4], Rafaela Nicolau[5], Filipe Oliveira Pinheiro[2], Maria Seabra Rato[2], Miguel Bernardes[2], Lúcia Costa[2]

[1]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [2]Rheumatology Department, 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, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [5]Rheumatology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Background: SARS-Cov-2 infection had a major impact on patients with inflammatory rheumatic diseases. Spondyloarthritis (SpA) patients were one of the most affected groups of these patients.

Objectives: To assess the impact of Covid19 in spondyloarthritis patients under biological disease modifying antirheumatic drugs (bDMARDs).

Methods: A retrospective observational study was conducted using registry data of patients with SpA under bDMARD therapy, followed at a tertiary level hospital, that have been diagnosed with COVID19 from March 2019 to December 2021. At least one evaluation previous (t0) and two evaluations after SARSCoV-2 infection (t1, t2) were included in our analysis. Sociodemographic, clinical and laboratorial data were collected. Statistical analysis (significance at $p < 0.05$) was performed using paired T-test, Wilcoxon test and McNemar tests for paired samples. Linear and logistic regression models were performed to assess direction and strength of association.

Results: Thirty-two patients with SpA under bDMARD had COVID19, mostly women (20, 62.5%), with a average disease course of 18.65 (± 9.69) years, mainly with axial involvement (19, 59.4%) and HLA-B27 positive (11, 64.7%). Thirty (93.75%) were under-TNF inhibitors, golimumab being the most used (9, 28.1%), with a median bDMARD persistence of 2.63 (5.09) years. Seven (21.9%) were under a cDMARD, 3 (9.4%) under NSAID and 18 (56.3%) under corticosteroids. Three (9.4%) were already vaccinated against SARS-CoV-2, 2 (66.6%) with the mRNA1273 vaccine. Arterial hypertension was uncommon (5, 15.6%) and one patient (3.1%) had a previous diagnosis of type 2 diabetes. Most were never-smokers (17, 53.1%) and never-drinkers (29, 90.6%). Age at infection was 40.97 (± 6.15) years and the most common symptom was cough (22, 68.8%). Event tree analysis didn't show association between infection and SpA subtype, habits or sociodemographic variables. One patient needed hospital admission, without need of oxygen therapy. One patient had an overlaid bacterial infection and no thromboembolic complications were observed. Two patients needed specific SARS CoV-2 infection treatment. Twelve (37.5%) patients suspended bDMARD, only 2 (6.3%) maintaining suspension after infection. Higher BASDAI at t1 was observed, without statistical significance. Higher VAS scores for all domains were observed at t1, but not at t2, also without statistical significance; moreover, physical function didn't change significantly. No differences were observed according to gender or SpA subtype or use of other therapies. The only statistically significant difference concerned MASES score between t0 and t1 (1 ± 4 vs. 2 ± 6, p=0.04). Higher baseline tender joint score ($p < 0.01$) and higher baseline LEI (p=0.03) negatively correlated with MASES score variation. Several baseline variables correlated positively with MASES at t1, including female gender ($p < 0.01$), corticosteroid use ($p = 0.04$), BASDAI ($p < 0.01$), ASDAS-ESR ($p < 0.01$), ASDAS-CRP ($p < 0.01$), DAS28 ($p < 0.01$), SPARCC ($p = 0.04$), physician VAS ($p = 0.03$) and total spine VAS ($p = 0.01$). Working status varied significantly after SARS-Cov-2 infection (at least part-time - 29, 90.6% vs. 22, 68.8%, p= 0.016).

Conclusion: SpA patients on bDMARD had a mild course of SARS-CoV-2 infection, with slight changes in enthesitis score in the short term, the latter particularly in those with higher disease activity in the pre-infection period. Long-term effects on work status could represent confounding factors related to the economic constraints of the pandemic.

061 - CLINICAL AND IMMUNOLOGICAL CHARACTERISATION OF A (VERY) EARLY DIAGNOSIS OF SYSTEMIC SCLEROSIS COHORT: CAN AUTOANTIBODIES ADD PROGNOSTIC VALUE?

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], Inês Genrinho[10], Couto

M[10], Marília Rodrigues[9], Maria João Salvador[4, 5], Ana Catarina Duarte[3], Ana Cordeiro[3], Maria José Santos[2, 3], JE Fonseca[1, 2], Inês Cordeiro[1, 2], Catarina Resende[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]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [4]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [5]Faculdade de Medicina, Universidade de 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, Centro Hospitalar de Leiria, Leiria, Portugal, [10]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Introduction: Diagnosis of systemic sclerosis (SSc) in its earliest stages is critical due to the importance of an early follow-up and treatment. However, early diagnosis is still challenging. Three sets of (very) early diagnosis of SSc (VEDOSS) classification criteria have been developed. All include Raynaud's phenomenon, the presence of SSc-specific autoantibodies and changes in nailfold capillaroscopy (NC) as key VEDOSS features.
Methods: Multicentre open cohort study including adult VEDOSS patients registered in the Rheumatic Diseases Portuguese Register (Reuma.pt) up to February 2021. Patients who fulfilled the 2013 ACR/EULAR classification criteria for SSc were excluded. The associations between autoantibody expression and clinical data were established using Chi-Square, Fischer's Exact or Mann-Whitney U tests.
Results: 133 patients were initially identified, of whom 124 were included (9 fulfilled the 2013 ACR/EULAR classification criteria for SSc). These patients had a mean age and disease duration (since Raynaud's phenomenon onset) of 53.1 ± 14.9 and 8.5 ± 7.0 years, respectively. Most were females (N=115/124, 92.7%) and had European ancestry (N=49/51, 96.1%). The most common clinical manifestations were Raynaud's phenomenon (N=108/122, 88.5%), joint involvement (N=30/117, 25.6%), puffy fingers (N=12/92, 13.0%), telangiectasia (N=14/122, 11.5%), digital ulcers (DU, N=11/120, 9.2%), and oesophageal involvement (N=8/116, 6.9%). Three patients (3.3%) had interstitial lung disease with ground-glass opacities (N=3/65, 4.6%) but not honeycombing (N=0/65) on the chest CT scan. None of the patients had skin

Table I. Demographic and clinical data from the 124 patients included in the study

Variable	NPAD	Total
Demographic data		
Females, N (%)	124	115 (92.7)
Ancestry		
European white, N (%)	51	49 (96.1)
Non-European white, N (%)	51	1 (2.0)
Romani, N (%)	51	1 (2.0)
Age at the symptom onset, mean (SD)	104	44.6 (15.8)
Age at diagnosis, mean (SD)	109	48.9 (15.1)
Age at the last appointment, mean (SD)	124	53.1 (14.9)
Diagnosis delay, mean (SD)	103	4.0 (6.1)
Disease duration (since Raynaud onset), mean (SD)	104	8.5 (7.0)
Death, N (%)	124	2 (1.6)
Classification criteria		
SSc classification criteria		
2013 ACR/EULAR, N (%)	115	0 (0.0)
1980 ACR, N (%)	91	0 (0.0)
VEDOSS classification criteria		
2010 EUSTAR, N (%)	124	94 (75.8)
2001 LeRoy, N (%)	124	25 (20.2)
Clinical manifestations		
Skin disease		
Skin thickening, N (%)	121	1 (0.8)
Skin thickening proximal to the MCP in any location, N (%)	121	0 (0.0)
Skin thickening proximal to the MCP in hand, N (%)	121	0 (0.0)
Sclerodactyly without skin thickening proximal to the MCP, N (%)	121	0 (0.0)
Puffy fingers, N (%)	92	12 (13.0)
mRSS in the last appointment, mean (SD)	123	0 (0.0)
Calcinosis, N (%)	117	2 (1.7)
Musculoskeletal disease		
Joints involvement (arthritis or arthralgia), N (%)	117	30 (25.6)
Flexion contractures, N (%)	114	0 (0.0)
Tendon friction rubs, N (%)	114	0 (0.0)
Myositis, N (%)	117	0 (0.0)
Vascular disease		
Raynaud phenomenon, N (%)	122	108 (88.5)
Digital ulcers, N (%)	120	11 (9.2)
Pitting scars, N (%)	92	3 (3.3)
Telangiectasia, N (%)	122	14 (11.5)
Pulmonary arterial hypertension, N (%)	119	0 (0.0)
Capillaroscopy changes, N (%)	87	47 (54.0)
Early scleroderma pattern, N (%)	92	24 (26.1)
Active scleroderma pattern, N (%)	92	5 (5.4)
Late scleroderma pattern, N (%)	92	0 (0.0)

continua na página seguinte

Table I. Demographic and clinical data from the 124 patients included in the study

Variable	NPAD	Total
Lung disease		
Interstitial lung disease, N (%)	90	3 (3.3)
Restrictive syndrome, N (%)	66	0 (0.0)
Lung fibrosis on chest radiograph, N (%)	32	1 (3.1)
Fibrosis on chest CT scan, N (%)	65	1 (1.5)
Ground glass opacities on chest CT scan, N (%)	65	3 (4.6)
Honeycombing on chest CT scan, N (%)	65	0 (0.0)
Gastrointestinal disease		
Oesophageal involvement, N (%)	116	8 (6.9)
Gastric involvement, N (%)	116	1 (0.9)
Intestinal involvement, N (%)	116	1 (0.9)
Heart involvement, N (%)	115	2 (1.7)
Renal involvement, N (%)	114	1 (0.9)
Autoantibodies		
Anti-nuclear antibodies, N (%)	121	110 (88.7)
Anti-centromere antibodies, N (%)	124	80 (64.5)
Anti-topoisomerase I antibodies, N (%)	123	15 (12.2)
Anti-Pm/Scl antibodies, N (%)	108	5 (4.6)
Anti-Th/To antibodies, N (%)	97	3 (3.1)
Anti-U11/U12-RNP antibodies, N (%)	99	1 (1.0)
Anti-U3-RNP antibodies, N (%)	103	1 (1.0)
Anti-RNA polymerase III antibodies, N (%)	120	1 (0.8)
Anti-Ku antibodies, N (%)	100	0 (0.0)
Anti-U1-RNP antibodies, N (%)	109	0 (0.0)

These recommendations should be interpreted in the light of the clarifications provided in the body of the text and by the supporting SLR. †Numbers in column 'LoA' indicate the mean and SD (in parentheses) of the LoA, as well as the percentage of task force members with an agreement ≥8. LoE, level of evidence; GoR, Grade of recommendation; LoA, level of agreement; n.a., not applicable. SSc, systemic sclerosis; EULAR, European Alliance of Associations for Rheumatology.

thickening, flexion contractures, tendon friction rubs, or myositis. Most patients expressed antinuclear antibodies (N=100/121, 88.7%), and the most frequent were anti-centromere (ACA, N=80/124, 64.5%), anti-topoisomerase I (N=15/123, 12.2%), anti-Pm/Scl (N=5/108, 4.6%), and anti-Th/To (N=3/97, 3.1%) antibodies. Most patients had NC changes (N=47/87, 54.0%), including early (N=24/92, 26.1%) and active (N=5/92, 5.4%) but not late (N=0/92) scleroderma pattern. Most included patients fulfilled the 2010 EUSTAR VEDOSS classification criteria (N=94/124, 75.8%) but not the 2001 LeRoy classification criteria (N=25/124, 20.2%). Patients that were positive for ACA were less likely to present DU [odds ratio (OR)=0.09, 95% confidence interval (95%CI):0.02-0.45, p=0.001) or pitting scars (p=0.043, OR not calculable), and patients positive for anti-Pm/Scl antibodies patients had higher risk of oesophageal involvement (OR=12.40, 95%CI:1.67-91.87, p=0.036).

Conclusion: Most clinical findings were similar to what has been described, but puffy fingers and oesophageal involvement were less prevalent, and joint involvement was more prevalent than previously reported. To the best of our knowledge, anti-Pm/Scl and anti-Th/To autoantibodies prevalence in VEDOSS had not been previously reported. Our results suggest that autoantibodies can have a prognostic role in VEDOSS. Of note, our VEDOSS cohort has a longer mean disease duration than those previously reported.

064 - ANTI-RO/SSA ANTIBODIES POSITIVITY - THE CLINICAL RELEVANCE!

Carolina Mazeda[1, 2], Susana P. Silva[1, 2], Rita Monteiro[2, 3], Renata Aguiar[1, 2], Barcelos A.[1, 2]

[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz, Aveiro, Portugal, [3]Clinical Pathology, Centro Hospitalar Baixo Vouga, Aveiro, Portugal

Background: Anti-Ro/SSA antibodies are directed against different proteins of intracellular small ribonucleic acid (RNA)-protein complexes, Ro60 (60 kDa) and Ro52 (52 kDa) and are the most frequently detected autoantibodies in patients´ sera. The clinical associations with antibodies to the Ro60 protein is well documented and includes systemic lupus erythematosus (SLE) and Sjögren syndrome (SS).1 The presence of anti-Ro52 antibodies has been reported in a large variety of diseases, such as neoplastic diseases or viral infections.1,2

Objectives: The aim of this study was to analyze the clinical relevance and the disease phenotype of patients with anti-Ro52 and/or anti-Ro60 autoantibodies in a single portuguese secondary hospital.

Methods: Retrospective and descriptive study included all patients screened for anti-nuclear antibodies (ANA) in the database of the immunology laboratory of our Hospital between 2017 and 2021. Only adult patients (≥ 18-years-old) with positive anti-Ro52 and/or Ro60 were included in the study and divided into three groups: group 1 – Ro52+/Ro60-, group 2 – Ro52+/Ro60+ and group 3 – Ro52-/Ro60+. The presence of ANA was tested by indirect immunofluorescence (threshold of positivity established at titer 1/160) and further screened for anti-extractable nuclear antigen (ENA) antibodies. Socio-demographic and clinical data were collected. Descriptive analysis was performed, p-value≤0.05 was statistically significant.

Results: Among 1210 patients, 277 were positive for

Table I. Clinical data of three groups

Variable	Group 1 Ro52+/ Ro60- (n=107)	Group 2 Ro52+/ Ro60+ (n=86)	Group 3 Ro52-/ Ro60+ (n=84)	p
Immune-mediated rheumatologic diseases (n)	51	69	53	0.004
Systemic lupus erythematosus	10	16	7	0.396
Sjögren syndrome	21	40	20	0.050
Systemic sclerosis	2	1	2	0.618
Inflammatory myositis	4	0	1	0.024
Rheumatoid arthritis	9	2	7	0.020
Undifferentiated connective tissue disease	4	9	14	0.025
Other diseases	1	1	2	0.824
Non-rheumatologic disease (n)	56	17	31	0.369
Infections	3	1	2	0.978
Neoplasms	7	2	1	0.319
Liver cirrhosis	6	2	8	0.178
Interstitial lung disease	2	0	2	0.637
Chronic kidney disease	2	0	1	0.736
Unclassified	36	12	17	0.444

the antibody anti-Ro52 and/or Ro60. Most of the patients included were women (79.8%) with a mean age of 58.87±16.12. Immune-mediated rheumatologic diseases were diagnosed in 173 (62.5%) patients, while 104 (37.5%) had non-rheumatologic disease. About 38.6% were exclusively anti-Ro52 positive, 30.3% of patients were anti-Ro60 positive alone and 31% tested positive for both. The clinical data of the patients in these three groups is represented in table 1. In each of the three groups, the most frequent pattern was nuclear fine speckled (AC-4). In group 2, SS was the most frequent diagnosis (46.5%), followed by SLE (19.6%). Regarding ANA titers, a higher amount was observed in this group ($p<0.05$). The most frequently associated autoantibodies were anti-La (15.1%). In group 3, the diagnosis of SS was the most frequent (23.8%) followed by undifferentiated connective tissue disease (16.7%). Lupus anticoagulant (7.1%) and anti-RNP (6.0%) were more represented in this group. Finally, in group 1, non-rheumatologic disease was the most represented group of disease. In the case of SLE and systemic sclerosis, there were no statistically significant differences between the three groups and the different types of organ involvement. In SS, sicca symptoms were more prevalent in group 2, with statistically significant differences between the groups ($p=0.027$).

Conclusion: In line with published studies, our work demonstrated that, among the sera tested, autoreactivity to anti-Ro52 alone was the most often observed and in most patients the presence of this antibody alone was not associated with immune-mediated rheumatologic disease, but mainly with malignancies. We also identified the clinical phenotype of patients with positive anti-Ro52 and/or anti-Ro60. Thus, we reinforce the importance of specific detection such as Ro-52 or Ro60 as they present individual diagnostic utility.

REFERENCE

1. Wu S et al. Int J Clin Pract. 2021 Mar;75(3):e13679.; 2-Robbins A et al. Front Immunol. 2019 Mar 12;10:444.

071 - APPLICATION OF ARTIFICIAL INTELLIGENCE TECHNIQUES IN RHEUMATOID ARTHRITIS - A NARRATIVE REVIEW

Filipe Cunha Santos[1], Ana Sofia Pinto[1], Sara Paiva Dinis [1], Emanuel Costa[2], Nathalie Madeira[1], Joana Fonseca Ferreira[1, 3], Cláudia Vaz[1, 3]

[1]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, [2]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal, [3]Faculdade de Ciências da Saúde, Universidade da Beira Interior, Covilhã, Portugal

Background: Artificial intelligence (AI) is a broad term which refers to the performance of a human being's task with the help of machine and technology and it can be used to predict or analyse complex medical data. Rheumatoid arthritis (RA) is a chronic inflammatory disease, multifaceted, which often represents a challenge from a diagnostic and disease control point-of-view.

Objectives: The objective of this review was to provide a comprehensive overview of the use of AI in clinical practice of RA.

Methods: An electronic search was performed on May 23th, 2022 in PubMed/MEDLINE (National Library of Medicine) database using MeSH keywords and the following string: "Arthritis, Rheumatoid"[Mesh] AND (("Artificial Intelligence"[Mesh]) OR ("Deep Learning"[Mesh]) OR ("Neural Networks, Computer"[Mesh]) OR ("Machine Learning"[Mesh])). Only articles written in English were considered.

Results: From 197 screened articles, 49 were considered and analysed; most were published after 2016. 18 articles were related to genetic and transcriptic biomarkers, 13 to prediction or assessment of disease activity, 12 to diagnosis and 6 to treatment response. The number of patients included ranged from 8 to 200900. Overall, 56% of the articles described the use of machine learning, 26% neural networks, 12% imaging analyses and 6% deep learning. External validation was performed in 32% of the articles. In regards to genetic

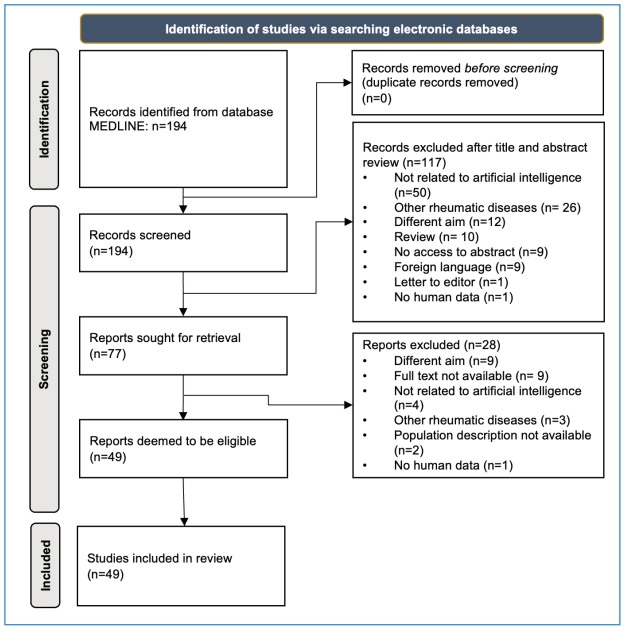


Figure 1.

and transcriptic biomarkers, the main applications described were the ability to differenciate patients with RA and osteoarthritis; algorithms using molecular signatures and specific gene polymorphisms in order to differenciate healthy controls and RA patients; prediction of clinical response before initiating treatment with methotrexate and anti-TNF; identification of possible biomarkers related to disease activity and postulate scores to predict that activity. In regards to prediction or assessment of disease activity, articles considered described the use of computer-assisted imaging analysis (magnetic resonance imaging, ultrasound and x-ray) to detect subclinical synovitis, presence of intersticial lung disease, erosions and automatic classification of ultrasound images; prediction of next appointment's disease activity based on electronic records data; prediction of disease flares using physical activity data from wearables; validation of an estimated Clinical Disease Activity Index (CDAI) score. In regards to diagnosis, authors described algorithms capable of accurately diagnosing RA and defining disease phenotypes from electronic data registry and find potencial risk factors related to disease progression in patients with early arthritis. Lastly, in regards to treatment response, articles described the use of AI techniques to model the individual risk of flares in anti-TNF treated patients in remission, under a tapering regimen; identification and prediction of difficult-to-treat RA patients; modelling responses to various biologic disease-modifying anti-rheumatic drugs (bDMARDs) and analyse gut microbiome to predict clinical response to methotrexate.

Conclusions: This review provides an up-to-date overview of the usefulness of AI approaches in various aspects of RA. In the future, development of this field should aid rheumatologists in the diagnosis, clinical decision and overall management of rheumatoid arthritis.

073 - THE IMPACT OF SARS-Cov-2 INFECTION ON DISEASE ACTIVITY AND CLINICAL RESPONSE TO BIOLOGICAL DMARDS IN PATIENTS WITH RHEUMATOID ARTHRITIS

Beatriz Samões[1], Ana Martins[2], Daniela Santos Oliveira[2, 3], Frederico Rajão Martins[4], Rafaela Nicolau[5], Maria Seabra Rato[2], Filipe Oliveira Pinheiro[2], Miguel Bernardes[2, 6], Lúcia Costa[2]

[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [2]Rheumatology Department, 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]Rheumatology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [6]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal

Background: Infections are a known trigger for Rheumatoid Arthritis (RA) flares.1 It is still unclear whether SARS-Cov-2 infection affects RA disease activity and the clinical response to biological disease-modifying antirheumatic drugs (bDMARDs).

Objectives: To evaluate the effect of SARS-Cov-2 infection on disease activity and bDMARD responses in patients with RA.

Methods: A retrospective study was carried out in a cohort of RA patients treated with bDMARDs from a tertiary hospital centre. Demographic and clinical data, including occurrence of SARS-Cov-2 infection, were obtained through medical records. Disease activity (DAS28, DAS28-CRP, CDAI and SDAI) and ACR and EULAR bDMARD responses were evaluated at four time points: baseline (t1 - last evaluation before Covid-19 pandemic), before (t2) and after (t3) SARS-Cov-2 infection and at the end of follow-up (t4 - last appointment of 2021). In patients with no record of SARS-Cov-2 infection the middle evaluations were obtained from two random consecutive appointments during Covid-19 pandemic. Statistical analysis (significance at $p<0.05$) was performed using paired t-test, Wilcoxon and McNemar tests for paired samples and unpaired t-test, Mann-Whitney, Fisher and $\chi 2$ tests for independent samples according to the type of variable and the presence of normal distribution.

Results: Of the 237 patients included, most of them

was women [n = 195 (82.3%)], with a mean age of 59.6 ± 10.1 years old and a median [min, max] disease duration of 18 [2, 50] years. The majority presented rheumatoid factor (RF) and/or anti-citrullinated protein antibodies (ACPA) positivity [n = 204 (87.9%)] and radiographic erosions [n = 119 (72.6%)]. The prevalence of SARS-Cov-2 infection was 11.4% (n=27). Mean disease activity was lower after SARS-Cov-2 infection compared to the previous evaluation on all scores used; however, this difference was not statistically significant. Nevertheless, when compared to the mean disease activity at the end of follow-up, there were statistically significant differences in DAS28-CRP (t2 3.2±1.0 vs. t4 2.8±1.1, p=0.017) and CDAI (t2 11.1±8.1 vs. t4 8.7±6.2, p=0.05) scores. The relative number of patients with no ACR or EULAR bDMARD responses before SARS-Cov-2 infection wasn't different from post infection and at the end of follow-up. At baseline, the infected and uninfected groups were similar regarding gender, age, RF and/or ACPA positivity, erosive disease, disease and biologic treatment durations, baseline disease activity and ACR and EULAR response. The variation in disease activity and the relative number of patients with worsening or improving EULAR and ACR bDMARD responses between t2 and t3 were not significantly different in the two groups, as well as between t2 and t4. The prevalence of patients who switched to another bDMARD was significantly higher in the group of patients who had Covid-19 [n=4 (14.8%) vs. 9 (4.3%), p=0.047]. The main reason for switching was the ineffectiveness of the therapy (n=11).

Conclusions: No worsening of disease activity or ACR and EULAR bDMARD responses was found after SARS-Cov-2 infection in RA patients under bDMARD. However, the later can be explained by the small sample size. Indeed, these patients exhibited a higher rate of switch due to ineffectiveness of therapy, suggesting a negative impact of SARS-Cov2 infection on the disease course.

REFERENCES

1. Rashmi Roongta, Alakendu Ghosh. Managing rheumatoid arthritis during COVID-19. Clinical Rheumatology (2020); 39(11):3237-3244

074 - RELIABILITY AND CONTENT VALIDITY OF THE PORTUGUESE VERSION OF THE COMMISSIONING FOR QUALITY IN RHEUMATOID ARTHRITIS PATIENT--REPORTED EXPERIENCE MEASURE (CQRA-PREM): PRELIMINARY RESULTS

Daniela Santos Oliveira[1, 2, 3], Raquel Rainho[3], Miguel Bernardes[1, 3], Carlos Vaz[1, 2, 3], Lúcia Costa[1], João Almeida Fonseca[2, 4, 5], Cristina Jácome[2, 5]

[1]Rheumatology Department, Centro Hospitalar Universitário 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]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal, [4]MEDIDA – Medicina, Educação, Investigação, Desenvolvimento e Avaliação, Porto, Portugal, [5]Department of Community Medicine, Information and Health Decision Sciences (MEDCIDS), Faculty of Medicine, University of Porto, Porto, Portugal, Porto, Portugal

Background: The CQRA-PREM has been developed in the United Kingdom to evaluate the perspective of patients with rheumatoid arthritis (RA) about the care provided in rheumatology units of the National Health Service (1). This PREM might also be feasible to be used in Portugal, yet an adaptation and validation process is needed.

Objectives: We aimed to translate and cross-cultural adapt CQRA-PREM to Portuguese and evaluate the reliability and content validity of this version.

Methods: A study combining qualitative and quantitative approaches was conducted with patients with RA from a single rheumatology center. The translation and cultural adaptation of Portuguese CQRA-PREM included initial translation and cultural adaptation by 2 native Portuguese researchers, evaluation of this initial translation by a panel of 5 experts during an online meeting and back translation by 1 bilingual researcher. Authors of the original version approved this initial translation. CQRA-PREM includes 7 domains for patient-centered care: Needs and preferences (5 items); Coordination of care and communication (4 items); Information, education, and self-care (4 items); Daily living and physical comfort (2 items); Emotional support (2 items); Family and friends (1 item); Access to care (5 items) and 1 question for the overall experience with the care provided. Answers are given on a 5-point Likert scale (strongly disagree-1 to strongly agree-5). A total of 21 patients were invited to participate in online focus groups and 14 accepted. Patients were then asked to fill in a consent form and the preliminary version of the CQRA-PREM. The focus groups were transcribed and analyzed using thematic analysis. Questionnaire responses were analyzed with descriptive statistics and reliability (internal consistency) with the Cronbach´s alpha (α).

Results: A total of 12 patients (53±9y; 92% female) with a mean disease duration of 14±9 years participated in 2 focus groups (duration 95±7min). The focus groups revealed that patients considered CQRA-PREM "simple" and "objective" and that all questions were easy to understand. Nevertheless, patients suggested the addition of synonyms for certain terms and of daily living examples to clarify some items. The "Needs and preferences" and "Access to care" were the domains

Table I. Median's responses and quartile (Q) for each domain

CQRA-PREM domains	Median (Q1-Q3)
1. Needs and preferences	4.5 (4.25-4.5)
2. Coordination of care and communication	4 (3.25-4.5)
3. Information, Education and Self-care	3 (2-4)
4. Daily living and physical comfort	3.25
5. Emotional support	3.25
6. Family and friends	4 (3-4)
7. Access to care	4.5 (2.5-5)

1-Strongly disagree; 2-Disagree; 3-Neither agree nor disagree; 4-Agree; 5-Strongly agree

with better experience (Table 1). The Cronbach's alpha was 0.94 for the total questionnaire and between 0.71 and 0.91 for the domains.

Conclusions: Our findings suggest that both content validity and internal consistency of the Portuguese version of CQRA-PREM are acceptable. In future, a field-testing study to assess other psychometric properties (e.g., test-reliability and validity) should be conducted. We believe this PREM will contribute to optimize patient-centered care in Portugal.

REFERENCES

1. Bosworth A, et al. Curr. Rheumatol. Rev. 2015;11.

080 - CLINICAL AND IMMUNOLOGICAL CHARACTERISTICS OF A SINGLE-CENTRE COHORT OF MILD SYSTEMIC LUPUS ERYTHEMATOSUS: HAS IT ALWAYS BEEN MILD?

Francesca Bottazzi[1], Eduardo Dourado[1, 2, 3], Bianca Lucia Palermo[1], Carlomaurizio Montecucco[1], Giovanni Zanframundo[1], Lorenzo Cavagna[1]

[1]Rheumatology Unit, University and IRCCS Policlinico S. Matteo Foundation, Pavia, Italy, [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

Background: The improved early recognition and treatment of systemic lupus erythematosus (SLE) allowed the increase of patients with mild disease. The phenotype of mild SLE has not been fully characterised.

Objective: We aimed to characterise a cohort of mild SLE patients.

Methods: The clinical information of adult patients with mild SLE referring to our centre from January to December 2021 was retrospectively collected and analysed. Mild SLE was defined as the occurrence of low disease activity (LDA) or remission for at least 24 consecutive months, according to the Systemic Lupus Erythematosus Disease Activity Index 2000 (SLEDAI-2k) and the Systemic Lupus Erythematosus Disease Activity Score (SLEDAS) scores, using only hydroxychloroquine and/or prednisolone up to 5 mg/day. Descriptive data were presented as absolute frequencies (percentage) for categorical variables and median (interquartile range) for continuous variables.

Results: The clinical information of 240 SLE patients was reviewed. About a third of the patients (N=81/240, 33.8%) were included (Table 1). Most included patients were females (N=76/81, 93.8%), with a median age of 35.0 (23.5-48.5) years at diagnosis and 54.0 (44.0-67.0) years at the last follow-up. The vast majority of patients met the Systemic Lupus International Collaborating Clinics (SLICC, N=65/80, 81.3%) and the 2019 European Alliance of Associations for Rheumatology/ American College of Rheumatology (EULAR/ACR, N=61/81, 75.3%) SLE classification criteria. According to SLEDAI-2k and SLEDAS, 36/80 (45.0%) and 75/79 (95.0%) of the patients were in remission, and 44/80 (55.0%) and 4/79 (5.0%) were in LDA at the last follow-up, respectively. All patients were positive for antinuclear antibodies (ANA, N=81/81, 100%), and anti-Ro antibodies were the most commonly found antibodies (N=40/81, 49.4%), followed by the anti-dsDNA antibodies (N=23/81, 28.4%). Included patients most commonly had a history of cutaneous lupus (N=60/81, 74.1%), inflammatory arthralgia (N=51/81, 71.6%), or arthritis (N=36/81 (44.4%). Also, severe internal organ manifestations such as lupus nephritis (N=11/81, 13.6%) and central nervous system involvement (N=4/81, 4.9%) were present. The most commonly used treatments were hydroxychloroquine (N=80/80, 100%) and prednisolone (N=60/79, 75.9%). However, previous treatments also included cyclophosphamide (N=9/77, 11.7%), mycophenolate mofetil (N=5/77, 6.5%), and rituximab (N=2/81, 2.5%). Maximum SLEDAI-2k and SLEDAS scores were 7 (5-8) and 8.00 (5.59-11.72), respectively, generally recorded at disease onset. On the last follow-up, the median SLEDAI-2k was 0.0 (0.0-2.0), and the median SLEDAS was 0.37 (0.37-1.12).

Discussion: Mild SLE is phenotypically characterised by the common occurrence of joint and skin involvement but is also achievable in patients with previous kidney and central nervous system involvements and patients that were highly immunosuppressed.

Table I. Clinical and serological characterisation of the cohort (N=81)

Variable	Data
Demographic data	
Females, n/N (%)	76/81 (93.8)
Age at diagnosis, median (IQR)/N	35.0 (23.5-48.5)/81
Age at last follow up, median (IQR)/N	54.0 (44.0-67.0)/81
Disease duration (in months), median (IQR)/N	170 (90-289)
Clinical data	
Joint involvement	
Inflammatory arthralgia	
Inflammatory arthralgia (ever), n/N (%)	51/81 (71.6)
Inflammatory arthralgia (current), n/N (%)	3/81 (3.7)
Arthritis	
Arthritis (ever), n/N (%)	36/81 (44.4)
Arthritis (current), n/N (%)	0/81 (0)
Jaccoud arthritis	
Jaccoud arthritis (ever), n/N (%)	4/81 (4.9)
Serosal involvement	
Pleuritis	
Pleuritis (ever), n/N (%)	4/81 (4.9)
Pleuritis (current), n/N (%)	0/81 (0)
Pericarditis	
Pericarditis (ever), n/N (%)	8/81 (9.9)
Pericarditis (current), n/N (%)	0/81 (0)
Mucocutaneous involvement	
Oral aphthae	
Oral aphthae (ever), n/N (%)	30/81 (37.0)
Oral aphthae (current), n/N (%)	4/81 (4.9)
Cutaneous lupus	
Cutaneous lupus (ever), n/N (%)	60/81 (74.1)
Cutaneous lupus (current), n/N (%)	1/81 (1.2)
Lupus nephritis	11/81 (13.6)
Lupus nephritis class II, n/N (%)	1/9 (11.1)
Lupus nephritis class III+V, n/N (%)	1/9 (11.1)
Lupus nephritis class IV, n/N (%)	3/9 (33.3)
Lupus nephritis class V, n/N (%)	4/9 (44.4)
Lupus nephritis, no kidney biopsy, n/N (%)	2/72 (2.8)
Central nervous system involvement, n/N (%)	4/81 (4.9)
Seizures, n/N (%)	2/81 (2.5)
Stroke, n/N (%)	1/81 (1.2)
Not characterised, n/N (%)	1/81 (1.2)

continues on the next page

Table I. Continuation

Variable	Data
Serological data	
ANA positivity, n/N (%)	81/81 (100)
ANA titer	
1:80, n/N (%)	10/81 (12.3)
1:160, n/N (%)	18/81 (22.2)
1:320, n/N (%)	23/81 (28.4)
1:640 or higher, n/N (%)	30/81 (37.0)
ANA pattern	
Homogeneous, n/N (%)	33/81 (40.7)
Speckled, n/N (%)	42/81 (51.9)
Nucleolar, n/N (%)	3/81 (3.7)
Others, n/N (%)	3/81 (3.7)
Autoantibody specificity	
Anti-Sm, n/N (%)	10/81 (12.3)
Anti-Ro, n/N (%)	40/81 (49.4)
Anti-La, n/N (%)	11/81 (13.6)
Anti-dsDNA (ever) , n/N (%)	23/81 (28.4)
Anti-dsDNA (current) , n/N (%)	7/81 (8.6)
Anti-cardiolipin, n/N (%)	19/81 (23.5)
Anti-cardiolipin IgG, n/N (%)	15/81 (18.5)
Anti-cardiolipin IgM, n/N (%)	7/81 (8.6)
Anti-beta-2-glycoprotein I, n/N (%)	12/81 (14.8)
Anti-beta-2-glycoprotein I IgG, n/N (%)	9/81 (11.1)
Anti-beta-2-glycoprotein I IgM, n/N (%)	3/81 (3.7)
Lupus anti-coagulant, n/N (%)	6/81 (7.4)
Anti-c1q, n/N (%)	2/39 (5.1)
Anti-ribosomal phosphoprotein, n/N (%)	2/27 (7.4)
Other serological tests	
Direct Coombs test	
Direct Coombs test (ever), n/N (%)	11/67 (16.4)
Direct Coombs test (current), n/N (%)	1/67 (1.5)
Leukopenia (<3.000/mm3)	
Leukopenia (ever), n/N (%)	15/81 (18.5)
Leukopenia (current), n/N (%)	5/81 (6.2)
Thrombocytopaenia (<100.000/mm3)	
Thrombocytopaenia (ever), n/N (%)	5/81 (6.2)
Thrombocytopaenia (current), n/N (%)	0/81 (0)
Haemolytic anaemia	
Haemolytic anaemia (ever), n/N (%)	2/81 (2.5)
Haemolytic anaemia (current), n/N (%)	0/81 (0)
Hypocomplemmentaemia	
Hypocomplemmentaemia (ever), n/N (%)	52/81 (64.2)
Hypocomplemmentaemia (current), n/N (%)	25/81 (30.9)

continues on the next page

Table I. Continuation

Variable	Data
Classification criteria	
SLICC criteria, n/N (%)	65/80 (81.3)
2019 EULAR/ACR criteria, n/N (%)	61/81 (75.3)
Disease activity scores	
SLEDAI-2K	
SLEDAI-2K (disease onset), median (IQR)/N	7.0 (5.0-8.0)/80
SLEDAI-2K (maximum score), median (IQR)/N	7.0 (5.0-8.0)/80
SLEDAI-2K (last follow-up), median (IQR)/N	0.0 (0.0-2.0)/80
SLEDAI-2K (last follow-up) =0 (remission), n/N (%)	36/80 (45.0)
SLEDAI-2K (last follow-up) 1 to 4 (LDA), n/N (%)	44/80 (55.0)
SLEDAI-2K (last follow-up) > 4, n/N (%)	0/80 (0.0)
SLE-DAS	
SLE-DAS (disease onset), median (IQR)/N	8.00 (5.58-1.59)/79
SLE-DAS (maximum score), median (IQR)/N	8.00 (5.59-1.72)/79
SLE-DAS (last follow-up), median (IQR)/N	0.37 (0.37-1.12)/79
SLE-DAS (last follow-up) ≤ 2.08 (remission), n/N (%)	75/79 (95.0)
SLE-DAS (last follow-up) 2.09 to 7.64 (LDA), n/N (%)	4/79 (5.0)
SLE-DAS (last follow-up) > 7.64, n/N (%)	0/79 (0.0)
Treatments	
Methotrexate	
Methotrexate (ever), n/N (%)	10/77 (13.0)
Methotrexate (current), n/N (%)	0/77 (0)
Azathioprine	
Azathioprine (ever), n/N (%)	8/81 (9.9)
Azathioprine (current), n/N (%)	0/81 (0)
Cyclophosphamide	
Cyclophosphamide (ever), n/N (%)	9/77 (11.7)
Cyclophosphamide (current), n/N (%)	0/77 (0)
Cyclosporine	
Cyclosporine (ever), n/N (%)	2/77 (2.6)
Cyclosporine (current), n/N (%)	0/77 (0)
Mycophenolate mofetil	
Mycophenolate mofetil (ever), n/N (%)	5/77 (6.5)
Mycophenolate mofetil (current), n/N (%)	0/77 (0)
Belimumab	
Belimumab (ever), n/N (%)	0/77 (0)
Belimumab (current), n/N (%)	0/77 (0)
Tocilizumab	
Tocilizumab (ever), n/N (%)	1/81 (1.2)
Tocilizumab (current), n/N (%)	0/81 (0)
Rituximab	
Rituximab (ever), n/N (%)	2/81 (2.5)
Rituximab (current), n/N (%)	0/81 (0)

continues on the next page

Table I. Continuation

Variable	Data
Plasmapheresis	
Plasmapheresis (ever), n/N (%)	2/81 (2.5)
Plasmapheresis (current), n/N (%)	0/81 (0)
Intravenous immunoglobulin	
Intravenous immunoglobulin (ever), n/N (%)	2/81 (2.5)
Intravenous immunoglobulin (current), n/N (%)	0/81 (0)
Oral anticoagulants	
Oral anticoagulants (ever), n/N (%)	8/77 (10.4)
Oral anticoagulants (current), n/N (%)	8/77 (10.4)
Prednisolone	
Prednisolone (ever), n/N (%)	60/79 (75.9)
Prednisolone (current), n/N (%)	28/79 (35.4)
Prednisolone maximum dose (mg/day), median (IQR)/N	10.0 (5.0-25.0)/57
Prednisolone current dose (mg/day), median (IQR)/N	0.0 (0.0-1.0)/78
Hydroxychloroquine	
Hydroxychloroquine (ever), n/N (%)	80/80 (100)
Hydroxychloroquine (current), n/N (%)	80/80 (100)
Hydroxychloroquine current dose (mg/day), median (IQR)/N	200 (200-200)/79
Hydroxychloroquine treatment duration (months), median (IQR)/N	163 (87-253)/80

Abbreviations: ACR – American College of Rheumatology, ANA – antinuclear antibodies, EULAR – European Alliance of Associations for Rheumatology, dsDNA – double-stranded deoxyribonucleic acid, IQR – interquartile range, LDA – low disease activity, mg – milligrams, n – number of patients positive for the variable of interest, N – number of patients without missing information regarding the variable of interest, SLEDAI-2K – Systemic Lupus Erythematosus Disease Activity Index 2000, SLE-DAS – Systemic Lupus Erythematosus Disease Activity Score, SLICC – Systemic Lupus Erythematosus International Collaborating Clinics.

081 - ANTI-NOR90 ANTIBODIES IN THE SETTING OF CONNECTIVE TISSUE DISEASE: CLINICAL SIGNIFICANCE AND COMPARISON WITH A COHORT OF SYSTEMIC SCLEROSIS PATIENTS

Alessandro Biglia[1], Eduardo Dourado[2, 3], Boaz Palterer[4], Paola Parronchi[4], Maria Rosa Pellico[5], Giovanni Zanframundo[1], Pedro Abisay Rivera Matias[6], Patrícia Martins[2, 3], Ana Miranda[7], JE Fonseca[2, 3], Ester Di Agosta[4], Daniele Cammelli[4], Giacomo Emmi[4], Elisabetta Rosi[4], Riccardo Bixio[8], Edoardo Conticini[9], Elisa Bellis[1], Cosimo Bruni[4], Carlomaurizio Montecucco[1], Marco Matucci[4], Jorge Rojas-Serrano[6], Lorenzo Cavagna[1]

[1]Rheumatology Unit, University and IRCCS Policlinico S. Matteo Foundation, Pavia, Italy, [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]Department of Clinical and Experimental Medicine, University of Florence, Firenze, Italy, [5]Dermatology Unit, University and IRCCS Policlinico S. Matteo Foundation, Pavia, Italy, [6]Unidad de Enfermedades del Intersticio Pulmonar y Reumatologia, Istituto Nacional de Enfermedades Respiratorias Ismael Cosiò Villegas, Mexico City, Mexico, [7]Clinical Pathology Department, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [8]Rheumatology Section, Department of Medicine, Policlinico G.B. Rossi, University of Verona, Verona, Italy, [9]Rheumatology Unit, Department of Medicine, Surgery and Neurosciences, University of Siena, Firenze, Italy

Background: Anti-NOR90 antibodies are directed against a 90-kD nucleolar protein located in the nucleolus organising regions (NORs) and have mainly been described in systemic sclerosis (SSc) patients. However, anti-NOR90 antibodies have also been reported in other rheumatic and neoplastic diseases. The clinical correlates of anti-NOR90 antibodies are still undefined due to the rarity of this autoantibody and the very small size of previously reported cohorts.

Objective: We aimed to describe the characteristics of a multicentric international cohort of anti-NOR90-positive patients and compare them with a matched cohort of anti-NOR90-negative SSc patients.

Methods: We performed a case-control study, including in the interest group patients positive for anti-NOR90 antibodies referring to the six participating centres from three different countries. The control group was composed of SSc patients negative for anti-NOR90 antibodies randomly chosen from our cohort of SSc patients with regular follow-ups. The primary diagnosis, demographical and clinical data were retrospectively collected for all cases (cases and controls) from clinical files reviewed in each participating centre. The EUROLINE Systemic Sclerosis Profile kit from Euroimmun (Lübeck, Germany) was used to detect anti-NOR90 antibodies. The frequency of each clinical manifestation was compared between cases and controls using the Chi-square test.

Results: We included 101 patients positive for anti-NOR90 antibodies and 242 controls. Most anti-NOR90-positive patients were females (N=88/101, 87.1%), with a mean age of 52.5 years. The primary diagnosis of anti-NOR90-positive patients was SSc in 38/101 cases (38%), undifferentiated connective tissue disease (UCTD) in 21/101 (21%), interstitial pneumonia with autoimmune features (IPAF) in 11/101 (11%), systemic lupus erythematosus (SLE) in 8/101 (8%), and less frequently rheumatoid arthritis (N=5/101, 5%), Sjögren's syndrome (N=3/101, 3%) and other rheumatic diseases (N=11/101, 11%). The most frequent clinical manifestations were arthralgia (N=72/101, 71%), Raynaud's phenomenon (RP, N=58/101, 57%), sicca syndrome (N=49/101, 49%), interstitial lung disease (ILD, N=40/101, 40%), puffy fingers (N=32/101, 32%), arthritis (N=30/101, 30%), and skin fibrosis (N=24/101, 24%). Anti-NOR90 antibodies were co-positive with other autoantibodies in 53/101 cases (53%), mostly anti-Ro52 (21%), rheumatoid factor (13%), anti-RNP (11%), anti-dsDNA (10%), anti-centromere (9%) and anti-Scl70 (7%).

Compared with the matched controls, anti-NOR90-positive patients more frequently had arthritis (30% vs 11%, p<0.01) and sicca syndrome (49% vs 19%, p<0.01) and less frequently had RP (58% vs 89%, p<0.01), scleroderma pattern on nail fold capillaroscopy (27% vs 81%, p<0.01), digital ulcers (7% vs 23%, p<0.01), pitting scars (5% vs 25%, p<0.01), telangiectasia (11% vs 27%, p<0.01), and dysphagia (12% vs 26%, p<0.01, Table 1). There were no significant differences in the prevalence of ILD or pulmonary arterial hypertension.

Discussion/Conclusions: In our cohort, anti-NOR90 antibodies were found in a heterogeneous population of connective tissue disease patients. Most patients had co-positivity of anti-NOR90 and other autoantibodies, including SSc-specific antibodies. Most patients positive for anti-NOR90 antibodies had arthralgia and RP, common SSc manifestations. However, these patients

Table I. Clinical manifestations in the interest and control groups.

Clinical manifestation	Anti-NOR90 positive (N=101)	Anti-NOR90 negative	χ^2	Descriptive statistic
Skin fibrosis	24 (24%)	123 (51%)	21,3	<0,001
Puffy fingers	32 (32%)	60 (37%)	1,72	0,189
RP	58 (58%)	207 (89%)	43,8	<0,001
Scl pattern (NVC)	27 (27%)	140 (81%)	65,1	<0,001
Teleangiectasia	11 (11%)	60 (27%)	10,5	0,001
Pitting scars	5 (5%)	50 (25%)	18,1	<0,001
Acral ulcers	7 (7%)	48 (23%)	11,5	<0,001
Arthritis	30 (30%)	22 (11%)	16,9	<0,001
ILD	40 (40%)	95 (39%)	0,004	0,952
PAH	8 (8%)	34 (19%)	6,48	0,011
Fibromyalgia	8 (8%)	34 (21%)	7,47	0,006
Dysphagia	12 (12%)	59 (26%)	6,78	0,009
Sicca syndrome	49 (49%)	35 (19%)	26,4	<0,001

Abbreviations: ILD – interstitial lung disease, NVC – nail fold video capillaroscopy, PAH – pulmonary arterial hypertension, RP – Raynaud's phenomenon, Scl pattern – scleroderma pattern.

more frequently had arthritis and sicca syndrome and less frequently had vascular and gastrointestinal manifestations than SSc patients negative for this autoantibody.

083 - THE LONG-TERM IMPACT OF FATIGUE ON DISEASE ACTIVITY IN PATIENTS WITH RHEUMATOID ARTHRITIS TREATED WITH BIOLOGICAL DISEASE-MODIFYING ANTIRHEUMATIC DRUGS

Daniela Santos Oliveira[1, 2, 3], Ana Martins[2, 3], Frederico Rajão Martins[4], Beatriz Samões[5], Maria Seabra Rato[2, 3], Filipe Oliveira Pinheiro[2, 3], Rafaela Nicolau[6], Carlos Vaz[1, 2, 3], Miguel Bernardes[2, 3], Lúcia Costa[3]

[1]Center for Health Technology and Services Research (CINTESIS),, Faculdade de Medicina da Universidade do Porto, Porto, Portugal, Porto, Portugal, [2]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal, [3]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [4]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [5]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [6]Rheumatology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Background: Fatigue is common among patients with rheumatoid arthritis (RA) with a substantial impact on quality of life (1). Biological disease-modifying antirheumatic drugs (bDMARDs) have been shown to significantly improve fatigue in these patients. However, fatigue is under-assessed by the physicians and evidence is still scarce regarding a possible impact of fatigue on disease activity over time.

Objectives: To explore the long-term impact of fatigue on the disease activity in patients with RA treated with bDMARDs.

Methods: A monocentric observational retrospective cohort study was conducted with 24 months (M) of follow-up. Patients diagnosed with RA, according to the 2010 American College of Rheumatology (ACR)/ European League Against Rheumatism (EULAR) criteria, and registered on the Rheumatic Diseases Portuguese Register (Reuma.pt) who started their first bDMARD between 2015 and 2021 were included. Demographic, clinical and laboratory data were obtained by consulting Reuma.pt. Fatigue was monitored at baseline, 12 and 24M using Functional Assessment of Chronic Illness Therapy-Fatigue (FACIT-F). This scale is a 13-item self-reported questionnaire with a total score ranging from 0 to 52. A score ≤ 39 indicates the presence of clinically significant fatigue. Disease Activity Score for 28 joints with erythrocyte sedimentation rate (DAS28) or with C-reactive Protein [(DAS28(CRP)], DAS28 delta, Clinical Disease Activity Index (CDAI) and Simplified Disease Activity Index (SDAI) were calculated to measure disease activity. Clinical response was evaluated by EULAR criteria in three response categories- good, mild and no response- and by ACR criteria (0 to 100%) at 12 and 24M. To correlate fatigue score with EULAR clinical response Chi-square test was used. Multivariate linear regression models adjusted for age, gender and disease duration were used to assess the effect of fatigue on disease activity and ACR clinical response over time.

Table I. Baseline characteristics of sample

Variable	Descriptive statistic
Treatment options	n (%)
Glucocorticoids	37 (92.5)
Conventional DMARDs	37 (92.5)
Anti TNF	19 (47.5)
Etanercept	13 (32.5)
Rituximab	4 (10)
Tocilizumab	4 (10)
Disease activity score	Mean ±SD
DAS-28	5.4±0.87
DAS-28(CRP)	4.9±0.69
CDAI	27.6±8.5
SDAI	28.9±8.7

DMARDs: disease-modifying antirheumatic drugs; DAS-28: Disease Activity Score for 28 joints; CRP: C-reactive Protein; CDAI: Clinical Disease Activity Index; SDAI: Simplified Disease Activity Index.

Results: A total of 40 patients with RA were included, with a mean age of 47.4±11.4 years old and disease duration of 10.4±5.6 years. Most patients were female (90.2%). Rheumatoid Factor was positive in 70% of patients. The majority of patients (85%) had clinically significant fatigue at baseline moment (FACIT-F 26.9±11.8). Treatment characteristics and baseline disease activity scores are described in Table 1. Fatigue at baseline moment predicted DAS28(CRP) (β=-0.061, 95%CI [-0.12; -0.003]) and CDAI (β=- 0.30, 95%CI [-0.57; -0.029]) at 12M. Additionally, fatigue predicted SDAI (β=-0.38, 95%CI [-0.72; -0.047]) and CDAI (β=-0.39, 95%CI [-0.73; -0.051]) at 24M. In general, for these models, fatigue as a symptom was shown to have negative effects on the different outcomes analysed. Fatigue did not associate with EULAR and ACR responses over time. **Conclusions:** These findings showed that, in our sample, the most RA patients had severe fatigue and that its presence may be a predictor of increased disease activity. Indeed, previous research observed a positive association between fatigue and disease activity over the time in patients with RA (2). Therefore, fatigue

should be regularly monitored in patients with RA and its impact on treatment must be considered. Moreover, further research with larger samples is needed to explore the impact of fatigue on clinical response and the potential of fatigue relief as an outcome measure of RA treatment.

REFERENCES:

1. Katz P. Curr Rheumatol Rep. 2017;19(5):25; 2. Holten K, et al. Ann. Rheum. Dis. 2021; 0:1–7.

085 - THE USE OF ABATACEPT TO MANAGE RHEUMATOID ARTHRITIS: A REAL-WORLD STUDY FROM A SINGLE NATIONAL REGISTER

Daniela Santos Oliveira[1, 2, 3], Ana Martins[1, 3], Rafaela Nicolau[4], Maria Seabra Rato[1, 3], Filipe Oliveira Pinheiro[1, 3], Bruna Silva[1], Carlos Vaz[1, 2, 3], Miguel Bernardes[1, 3], Lúcia Costa[1]

[1]Rheumatology Department, 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, [3]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal, [4]Rheumatology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Introduction: Abatacept is a biologic Disease Modifying Anti-Rheumatic Drug (bDMARD) approved for rheumatoid arthritis (RA) that works by selectively inhibiting T cell co-stimulation signals. However, there are limited data on the real-world effectiveness and safety of abatacept, namely derived from a national register.

Objectives: To characterize the effectiveness and safety of abatacept in patients with RA over 2 years of follow-up. **Methods:** A monocentric observational retrospective cohort study was conducted. Patients diagnosed with RA, according to the 2010 American College of Rheumatology (ACR)/European League Against Rheumatism (EULAR) criteria, treated currently with abatacept and registered on the Rheumatic Diseases Portuguese Register (Reuma.pt) were included. Demographic, clinical and laboratory data were obtained at baseline, 6, 12 and 24 months (M) by consulting Reuma.pt. Disease Activity Score for 28 joints with erythrocyte sedimentation rate (DAS28) or with C-reactive Protein [(DAS28(CRP)] as well as the change in DAS28 were calculated to measure disease activity. Clinical effectiveness of abatacept was translated by a moderate or good EULAR response. Remission was defined by DAS28<2.6 and low disease activity by DAS28 ≤ 3.2. Safety was evaluated by the number of adverse events.

Table I. Sample baseline clinical characteristics and disease activity score over time

Sample characteristics	Descriptive statistic (N=16)
Baseline clinical characteristics	n (%)
Rheumatoid factor positive	9 (56.3)
Anti-cyclic citrullinated peptide antibody positive	14 (87.5)
Erosive disease	8 (50)
Extra-articular manifestations:	13 (32.5)
Sjogren syndrome	4 (10)
Rheumatoid nodules	4 (10)
Interstitial pulmonary disease	Mean ±SD
Caplan syndrome	4 (25)
Methotrexate	5 (31.3)
Other csDMARD	11 (68.8)
Glucocorticoid	13 (81.3)
Patient reported outcome	Mean ±SD
HAQ at baseline moment	1.8±0.4
HAQ 6M	1.6±0.8
HAQ 12M	1.4±0.9
HAQ 24M	1.9±0.8
Disease activity scores	Mean ±SD
DAS-28 at Baseline moment	4.5±1.4
DAS-28 at 6M	3.8±1.3
DAS-28 at 12M	4.2±1.5
DAS-28 at 24M	4±1.2
DAS-28(CRP) at baseline moment	4.0±1.2
DAS-28(CRP) at 6M	3.6±1.2
DAS-28(CRP) at 12M	3.6±1.6
DAS-28(CRP) at 24M	4.0±1.0

csDMARD: conventional synthetic DMARD; HAQ: Health Assessment Questionnaire; SD: standard deviation; M: months; DAS-28: Disease Activity Score for 28 joints; CRP: C-reactive Protein.

Results: A total of 16 patients with RA currently treated with abatacept were included, with a mean age of 58.7±11.4 years old and disease duration of 20.6±9.9 years at the beginning of abatacept. Most patients were female (n=14, 87.5%). In the majority of patients (n=12, 75%) abatacept was not the first biologic drug. Sample baseline clinical characteristics and disease activity scores at baseline moment, 6M, 12M and 24M (mean ± standard deviation) are described in Table 1. Twenty-three percent of the patients achieved a EULAR good response at 6M, 16.7% at 12M and 18.2% at 24M. The proportions of patients with a EULAR good or moderate response were 76.9%, 50% and 54.5% at 6, 12 and 24M, respectively. Remission was achieved

in 6.3%, 18.8% and 12.5% at 6, 12 and 24M of abatacept treatment, respectively. Low disease activity was achieved in 18.8%, 25% and 6.3%, respectively. No adverse events were reported during 2 years of follow-up.
Conclusions: Our study confirmed that in our centre abatacept is an effective and safe option to reduce disease activity in patients with active RA. Multicentric studies with larger samples are needed to explore these results.

086 - THE LONG-TERM PREDICTORS OF FATIGUE IN PATIENTS WITH RHEUMATOID ARTHRITIS TREATED BIOLOGICAL DISEASE-MODIFYING ANTIRHEUMATIC DRUGS

Daniela Santos Oliveira[1, 2, 3], Ana Martins[1, 3], Beatriz Samões[4], Frederico Rajão Martins[5], Filipe Oliveira Pinheiro[1, 3], Maria Seabra Rato[1, 3], Rafaela Nicolau[6], Carlos Vaz[1, 2, 3], Miguel Bernardes[1, 3], Lúcia Costa[1]
[1]Rheumatology Department, 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, [3]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal, [4]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/ Espinho, Vila Nova de Gaia, Portugal, [5]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [6]Rheumatology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Background: Fatigue is one of the major complaints of patients with rheumatoid arthritis (RA). However, the literature relating fatigue to other disease-related parameters has presented discrepant results and longitudinal studies with multivariate analyses are scarce.
Objectives: To explore potential associations between fatigue and demographic variables and other patient-reported outcomes (PROMs) in patients with RA treated with biological disease-modifying antirheumatic drugs (bDMARDs) over the time.
Methods: A 24-month (24M) monocentric observational retrospective cohort study was conducted. Patients diagnosed with RA, according to the 2010 American College of Rheumatology (ACR)/ European League Against Rheumatism (EULAR) criteria, and registered on the Rheumatic Diseases Portuguese Register (Reuma.pt) who started their first bDMARD between 2015 and 2021 were included. Age, gender, disease duration, body mass index (BMI) and PROMs were obtained by consulting Reuma.pt. Fatigue was monitored at baseline, 12 and 24M using Functional Assessment of Chronic Illness Therapy-Fatigue (FACIT-F), a 13-item questionnaire with a total score ranging from 0 to 52. A score ≤ 39 indicates the presence of clinically significant fatigue. Health Assessment Questionnaire (HAQ), EuroQol-5D (EQ-5D), 36-Item Short Form Survey (SF-36), patient global assessment visual analogue scale (VAS) and pain VAS were assessed at baseline, 12 and 24M. Multivariate linear regression models were conducted with FACIT-F as the dependent variable.
Results: A total of 40 patients (47.4±11.4 years; 90.2% female) with a BMI of 29.87±8.54 kg/m² and a mean disease duration of 10.4±5.6 years were included. A total of 47.5% of patients were treated with an anti TNF. About 85% of patients had clinically significant fatigue at baseline (FACIT-F 26.9±11.8). At baseline, patient global assessment VAS (β=-0.4, 95%CI [- 0.68; -0.095]) and pain VAS (β=- 0.34, 95%CI [-0.6; -0.068]) predicted fatigue. SF-36 predicted fatigue at baseline (β=0.35, 95%CI [0.14;0.56]), 12M (β=0.23, 95%CI [0.084;0.37]) and 24M (β=0.26, 95%CI [0.13;0.39]). HAQ predicted fatigue at baseline (β=- 12.2, 95%CI [- 19.8; - 4.5]) and 24M (β=- 11.4, 95%CI [- 17.47; - 5.38]). EQ-5D (β=39.5, 95%CI [15.84; 63.22]) predicted fatigue at 24M.
Conclusions: Our results showed that pain levels and patient global assessment of disease activity predicted a higher level of fatigue at baseline. The decrease in physical function and a worse overall health status perceived by patient predicted higher fatigue over the time. Previous research has suggested that disease-related factors, such as inflammation, pain or decreased physical function are associated with greater fatigue in RA (1). These findings encourage the pre-treatment screening of fatigue in patients with RA in order to design individualized non-pharmacological approaches in addition to bDMARDs therapy.

REFERENCES:

1. Katz P. Curr Rheumatol Rep. 2017;19(5):25.

089 - THE PHENOTYPE AND PROGNOSIS OF ANTI-NEUTROPHIL CYTOPLASMATIC ANTIBODY-ASSOCIATED VASCULITIS

Mariana Santos[1], Maria João Gonçalves[1], Maria Manuela Costa[1], Jaime C. Branco[1, 2], Alexandre Sepriano[1, 2]
[1]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [2]CEDOC, NOVA Medical School, Lisboa, Portugal

Aim: The 'umbrella-term', anti-neutrophil cytoplasmatic antibody (ANCA)-associated vasculitis (AAV) is used to describe a group of 3 rare diseases: granulomatous with polyangiitis (GPA), eosinophilic granulomatous with polyangiitis (EGPA) and microscopic polyangiitis (MPA). All AAVs cause inflammation of small vessels,

Table I. Phenotypes of Anti-neutrophil cytoplasmatic antibody-associated vasculitis

	All (n=19)	GPA (n=7)	EGPA (n=5)	MPA (n=2)	UAAV (n=5)
Female, n (%)	14 (74)	7 (100)	2 (40)	1 (50)	4 (80)
Age at symptom onset, median (IQR)	54 (50)	43 (33)	54 (36)	59 (5)	70 (38)
Age at diagnosis, median (IQR)	55 (49)	44 (32)	53 (32)	61 (0)	70 (20)
Diagnostic delay, median, years (IQR)	1 (37)	1 (2)	0.6 (37)	3 (6)	0.2 (26)
Follow-up, median, years (IQR)	4 (18)	12 (14)	1 (18)	10 (12)	2 (1)
Hospitalizations at diagnosis, n (%)					
GCU	4 (27)	2 (50)	4 (80)	1 (50)	1 (35)
ICU	11 (73)	2 (50)	1 (20)	1 (50)	3 (75)
Total	15 (79)	4 (57)	5 (100)	2 (100)	4 (80)
ANCAs at diagnosis, n (%)					
Negative	6 (32)	4 (57)	2 (40)	0 (0)	0 (0)
c-ANCA + anti-PR3	5 (38)	3 (100)	0 (0)	1 (50)	1 (20)
c-ANCA + anti-MPO	2 (15)	0 (0)	1 (33)	0 (0)	1 (20)
p-ANCA + anti-PR3	2 (15)	0 (0)	1 (33)	1 (50)	1 (20)
p-ANCA + anti-MPO	4 (31)	0 (0)	1 (33)	0 (0)	2 (40)
Main organ affected at onset[1], n (%)					
ENT	4 (21)	4 (57)	0 (0)	0 (0)	0 (0)
Lungs	9 (47)	2 (29)	2 (40)	2 (100)	3 (60)
Skin	1 (5)	0 (0)	0 (0)	0 (0)	1 (20)
Kidney	0 (0)	0 (0)	0 (0)	0 (0)	0 (0)
Nervous system	2 (11)	0 (0)	1 (20)	0 (0)	1 (20)
Systemic[2]	2 (11)	1 (14)	1 (20)	0 (0)	0 (0)
Cardiovascular	1 (5)	0 (0)	1 (20)	0 (0)	0 (0)
Others[3]	0 (0)	0 (0)	0 (0)	0 (0)	0 (0)
Organs affected during follow-up4, n (%)					
ENT	13 (68)	7 (100)	4 (80)	1 (50)	1 (20)
Lungs	15 (79)	4 (57)	5 (100)	2 (100)	4 (80)
Skin	10 (53)	1 (14)	3 (60)	1 (50)	5 (100)
Kidney	10 (53)	4 (57)	1 (20)	1 (50)	4 (80)
Nervous system	6 (32)	1 (14)	3 (60)	0 (0)	2 (40)
Systemic2	18 (95)	6 (86)	4 (80)	2 (100)	5 (100)
Cardiovascular	3 (16)	1 (14)	1 (20)	1 (50)	0 (0)
Others3	3 (16)	1 (14)	1 (20)	0 (0)	1 (20)
Flares[5], n (%)	11 (58)	4 (57)	2 (40)	2 (100)	3 (60)
Irreversible organ damage, n (%)	5 (26)	3 (29)*	1 (20)†	1 (20)‡	0 (0)
VDI[6], median (IQR)	1 (4)	1 (3)	1 (2)	2.5 (3)	2 (4)
FFS[6], median (IQR)	1 (3)	0 (1)	1 (1)	2 (2)	2 (2)
Mortality, n (%)	2 (11)	0 (0)	1 (20)Ω	1 (50)¤	0 (0)

GPA, granulomatous with polyangiitis; EGPA, eosinophilic granulomatous with polyangiitis; MPA, microscopic polyangiitis; UAAV, undifferentiated AAV; IQR, interquartile range; ENT, ear, nose and throat; GCU, general care unit; ICU, intensive care unit; ANCA, anti-neutrophil cytoplasmatic antibody; VDI, vasculitis damage index; FFS, five factor score; 1 Organ involvement categories were defined according to the Birmingham Vasculitis Activity Score (BVAS) divisions.; 2 Systemic involvement: myalgia, arthralgia/arthritis, fever ≥38° C, weight loss ≥2 kg; 3 Other categories considered in BVAS but with less expression in our patients, including mucous membranes/eyes and abdominal involvements; 4 Including organs involved at disease onset; 5 New or aggravated symptoms requiring therapeutic intervention. 6 Calculated accordingly with data of last evaluation; *Deafness, tracheostomy because of subglottic stenosis and saddle nose; † Heart failure; ‡ Renal failure with kidney substitution therapy; Ω Related with terminal chronic kidney disease; ¤ Subsequent to cerebral hemorrhage with no clear AAV association

but their presentation can differ resulting in diagnostic and prognostic differences. We aimed at comparing the phenotype and prognosis of the AAVs.
Methods: Retrospective cohort study of all patients with the diagnosis of AAV according to their treating rheumatologist followed in the rheumatology department of a tertiary academic centre, from 2004 until April 2022. Demographic, clinical, histologic and analytical features were retrieved from electronic medical records and described overall and separately per each AAV.
Results: In total, 19 patients followed over a median of 4 years, were included. Seven patients (37%) were diagnosed as GPA, 5 (26%) as EGPA and 2 (11%) as MPA. Five (26%) patients were not considered to fit any of the 3 diagnosis and were therefore labelled as undifferentiated AAV (UAAV). Patients with GPA were more often female (100%) and younger than those of the other groups (table 1). Diagnostic delay was higher for MPA and GPA. The majority (79%) of the patients were hospitalized at diagnosis, including all patients with EGPA and MPA.
Biopsy was performed in 18 patients (95%) and histology was compatible with vasculitis in 10 (56%) of these patients, including 3 with GPA, 4 with EGPA and 3 with UAAV. ANCAs were negative in 6 (32%) patients at the time of diagnosis, and 3 of these (all GPA) were re-evaluated during follow-up, remained negative. At diagnosis, the two most common ANCA patterns were c-ANCA with anti-PR3 (38%) and p-ANCA with anti-MPO (31%). All seropositive GPA patients were positive for c-ANCA with anti-PR3, while no clear pattern was found for the others AAV. Of the 13 initially positive patients reassessed during follow-up, 12 had lower and 1 the same titles of ANCA.
The most common clinical feature at diagnosis was lung involvement (47%), which was present in all AAV subtypes. Ear, nose and throat (ENT) involvement was the leading presenting manifestation in GPA (57%), while EGPA had a more heterogeneous phenotype. During follow-up, all patients had multiorgan involvement. All patients with GPA eventually developed ENT symptoms (but no cases of localized GPA were observed), which was also common in EGPA (80%). However, in the latter, lung (100%) and nervous system involvement (60%) were the dominant clinical features. Lung and kidney disease was the hallmark of the MPA patients. The median values of the vasculitis damage index and of the five-factor score are shown in table 1. Irreversible damage of the ENT was the most common complication, exclusively in GPA. The EGPA patient with cardiac involvement at presentation developed heart failure. Only one patient with MPA, out of 10 patients with kidney disease, developed terminal chronic kidney disease (TCKD), having been the cause of death.
Conclusions: We found important differences in the phenotype and prognosis of patients with AAV. GPA mostly affects and often leads to irreversible damage of the upper respiratory system. EGPA affects both the upper and lower respiratory system and with cardiac involvement leading to worse prognosis in this group. MPA is the least common AAV in this cohort and the only in which a patient developed TCKD.

098 - RHEUMATOID FACTOR IS STRONGLY ASSOCIATED WITH PULMONARY INVOLVEMENT IN PATIENTS WITH PRIMARY SJÖGREN'S SYNDROME

Matilde Bandeira[1, 2], Manuel Silvério-António[1, 2], Nikita Khmelinskii[1, 2], JE Fonseca[1, 2], Vasco C Romão[1, 2]
[1]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Background:Purpose: Sjögren's syndrome (SS) is a systemic autoimmune disease targeting mainly exocrine glands clinically characterized by ocular and oral dryness. Systemic extra-glandular involvement is present in one third of patients. It is of major importance to uncover easily available tools to predict disease phenotypes and outcomes. Rheumatoid factor (RF) has previously been correlated to higher disease activity as measured by EULAR Sjögren's syndrome disease activity index (ESSDAI). This work aims to understand whether RF can be a predictor for specific organ involvement in SS.
Methods: Retrospective cohort study including SS patients followed in our center until March 2022, who fulfilled 2016 ACR/EULAR classification criteria for SS and had available RF. Multiorgan involvement was defined according to ESSDAI definitions. Univariate analysis was performed using chi-square, Kruskal-Wallis or ANOVA tests, as appropriate. Independent predictors of systemic involvement were identified through binomial logistic regression modelling.
Results: 209 patients were included; 96.7% females (n=202), with a mean age at diagnosis of 51.38± 14.95 years. RF-positive patients (Table 1) had more frequently pulmonary involvement [14/110 (12.7%) vs 3/99 (3.0%), p=0.011] and biological involvement [71/110 (64.5%) vs 42/99 (42.4%), p=0.001], and were more frequently positive for anti-SSB antibody [74/110 (67.3%) vs 39/99 (39.4%), p<0.001]. RF-positive patients also had a higher focus score mean when compared to RF-negative patients (2.88±1.68 vs

Table I. Clinical and serological characteristics of patients rheumatoid factor positive and negative

	RF-positive (n=110)	RF-negative (n=99)	Univariate analysis
Age at diagnosis, mean ± SD (N)	49.54±15.22 (110)	53.42±14.45 (99)	p=0.060
Disease duration (in years), mean ± SD (N)	9.05±7.74 (110)	8.66±7.35 (99)	p=0.704
Females, n/N (%)	107/110 (97.3)	95/99 (96.0)	p=0.710
Deceased patients, n/N (%)	3/110 (2.7)	4/99 (4.0)	p=0.710
Ocular dryness, n/N (%)	100/110 (90.9)	91/99 (91.9)	p=0.811
Oral dryness, n/N (%)	104/110 (94.5)	92/99 (92.9)	p=0.776
Schirmer's test < 5mm in 5 minutes, n/N (%)	69/91 (75.8)	57/90 (63.3)	p=0.077
Unstimulated salivary flow < 1.5mL in 15 minutes, n/N (%)	30/60 (50.0)	22/60 (36.7)	p=0.197
Positive salivary biopsy, n/N (%)	65/90 (72.2)	55/86 (64.0)	p=0.260
Focus score, mean ± SD (N)	**2.88±1.68 (40)**	**2.17±1.10 (33)**	**p=0.034**
Low C3, n/N (%)	8/106 (7.5)	12/94 (12.8)	p=0.245
Low C4, n/N (%)	11/106 (10.4)	4/94 (4.3)	p=0.115
Cryoglobulinemia, n/N (%)	4/47 (8.5)	1/18 (5.6)	p=1.000
Positive anti-SSA/Ro, n/N (%)	104/110 (94.5)	87/99 (87.9)	p=0.137
Positive anti-SSB/La, n/N (%)	**74/100 (67.3)**	**39/99 (39.4)**	**p<0.001**
Pulmonary involvement, n/N (%)	**14/110 (12.7)**	**3/99 (3.0)**	**p=0.011**
Neurologic involvement, n/N (%)	7/110 (6.4)	6/99 (6.1)	p=1.000
Hematologic involvement, n/N (%)	36/110 (32.7)	24/99 (24.2)	p=0.221
Glandular involvement, n/N (%)	37/110 (33.6)	23/99 (23.2)	p=0.125
Constitutional involvement, n/N (%)	29/110 (26.4)	17/99 (17.2)	p=0.133
Lymphadenopathic involvement, n/N (%)	20/110 (18.2)	13/99 (13.1)	p=0.347
Articular involvement, n/N (%)	52/110 (47.3)	42/99 (42.4)	p=0.490
Cutaneous involvement, n/N (%)	27/110 (24.5)	22/99 (22.2)	p=0.745
Renal involvement, n/N (%)	3/110 (2.7)	2/99 (2.0)	p=1.000
Muscular involvement, n/N (%)	2/110 (1.8)	1/99 (1.0)	p=1.000
Biological involvement, n/N (%)	**71/110 (64.5)**	**42/99 (42.4)**	**p=0.001**
Hematologic neoplasia, n/N (%)	3/110 (2.7)	6/99 (6.1)	p=0.313

Abbreviations: RF – Rheumatoid Factor; SD – Standard Deviation

Table II. Clinical and immunological characteristics of SS patients with pulmonary involvement

	Patients with lung involvement (n=18)	Patients without lung involvement throughout follow-up (n=198)	Univariate analysis
Age at diagnosis, mean ± SD (N)	53.22±13.68 (18)	51.15±14.98 (198)	p=0.774
Disease duration (in years), mean ± SD (N)	11.61±9.57 (18)	8.71±7.54 (198)	p=0.181
Females, n/N (%)	18/18 (100.0)	191/198 (96.5)	p=1.000
Deceased patients, n/N (%)	2/18 (11.1)	6/198 (3.0)	p=0.136
Ocular dryness, n/N (%)	18/18 (100.0)	180/198 (90.9)	p=0.374
Oral dryness, n/N (%)	18/18 (100.0)	185/198 (93.4)	p=0.608
Schirmer's test < 5mm in 5 minutes, n/N (%)	12/14 (85.7)	117/172 (68.0)	p=0.233
Unstimulated salivary flow < 1.5mL in 15 minutes, n/N (%)	5/11 (45.5))	47/110 (42.7)	p=1.000
Positive salivary biopsy, n/N (%)	13/17 (76.5)	109/162 (67.3)	p=0.587
Focus score, mean ± SD (N)	2.94±2.05 (6)	2.51±1.43 (68)	p=0.704
Rheumatoid factor, n/N (%)	**14/17 (82.4)**	**96/192 (50.0)**	**p=0.011**
Low C3, n/N (%)	0/16 (0.0)	21/189 (11.1)	p=0.381
Low C4, n/N (%)	0/16 (0.0)	15/189 (7.9)	p=0.613
Positive anti-SSA/Ro, n/N (%)	18/18 (100.0)	180/198 (90.9)	p=0.374
Positive anti-SSB/La, n/N (%)	12/17 (70.6)	105/198 (53.0)	p=0.208
Neurologic involvement, n/N (%)	2/18 (11.1)	12/198 (6.1)	p=0.329
Hematologic involvement, n/N (%)	4/18 (22.2)	57/198 (28.8))	p=0.785
Glandular involvement, n/N (%)	5/18 (27.8)	56/198 (28.3)	p=1.000
Constitutional involvement, n/N (%)	5/18 (27.8)	42/198 (21.2)	p=0.552
Lymphadenopathic involvement, n/N (%)	3/18 (16.7)	31/198 (15.7)	p=1.000
Articular involvement, n/N (%)	7/18 (38.9)	89/198 (44.9)	p=0.805
Cutaneous involvement, n/N (%)	5/18 (27.8)	46/198 (23.2)	p=0.772
Renal involvement, n/N (%)	1/18 (5.6)	4/198 (2.0)	p=0.356
Muscular involvement, n/N (%)	1/18 (5.6)	2/198 (1.0)	p=0.231
Biological involvement, n/N (%)	12/18 (66.7)	103/198 (52.0)	p=0.325
Hematologic neoplasia, n/N (%)	2/18 (11.1)	9/198 (4.5)	p=0.230

Abbreviations: SD – Standard Deviation

2.17±1.10, p=0.043).

In the multivariate analysis, RF was associated with the presence of anti-SSB (OR 2.491, 95%CI: 1.363-4.552, p=0.003) and pulmonary involvement (OR 4.061, 95%CI: 1.081-15.254, p=0.038), independently of sex, age at diagnosis and biological involvement.

Patients with pulmonary involvement were more likely to be RF-positive [14/17 (82.4%) vs 96/192 (50.0%), p=0.011] than those without lung disease throughout follow-up (Table 2). On multivariate analysis, RF (OR 4.387, 95%CI: 1.183-16.274, p=0.027) was an independent predictor of pulmonary involvement, accounting for sex, age at diagnosis and biological involvement.

Conclusions: In our cohort, RF-positive patients were more likely to have pulmonary and biological involvement, and were more frequently positive for anti-SSB antibody. RF was associated with both anti-SSB positivity and pulmonary involvement in SS and was identified as an independent predictor of this involvement. These results reinforce RF as a relevant prognostic factor in SS.

099 - RESPONSE TO CERTOLIZUMAB PEGOL IN PATIENTS WITH NON-RADIOGRAPHIC AXIAL SPONDYLOARTHRITIS BY BASELINE C-REACTIVE PROTEIN CUT-OFFS: POST-HOC ANALYSIS FROM A PHASE 3 MULTICENTER STUDY

Philip C. Robinson[1], Stephen Hall[2], Ana Lourenço[3], Bengt Hoepken[4], Lars Bauer[4], Eleni Demas[5], Mindy Kim[6], Atulya Deodhar[7]

[1]University of Queensland School of Clinical Medicine, Brisbane, Queensland, Australia, [2]Cabrini Medical Centre, Monash University and Emeritus Research, Melbourne, Victoria, Australia, [3]UCB Pharma, Oeiras, Portugal, [4]UCB Pharma, Monheim am Rhein, Germany, [5]UCB Pharma,

Slough, United Kingdom, [6]UCB Pharma, Smyrna, United States, [7]Oregon Health & Science University, Portland, United States

Background: Certolizumab pegol (CZP), an Fc-free, PEGylated tumour necrosis factor (TNF) inhibitor, has previously demonstrated efficacy and safety in patients with radiographic (r) and non-radiographic (nr) axial spondyloarthritis (axSpA).1,2 In patients with nr-axSpA, CZP has demonstrated efficacy across all C reactive protein (CRP) subgroups, including patients with normal baseline CRP levels.3 However, the association between CZP efficacy and baseline CRP levels has not been investigated in the subset of nr-axSpA patients with positive magnetic resonance imaging (MRI+; defined as the presence of active sacroiliitis on MRI based on the Assessment of SpondyloArthritis international Society [ASAS] criteria).4 This post-hoc analysis explores the association between baseline CRP levels and CZP efficacy in MRI+ nr-axSpA patients from the C OPTIMISE trial.

Methods: C-OPTIMISE (NCT02505542) was a two-part, multicenter, phase 3b study in adult patients with r-axSpA or nr-axSpA.1,2 In the open-label run-in period (0–48 weeks), patients received CZP 400 mg at Weeks 0, 2, and 4, then CZP 200 mg every 2 weeks thereafter. This analysis included the subset of MRI+ nr axSpA patients in the C OPTIMISE cohort who received ≥1 dose of study medication in the open-label period. Efficacy outcomes were evaluated and stratified by baseline CRP levels (<5 mg/L, ≥5–<10 mg/L and ≥10 mg/L). The upper limit of normal of the CRP assay was defined as 9.99 mg/L by the central laboratory. The lower limit of quantification (LLoQ) was 4 mg/L; where CRP levels < LLoQ, a CRP value of 2 mg/L was used to calculate Ankylosing Spondylitis Disease Activity Score (ASDAS).5 Outcomes included ASAS ≥40% improvement (ASAS40), ASDAS – major improvement (ASDAS-MI), ASAS partial remission (ASAS PR), ASDAS, BASDAI, and BASFI.

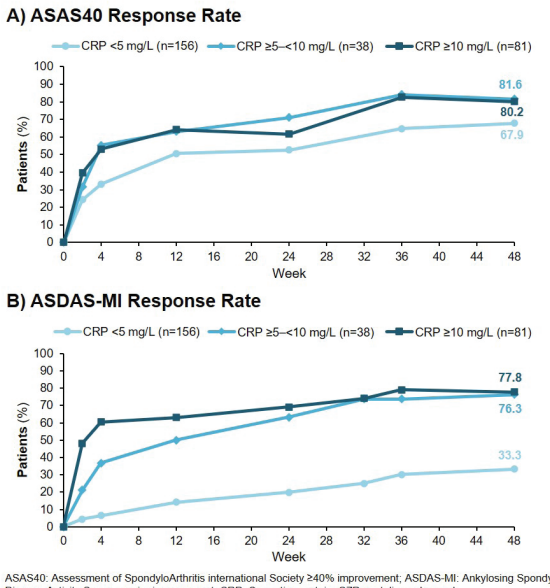
Results: In total, 275 MRI+ nr-axSpA patients were included in this analysis (CRP <5 mg/L: n=156; CRP ≥5–<10 mg/L: n=38; CRP ≥10 mg/L: n=81). Response rates for ASAS40 increased over the treatment period and were comparable across CRP subgroups (Figure 1A). Response rates for ASDAS-MI were higher in the CRP ≥10 mg/L and CRP ≥5–<10 mg/L subgroups than the CRP <5 mg/L subgroup (Figure 1B). Across other efficacy measures, improvements were observed at Week 48 compared with baseline in all CZP-treated CRP subgroups (Table 1).

Conclusions: Clinically relevant responses were observed in MRI+ nr-axSpA patients treated with CZP, across CRP subgroups and measured outcomes. The responses in each subgroup were consistent with those previously reported in total nr-axSpA patient group.1 ASDAS-MI response rates were lower in the CRP <5 mg/L subgroup, however, CRP is a key factor in ASDAS derivation.6 It is unlikely that ASDAS-MI could be achieved in patients in the CRP <5 mg/L subgroup since most of them had CRP levels < LLoQ.

Table I. Clinical responses im CZP-treated patients by baseline CRP category

		CRP <5 mg/L (n=156)	CRP ≥5–<10 mg/L (n=38)	CRP ≥10 mg/L (n=81)
ASAS40 % (n)	Baseline	–	–	–
	Week 48	**67.9** (106)	**81.6** (31)	**80.2** (65)
ASDAS-MI % (n)	Baseline	–	–	–
	Week 48	**33.3** (52)	**76.3** (29)	**77.8** (63)
ASAS PR % (n)	Baseline	–	–	–
	Week 48	**50.0** (78)	**73.7** (28)	**67.9** (55)
ASDAS Mean (SD)	Baseline	**3.1** (0.5)	**3.6** (0.5)	**4.3** (0.6)
	Week 48	**1.6** (1.0)	**1.3** (1.0)	**1.5** (1.0)
BASDAI Mean (SD)	Baseline	**6.7** (1.4)	**6.5** (1.6)	**6.9** (1.3)
	Week 48	**2.6** (2.5)	**1.6** (2.4)	**1.7** (2.2)
BASFI Mean (SD)	Baseline	**5.0** (2.0)	**4.7** (2.3)	**5.5** (2.1)
	Week 48	**1.9** (2.3)	**1.4** (2.3)	**1.3** (1.9)

ASDAS-MI, ASAS40 and ASAS PR are reported using non-responder imputation.
ASDAS, BASDAI and BASFI are reported using the last observation carried forward.
ASAS: Assessment of SpondyloArthritis international Society; ASAS40: ASAS ≥40% improvement; ASDAS-MI: Ankylosing Spondylitis Disease Activity Score – major improvement; BASDAI: Bath Ankylosing Spondylitis Disease Activity Index; BASFI: Bath Ankylosing Spondylitis Functional Index; CRP: C-reactive protein; CZP: certolizumab pegol; PR: partial remission; SD: standard deviation.



ASAS40: Assessment of SpondyloArthritis international Society ≥40% improvement; ASDAS-MI: Ankylosing Spondylitis Disease Activity Score – major improvement; CRP: C-reactive protein; CZP: certolizumab pegol.

Figure 1. (A) ASAS40 and (B) ASDAS-MI response rates in CZP-treated patients by baseline CRP category (non-responder imputation)

REFERENCES

1. Landewé R et al. Rheumatol Ther 2020;7:581–99.
2. Landewé R et al. Ann Rheum Dis 2020;79:920–28.
3. Robinson P et al. Int J Rheum Dis 2020;23:67.
4. Rudwaleit M et al. Ann Rheum Dis 2009;68:1520–7.
5. Machado P et al. Arthritis Rheumatol 2015;67:408–13.
6. Machado P et al. Ann Rheum Dis 2011;70:47–53.

ACKNOWLEDGEMENTS

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

106 - "I WILL NEVER FORGET THE SHAME I FELT": A SURVEY TO PEOPLE WITH A RHEUMATIC DISEASE ABOUT INVALIDATION FROM HEALTH PROFESSIONALS AND OTHER PEOPLE

Joana Vicente[1], Armando Silva[2], Andrea Marques[2, 3], Elsa Mateus[4], Ricardo J. O. Ferreira[3]

[1]Associação Portuguesa de Fibromialgia (APJOF), Leiria, Portugal, [2]Health Sciences Research Unit: Nursing (UICISA: E), Nursing School of Coimbra, Coimbra, Portugal, [3]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [4]Liga Portuguesa Contra as Doenças Reumáticas, Lisboa, Portugal

Background: The term invalidation refers to the patients' perception that their medical condition is not recognised, either in denying, lecturing, not supporting or not acknowledging the condition. This may be the felt from health professionals themselves but also from family, friends, at work and in other social areas, imposing great suffering.[1] The European Alliance of Associations for Rheumatology (EULAR) has made efforts to raise awareness for the burden imposed by rheumatic and musculoskeletal conditions (RMDs) and promote the best quality of care, including recognition and psychosocial support. However, it is unclear how frequent and severe the problem remains nowadays.

Objectives: The aims of this national survey were: (i) to identify the levels of invalidation and lack of understanding felt by adults with RMDs from health professionals and other people, (ii) to investigate the relationship between invalidation, sociodemographic characteristics and disease; and (iii) to understand its impact on people's life and health outcomes.

Methods: An online survey was developed by the national health professionals in rheumatology and patients' organisations and opened between May and December of 2021. The questionnaire included demographic and disease information, the Illness Invalidation Inventory (3*I),[1] with additional questions in a Likert format and open questions for a detailed understanding of the phenomenon. The3*I is composed of 8 items, measured from 1 (=never) to 5 (=very often), forming two factors: Discounting (mean of 5 items; lower scores indicating more discounting) and Lack of understanding (mean of 3 items; Higher scores representing higher lack of understanding). Quantitative data were analysed using descriptive statistics. Associations were tested with a t-student and ANOVA one-way test (Bonferroni correction). Open responses were categorised using the content analysis technique, and themes were defined a posteriori.

Results: From the > 1500 responses obtained, 1410 responses were filled out completely (mean age of 46 years [SD=11], 95% females, 60% with FM, among which 59% were diagnosed in the last 5 years). Invalidation was reported by 86% of the participants and 70% rated ≤5 on a scale from 0 (nothing) to 10 (totally) on feeling understood by other people. Invalidation was mostly felt from family (56%), health professionals (48%), friends (39%) and social environment (38%). The impact of this invalidation is mainly on the psychological well-being (58%), also reducing seeking health care (41%) and therapeutic adherence (17%), affecting work (41%), and to a less extent, (family) relations (31%). Figure 1 shows the frequency of responses and means scores on the 3*I items and factors for participants with and without FM. The burden is greater for people with FM, which was statistically significant. People with higher education felt more discounting and more lack of understanding. No differences (p>0.05) were observed for gender or civil status. Elucidative expressions of invalidation were shared, mostly by people with FM, encompassing their ability to work and need for social support, faking pain and treatment efficacy, and even intimacy aspects. These emotionally uncomfortable situations can be linked to lesser engagement with healthcare and disease management, and therefore, with worse health outcomes.

Conclusion: Invalidation remains a source of suffer-

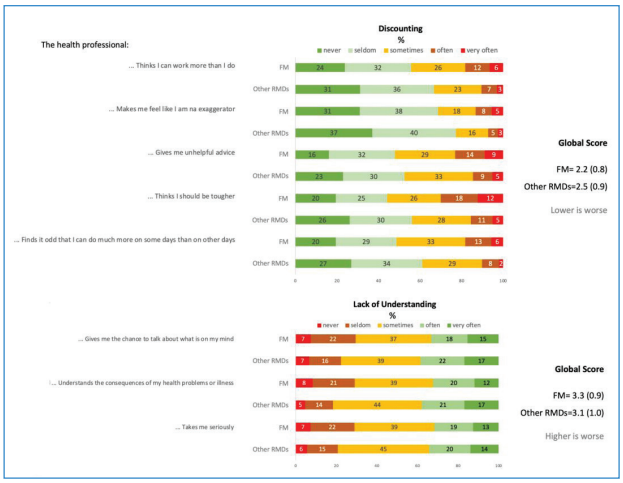


Figure 1. Percentages of responses per type of disease for the eight items of the Illness Invalidation Inventory

ing, affecting well-being and health outcomes. Specific awareness and educational campaigns are needed to target this problem on different play-actors.

111 - PSORIATIC ARTHRITIS RELATED FATIGUE: WHAT IS THE MAGNITUDE OF THIS PROBLEM AND THE CORRELATED FACTORS? A CROSS-SECTIONAL STUDY.

Ana Martins[1, 2], Daniela Santos Oliveira[1, 2, 3], Frederico Rajão Martins[4], Beatriz Samões[5], Rafaela Nicolau[6], Maria Seabra Rato[1, 2], Filipe Oliveira Pinheiro[1, 2], Sofia Pimenta[1, 2], 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, [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]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [6]Rheumatology Unit, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Background: Fatigue is a common constitutional feature and has a significant impact on quality of life in patients with chronic inflammatory rheumatic diseases, such as psoriatic arthritis (PsA). It is a complex phenomenon and its pathogenesis remains unclear. Despite being a common symptom, it is largely ignored and rarely assessed in clinical practice.

Objectives: This study aims to evaluate the incidence and severity of fatigue in PsA patients under biological agents and to assess the influence of several clinical and demographic features on PsA related fatigue.

Methods: We conducted a cross-sectional study including patients with PsA, according to CASPAR criteria, treated with biological agents, from our University Hospital and registered in the national database (reuma.pt). Fatigue was assessed by a 13-item self-administered questionnaire (Functional Assessment of Chronic Illness Therapy-Fatigue [FACIT-F]). Data collected and analyzed included: demographic data, disease activity data, functional status, comorbidities and therapies. Student's t-test, ANOVA and Pearson's correlation were performed to compare data, as appropriate. A p-value <0.05 was considered statistically significant.

Results: 60 PsA patients were included, 61.7% were males, with a mean age at diagnosis of 38.1±10.5 years and a median disease duration of 13.0 (8.75-19.25) years. Most of the included patients had a predominant polyarticular pattern (n=33, 55.0%). The mean FACIT-F score was 34.48±11.61. No differences were found in the FACIT-F score according to gender, pattern of joint involvement, presence or absence of cutaneous involvement, nail dystrophy and/ or dactylitis. Patients with depression and enthesitis exhibited a lower FACIT-F score (p=0.017 and p=0.007, respectively). Patients treated with tofacitinib had a lower FACIT-F score than patients treated with adalimumab (p=0.025). No differences were found among the other biological agents. Patients in remission (according to EULAR response criteria) had a higher FACIT-T score than patients with moderate disease activity (p=0.032). In patients with a predominant axial involvement, inactive disease (according to ASDAS) was associated with a higher FACIT-T score, when compared to very high disease activity (p=0.02). Also, patients with moderate disease activity had a higher FACIT-T score than patients with very high (p=0.003) and high (p=0.008) disease activity.

FACIT-F showed a significant correlation with disease activity scores as BASDAI (r=-0.546, p<0.001), DAS 28 CRP (r=-0.506, p<0.001), CDAI (r=-0.672, p<0.001), SDAI (r=-0.641, p<0.001) and ASDAS CRP (r=-0.500, p<0.001). FACIT-F was also correlated with BASFI (r=-0.598, p<0.001), HAQ (r=-0.701, p<0.001), BASMI (r=-0.431, p<0.001) and MASES (r=-0.401, p<0.001). The authors found strong correlations between FACIT-F and HADS domains (Depression and Anxiety domains; r=-0.850, p<0.001 and r=-0.748, p<0.001, respectively). A strong correlation was also found between the FACIT-F and the 8 domains of health of the SF36 (p<0.001).

Conclusions: Fatigue was a common symptom in our sample of PsA patients and, its magnitude was closely related with disease activity, physical function, depression and anxiety status, indicating its multifactorial nature. We can speculate that achieving disease remission could significantly alleviate the fatigue intensity and vice versa. As recommended by EULAR/ACR task force in 2008, fatigue should be measured in clinical practice and should be part of the multidisciplinary approach, in addition to controlling disease activity.

112 - ANTI-FIBRÓTICOS NO TRATAMENTO DA DOENÇA INTERSTICIAL PULMONAR SECUNDÁRIA ÀS DOENÇAS REUMÁTICAS SISTÉMICAS – A EXPERIÊNCIA DE UM CENTRO

Ana Catarina Duarte[1], Ana Cordeiro[1], Miguel Lopes[2], Jorge Soares[2], Maria José Santos[1, 3]

[1]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal, [2]Serviço de Pneumologia, 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

Introdução: Os anti-fibróticos (pirfenidona e nintedanib) foram inicialmente aprovados para o tratamento da fibrose pulmonar idiopática (FPI). Pelas semelhanças fisiopatológicas entre a FPI e a doença pulmonar intersticial, secundária às doenças reumáticas sistémicas (DPI-DRS), sobretudo com padrão intersticial usual (UIP), têm tido utilização crescente no tratamento da DPI-DRS. Este trabalho tem como objetivo descrever a experiência de um centro de Reumatologia na utilização de anti-fibróticos na DPI-DRS, com foco na tolerância e efetividade.

Métodos: Análise retrospetiva de todos os doentes com DPI-DRS sob anti-fibrótico seguidos em consulta de Reumatologia. A prescrição destes fármacos foi discutida/aprovada em consulta multidisciplinar de interstício pulmonar, com a Pneumologia.

Foram colhidos dados socio-demográficos, diagnóstico reumatológico e terapêutica DMARD em curso, comorbilidades de relevo, padrão imagiológico/histopatológico da DPI e resultados das provas funcionais respiratórias.

Resultados: De Julho de 2016 a Maio de 2022, foram tratados com anti-fibróticos 14 doentes. Destes, 9 (64.3%) eram do sexo feminino, com uma mediana de idades à data da última consulta de 67.5 anos. Seis (42.9%) doentes tinham artrite reumatoide (AR; todos com fator reumatoide e anticorpos anti-péptidos citrulinados cíclicos positivos), 6 esclerose sistémica (SSc; 5 com forma cutânea limitada e 1 com forma cutânea difusa; todos com anticorpos antinucleares positivos e 2 com anticorpos anti-topoisomerase I positivos), 1 doente com dermatomiosite (DM) e outro com síndrome de Sjögren primária (SSp).

Nove (64.3%) doentes apresentavam padrão UIP, 4 (28.6%) pneumonia intersticial não específica (NSIP) fibrótica e 1 pneumonia intersticial descamativa, embora com descrição de pulmão em favo de mel.

A duração mediana do tratamento com anti-fibrótico era de 16 meses (mínimo 1; máximo 51). A pirfenidona foi inicialmente prescrita a 8 (57.1%) doentes. Dois doentes apresentaram intolerância gastro-intestinal , não resolvida com redução da dose ou terapêutica sintomática, tendo um feito switch para nintedanib e o outro optado por suspender tratamento anti-fibrótico.

Três dos 6 doentes medicados inicialmente com nintedanib tiveram necessidade de reduzir a dose (para 100mg bid) por intolerância gastro-intestinal. Contudo, num doente as queixas persistiram, e por progressão imagiológica e funcional foi alterado para pirfenidona.

Todos os doentes com AR estavam sob terapêutica

Table I. Histomorphometric parameters of bone biopsy.

DRS	Padrão	Anti-fibrótico	Duração tratamento anti-fibrótico (meses)	Imunossupressão concomitante	PFR prévia a anti-fibrótico		PFR ≥ 9 meses anti-fibrótico	
						DLCO (%)	FVC (%)	DLCO (%)
AR	UIP	Nintedanib	13	MTX	69.5	68.7	61	81
AR	UIP	Pirfenidona	19	LFN, ABA	63.4	47.2	59.8	32
AR	UIP	Pirfenidona	22	RTX	51.5	30.4	54	22
AR	UIP	Pirfenidona	1	LFN				
AR	UIP	Pirfenidona	1	MTX, TCZ				
AR	UIP	Pirfenidona	29	LFN, IFN	70.1	44	74	57.9
SSc	NSIP fibrótica	Nintedanib	43	MMF	61.5	-	60.6	-
SSc	NSIP fibrótica (posterior progressão para UIP)	Nintedanib/ Pirfenidona	41 / 1	-	80	45	67	-
SSc	UIP	Nintedanib	1	-				
SSc	DIP (favo de mel)	Nintedanib	64	-	65	35	57	30
SSc	UIP	Nintedanib	13	MMF	40.5	29.2	52.1	18.1
SSc	UIP	Pirfenidona	4	AZA				
DM	NSIP fibrótica	Pirfenidona/ Nintedanib	6 / 4	RTX	61.7	43.6	63	60
SSp	NSIP fibrótica	Pirfenidona	51	RTX	47.3	26.6	56	54

Legenda: DRS – doença reumática sistémica; DPI – doença pulmonar intersticial; PFR – provas de função respiratória; FVC – capacidade vital forçada; DLCO – capacidade de difusão do monóxido de carbono; AR – artrite reumatoide; SSc – esclerose sistémica; DM – dermatomiosite; SSp – síndrome de Sjögren primária; UIP – pneumonia intersticial usual; NSIP – pneumonia intersticial não específica; DIP – pneumonia intersticial descamativa; MTX – metotrexato; LFN – leflunomida; ABA– abatacept; RTX – rituximab; TCZ – tocilizumab; IFN – infliximab; MMF – micofenolato de mofetil; AZA - azatioprina

DMARD para controlo da sua doença articular, sendo que em 2 deles a escolha do biológico (rituximab e abatacept) foi influenciada pela presença de DPI. Dois doentes com SSc estavam sob micofenolato de mofetil e os doentes com DM e SSp estavam ambos sob rituximab.
Na tabela 1 é feita uma caracterização dos doentes sob anti-fibrótico, com indicação da evolução em termos funcionais sempre que o tratamento dure há pelo menos 9 meses.
Dois dos doentes com SScmorreram, um por progressão de neoplasia do pulmão e o outro por insuficiência respiratória parcial grave, com radiografia mostrando infiltrados pulmonares difusos de novo.
Conclusão: A experiência do nosso centro corrobora a estabilização da DPI nos doentes tratados com anti-fibróticos em alguns casos em associação com DMARD. A escolha do anti-fibrótico tem muitas vezes em conta as queixas gastro-intestinais, frequentes com esta classe de fármacos, bem como o uso concomitante de anticoagulantes orais.

118 - VISION-RELATED QUALITY OF LIFE IN SPONDYLOARTHRITIS PATIENTS WITH A HISTORY OF ACUTE ANTERIOR UVEITIS UNDER TREATMENT WITH GOLIMUMAB: 12 MONTHS RESULTS OF THE GO-VISION OBSERVATIONAL STUDY

Inês Leal[1, 2], Ana Teresa Melo[3, 4], Carolina Ochôa Matos[3], Catarina Tenazinha[3, 4], Cláudia Vaz[5], Eduardo Dourado[3, 4], Fernando Pimentel-Santos[6], Filipe Cunha Santos[3, 5], Henrique Fernandes[7], Joana Fonseca Ferreira[5], Manuel Silvério-António[3, 4], Margarida Faria[3, 8], Marta Guedes[9], Miguel Cordeiro[9], Miguel Santos[1, 2], Nikita Khmelinskii[3, 4], Patricia Jose[1, 2], Rafael Barão[1, 2], Sara Paiva Dinis [5], Sofia Sousa Mano[1, 2], 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]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [4]Rheumatology Research Unit, Instituto de Medicina Molecular, Faculdade de Medicina da Universidade de Lisboa, Lisboa, Portugal, [5]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, [6]Rheumatology Department, Centro Hospitalar Lisboa Ocidental, Lisboa, Portugal, [7]Oftalmologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, [8]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [9]Oftalmologia, Centro Hospitalar de Lisboa Ocidental - Hospital Egas Moniz, Lisboa, Portugal

Background:Objective: Acute anterior uveitis (AAU) is one of the most common extra-articular manifestations of spondyloarthritis (SpA), causing a significant burden on quality of life (QoL). Golimumab (GLM) is a tumour 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. We aim to study the impact of GLM on 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 a history of AAU treated with GLM followed-up for 12 months. We intend to recruit 30 patients, and we report herein the outcomes for the first 10 patients enrolled that completed the 12 months follow-up. The occurrence of AAU was assessed in the 2 years before GLM treatment onset and the 12 months of follow-up. The risk for a new AAU was calculated for each period. VR QoL was assessed with the self-administered National Eye Institute Visual Functioning Questionnaire-25 (NEI VFQ-25). Patients had assessments in Ophthalmology and Rheumatology departments. Adverse events were noted.
Results: 10 patients (50% female, 70% TNFi-naive, mean age 45.8±11.1 years [range 22-65]) have completed the 12 months of follow-up. Five patients (50%) were also under oral methotrexate. The total number of AAU flares in the 2 years preceding the start of GLM was 10, a number reduced to 1 during the 12 months of treatment. The AAU incidence rate was reduced from 1.52 to 0.10 per 100 patient-years (incidence ratio-ratio 14.79 [2.39;610.52], p<0.01). At baseline, 24 weeks and 48 weeks after GLM onset, the mean overall index NEI VFQ-25 total score was 72.4±12.0, 84.6±17.2, and 89.4±8.6, respectively. Improvement in the NEI VFQ-25 total score between baseline and 48 weeks was +17. The difference across the three measurements was assessed with generalized estimating equations and adjusted to age and the baseline dose of oral prednisolone and was <0.001. No significant or new adverse events occurred.
Conclusion: The GO-VISION study is the first prospective study in the uveitis setting designed to have a patient-reported outcome measure as the primary outcome. Data from the GO-VISION study suggests that GLM has an acceptable safety profile and effective in patients with SpA and history of AAU, reducing the AAU occurrence rate and potentially increasing VR QoL.

119 - PERFORMANCE OF CLINICAL, LABORATORY AND IMAGING FEATURES FOR THE DIAGNOSIS OF AXIAL SPONDYLOARTHRITIS - A SYSTEMATIC LITERATURE REVIEW

Ana Bento da Silva[1, 2], Maria Helena Lourenço[1, 2], Sofia Ramiro[3, 4], Louise Falzon[5], Jaime C. Branco[1, 6], Désirée van der Heijde[3], Robert Landewé[4, 7], Alexandre Sepriano[1, 3, 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]Department of Rheumatology, Leiden University Medical Center, Leiden, Netherlands, [4]Rheumatology, Zuyderland Medical Center, Heerlen, Netherlands, [5]Health Economics and Decision Science, School of Health and Related Research, University of Sheffield, Sheffield, United Kingdom, [6]NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [7]Department of Rheumatology & Clinical Immunology, Amsterdam University Medical Center, Amsterdam, Netherlands

Background:Purpose: The Berlin algorithm for the diagnosis of axial spondyloarthritis (axSpA) was developed more than 15 years ago (2004). Since then, new studies suggest that the diagnostic performance of some SpA features might not be as good as initially thought. We aimed at reviewing the evidence on the performance of clinical, laboratory and imaging SpA features in discriminating between a clinical diagnosis of axSpA and no axSpA.

Methods: Systematic literature review of cross-sectional, longitudinal and case-control studies (2004-2021) reporting data that could be used to evaluate ≥1 SpA feature. The population was defined as adults (≥16 years) with a suspicion or definite clinical diagnosis of axSpA. The diagnostic performance of each SpA feature was tested using the clinical diagnosis of axSpA as reference standard. Studies in which classification criteria were used to define axSpA were excluded. Sensitivity, specificity, positive and negative likelihood ratio (LR+ and LR-) were calculated. Risk of bias was assessed using the QUADAS 2 tool.

Results: Of 11,420 screened articles, 20 studies evaluating patients with axSpA fulfilled the inclusion criteria. Five of these studies (n=3,366 patients; axSpA prevalence range 41-78%) were at low risk of bias and evaluated the same SpA features (n=10/20) allowing across-feature comparisons (Table 1). Three of these 5 studies (n=2,204) also evaluated bone marrow edema (BME) on MRI of the sacroiliac joints (MRI-SIJ) and definite damage on pelvic radiographs (X-SIJ). A good balance between sensitivity and specificity was found for HLA-B27, BME on MRI-SIJ and damage on X-SIJ which resulted in high LR+ and low LR-. For instance, patients with axSpA where 2 to 7 times more likely to be HLA-B27+ than patients without axSpA (LR+ range 2-7). Similarly, patients with axSpA were less likely to be HLA-B27 negative than patients without axSpA (LR- range 0.4-0.6). Peripheral manifestations and extra-musculoskeletal manifestations (EMM) were highly specific for axSpA but much less sensitive than HLA-B27 and imaging findings. Among peripheral manifestations, dactylitis had the highest LR+ (2.8-16.7), but was uncommon in axSpA (sensitivity range 5-9%), meaning that no dactylitis is almost as likely in axSpA as in no axSpA (LR- range 0.9-1). Similarly

Table I. Performance of each SpA feature for the diagnosis of axial spondyloarthritis

Number of Studies (Cohorts)	5 (4)			
Number of Patients (range of prevalence of axSpA-%)	3,366 (41 – 78%)			
	LR + (range)	LR - (range)	Sensitivity % (range)	Specificity % (range)
Inflammatory back pain	1.0 - 2.9	0.2 - 1.4	28 - 93	5 - 88
Peripheral arthritis	1.2 - 8.9	0.8 - 0.9	18 - 41	75 - 98
Dactylitis	2.8 - 16.7	0.9 - 1.0	5 - 9	98 - 100
Acute anterior uveitis	1.3 - 3.3	0.9 - 1.0	6 - 15	92 - 98
Psoriasis	0.8 - 3.0	0.9 - 1.0	8 - 15	89 - 95
Inflammatory bowel disease	0.9 - 3.5	1.0 - 1.0	3 - 8	95 - 99
Family history of spondyloarthritis	1.0 - 1.8	0.7 - 1.0	20 - 50	68 - 82
Good response to NSAIDs	1.2 - 2.7	0.5 - 0.8	42 - 67	56 - 80
Elevated CRP/ESR	1.3 - 2.6	0.7 - 0.9	12 - 40	71 - 95
HLA-B27	2.3 - 6.9	0.4 - 0.6	47 - 68	72 - 90
BME on MRI-SIJ (ASAS definition) *	6.7 - 19.6	0.5 - 0.6	42 - 53	93 - 100
Definitive damage on X-SIJ (mNY criteria) *	8.6 - 31.6	0.7 - 0.8	17 - 34	97 - 99

* Evaluated in 3 of the 5 studies (total population 2,204 patients; axSpA prevalence range 41-78%). axSpA: axial spondyloarthritis; BME on MRI-SIJ: bone marrow edema on magnetic resonance imaging of sacroiliac joints; CRP: C-Reactive Protein; ESR: Erythrocyte Sedimentation Rate; LR+: positive likelihood ration; LR-: negative likelihood ration; mNY criteria: modified New York criteria; NSAIDs: nonsteroid anti-inflammatory drugs; X-SIJ: pelvic radiographs.

poor LR- were found for the other peripheral features and EMM. The performance of IBP, good response to NSAIDs and family history was variable, but mostly poor, with LR+ and LR- close to 1.
Conclusion: The results for HLA-B27 and imaging findings are in agreement with the literature informing the Berlin algorithm and reflect good diagnostic performance for axSpA, even though circularity cannot be ruled out. New evidence confirms that peripheral features and EMM, when present, are suggestive of axSpA, but that their absence does not help in ruling out the diagnosis. The low diagnostic value of IBP, good response to NSAIDs and family history of SpA, compared to data previously informing the Berlin algorithm, deserve further consideration and may justify an adaptation of the algorithm.

130 - ASSESSMENT OF CALCINOSIS IN PORTUGUESE PATIENTS WITH SYSTEMIC SCLEROSIS – A MULTICENTER STUDY

Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Tiago Beirão[1], Flávio Campos Costa[1], Romana Vieira[1], Georgina Terroso[2], Raquel Miriam Ferreira[2], Rafaela Nicolau[3], André Saraiva[4], Maria João Salvador[4, 5], Ana Catarina Duarte[6], Ana Cordeiro[6], Joao Paulo Vilas-Boas[7], Inês Genrinho[3], Ana Bento da Silva[8, 9], Laura Gago[8, 9], Catarina Resende[10], Patrícia Martins[10, 11], Nathalie Madeira[12], Sara Paiva Dinis [12], Couto M[3], Inês Santos[3], Filipe Araújo[13], Ana Filipa Mourão[8, 9, 13], Miguel Guerra[14], Margarida Oliveira[14], Alexandra Daniel[15], Marília Rodrigues[15], Catarina Soares[16], Hugo Parente[16], Carolina Furtado[17], Tomás Fontes[17], Joana Aleixo[1]

[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [2]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [3]Rheumatology Unit, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [4]Rheumatology Department, Centro Hospitalar Universitário de Coimbra, Coimbra, Portugal, [5]Coimbra Institute for Clinical and Biomedical Research (iCBR) - Faculty of Medicine, University of Coimbra, Coimbra, Portugal, [6]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [7]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [8]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [9]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [10]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [11]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal, [12]Rheumatology Department, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, [13]Hospital Ortopédico de Sant 'Ana, Cascais, Portugal, [14]Rheumatology Department, Centro Hospitalar e Universitário Cova da Beira, Covilhã, Portugal, [15]Serviço de Reumatologia, Centro Hospitalar de Leiria, Leiria, Portugal, [16]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [17]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal

Background:Purpose: Calcinosis is a challenging problem among Systemic Sclerosis (SSc) patients with a reported prevalence of 18-49%. We aim to define the prevalence of clinical and subclinical calcinosis among Portuguese SSc patients, currently unknown, as well as the most sensitive anatomical location and the sensitivity of the clinical method for its diagnosis. We also aim to clarify the phenotype of SSc patients with calcinosis.
Methods: A cross-sectional multicenter study was conducted evaluating SSc patients from 14 Portuguese centers, registered in the Rheumatic Diseases Portuguese Registry (Reuma.pt), that fulfilled Leroy/Medsger 2001 or ACR/EULAR classification criteria for SSc. The assessment of calcinosis was made systematically through clinical examination and plain radiographs of hands, elbows, knees and feet, analyzed by 2 rheumatologists of each center. Statistical analysis with independent parametric or non-parametric tests, multivariate logistic regression of the significant variables in univariate analysis and sensitivity calculation of each radiographed site and of the clinical method for the diagnosis of calcinosis were performed.
Results: 226 patients were included, of whom 191 were female (84.5%), with a median [min, max] age of 64 [20, 90] years-old and a median [min, max] disease duration of 11.47 [1, 62] years. 172 patients (76.4%) had limited SSc, 36 (16%) diffuse SSc, 9 (4%) sine scleroderma SSc and 8 (3.6%) early SSc. Clinical calcinosis was described in 63 (28.1%) patients, 10 of which was not radiologically confirmed. 91 (40.3%) patients had radiological calcinosis in at least one site [hand in 68 (74.7%), knee in 32 (35.2%), elbow in 30 (34.1%) and foot in 21 (23.9%)], of which 37 (40.7%) were subclinical. The most sensitive location to detect calcinosis was the hand (74.7%) while the sensitivity of the clinical method was 58.2%. Table 1 summarizes the clinical characteristics of patients with and without calcinosis. Patients with calcinosis were more often female ($p=0.008$), older ($p<0.001$) and with longer disease duration ($p<0.001$). The presence of calcinosis was significantly associated with limited SSc ($p=0.017$), telangiectasia ($p=0.039$), digital ulcers ($p=0.001$), oesophageal ($p<0.001$) and intestinal ($p=0.003$) involvements. Os-

Table I. Demographic and clinical features of SSc patients with and without calcinosis.

Variable	Calcinosis (N=91)	No calcinosis (N=135)	p-value
Age, median [min, max]	69 [28, 87]	61 [20, 90]	<0.001
Age at diagnosis, mean ± SD	55.01 ± 12.83	53.71 ± 11.86	0.442
Female, n (%)	84 (92.3)	107 (79.3)	0.008
Disease duration, median [min, max]	11 [1, 40]	7 [0, 53]	<0.001
Diagnosis delay, median [min, max]	4.06 [0, 54.71]	2 [0, 48]	0.180
BMI, median [min, max]	24.73 [17.48, 41.09]	25.44 [15.92, 36.48]	0.142
Disease subsets			
- Limited, n (%)	77 (84.6)	95 (70.9)	0.017
- Diffuse, n (%)	11 (12.1)	25 (18.7)	0.187
- Sine scleroderma, n (%)	2 (2.2)	7 (5.2)	0.318
- Early, n (%)	1 (1.1)	7 85.2)	0.147
Clinical manifestations			
- Raynaud phenomenon, n (%)	89 (97.8)	127 (95.5)	0.478
- Telangiectasia, n (%)	61 (68.5)	71 (54.6)	0.039
- Digital ulcers, n (%)	46 (50.5)	38 (28.8)	0.001
- Flexion contractures, n (%)	12 (14.5)	12 (9.9)	0.323
- Tendon friction rubs, n (%)	6 (7.3)	7 (5.9)	0.684
- Arthritis or arthralgia, n (%)	35 (39.3)	66 (50.8)	0.095
- Miositis, n (%)	2 (2.2)	3 (2.3)	1.000
- Oesophageal involvement, n (%)	52 (59.8)	41 (32.3)	<0.001
- Gastric involvement, n (%)	14 (16.3)	16 (13)	0.507
- Intestinal involvement, n (%)	11 (12.9)	3 (2.4)	0.003
- Cardiac involvement, n (%)	11 (12.5)	12 (9.7)	0.515
- Lung involvement, n (%)	27 (31)	40 (32.3)	0.851
- Renal involvement, n (%)	4 (4.4)	2 (1.6)	0.239
Autoantibodies			
- Anti-nuclear, n (%)	86 (94.5)	126 (94.7)	1.000
- Anti-centromere, n (%)	62 (68.1)	75 (56.4)	0.077
- Anti-topoisomerase I, n (%)	13 (14.3)	30 (23.1)	0.104
- Anti-RNA polymerase III, n (%)	2 (3.3)	5 (4.9)	1.000
- Anti-Th/To, n (%)	0 (0)	1 (1.1)	1.000
- Anti-U3-RNP, n (%)	2 (3.4)	2 (2.1)	0.636
- Anti-Pm/Scl, n (%)	4 (6.1)	11 (10.4)	0.329
- Anti-Ku, n (%)	1 (1.7)	0 (0)	0.371
- Anti-U1-RNP, n (%)	0 (0)	2 (1.8)	0.527
mRSS in the last appointment, median [min, max]	5.5 [0, 33]	4 [0, 37]	0.114
Nailfold capillaroscopy scleroderma pattern, n (%)	60 (90.9%)	82 (75.9)	0.013
- Early, n (%)	9 (15)	30 (38)	0.003
- Active, n (%)	26 (43.3)	42 (53.2)	0.251
- Late, n (%)	24 (40)	7 (8.9)	<0.001
Osteoporosis, n (%)	20 (22)	15 (11.2)	0.028
Current or past steroid use, n (%)	36 (39.6)	42 (31.6)	0.218

BV – bone volume; TV – tissue volume; OV – osteoide volume; OS – osteoide surface; BS – bone surface; OTh – osteoide thickness; TbN – trabecular number; Tb.sp – trabecular separation; ObS – osteoblast surface; OcS – osteoclast surface; MS – mineralizing surface; sL.S – single labelled surface; MLT- mineralization lag time. NS – not specified. Note: BFR/BS and MLT were not calculated due to absence of tetracycline double labels.

teoporosis and late scleroderma capillaroscopic pattern (p<0.001) were significantly more often on calcinosis group (p=0.028). In multivariate analysis, digital ulcers (OR 2.631, 95%CI 1.022-6.775, p=0.045) were found to be predictors of overall calcinosis, oesophageal involvement (OR 3.521, 95%CI 1.282-9.668, p=0.015) and osteoporosis (OR 4.083, 95%CI 1.176-14.176, p=0.027) of hand calcinosis and late capillaroscopic pattern (OR 7.608, 95%CI 1.661-34.858, p=0.009) of knee calcinosis.
Conclusion: The prevalence of overall calcinosis was in the range of that previously reported. The higher prevalence of subclinical calcinosis suggests that calcinosis might be underdiagnosed and that radiographic screening in SSc patients might be relevant. The differences between patients with and without calcinosis are in agreement with the literature. Calcinosis predictors appear to vary according to location and suggest a multifactorial pathogenesis.

133 - PREVALENCE OF FOOT INVOLVEMENT AT THE INITIAL PRESENTATION OF RHEUMATOID ARTHRITIS

C. Pinto Oliveira[1, 2], Susana P. Silva[1, 2], Sofia Ferreira Azevedo[1, 2], Carolina Mazeda[1, 2], Anabela Barcelos[1, 2]
[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, EPE, Aveiro, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz, Aveiro, Portugal

Rheumatoid arthritis (RA) frequently affects the feet, inducing pain and incapacity.
Objective: To evaluate the prevalence of foot involvement at the initial presentation of RA.
Methods: We conducted a retrospective study of 119 patients followed in our Rheumatology Department with the diagnosis of RA over a period of 3 months, from January to March 2022. Patient records were searched for demographic data, medical comorbidities, and clinical details at the time of RA's initial presentation. Descriptive analysis used mean and standard deviation for continuous data, as well as frequency counts and percentages for categorical variables. Parametric and nonparametric tests were used for statistical analysis with SPSS® software. p-values ≤ 0.05 were considered statistically significant.
Results: 119 patients were included, from which 61 (51.3%) had foot involvement at the initial presentation of RA, either detected clinically or by ultrasound. In this group, 41 patients were women (67.2%). The mean age at diagnosis was 55.9 years. The tibiotarsal joint was most frequently affected (40 patients; 65.6%), followed by forefoot (26 patients; 42.6%) and midfoot involvement (20 patients; 32.8%). Patients with foot involvement were more likely to show compromise of small joints and polyarthritis at the initial presentation of RA (p < 0.05). We found no statistically significant differences between patients with or without foot involvement regarding age at diagnosis, rheumatoid factor or anti-citrullinated protein antibodies positivity, morning stiffness, comorbidities, inflammatory markers levels or mean Disease Activity Score in 28 Joints.
Conclusion: Foot involvement at the initial presentation of RA was common in our population, affecting approximately one-half of the patients. This finding is consistent with other studies available in scientific literature. Furthermore, it reinforces the importance of proper foot evaluation as part of RA's management.

134 - INFEÇÃO POR SARS-COV2 EM DOENTES COM PATOLOGIA REUMÁTICA SOB TERAPÊUTICA COM RITUXIMAB

Ana Margarida Correia[1], Carla Campinho Ferreira[1], Emanuel Costa[1], Diogo Esperança Almeida[1], Joana Leite Silva[1], Joana Sousa-Neves[1], Marcos Cerqueira[1], José Redondo[1], Ana Ribeiro[1]
[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

Introdução: O rituximab (RTX) é utilizado no tratamento de doenças reumáticas geralmente associadas a pior prognóstico clínico, como vasculites e conetivites, principalmente com envolvimento intersticial pulmonar. Com o surgimento da pandemia por COVID19, alguns estudos demonstraram a associação entre a terapêutica com RTX e a maior severidade da infeção por SARS-CoV2, embora não tenham ficado claros os fatores que justificam essa associação.
Objetivos: Determinar quais os fatores associados a maior gravidade da infeção por SARS-CoV2 em doentes com patologia reumática sob terapêutica com RTX.
Metodologia: Estudo retrospetivo, realizado em centro terciário, que incluiu pacientes com patologia reumática seguidos em Consulta de Reumatologia sob terapêutica com RTX e que tiveram infeção por SARS-CoV2. A análise estatística dos dados foi feita com recurso ao SPSS, tendo sido utilizados os testes Qui-quadrado, Exato de Fisher e Mann-Whitney, conforme apropriado. Foi considerado estatisticamente significativo um valor de p < 0,05.
Resultados: Foram incluídos um total de 10 doentes com uma idade média de 54±12,3 anos, a maioria do sexo feminino (60%). As conetivites foram o diagnóstico mais prevalente (60%), salientando-se neste grupo a Artrite Reumatóide (n=4), tendo os restantes doentes o diagnóstico de vasculite. Verificou-se a presença de envolvimento pulmonar em 4 doentes (40%), metade sob a forma de hemorragia alveolar e metade sobre a forma de doença intersticial fibrosante. 60% dos doen-

Tabela I. Caracterização da amostra e valor de p dos testes utilizados para avaliar a associação entre a gravidade da infeção por SARS-Cov2 em doentes sob RTX e as diferentes variáveis em estudo.

Variáveis	Caracterização (n/N)	Análise univariada
Ex-fumador	3/10	p=0,530
Excesso de peso/Obesidade	2/10	p=0,435
Diabetes Mellitus tipo 2	1/10	p=0,108
Hemodiálise	2/10	p=0,435
Patologia Reumática		
Vasculite	4/10	p=0,108
Conetivite	6/10	p=0,108
Envolvimento Pulmonar		
Hemorragia alveolar	2/10	p=0,435
Doença intersticial fibrosante	2/10	p=0,007
Tratamento com cDMARD	6/10	p=0,108
Dose de PDN ≥ 15 mg/dia	2/10	p=0,007
Última perfusão de RTX		
RTX ≤ 1 mês	3/10	p=0,049
RTX > 1 mês e < 6 meses	5/10	p=0,135
RTX ≥ 6 meses	2/10	p=0,435
Esquema de vacinação		
Sem vacinação	2/10	p=0,435
Vacina ≤ 1 mês	4/10	p=0,870
Vacina > 1 mês e ≤ 6 meses	2/10	p=0,435
Vacina > 6 meses	2/10	p=0,435
Internamento	4/10	p=0,007
Mortalidade	2/10	p=0,007

tes encontravam-se sob terapêutica concomitante com fármacos reumáticos modificadores de doença convencionais (cDMARD) e 20% estavam sob doses de prednisolona (PDN) = ou > 15 mg/dia. 30% dos doentes tinham cumprido a última perfusão de RTX 1 mês ou menos antes do diagnóstico da infeção. No que concerne à vacinação, a maioria dos doentes tinha pelo menos 1 dose da vacina contra a COVID19 (80%), sendo que 40% dos doentes tinha feito a última inoculação até 1 mês antes do diagnóstico da infeção. A infeção por SARS-CoV2, na maioria dos casos, foi de gravidade ligeira (60%), tendo-se verificado 2 casos de infeção moderada (20%) e 2 de infeção grave (20%). Os doentes com infeção COVID19 moderada a grave necessitaram de internamento hospitalar e verificou-se que, aqueles com apresentação grave da doença, faleceram (20%). Verificou-se uma associação estatisticamente significativa entre a forma grave da infeção por SARS-CoV2 em doentes com patologia reumática sob RTX e as seguintes variáveis: presença de doença intersticial fibrosante, dose de PDN = ou > 15 mg/dia e perfusão de RTX < ou = 1 mês (p=0,007; 0,007 e 0,049 respetivamente), assim como uma associação entre a gravidade da infeção e a taxa de internamento e de mortalidade (p=0,007). Documentou-se ainda uma associação entre a presença de doença intersticial fibrosante e a mortalidade pela COVID19 em doentes com patologia reumática sob tratamento com RTX (p=0,002).

Conclusão: Neste estudo, verificou-se que os doentes com patologia reumática com doença intersticial fibrosante sob terapêutica com RTX apresentaram maior gravidade de infeção por SARS-CoV2 e maior mortalidade associada, o que poderá ser explicado não só pela imunossupressão, bem como pelas alterações estruturais pulmonares e ventilatórias pré-existentes. Para além disso, verificou-se que a gravidade da infeção por SARS-CoV2 em doentes sob terapêutica com RTX estava associada à realização recente da perfusão (< ou = 1 mês), sugerindo uma relação dose-dependente, e à terapêutica com PDN = ou > 15 mg/dia, que confere um maior grau de imunossupressão.

139 - IMMUNE-MEDIATED SKIN LESIONS RELATED TO BIOLOGICAL DISEASE-MODIFYING ANTIRHEUMATIC DRUGS: A 22-YEAR EXPERIENCE OF A TERTIARY CENTER

Ana Martins[1,2], Daniela Santos Oliveira[1,2,3], Frederico Rajão Martins[4], Rafaela Nicolau[5], Filipe Oliveira Pinheiro[1,2], Maria Seabra Rato[1,2], Sofia Pimenta[1,2], Miguel Bernardes[1,2], Lúcia Costa[2]

[1]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, 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]Rheumatology Unit, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Background: Biological disease-modifying antirheumatic drugs (bDMARDs) have revolutionized the treatment of chronic inflammatory rheumatic diseases. However, the physician and the patient should be aware of possible adverse reactions. Skin is one of the most frequent organs involved in bDMARD adverse reactions and immune-mediated skin lesions (IMSL) have

rarely been described before in cohort studies and their incidence is unknown.

Objectives: To explore the cumulative incidence, type of lesions, management and outcomes of IMSL related to bDMARD in a large cohort of patients with rheumatoid arthritis (RA), axial spondyloarthritis (axSpA) and psoriatic arthritis (PA).

Methods: We conducted a retrospective single-center study including patients with RA, axSpA and PA followed at the Department of Rheumatology of a University Hospital Center between April 2000 and December 2021, treated with at least one bDMARD for at least 6 months. Data were collected from reuma.pt and medical records. Sociodemographic characteristics, disease duration, age at diagnosis, comorbidities, smoking and drinking habits, concomitant immunosuppressive medications, type and duration of the treatment with bDMARD and number of previous bDMARD were collected. For all patients with IMSL, age at onset, disease duration at the time of the IMSL, culprit bDMARD and duration of the treatment, specific management and outcomes were collected. Descriptive statistics for continuous variables were presented with mean and standard deviation and categorical variables were presented with absolute and relative frequencies.

Results: A total of 441 patients with RA, 386 with axSpA and 162 with PsA were included. The majority were female (63.4%), with a mean age of 54.3 ± 12.8 years. An important proportion of patients (47.6%, n=471) were taking csDMARDs and the most prescribed bDMARD was adalimumab (21.8 %), followed by etanercept (16.5%). Twenty-seven (2.7%) patients presented IMSL potentially related to the bDMARD. Regarding the patients with IMSL, 55.6% were females, mean age at the onset of IMSL was 48.4 ± 12.0 years, mean duration of the treatment with bDMARDs was 4.3 ± 4.5 years and mean duration of the treatment with the culprit bDMARD was 2.3 ± 2.1 years. The majority of patients had SpA (n= 14), followed by RA (n=10) and PsA (n=3). Adalimumab was the culprit agent in half of the patients (n=14), followed by etanercept (n=4), golimumab (n=3), infliximab (n=3), rituximab (n=2) and tocilizumab (n=1). Four patients (14.8%) needed hospitalization with the purpose of performing a clinical, laboratorial and histological investigation. In most patients, skin lesions resolved completely with topical (n=12) or systemic (n=6) treatment. IMSL led to withdrawal of bDMARD in 18 patients (66.7%). More in-

Table I. Description of number of cases, age at IMSL onset, disease duration and duration of treatment with the culprit bDMARD for each type of IMSL

Type of IMSL	Number of patients, n (%)	Age at IMSL onset, mean ± SD, years	Female, n (%)	Disease duration, mean ± SD, years	Duration of treatment with culprit bDMARD, mean ± SD, years
Psoriasis	12	49.3 ± 14.5	6 (50.0)	19.5 ± 15.3	1.9 ± 1.7
Plaque psoriasis	5 (41.7)				
Palmoplantar pustulosis	4 (33.3)				
Guttate psoriasis	1 (8.3)				
Inverse psoriasis	1 (8.3)				
Undefined	1 (8.3)				
DILE	6	40.8 ± 2.9	4 (66.7)	15.2 ± 7.8	2.4 ± 1.4
Malar Rash	1 (16.7)				
Alopecia	2 (33.3)				
Chilblains	2 (33.3)				
Subacute cutaneous LE	1 (16.7)				
LE tumidus	1 (16.7)				
Alopecia areata	3	4.4 ± 6.7	1 (33.3)	12.2 ± 627	1.2 ± 0.6
Leukocytoclastic vasculitis	2	60.9 ± 2.8	0 (0)	31.9	2.9 ± 3.6
Urticaria	2	57.3 ± 15.9	2 (100)	27.8 ± 22.1	0.9 ± 1.2
Rosacea	1	48	1 (100)	18.5	9.7
Erythema nodosum	1	60	1 (100)	40.7	2.9

Legend: IMSL: Immune-mediated skin lesions, DILE: drug-induced lupus erythematosus.

formation about the type of IMSL, age at IMSL onset, disease duration and duration of the treatment with bDMARD for each type of IMSL was described in table 1.
Conclusions: IMSL related to bDMARDs are unusual events with an estimated cumulative incidence of 2.9%, in our sample. The most frequent IMSL were psoriasis and cutaneous manifestations of DILE and the most frequent culprit bDMARD was adalimumab. The majority of patients didn't need hospitalization and presented complete resolution of IMSL. IMSL led to withdrawal of bDMARD in 2/3 of patients.

140 - ADALIMUMAB AND NUMBER OF PREVIOUS BIOLOGICAL DISEASE-MODIFYING ANTIRHEUMATIC DRUGS AS PREDICTIVE FACTORS FOR THE DEVELOPMENT OF IMMUNE-MEDIATED SKIN LESIONS

Ana Martins[1, 2], Daniela Santos Oliveira[1, 2, 3], Frederico Rajão Martins[4], Rafaela Nicolau[5], Filipe Oliveira Pinheiro[1, 2], Maria Seabra Rato[1, 2], Sofia Pimenta[1, 2], 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, [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]Rheumatology Unit, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Adalimumab and number of previous biological disease-modifying antirheumatic drugs as predictive factors for the development of immune-mediated skin lesions
Background: Risk factors associated with the occurrence of immune-mediated skin lesions (IMSL) in rheumatic patients under biological disease-modifying antirheumatic drugs (bDMARDs) are poorly known and studied. Therefore, we conducted a retrospective cohort single-center study and we aim to identify predictive factors for IMSL in patients with rheumatoid arthritis (RA), axial spondyloarthritis (axSpA) and psoriatic arthritis (PsA) under bDMARD therapy.
Methods: A retrospective single-center study including patients with RA, axSpA and PsA followed at the Department of Rheumatology of a University Hospital Center between April 2000 and December 2021, treated with at least one bDMARD for at least 6 months was conducted. Data were collected from reuma.pt and medical records. Sociodemographic characteristics, disease duration, age at diagnosis, smoking and drinking habits, concomitant immunosuppressive medications, type and duration of the bDMARD treatment and number of previous bDMARD were collected. IMSL development were also collected. To examine the differences between groups with and without IMSL we performed independent samples t-test for normally distributed continuous data and chi-square tests for categorical variables. Also, a multivariate logistic regression analysis was performed to identify possible predictive factors for the occurrence of IMSL. Odds ratios (ORs) and 95% confidence intervals (CIs) were calculated. Statistical significance was set at a p-value <0.05.
Results: From a total of 441 patients with RA, 386 with axSpA and 162 with PsA, 27 developed IMSL related to bDMARD (2.7%). Comparing the groups with and without IMSL, no differences were found regarding age, gender, BMI, presence of concomitant csDMARD or type of rheumatic disease. Patients with IMSL have a significant younger age at diagnosis (p=0.038), a longer disease duration (p=0.018) and a longer duration of bDMARD treatment (p=0.008). Patients with IMSL also have a higher number of previous bDMARDs (p<0.001) and adalimumab was the bDMARD with the higher risk of IMSL development (p<0.001). Table 1 describe the demographic and clinical features of these two groups. In a multivariate regression model, number of previous bDMARDs (OR 2.13, 95% CI 1.47 to 3.10, p<0.001) and adalimumab vs other bDMARD (OR 4.60, 95% CI 1.96 to 10.80, p<0.001) were statistically significant predictive factors for IMSL development.
Conclusion: In our cohort, we found that a younger age at diagnosis, longer disease duration, longer duration of bDMARD treatment, higher number of previous bDMARDs and treatment with adalimumab were independently associated with an increased risk of IMSL development. In the multivariate regression model, number of previous bDMARDs and adalimumab administration were statistically significant predictive factors for IMSL development. Further research is required to better understand and recognize the risk factors for IMSL.

141 - IS TENDER TO SWOLLEN JOINT COUNT RATIO A USEFUL CLINICAL MARKER OF RESPONSE TO BIOLOGICAL TREATMENT IN PSORIATIC ARTHRITIS PATIENTS?

Maria Seabra Rato[1], Filipe Oliveira Pinheiro[1], Ana Martins[1], Daniela Santos Oliveira[1], Frederico Rajão Martins[2], Rafaela Nicolau[3], Beatriz Samões[4], Alexandra Bernardo[1], Rita Gaio [5], Miguel Bernardes[1, 6], Lúcia Costa[1]
[1]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [3]Rheumatology Unit, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [4]Rheumatology Department, Centro Hospitalar

de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [5]Departamento de Matemática, Faculdade de Ciências da Universidade do Porto & Centro de Matemática da Universidade do Porto, Porto, Portugal, [6]Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Background: Chronic inflammation in psoriatic arthritis (PsA) may trigger both peripheral and central sensitization via central modifications of pain pathways that can lead to disconnection between tender and swollen joint count. This can result in increased difficulties for the clinician in the assessment of the disease and response to treatment.

Objective: To study the impact of tender to swollen joint count ratio (TSR) on treatment response to a first course of biologic DMARD (bDMARD) therapy in PsA patients.

Methods: Observational study including PsA patients under bDMARD, followed with clinical and laboratory examination at baseline, 6 and 12 months of treatment. All patients meet the CASPAR classification criteria. TSR was defined as the tender joint count divided by the swollen joint count, using the 68/66 joint assessment. Patients with no tender nor swollen joints at baseline were excluded. TSR was categorized into 3 groups, based on the empirical distribution, with cuts corresponding to the 20th and 70th percentiles. Disease activity was assessed using the CDAI, SDAI and DAS28-CRP(4). Individual time profiles were plotted within each TSR group. CDAI, SDAI and DAS28-CRP(4) individual time profiles within each TSR group were modelled by mixed-effects linear regression using the TSR group and time as fixed factors and a random factor at the intercept level (accounting for the intra-individual correlation structure). The identification of the statistically significant pairwise differences was obtained from the Tukey's method for multiple comparisons.

Results: We included 113 patients, 62 (54.0% females) with a mean age of 48.1±10.8 years-old at the start of the first bDMARD. Sixty-four patients (56.6%) had symmetric polyarthritis, 19 (16.8%) spondyloarthritis, 25 (22.1%) asymmetric oligoarthritis, 2 (1.8%) distal arthritis and 1 (0.9%) arthritis mutilans. Forty-three percent were under corticosteroid therapy and 57.5% under conventional synthetic DMARD therapy at baseline. Etanercept (n=35, 31.0%), adalimumab (n=34, 30.1%), golimumab (n=25, 22.1%), infliximab (n=6, 5.3%), certolizumab (5, 4.4%), secukinumab (n=8, 7.1%) were the bDMARD started in these patients. TSR was categorized into 3 groups, namely low [TSR < 1], moderate [1 ≤ TSR ≤ 2.2] and high [TSR > 2.2], with

Table I. Mean values (SD) of CDAI, SDAI and DAS28-CRP(4) during follow-up of PsA patients

	Low (TSR < 1) (n=15)			Moderate (1 ≤ TSR ≤ 2,2) (n=66)			High (TSR > 2,2) (n=32)		
	Baseline	6M	12M	Baseline	6M	12M	Baseline	6M	12M
CDAI	20.1 (2.8)	6.4 (6.4)	5.9 (2,9)	26.7(1.3)	10.8(1.3)	10.3 (1.3)	21.2 (1.9)	12.2 (1.9)	10.1(2.1)
SDAI	22.2(3.0)	6.6 (3.3)	6.2 (3.1)	29.3 (1.3)	11.6 (1.4)	10.8 (1.4)	22.2(11.6)	13.1 (2.0)	10.6 (2.3)
DAS28-CRP(4)	4.2 (0.3)	2.4(0.4)	2.3(0.3)	4.8(0.2)	2.8 (0.2)	2.8 (0,2)	4.2 (0.2)	3.2 (0.2)	2.9 (0.3)

Legend: IMSL: Immune-mediated skin lesions, DILE: drug-induced lupus erythematosus.

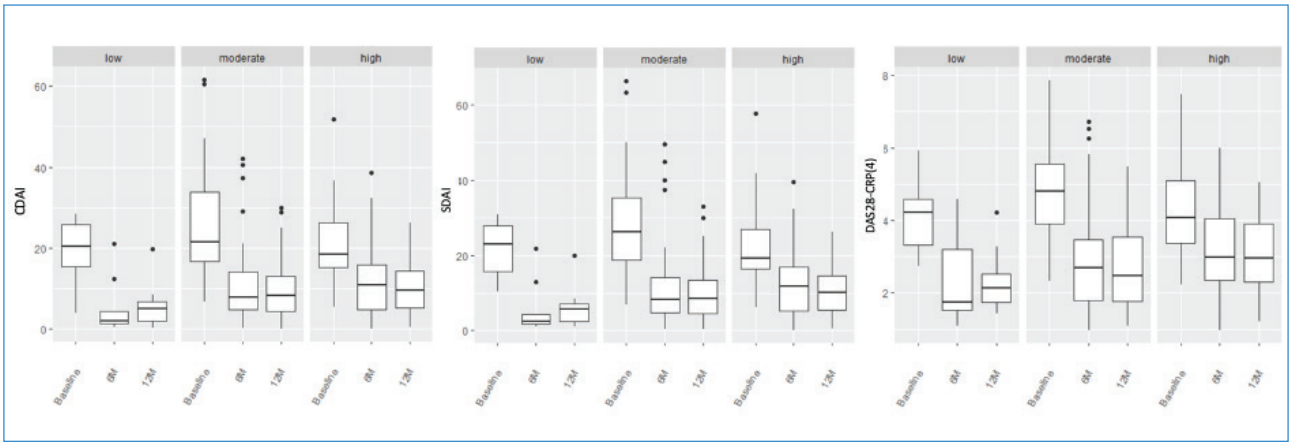


Figure 1. CRP(4) for each TSR group (low, moderate and high) at baseline, 6 and 12 months

frequencies 15 (13.3%), 66 (58.4%) and 32 (28.3%), respectively. Whenever the number of tender joints was different from 0 and that of swollen joints equal to 0, patients were included in the group high TSR. All TSR groups, with initiation of bDMARD, showed significantly decreases at 6 months in CDAI (low: p=0.006, moderate: p<0.001, high: p<0.001), SDAI (low: p<0.001, moderate: p<0,001, high: p<0.001) and DAS28-CRP(4) (low: p<0.001, moderate: p<0.001, high: p<0.001). From 6 to 12 months of treatment, the differences were not significant in any of the groups (p>0.05). At baseline, CDAI, SDAI and DAS28-CRP(4) means did not differ between groups (p>0.05). There were also no differences in the means of outcome measures at 6 months as well as at 12 months of treatment (p>0.05). Despite this, patients with low baseline TSR had lower mean values of CDAI, SDAI and DAS28-CRP(4) at 6 and 12 months of treatment, consistent with a low disease activity.

Conclusions: To our knowledge this is the first study exploring the TSR on treatment response in samples of patients exclusively with PsA. All patients benefited from bDMARD therapy, regardless of the group, suggesting that TSR might not be a good predictor of treatment response in patients with PsA.

146 - GASTROINTESTINAL INVOLVEMENT IN SYSTEMIC SCLEROSIS – PREVALENCE AND HEALTH-RELATED QUALITY OF LIFE IMPACT

Inês Genrinho[1, 2], Inês Santos[1], Maura Couto[1], Carolina Mazeda[2], Anabela Barcelos[2], Adriana Carones[3], Tânia Santiago[3], Maria Rato[4], Georgina Terroso[4], Tiago Beirão[5], Flávio Campos[5], Pedro Ferreira[6], Paulo Monteiro[1]

[1]Serviço de Reumatologia do Centro Hospitalar Tondela Viseu, [2]Serviço de Reumatologia do Centro Hospitalar Baixo Vouga, [3]Serviço de Reumatologia do Centro Hospitalar e Universitário de Coimbra, [4]Serviço de Reumatologia do Centro Hospitalar e Universitário São João, [5]Serviço de Reumatologia do Centro Hospitalar Vila Nova de Gaia/Espinho, [6]Faculdade de Economia da Universidade de Coimbra

Introduction/Aim: Gastrointestinal (GI) disease is a major cause of morbidity and mortality in systemic sclerosis (SSc) and affects up to 90% of patients. Due to its clinical heterogeneity, GI manifestations are frequently unrecognized in early phases resulting in a significant impairment on health-related quality of life (HRQoL). The aim of this study was to determine GI involvement in patients with SSc and its impact on HRQoL.

Methods: A cross-sectional multicenter study was conducted, enrolling SSc patients from five Rheumatology centers that fulfilled ACR/EULAR 2013 classification criteria. Patients were requested to answer UCLA Scleroderma Clinical Trial Consortium Gastrointestinal Tract (UCLA SCTC GIT 2.0), Scleroderma Health Assessment (SHAQ) and Short Form Health Survey (SF36) v2 questionnaires. HRQoL was assessed by the European Quality of Life-5 Dimensions (EQ5D) questionnaire. Social-demographic and clinical data were collected.

The GI involvement was determined by the presence of at least one of the following criteria: GI symptoms for at least 3 of the last 7 days, abnormalities in GI exams and/or use of pharmacological therapies to manage GI symptoms.

General descriptive analysis and independent parametric or non-parametric tests were performed using SPSS Statistics v26.

Results: Eighty nine patients were included, being 73 female (82%), with a mean age of 57.1 ± 12.38 years-old and a mean duration of disease 79.0 ± 75.8 months. 59 patients (66.3%) had limited SSc and 30 (33.7%) diffuse SSc.

Sixty three patients with SSc (70.8%) had GI involvement. Clinical manifestations were present in 47 (52.8%) patients, abnormality GI exams were reported in 29 (32.6%) and pharmacotherapy was used in 33 (37.1%) patients. Thirty three patients (52.4.2%) present more than one criteria for GI involvement.

The most frequent UCLA SCTC domains involved were reflux (54%), distension (44.4%), constipation (33.3%), diarrhea (12.7%) and faecal soilage (4.8%). GI exams determine the presence of dysmotility in 33.3% patients, esophagitis in 22.2%, lower oesophageal sphincter disfunction in 11.1% and gastric antral vascular ectasia in 7.8%. Thirty three patients used GI pharmacotherapy, being proton pump inhibitors the most frequently drug, followed by prokinetics, laxatives and antibiotherapy for small intestinal bacterial overgrowth. Five patients needed more than one pharmacological approach.

Patients with GIT involvement presented higher UCLA scores in all domains, with exception of faecal soilage. Severe disease was found in 10 patients (15,9%). EQ5D, SF36 and several SHAQ domains (VAS for pain, intestinal problems, Raynaud and disease gravity) presented higher scores when GIT were involved. No differences were found in disease duration between groups.

Discussion/Conclusion: The overall prevalence of GI involvement in SSc patients was 70,8%, similar from other studies reported in literature. GI manifestations negatively affected HRQoL in physical and mental domains, increased pain VAS and were associated to severe disease.

157 - FOOT INVOLVEMENT IN PSORIATIC ARTHRITIS- A RETROSPECTIVE STUDY

Sofia Ferreira Azevedo[1], Carolina Mazeda[1], Susana P. Silva[1], C. Pinto Oliveira[1], Anabela Barcelos[1]
[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal

Introduction: Foot involvement is well recognized in many inflammatory rheumatic diseases, namely in Spondylarthritis (SpA), where its early recognition is of utmost importance. Psoriatic arthritis (PsA), although included in SpA group, is an individual entity with characteristic features. Foot involvement in PsA is frequent and often develops early in the disease's course, proving diagnostic and prognostic information.
The purpose of this study was to evaluate and describe foot involvement in a group of patients with PsA.
Materials and Methods: Single-center, retrospective study including patients with PsA (all patients fulfill CASPAR criteria) followed in our center over a period of five months (from January-May 2022). Patients were divided into two groups: with current or previous foot involvement (assessed clinically or by ultrasound) (group 1) and without current or previous foot involvement (group 2). Sociodemographic, clinical, laboratory, and radiological data were collected.
Descriptive analysis was performed using means and standard deviation (SD), medians and Interquartile range (IQR) for continuous data, and frequencies and percentages for qualitative variables.
Clinical, laboratory and radiological findings were compared between patients with and without foot involvement using parametric and non-parametric tests, with a p-value ≤ 0.05, with SPSS® software.
Results: 150 patients were enrolled. The mean age was 56.92 years, and 38.7% were women, without statistically significant differences between groups. Foot involvement was found in 104 patients (69.3%). Arthritis was found in 93 patients (62.0%), with the tibiotarsal joint as the most frequent site (30.0% of patients). Enthesitis was found in 28.0% (42 patients), with calcaneal tendonitis as the most frequent manifestation (10.6% of patients). 18.6% (28 patients) had current/previous dactylitis. Radiological findings showed osteopenia in 26.6% of patients, symmetrical joint space narrowing in 12.0%, 20.0%, and 10.0% in tibiotarsal, metatarsophalangeal and Interphalangeal joints, respectively. Erosions were found in 28.6% of patients.
Extra-articular manifestations were significantly more prevalent in the group with foot involvement (p=0.03)
We found statistically significant higher HAQ disability index values in group 1 [median 1.00, IQR 0.875 (group 1) VS median 0,0625, IQR 0.875 (group 2); p<0.01]; Multimorbidity (defined as 2 or more comorbidities) was also more frequent in group 1 [58.65 % (group 1) VS 37.70% (group 2); p=0.02];
Patients with foot involvement had higher C-reactive protein (CRP) [median CRP 1.17, IQR 1.58 (group 1) VS median CRP 0.26, IQR 0.56 (group 2); p=0.01] and erythrocyte sedimentation rate (ESR) [Median ESR 28.00, IQR 21.00 (group 1) VS Median ESR 8.00, IQR 17.00; p=0.01)] levels, and were more frequently under corticosteroids (p=0.01) and non-steroidal anti-inflammatory drugs (NSAIDs) (p=0.02).
Conclusion: Our results suggest that foot involvement is frequent in patients with PsA and is associated with the presence of multimorbidity and extra-articular manifestations, as well as with higher rates of steroids and NSAIDs.
Patients with foot involvement had higher HAQ disability index levels, reflecting the negative impact of foot involvement in daily functionality in these patients. Considering that, our study highlights the importance of using activity indices that include foot involvement to better represent the disease's activity in PsA.

160 - LOW DOSE COMPUTED TOMOGRAPHY HOUNSFIELD UNITS: A RELIABLE METHODOLOGY FOR ASSESSING CHANGES IN VERTEBRAL BONE DENSITY IN RADIOGRAPHIC AXIAL SPONDYLOARTHRITIS

Mary Lucy Marques[1, 2], Nuno Pereira da Silva[3], Désirée van der Heijde[1], Monique Reijnierse[4], Juergen Braun[5], Xenofon Baraliakos[5], F vanGaalen[1], Sofia Ramiro[1, 6]
[1]Rheumatology, Leiden University Medical Center, Leiden, Netherlands, [2]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [3]Serviço de Radiologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [4]Radiology, Leiden University Medical Center, Leiden, Netherlands, [5]Department of Rheumatology, Rheumazentrum Ruhrgebiet, Herne, and Ruhr-University Bochum, Bochum, Germany, [6]Rheumatology Department, Zuyderland Medical Center, Heerlen, Netherlands

Background: In radiographic axial Spondyloarthritis (r-axSpA), low dose Computed Tomography (ldCT) Hounsfield Units (HU) were shown to cross-sectionally reliably assess bone density at each vertebra from C3 to L5.[1] However, HU change scores have never been studied.
Aims: In the present study we aimed to describe ldCT HU 2-year change scores and analyse inter-reader reliability per vertebra.
Methods: We used 49 patients with r-axSpA from the multicentre 2-year Sensitive Imaging in Ankylosing Spondylitis (SIAS) study. A standardized protocol

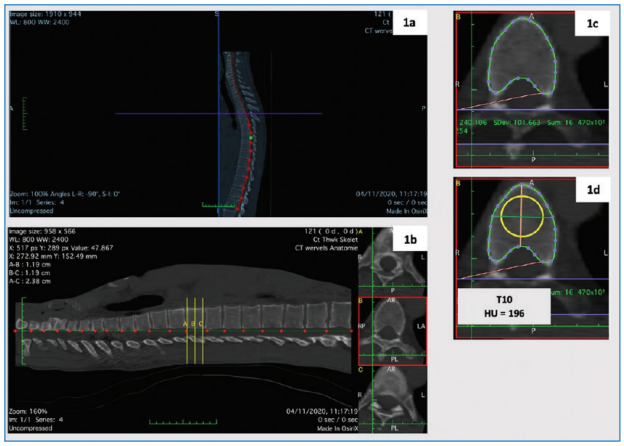


Figure 1. Methodology of low dose Computed Tomography Hounsfield Units (HU) measurement la: Using a three-dimensional curved-multiplanar reconstruction, the curve of the spine adjacent to the spinal canal was delimited. lb: On the obtained sagittal image, each vertebra (from C3 to L5) was identifiable. At each vertebra, two lines of reference were 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 was automatically positioned by the software at the center of the vertebral body. lc: In the reconstructed cross-sectional slice, the vertebral body was manually delimited. 1d: A region of Interest was manually selected, having a diameter equal to 75% of the average of anteroposterior and transverse diameters. The density of the vertebra was displayed by the software as the average image intensity within the sample region, reported in HU.

and automatic exposure control calibration in ldCT imaging acquisition were used. HU measurements were independently assessed by two trained readers at baseline and two years, at each vertebra from C3 to L5 (independent reading sessions ≥3 months apart) – Figure 1. Mean (standard deviation, SD) for the change-from-baseline HU scores were provided per vertebra by reader. Intraclass correlation coefficients (ICC; absolute agreement, two-way random effects), Bland-Altman plots and smallest detectable change (SDC) were obtained. Percentages of vertebrae in which readers agreed on direction of change and on change scores >|SDC| were computed.

Results: Overall, 1,053 (98% of all possible) vertebrae were assessed at both time-points by each reader. Over two years, HU mean change values varied from -23 to 28 and 29 for reader 1 and 2, respectively – Table 1. Inter-reader reliability of the change scores per vertebra was excellent: ICC: 0.91 to 0.99; SDC: 6 to 10; Bland-Altman plots were homoscedastic, with negligible systematic error between readers. Readers agreed on the direction of change-score in 88-96% and on change-scores >|SDC| in 58-94% of vertebrae, per vertebral level, from C3 to L5. Overall, similar results were obtained across all vertebrae throughout the spine.

Conclusion. LdCT measurement of HU is a reliable method to assess changes in bone density at each vertebra from C3 to L5. Being reliable across all vertebrae, this methodology can aid the study of bone density changes in r-axSpA, a disease affecting the whole spine.

REFERENCES:

1. Marques ML, et al. Arthritis Rheumatol. 2021; 73 (suppl 10).

SIAS study was funded by the Dutch Rheumatism Association ("ReumaNederland"). MLM is supported by the Fundação para a Ciência e Tecnologia (FCT) grant SFRH/BD/143744/2019.

162 - ARTRITE IDIOPÁTICA JUVENIL – UMA DÉCADA DE SEGUIMENTO NO HOSPITAL DE FARO

Ana Teodósio Chícharo[1], Graça Sequeira[1]

[1]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal

A artrite idiopática juvenil (AIJ) integra um grupo heterogéneo de distúrbios inflamatórios, ainda sem etiologia conhecida, com envolvimento primordial das articulações. O diagnóstico assenta na persistência de artrite, por mais de 6 semanas, numa criança com menos de 16 anos e na exclusão de outras causas de artrite na infância (artrite reativa, artrite viral, lúpus eritematoso sistémico, entre outras). De acordo com a classificação ILAR, a AIJ pode ser subdividida em várias entidades diferentes: a AIJ sistémica, oligoarticular, poliarticular seropositiva para FR (fator reumatóide), poliarticular seronegativa para FR, psoriática, artrite relacionada com entesite e artrite indiferenciada. Cada uma destas condições implica uma referenciação precoce a consulta de Reumatologia, para um seguimento e tratamento adequado. A AIJ é uma doença rara, mas representa o diagnóstico reumatológico mais comum na idade pediátrica, com uma prevalência de 0.07-4.01 por 1000 crianças, e a incidência anual estimada de 0.008–0.226 por cada 1000 crianças.

Neste trabalho, as autoras propõem-se a caracterizar a população de crianças com o diagnóstico de AIJ, de acordo com os critérios mencionados acima, que foi seguida no Hospital de Faro entre 2011 e 2021.

Trata-se de um estudo observacional com uma análise retrospetiva, tendo os dados sido obtidos através da consulta de registos clínicos hospitalares e do Reuma.pt. O estudo obteve o parecer favorável da Comissão de Ética e do Conselho de Administração do CHUA.

Foram incluídas 35 crianças com o diagnóstico de AIJ, 20 (57.1%) do sexo feminino e 15 (42.9%) do sexo masculino. Em relação aos subtipos de AIJ, o mais frequente foi o oligoarticular (19, 54.3%), seguido da AIJ relacionada com entesite (8, 22.9%), da AIJ sistémica (4, 11.4%), da AIJ poliarticular seronegativa (3, 8.6%)

Tabela I. Características clínicas e demográficas das crianças com diagnóstico de Artrite Idiopática Juvenil seguidas no Hospital de Faro entre 2011 e 2021.

	AIJ oligoarticular	AIJ poliarticular seropositiva	AIJ poliarticular seronegativa	AIJ sistémica	AIJ relacionada com entesite	Total
Dados demográficos						
Frequência - N (%)	19 (54.3)	1 (2.9)	3 (8.6)	4 (11.4)	8 (22.9)	35 (100)
Idade 1os sintomas - média em anos (DP)	5.8 (4.3)	4.3 (-)	14.1 (3.0)	5.1 (3.3)	12.6 (3.5)	8.0 (5.12)
Idade ao diagnóstico- média em anos (DP)	6.4 (4.6)	11.3(-)	14.8 (3.0)	5.6 (3.3)	13.0 (3.5)	8.6 (5.2)
Sexo feminino – N (%)	14 (73.7)	1 (100)	3 (66.7)	2 (50.0)	1 (12.5)	20 (57.1)
Manifestações extra-articulares						
uveíte – N (%)	1 (5.2)	0	0	0	1 (12.5)	2 (5.7)
dactilite – N (%)	2 (10.5)	0	0	0	2 (25.0)	4 (11.4)
colite ulcerosa – N (%)	2 (10.5)	0	0	0	0	2 (5.7)
colite inespecífica-N(%)	0	0	0	0	1 (12.5)	1 (2.9)
febre+ exantema-N (%)	0	0	0	4 (100)	0	4 (11.4)
Anticorpos - ANA						
Positivo	8 (42.1)	0	0	0	1 (12.5)	9 (25.7)
Negativo	9 (47.4)	1 (100)	1 (33.3)	2 (50.0)	7 (87.5)	20 (57.1)
Desconhecido	2 (4.84)	0	2 (66.7)	2 (50)	0	6 (17.2)
Anticorpos – Fator Reumatóide						
Positivo	0	1 (100)	0	0	0	1 (2.9)
Negativo	15 (78.9)	0	3 (100)	2 (50.0)	8 (100)	28 (80.0)
Desconhecido	4 (21.1)	0	0	2 (50.0)	0	6 (17.1)
Anticorpos – anti-CCP						
Positivo	0	1 (100)	1 (33.3)	0	0	2 (5.7)
Negativo	11 (57.9)	0	2 (66.7)	2 (50.0)	3 (37.5)	18 (51.4)
Desconhecido	8 (42.1)	0	0	2 (50.0)	5 (62.5)	15 (42.9)
Anticorpos – HLA-B27						
Positivo	1 (5.2)	0	0	0	7 (87.5)	8 (22.8)
Negativo	7 (36.8)	1 (100)	1 (33.3)	2 (50.0)	1 (12.5)	12 (34.3)
Desconhecido	11 (57.9)	0	2 (66.7)	2 (50.0)	0	15 (42.9)
Terapêutica com Biológicos						
Sim	3 (15.8)	0	0	0	1 (12.5)	4 (11.4)
Não	16 (84.2)	1 (100)	3 (100)	4 (100)	7 (87.5)	31 (88.6)
Terapêutica com cDMARDs						
Sim	11 (57.9)	1 (100)	2 (66.7)	2 (50.0)	2 (50.0)	20 (57.1)
Não	8 (42.1)	0	1 (33.3)	2 (50.0)	2 (50.0)	15 (42.9)

DP = desvio padrão; cDMARDs = conventional disease-modifying antirheumatic drugs; Nota: Na AIJ poliartricular seropositiva não se apresenta DP na idade dos primeiros sintomas ou idade ao diagnóstico por haver apenas 1 doente nestas condições.

e da AIJ poliarticular seropositiva (1, 2.9%). Não se observaram casos de AIJ psoriática ou indiferenciada. A média de idade de início de sintomas foi 7.97 anos, enquanto a idade média de diagnóstico foi 8.65 anos.

A maioria das crianças foi referenciada para Reumatologia Pediátrica através do seu médico de família (11, 31.4%) ou da Consulta de Pediatria Geral do Hospital de Faro (7, 20.0%).

Foram reportadas as características clínicas, demográficas e laboratoriais destes doentes, incluindo as manifestações articulares e extra-articulares e a presença de anticorpos específicos (tabela 1).

A revisão da terapêutica administrada a estes doentes mostrou que uma minoria das crianças (4, 11.4%) re-

Origem da referenciação	
MGF– N (%)	11, 31.4
CE Pediatria Geral– N (%)	7, 20.0
Outra CE– N (%)	5, 14.3
Internamento Pediatria– N (%)	
Urgência Pediatria– N (%)	5, 14.3
Outro Hospital– N (%)	2, 5.7
Seguimento	
Em CE Reumatologia Pediátrica– N (%)	13 (37.1)
Em CE Reumatologia Geral– N (%)	
Transferência para outro hospital– N (%)	1 (2.9)
Alta– N (%)	2 (5.7)
Perda de seguimento– N (%)	11 (31.4)

cebeu terapêutica biológica, sendo o Etarnecept o mais utilizado como primeira escolha. No final de 2021, apenas 2 crianças se mantinham sob terapêutica biotecnológica, ambas sob Infliximab. Foram também identificadas outras terapêuticas administradas, nomeadamente DMARD's como o Metotrexato (19, 54.3%) e a Salazopirina (3, 8.6%), bem como outros imunomoduladores.

Realizou-se um levantamento da evolução destes doentes com o registo da atividade da doença no início e no final do seguimento. No final dos 10 anos de consultas, constatou-se que 13 (37.1 %), mantém seguimento em Reumatologia Pediátrica, 8 (22.9%) foram transferidos para consulta de Reumatologia Geral por terem atingido os 18 anos, 2 tiveram alta (5.7%), 1 foi transferido para outro Hospital (2.9%) e 11 (31,4%) perderam seguimento por faltas consecutivas.

Este trabalho constitui a primeira caracterização dos doentes seguidos com o diagnóstico de AIJ no Hospital de Faro, contribuindo para um melhor conhecimento da população pediátrica seguida neste centro.

166 - ANTI-NOR 90 ANTIBODIES: WHAT IS THE CLINICAL SIGNIFICANCE IN RHEUMATIC INFLAMMATORY CONDITIONS?

Joao Paulo Vilas-Boas[1, 2], Gisela Eugénio[1, 2], Anabela Barcelos[1, 2]

[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz Health Alliance, Portugal, Aveiro, Portugal

Background: Anti-NOR 90 antibodies have been detected in inflammatory conditions, neoplastic diseases and even in healthy individuals. Despite numerous epidemiologic studies, large scale studies are not available and the clinical relevance of the Anti-NOR 90 antibodies remains unclear. This antibody tends to be found in Systemic Sclerosis (SSc), overlap syndromes, as well as in other systemic autoimmune rheumatic diseases (SARD). Anti-NOR 90 can be found in 4.8% of patients with SSc and it may be considered a marker of limited cutaneous SSc and mild involvement of internal organs. Not all published studies agree about its specificity in SARD.

Objectives: The aim of this study is to identify the clinical and analytical manifestations associated to the presence of Anti-NOR90 in patients with SARD.

Methods: A retrospective study of patients with Anti-NOR90 positivity (detected by immunoblot assay), observed from january 2017 to december 2021, from a single Portuguese rheumatology outpatient centre, was included. Demographic, clinical, and immunological data at presentation and during follow-up were collected. A descriptive analysis was made.

Results: Forty patients were positive for anti-NOR90. SARD was diagnosed in 30% of the patients (SSc: n=3; undifferentiated connective tissue disease: n=3; spondyloarthritis: n=2; polymyalgia rheumatica: n=1; rheumatoid arthritis: n=1; sjogren´s disease: n=1; systemic lupus erythematous: n= 1), 7.5% had other autoimmune disease (primary biliary cholangitis: n=2; autoimmune tiroiditis: n=1) and 62.5% had no pathologic conditions. In the SARD group, 58.3% were female and the median age was 49.2 ± 11.2 years. The most frequent presenting clinical features was inflammatory arthralgias (50%), followed by Raynaud phenomenon (16%), arthritis (8.3%), photosensitivity (8.3%), sclerodactyly (8.3%) and xerostomia (8.3%). During follow-up, musculoskeletal involvement was present in 41.6% (n=5) of the patients, peripheral vascular involvement (detected by nailfold capillaroscopy) in 33% (n=4), gastrointestinal (esophageal hypomotility) and lung involvement (interstitial lung disease) was detected in 16% (n=2) and 8.3% (n=1) of the patients, respectively. Half of the SARD patients showed other antibodies specificities: anti-centromere (n=3), anti-Ku (n=1), anti-SSB (n=1), anti-fibrillarin (n=1), anti-Pm/Scl (n=1), anti- ribosomal P (n=1). Persistent elevated inflammatory markers were present in 58.3%, leucopenia in 16% and low complement levels in 8.3%.

Conclusion: Anti-NOR 90 was detected in diverse SARD (most common in SSc and undifferentiated connective tissue disease). Musculoskeletal and peripheral vascular symptoms were the most prevalent associated clinical features. A multicentre national study with a larger sample, to better understand the role of this antibody, is currently ongoing.

168 - COMPREHENSIVE ASSESSMENT OF PATIENTS WITH SUSPECTED SJÖGREN'S SYNDROME: 5-YEAR RESULTS OF A MULTIDISCIPLINARY SJÖGREN'S SYNDROME CLINIC

Vasco C Romão[1, 2], Matilde Bandeira[1, 2], Manuel Silvério-António[1, 2], Rita Simão[3], Joana Pinto[4], Ana Inês Gonçalves[4], Maria João Gonçalves[1, 2], Ana Silva Martins[3], Patricia Jose[4], Lila Morena Bueno Silva[1], Miguel Brito Lança[2], Gonçalo Coutinho[3], Raquel Esteves Marques[4], Carla Macieira[1], Nikita Khmelinskii[1, 2], Walter Rodrigues[4], Francisco Salvado[3], 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]Serviço de Estomatologia, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [4]Serviço de Oftalmologia, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Background: Primary Sjögren's syndrome (pSS) is a systemic rheumatic disease that affects several organ systems, most frequently the ocular, oral and musculoskeletal domains. Multidisciplinary care is thus crucial in the optimal management of SS patients.
Objectives: To report the clinical impact of a Multidisciplinary SS Clinic (MSSC) over a 5-year period.
Methods: We prospectively included patients assessed in the MSSC from September 2015 to October 2020. All patients had a full clinical evaluation, including disease-related questionnaires, specialized oral/ocular assessment, salivary gland biopsy (SGB) and ultrasound (SGUS), tear and salivary flow and ocular staining scores. We compared the results of patient-reported outcomes, comprehensive clinical assessments and specialized complementary exams in patients with pSS and other diagnoses.
Results: 445 patients (96% women, mean age 57±14 years) with sicca symptoms underwent complete multidisciplinary evaluation. Patients were most frequently referred from Rheumatology (91%), but also from Stomatology (n=5%), Ophthalmology (n=2%), Internal Medicine (1%) and other medical specialties (1%). Most patients were diagnosed with pSS (n=221; 50%), followed by non-Sjögren sicca syndrome (nSSS, n=134; 30%), secondary SS (sSS, n=60; 13%) and undifferentiated connective tissue disease (n=30; 7%). Positive sicca tests were present in 217/385 patients (56%): unstimulated salivary flow (USF) ≤0.1ml/min in 84/317 (27%); Schirmer's test ≤5mm/5min in 163/354 (46%); van Bijsterveld score ≥4 in 42/349 (12%); Ocular Staining Score (OSS) ≥5 in 36/343 (11%). Subjective complaints assessed by the EULAR Sjögren Syndrome Patient Reported Index (ESSPRI), the EULAR Sicca Score (ESS), the Profile in Fatigue and Dryness in SS Index (PROFAD-SSI), the Xerostomia Inventory (XI), and the Ocular Surface Disease Index (OSDI) did not differ between patients with pSS and other diagnoses. However, objective dryness measures such as USF (31% vs 20%, p=0.028), Schirmer's test (51% vs 40%, p=0.040), and OSS (14% vs 7%, p=0.048) were significantly associated with pSS. A positive SGB (focus score ≥1) was seen in 48% of patients with a clinical diagnosis of pSS (p<0.001 vs. other diagnoses), with a mean focus score of 1.1±1.6. Instead, 94% of patients with nSSS had grade 0-1 biopsies. Mean SGUS scores (p=0.006) and the frequency of moderate/severe changes (p<0.001) were higher in pSS patients.
Conclusions: Multidisciplinary evaluation was crucial in the assessment of patients with similar sicca complaints and in the management of ocular/oral/systemic involvement. Objective measurements and specialized complementary exams greatly contribute to establishing or confirming the diagnosis of pSS.

169 - I.FIT.NESSPROJECT: INTERNET INTERDISCIPLINARY EDUCATION FOR SYSTEMIC SCLEROSIS

Rúben Duarte Fernandes[1], Ricardo J. O. Ferreira[2], Paulo Costa[2], Tânia Santiago[1, 3]
[1]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [2]Health Sciences Research Unit: Nursing (UICiSA:E), Coimbra, Portugal, [3]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal

Introduction: People with Connective Tissue Diseases (CTD) including Systemic Sclerosis (SSc) often experience fatigue, pain, reduced mobility/range of motion, dyspnoea and decreased functional capacity. This leads to a significant decrease in quality of life (QoL) through its psychosocial, emotional, social and economic impact. Non-pharmacological interventions (NPI) contribute to better disease management. Telehealth interventions or remote consultations with a multidisciplinary team aimed at promoting physical activity (PA) may contribute to improving the QoL of these people with CTD.
Objectives: To develop a multidisciplinary online (telehealth) project on teaching healthy lifestyle habits to people with SSc, with a focus on PA promotion and to test its operationality in 4 European countries.
Methodology: Methodological study implementation of

care improvement project. Strategy developed in 3 phases. Phase 1: Scoping review (Suitability for e-health of non-pharmacological interventions in connective tissue diseases: scoping review with a descriptive analysis). This study adds pertinent information to the current landscape of treatment of CTDs, such as SSc, on the existence and importance of varied NPI and programs for the limitations of people with SSc, but not for the generality of CTDs. The most common interventions include patient education, self-management, PA/exercise and advice on healthy lifestyle.
Phase 2: programme development and video recording of content in English (subtitles in 4 languages), covering topics ranging from general health advice, to the specifics of developing an individual PA plan (range-of-motion gain, strength and aerobic exercises), and how to deal with everyday situations.
Phase 3: effectiveness and usefulness evaluation of the programme by means of satisfaction questionnaires and testimonials (patients and stakeholders).
Results: Contribute to answer the question regarding feasibility and suitability of telehealth services. Phase 1 allowed mapping interventions described in the literature in this thematic scope. It was possible for the project team to design and structure a digital intervention different from the existing educational offer, with innovative contributions.
Phase 2 is currently under development, dealing with specific NPIs for an individual PA plan (range of movement, strength and aerobic exercises) and how to deal with everyday situations.
Discussion: NPI are associated with low adherence rates because they involve significant changes to lifestyle habits and need to tailor these interventions to people's different particularities and clinical manifestations. A multidisciplinary approach using technological tools (telehealth) that allow a greater proximity and globality may be the necessary support to NPI complement of people with SSc. These interventions aim to meet the different needs of patients and allow users easy access to specialised health professionals.
Conclusion: The I.FIT.NESS project appears as an innovative digital NPI in the international context, aiming at promoting healthy lifestyles and disease management in people with SSc.
Its current discussion and continuous refinement, based on feedback from multidisciplinary experts and citizens with SSc, will allow the team to develop a program with applicability and meaning for its end-users.
Despite its structuring and development based on recent evidence and feedback from relevant stakeholders, it is essential to complete the last design phase in order to anticipate any weaknesses or gaps before making it available in open access.

172 - ACUTE PERICARDIAL AND MYOCARDIAL DISEASE IN SSC: LESSONS FROM A MULTICENTRE CASE SERIES

Marcelo Neto[1], Fernando Albuquerque[1], Maria Seabra Rato[2], Tiago Beirão[3], Ana Cordeiro[4], Carla Campinho Ferreira[5], Ana Teodósio Chícharo[6], Manuel Silvério-António[7, 8], Susana P. Silva[9], Dr[10], Raquel Miriam Ferreira[2], P Pinto[3], Joana Leite Silva[5], Graça Sequeira[6], Catarina Resende[7], Anabela Barcelos[9], Maria João Salvador[1], Tânia Santiago[1]
[1]Serviço de Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar de São João, Porto, Portugal, [3]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [4]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal, [5]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal, [6]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal, [7]Serviço de Reumatologia e Doenças Ósseas Metabólicas, 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, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [9]Serviço de Reumatologia, Centro Hospitalar do Baixo Vouga, E.P.E., Aveiro, Portugal, [10]Serviço de Reumatologia, Centro Hospitalar Lisboa Ocidental | Hospital Egas Moniz, Lisboa, Portugal

Background: Acute, life-threatening cardiac manifestations of systemic sclerosis (SSc) are rare and can present as severe pericardial effusion (with or without cardiac tamponade) and myocarditis. Early signs of cardiac involvement can be subtle, and distinguishing them from other causes of cardiomyopathy remains challenging.
Objectives: To describe the clinical features, treatment, and outcome of patients with SSc developing severe pericardial effusion or acute myocardial involvement.
Methods: A retrospective analysis of patients from 14 Portuguese tertiary centres, with SSc (fulfilling 2013 ACR/EULAR classification criteria), presenting with hemodynamically significant pericardial effusion or myocardial involvement was conducted. Diagnosis of the acute event was based on clinical, laboratory, and imaging features. Exclusion criteria were 1) a more probable cause than SSc for the acute event, including other autoimmune disorders, infections, neoplasms, or non-SSc-related heart disease or pulmonary hypertension; and 2) incomplete data regarding SSc diagnosis or the diagnostic workup of the heart involvement.
Results: Eight centres reported a total of 17 patients, of which 6 did not fulfil the eligibility criterion and 4 were excluded based on the exclusion criteria (1 by in-

Table I. Demographic and clinic characteristics of included patients

Patient	Gender	Age at SSc diagnosis (years)	Age at time of the event (years)	SSc subtype	Autoimmunity profile	Acute event
1	M	37	37	Diffuse	AC-29; Scl-70+	Cardiac tamponade
2	F	24	25	Limited	AC-1; Scl-70+	Cardiac tamponade
3	F	31	31	Diffuse	Nucleolar; Scl-70+	Acute myocarditis
4	F	35	45	Limited	Nuclear	Cardiac tamponade
5	F	27	29	Diffuse	AC-1; Scl-70+	Acute myocarditis + Moderate pericardial effusion
6	M	42	52	Limited	Nucleolar; Scl-70+	Severe pericardial effusion
7	M	61	64	Limited	Nucleolar; Scl-70+	Myocardial fibrosis

M – male; F – female; SSc – systemic sclerosis; AC-1 – nuclear homogenous AN ANA pattern; AC-29 – Topo-I like ANA pattern.

Table II. Demographic and clinical characteristics of the whole cohort

Female gender, N (%)	4 (57,1)
Age at SSc diagnosis, mean ± SD (years)	36,7±12,3
Age at time of the event, mean ±SD (years)	40,4±14,0
Inaugural manifestation, N (%)	2 (28,6)
SSc subtype	
Limited, N (%)	4 (57,1)
Diffuse, N (%)	3 (42,9)
Type or cardiac involvement	
Cardiac effusion, N (%)	4 (57,1)
Tamponade, N (%)	3 (75)
Myocardial, N (%)	3 (42,9)
Acute myocarditis, N (%)	2 (66,7)
Fibrosis, N (%)	1 (33,3)
Autoimmunity profile	
ANA positivity, N (%)	7 (100)
Nucleolar, N (%)	3 (42,9)
Nuclear homogeneous, N (%)	2 (28,6)
Topo-I like, N (%)	1 (14,3)
Nuclear, N (%)	1 (14,3)
Scl-70 positivity, N (%)	6 (85,7)

complete data, and 3 had a more probable cause for the acute event than primary SSc heart involvement). Six centres reported no patients fulfilling the eligibility criterion.

Demographic and clinical features of the patients included are presented in table 1 and 2. We included 7 patients: 57,1% female; mean age at SSc diagnosis 36,7±12,3 years, age at the acute cardiac event 40,4±14,0 years, 4 (57,1%) limited cutaneous SSc. All patients were positive for antinuclear antibodies, and 6/7 were Scl-70 positive. In 2 patients, the acute cardiac event was simultaneous to SSc diagnosis.

Pericardial effusion was present in 5/7 patients (71,4%), 3 of which presenting with cardiac tamponade. In these 3 cases, pericardial fluid analysis revealed an exudate.

Myocardial involvement was present in 3/7 patients (42,9%), 2 had an acute myocarditis and 1 had an extensive myocardial fibrosis, all confirmed by histopathology. One patient had concomitant pericardial effusion.

Cardiac MRI was performed in the 3 patients with myocardial involvement, revealing chamber enlargement and systolic dysfunction in all of them and late myocardial enhancement in 2. One patient with cardiac tamponade performed MRI 5 months after the event, revealing only mild systolic dysfunction.

All patients were hospitalized, with median stay of 17 days (IQR 10-23). Besides supportive therapy, 6/7 patients received corticosteroids (>10mg/d prednisolone or equivalent), and the 3 patients with cardiac tamponade also initiated colchicine. Induction treatment with cyclophosphamide was started on 2 patients (in one case oral and the other IV pulses). There was an improvement with no relapse in 6/7 patients, with follow-up ranging from 6 months to 8,6 years (median 13 months). One patient (with extensive myocardial fibrosis) was hospitalized 4 months after the acute event due to cardiac complications and died 2 months later.

Conclusions: Severe pericardial effusion and myocardial involvement are rare primary cardiac complications of SSc; however, they may occur at different stages of disease and can be the first manifestation. Early identification is crucial for a prompt intervention to avoid progression to heart failure. Optimal screening and treatment guidelines are lacking, representing an area of much-needed research.

ACKNOWLEDGEMENTS

The authors would like to thank the following members of the GEDRESIS/SPR group for their collaboration: Drª A. Neto, Drª Filipa Farinha, Dr. Filipe Araújo, Drª Marília Rodrigues and Drª Teresa Nóvoa.

179 - ENVOLVIMENTO PULMONAR NA ARTRITE REUMATOIDE

Constança Durão[1, 2, 3], Ana Catarina Duarte[1, 2, 3], Maria José Santos[1, 2, 3, 4]

[1]Faculdade de Medicina da Universidade de Lisboa, Lisboa, Portugal, [2]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal, [3]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [4]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 Artrite Reumatoide (AR) é uma doença reumática sistémica, de etiologia desconhecida, caracterizada, principalmente, pela inflamação crónica e pro-

Tabela I. Caracterização dos doentes com e sem envolvimento

	Variáveis	Sem envolvimento pleuropulmonar	p value
Idade atual (anos)			
(média ± desvio padrão)	65,6 ± 14,0	69,3 ± 10,5	0,006
Sexo masculino	105 (21,5%)	19 (24,4%)	0,573
Idade de início da AR (anos)			
(média ± desvio padrão)	47,6 ± 15,2	49,7 ± 14,4	0,454
Duração da AR (anos)			
(média ± desvio padrão)	18,1 ± 10,7	19,6 ± 9,3	0,093
IMC (Kg/m2)			
(média ± desvio padrão)	27,5 ± 5,4	28,2 ± 5,2	0,857
Fumador	127 (31,2%)	27 (36,0%)	0,413
Profissão de risco	11 (2,9%)	2 (2,7%)	0,914
Comorbilidades (≥1)	265 (63,7%)	45 (57,7%)	0,314
Doença pulmonar prévia	28 (5,7%)	9 (11,5%)	0,054
Seropositividade	378 (79,6%)	71 (92,2%)	0,008
Outras MEA	51 (10,4%)	17 (22,1%)	0,003
Doença erosiva	197 (53,8%)	44 (61,1%)	0,256
Terapêutica prévia com metotrexato	304 (62,2%)	57 (73,1%)	0,063
Terapêutica prévia com Anti-TNF	196 (40,1%)	27 (34,6%)	0,359

gressiva das articulações. O envolvimento pleuropulmonar é uma manifestação extra-articular frequente, com uma prevalência entre 5 a 30%.

Objetivos: Caracterizar a prevalência e o tipo de envolvimento pleuropulmonar; identificar diferenças nas características demográficas e clínicas, exposições prévias e comorbilidades entre doentes com e sem envolvimento pleuropulmonar, e entre os diferentes tipos de envolvimento pleuropulmonar; descrever achados imagiológicos e terapêutica instituída para a doença pulmonar; e descrever a evolução clínica dos doentes com envolvimento pleuropulmonar.

Metodologia: Estudo retrospetivo, realizado através da recolha de dados clínicos, laboratoriais e imagiológicos de doentes com diagnóstico de AR seguidos na Consulta de Reumatologia do Hospital Garcia de Orta. Aplicaram-se testes estatísticos, nomeadamente teste t de student, U de Mann-Whitney e qui-quadrado ou teste exato de Fisher, conforme apropriado. Os resultados foram considerados como estatisticamente significativos para um valor de $p<0,05$.

Resultados: A prevalência do envolvimento pleuropulmonar foi de 13,8%. Os doentes com envolvimento pulmonar são mais velhos (69,2 ± 10,5 anos vs 65,6 ± 14,0 anos; p=0,006), apresentam mais frequentemente manifestações extra-articulares (22,1% vs 10,4%; p=0,003) e positividade para FR e/ou ACPA (92,2% vs 79,6%; p=0,008). A doença pulmonar intersticial (DPI) foi a manifestação pulmonar mais comum, constituindo 49,0% dos casos. Dos doentes com DPI, 36,4% exibia padrão UIP e 11,4% padrão NSIP, e após o diagnóstico de DPI, 20,5% iniciou rituximab, 11,4% abatacept e 11,4% um fármaco anti-fibrótico. A taxa de letalidade da doença pulmonar foi de 7,5%.

Conclusões: A DPI foi a manifestação pulmonar mais prevalente, sendo o padrão UIP o mais comum. A idade avançada, a seropositividade para FR e/ou ACPA, bem como as MEA estão associadas ao envolvimento pulmonar. A combinação de terapêutica imunossupressora e anti-fibrótica pode apresentar benefício no tratamento da DPI associada à AR.

189 - EFETIVIDADE DO USTECINUMAB NAS ESPONDILARTRITES ASSOCIADAS À DOENÇA DE CROHN - DESCRIÇÃO DE 4 CASOS CLÍNICOS.

Bruna Silva[1], Bruno Miguel Fernandes[1], Rafaela Nicolau[2], Daniela Santos Oliveira[1], Frederico Rajão Martins[3], Miguel Bernardes[1, 4], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar de São João, Porto, Portugal, [2]Rheumatology Unit, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [3]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [4]Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Introdução: O Ustecinumab (UST) é um agente biotecnológico que bloqueia a subunidade p40 das inlerleucinas 12 e 23, encontrando-se aprovado para o tratamento da psoríase, da artrite psoriática e da doença de Crohn (DC). Os resultados dos ensaios clínicos que avaliaram o impacto do uso de UST na espondilartrite axial mostraram-se, no entanto, francamente desanimadores.

Objetivos: Descrever a resposta clínica ao UST em doentes com espondilartrite seguidos num Serviço de Reumatologia de um Centro Hospitalar Universitário.

Métodos: Foram selecionados todos os doentes registados no reuma.pt com diagnóstico de espondilartrite e sob tratamento actual com Ustecinumab. Para os doentes incluídos, descrevem-se aspetos da doença (diagnóstico, tempo de evolução, avaliação da atividade de doença antes e após o UST) e do seu tratamento (DMARDs prévios; tempo de seguimento, motivos de início e descontinuação; eventos adversos).

Resultados: Encontrámos 4 doentes com espondilartrite tratados com UST, todos com o diagnóstico confirmado de doença inflamatória intestinal associada. As doentes 1 e 4, do sexo feminino, iniciaram o UST aos 40 e 46 anos, respetivamente. Por outro lado, os doentes 2 e 3, do sexo masculino, iniciaram o biológico aos 73 e 71 anos, respetivamente.

Tabela I. Caracterização dos doentes com diagnóstico de espondilartrite tratados com ustecinumab e seguidos no Serviço de Reumatologia de um Centro Hospitalar Universitário. Dados recolhidos até Junho de 2022

		Doente 1	Doente 2	Doente 3	Doente 4
Doença Reumática		SpA associada a DC	SpA associada a DC	SpA associada a DC	SpA associada a DC
Sexo		Feminino	Masculino	Masculino	Feminino
À data de início de UST	**Idade (anos)**	40	73	71	46
	Duração da doença (meses)	51	12	312	60
	DMARDs prévios	MTX, AZT, ADA, IFX, GOL, CTZ	IFX, CTZ	IFX, ADA	IFX
DMARDs concomitantes		SZZ	0	0	0
Motivo de início		Atividade de DC	Atividade de DC	Neoplasia da próstata sob IFX	Tuberculose disseminada sob IFX
Tempo de UST (meses)		45	51	8	7
Dosagem UST		90 mg subcutâneo 8/8 semanas			
Δ atividade de doença desde o início de bDMARD para SpA (final – inicial)		**EVA doente:** -50mm **BASDAI:** -3.9 **ASDAS PCR:** -2.1 **PCR:** -21.8mg/L	**EVA doente:** -80mm **BASDAI:** -3.3 **ASDAS PCR:** -2.3 **PCR:** -46.1mg/L	**EVA doente:** -67mm **BASDAI:** -4.3 **ASDAS PCR:** -2.6 **PCR:** -36.3mg/L	**EVA doente:** -* **BASDAI:** -* **ASDAS PCR:** -* **PCR:** -*
Resposta clínica (face ao início de bDMARD para SpA)		**ASAS:** Resposta ASAS 70 **ASDAS:** Grande Melhoria Atividade elevada pelo ASDAS	**ASAS:** Sem resposta **ASDAS:** Grande melhoria Doença inativa pelo ASDAS	**ASAS:** Sem resposta **ASDAS:** Grande melhoria Atividade elevada pelo ASDAS	**ASAS:** -* **ASDAS:** -* Atividade elevada pelo ASDAS
Efeitos adversos		Nenhum identificado			
Descontinuação? (e motivo)		Não			

Legenda: ADA: Adalimumab; ASAS: Assessment of SpondyloArthritis International Society; ASDAS: Ankylosing Spondylitis Disease Activity Score; AZT: Azatioprina; BASDAI: Bath Ankylosing Spondylitis Disease Activity Index; CTZ: Certolizumab; DC: Doença de Crohn; DMARDs: Disease-Modifying Antirheumatic Drugs; ETN: Etanercept; EVA doente: escala visual analógica da atividade de doença pelo doente; GOL: Golimumab; IFX: Infliximab; SpA: Espondilartrite; SSZ: Sulfassalazina; UST: Ustecinumab. * Dados insuficientes

A doente 1 iniciou UST, após 51 meses de doença e falência de múltiplos fármacos biotecnológicos. O doente 2 iniciou-o após um período mais curto e com falência de apenas 2 agentes biotecnológicos. Ambos iniciaram UST por atividade da DC. O doente 3, iniciou o biológico após 26 anos de doença, na sequência do diagnóstico de neoplasia prostática sob tratamento com infliximab (IFX.) A doente 4, iniciou-o após 60 meses de doença e por intercorrência infeciosa (Tuberculose Disseminada) sob terapêutica com IFX.

Do ponto de vista da avaliação da doença articular, os doentes 1, 2 e 3 apresentaram grande melhoria ASDAS (Ankylosing Spondylitis Disease Activity Score), embora só o doente 2 tenha atingido o alvo terapêutico de doença inativa pelo ASDAS. Não obstante, a doente 1 apresentou resposta ASAS (Assessment of SpondyloArthritis International Society) 70. Todos estes três doentes apresentaram melhoria no BASDAI (Bath Ankylosing Spondylitis Disease Activity Index), na EVA (escala visual analógica) da doença pelo doente e na proteína c-reativa, face à avaliação basal pré-bioterapia.

Por fim, quanto à doente 4, dadas as insuficiências nos dados registados, não se conseguiu ainda aferir a resposta à terapêutica. Contudo, verificou-se uma discreta melhoria da PCR em 7 meses de terapêutica.

Todos estes dados podem ser observados na tabela 1.

Conclusão: Apesar da amostra exígua não permitir grandes inferências, os resultados da avaliação do uso de UST na espondilartrite no nosso centro mostraram uma resposta satisfatória em três dos quatro doentes. Contudo, apenas um doente alcançou doença inativa pelo ASDAS. Em termos de segurança, nenhum deles apresentou eventos adversos. Mais estudos são necessários para aferir a resposta clínica ao UST nas espondilartrites, particularmente na dose terapêutica para a doença inflamatória intestinal.

194 - TREATMENT AND OUTCOME OF IMMUNE-MEDIATED DIFFUSE ALVEOLAR HEMORRHAGE – A SINGLE CENTRE EXPERIENCE

Catarina Abreu[1], Vanessa Fraga[1], Alice Castro[1], Sandra Sousa[1], 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

Diffuse alveolar hemorrhage (DAH) is a severe complication of many immune-mediated disorders with a high mortality rate.[1,2] Although systemic glucocorticoids and additional immunosuppressive treatment proved to be beneficial, the efficacy of other therapeutic options has not yet been established.[2]

The objective is to describe the clinical manifestations, the underlying rheumatic disease diagnosis, the treatment approach, and evaluate the outcome of patients with DAH in our Rheumatology centre.

We performed a retrospective analysis of patients admitted to our Rheumatology centre between 2005 and 2021 with a diagnosis of DAH. Demographic characteristics, clinical presentation, underlying rheumatic disease, treatment and outcome was retrieved from medical records.

Eight patients with the diagnosis of DAH were identified, with a median age of 33.5 (IQR 25) and a female to male ratio of 5:3. The underlying rheumatic disease was systemic lupus erythematous (SLE) in 5 cases (62.5%), two of them with secondary antiphospholipid syndrome, and ANCA associated vasculitis in the remaining 3 (2 were positive for anti-proteinase 3 anti-neutrophil cytoplasmatic antibodies (PR3-ANCA) and 1 for myeloperoxidase anti-neutrophil cytoplasmatic antibodies (MPO-ANCA)). DAH was a presenting manifestation of the rheumatic disease in half of the cases. The most common symptoms were dyspnea (87.5%), fever (62.5%), fatigue (50%), cough (50%), and hemoptysis (37.5%). One patient (12,5%) had a subclinical presentation of DAH. Laboratory evaluation documented acute anemia in 7 patients (87.5%).

Six patients (75%) received methylprednisolone pulses (1g/day), the duration of which varied from 3 to 5 days, followed by a maintenance dose of oral prednisolone (1mg/kg/day) with progressive tapering. The other 2 patients, one of which was asymptomatic, were treated with oral prednisolone. Seven (87.5%) patients were treated with cyclophosphamide (CYC). The CYC protocol used was the one from National Institute of Health (NIH) in 5 patients and the Euro-lupus protocol in one. Four patients (50%) were treated with intravenous immunoglobulin (IVIg), three of them because an infectious etiology was suspected. Four patients (50%) underwent plasmapheresis (5 to 9 sessions).

Five patients required intensive care, 3 of whom depended on invasive mechanical ventilation. Most of them were treated with different combinations of triple therapy that included methylprednisolone, plasmapheresis, CYC and IVIg. Plasmapheresis and IVIg were used only in this setting.

DAH resolved in all patients and only one had a relapse before discharge (whilst under prednisolone 1mg/kg/day), after treatment with plasmapheresis and CYC. She was readmitted in the intensive care unit and retreated with plasmapheresis and CYC, with clinical and radiological recovery.

The survival rate after 1 year was 100% for the 7 patients that completed the 1-year follow-up (one has, so

far, only six months of follow-up).
This case series suggests that the treatment of immune-mediated DAH with immunosuppressants ± IVIg ± plasmapheresis is associated with a good outcome. Plasmapheresis and IVIg may be useful in the treatment of critically ill patients, in particular.

195 - FENÓMENO DE RAYNAUD PARANEOPLÁSICO - PADRÕES CAPILAROSCÓPIOS

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

Introdução: O fenómeno de Raynaud está presente em 5% da população adulta, podendo ser de etiologia primária ou secundária. Se o início for abrupto e associado a uma resposta à terapêutica limitada, suspeitamos da etiologia paraneoplásica.
Objetivo: Descrever os padrões capilaroscópicos do fenómeno de Raynaud paraneoplásico, bem como as características epidemiológicas e clínicas dos casos associados.
Métodos: Foram definidos como critérios de inclusão doentes com idade >18 anos, neoplasia primária identificada e respectivo padrão capilaroscópico. Foi realizado a revisão sistemática com base na pesquisa de dados MEDLINE com as palavras-chave "Raynaud's phenomenon" e "paraneoplastic" desde 1960 até 2022. Obtidos 78 artigos, tendo sido excluídos 19 após avaliação de resumos e do título. Posteriormente, foram excluídos mais 48 artigos após leitura completa.
Resultados: Foram incluídos 11 casos. 81,8% dos casos ocorreram no sexo feminino, com uma idade média de 54,0±19,9 anos. 72,2% dos casos apresentavam metástases à data do diagnóstico. A taxa de mortalidade foi de 72,2% aos 48 meses. As neoplasias associadas ao fenómeno de Raynaud foram a do pulmão (2 casos), da mama (2 casos), colorretal (2 casos), ovário (2 casos), cerebral (1 caso) e um caso de linfoma. Três padrões capilaroscópicos foram descritos – 4 casos apresentaram-se sem alterações, 5 casos apresentaram-se com alterações inespecíficas e 2 casos apresentaram-se com padrão esclerodérmico ativo.
Conclusão: O fenómeno de Raynaud paraneoplasico está associado a estádios avançados de doença neoplásica e com mau prognóstico, cursando com diferentes neoplasias. É de salientar que a maioria dos doentes se apresenta com capilaroscopia normal ou com alterações não específicas.

196 - FINDING THE CULPRIT IN PARANEOPLASTIC RAYNAUD'S PHENOMENON – DIFFERENCES BETWEEN MEN AND WOMEN

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

Background: Raynaud's phenomenon (RP) is a frequent condition, which is caused by a sudden arterial constriction, characterized by a tri-phasic reaction in which the fingers or toes turn white, blue and then red. Although most are primary Raynaud's, in a small but important proportion are secondary to an underlying disease, such as malignancy. It is important to know which tumours are more frequently associated with RP and if there are differences between men and women.
Aim: To identify the most frequent malignancies associated with paraneoplastic RP reported in literature between 1960 and 2022 and the differences observed between men and women.
Methods: Systematic review based on MEDLINE search with the keywords "Raynaud's phenomenon" and "paraneoplastic" between 1960 and 2022. Eligibility criteria were patients older than 18 years old, with primary malignancy identified.
Results: A total of 53 cases of secondary RP with primary malignancies identification were reported, 20 cases in men and 33 in women. The average age of men at diagnosis was 58.45±14.08 (23-78) years old and in females were 56.67±14.54 (25-86) years old. In 50.00% of men and 57.58% of women the neoplasia was already disseminated. The most frequent local of primary tumour was the lung (39.13%) in men and the ovary (26.67%) in females. In men, the most frequent anatomic area affected was the thoracic area (75.00%), while in women was abdominopelvic area (45.45%). The survival rate at 48 months of follow-up was 65.00% in male cases and 36.36% in female cases.
Conclusion: Paraneoplastic Raynaud's phenomenon is a signal associated with different malignancies, frequently disseminated and with poor prognosis. An abdominal-focus approach in females and a thoracic-focus approach in men is recommended to exclude the presence of malignancy. More studies are required to confirm these findings.

197 - YOUNG AND OLDER ONSET RHEUMATOID ARTHRITIS: ARE THERE TRULY DIFFERENCES?

Susana P. Silva[1, 2], C. Pinto Oliveira[1, 2], Sofia Ferreira Azevedo[1, 2], Carolina Mazeda[1, 2], Gisela Eugénio[1, 2],

Anabela Barcelos[1, 2]
[1]Rheumatology Department, Hospital Infante D. Pedro (CHBV), Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz Health Alliance, Portugal, Aveiro, Portugal

Background: Age at onset of rheumatoid arthritis (RA) has been implicated as a prognostic risk factor. Some studies describe young- and late-onset RA (YORA and LORA) as two separate entities with different prognoses, but the real impact of age on RA outcomes is still controversial.[1,2]

Objective: This study aims to determine if there are clinical and serological differences between YORA and LORA patients at baseline and after 12 months of initial therapy.

Material and Methods: We conducted a retrospective study on patients aged ≥ 18 years old, who fulfil the ACR/EULAR 2010 classification criteria for RA and excluded patients with overlap syndromes. Patients ≥65 years old at disease onset were considered to have LORA. YORA and LORA patients were compared, based on disease activity, severity parameters and drug history, with p-value ≤ 0.05 considered statistically significant.

Results: A total of 110 patients with RA were analyzed. Seventy-six patients (69.1%) had YORA and 34 (30.9%) LORA. Baseline DAS 28 scores were not different between groups (YORA 4.23 (1.60) vs 4.23 (2.04), p=0.163). Only 13 (38.2%) LORA patients had positive rheumatoid factor (RF) and/or anti citrullinated protein antibodies (ACPA), while 53 (69.7%) YORA patients had RF and 59 (77.6%) ACPA. In addition, 13 (38.2%) LORA patients were diagnosed with polymyalgia rheumatica (PMR) at presentation and developed chronic polyarthritis over time. Corticosteroids were used alone as first-line therapy in 38.3% of LORA patients, compared to 8.1% in YORA (p<0.001). On the other hand, DMARDs were used since baseline in 91.9% of YORA patients compared to 66.7% of the LORA patients. We observed at 6 and 12 months, a similar decrease in median DAS 28 scores, without statistically differences. LORA patients presented persistently higher erythrocyte sedimentation rate (ESR) and Health Assessment Questionnaire (HAQ) scores compared with YORA. After 1 year, the presence of erosions was similar comparing LORA and YORA (14 (18.4%) vs 3 (8.8%), p=0.198).

Conclusions: Both groups of patients presented at baseline, 6 and 12 months of follow-up with similar disease activity, although LORA was associated with higher ESR and HAQ scores but lower RF and ACPA positivity. Treatment choices were also different at baseline due to the initial clinical presentation of LORA as PMR in some patients. Given the increase in life expectancy and better health services, a consequent increase in the incidence of LORA is inevitable. Thus, the knowledge of the implications of age at disease onset is relevant to better understand the course and prognosis of the disease. Further studies with larger sample sizes and longer follow-up times are needed.

Table I. Comparison between YORA and LORA patients

	YORA	LORA	p value
N	76 (69.1%)	34 (30.9%)	
Age at beginning of symptoms	50 (19)	73 (10)	
Age at diagnosis	52.50 (18)	74.50 (11)	
Female gender	50 (65.8%)	21 (61.8%)	0.829
Rheumatoid factor	53 (69.7%)	13 (38.2%)	0.003
ACPA	59 (77.6%)	13 (38.2%)	<0.001
BASELINE			
TJC 28	5 (7)	4 (8)	0.868
SJC 28	4 (5)	3.50 (6)	0.671
TJC 68	5 (7)	3.50 (7)	0.220
SJC 66	6 (11)	5 (10)	0.373
ESR	29 (30)	46 (48)	0.004
CRP	1.47 (2.83)	1.83 (3.43)	0.445
DAS 28 3V	4.23 (1.60)	4.23 (2.04)	0.163
First-line treatment			
Corticosteroids	6 (8.1%)	11 (33.3%)	<0.001
DMARD	68 (91.9%)	22 (66.7%)	<0.001
6 MONTHS			
TJC 28	1 (3)	0.50 (2)	0.410
SJC 28	0 (2)	0 (2)	0.543
ESR	17 (19)	23.50 (26)	0.024
CRP	0.40 (0.98)	0.46 (1.80)	0.487
DAS 28 3V	2.80 (1.91)	2.89 (1.48)	0.506
12 MONTHS			
TJC 28	0 (2)	0 (1)	0.336
SJC 28	0 (1)	0 (0)	0.344
ESR	14 (11)	24 (28.3)	0.002
CRP	0.29 (0.72)	0.37 (1.20)	0.206
DAS28 3V	2.30 (1.13)	2.67 (0.92)	0.156
HAQ	0.625 (1.0)	1.875 (1.125)	0.020
Erosions at 12 months	14 (18.4%)	3 (8.8%)	0.198

Legends – ACPA - Anti citrullinated protein antibodies; TJC - tender joint count; SJC - swollen joint count; ESR - erythrocyte sedimentation rate; CRP - C-reactive protein; DAS 28 3 V - Disease Activity Score 28 3 Variables; DMARD – Disease-modifying antirheumatic drugs.

REFERENCES

1. Camacho EM et al. Influence of age and sex on functional out-

come over time in a cohort of patients with recent-onset inflammatory polyarthritis: results from the Norfolk Arthritis Register. Arthritis Care Res 2011; 63:1745-52; 2 - Mueller R B et al. Is radiographic progression of late-onset rheumatoid arthritis different from young-onset rheumatoid arthritis? **Results**:from the Swiss prospective observational cohort. Rheumatology 2014; 53:671-677.

198 - CARACTERIZAÇÃO DO ENVOLVIMENTO AXIAL NA ARTRITE PSORIÁTICA

Catarina Abreu[1], Vanessa Fraga[1], Alice Castro[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

A artrite psoriática (AP) é tradicionalmente classificada em cinco subtipos de acordo com Moll e Wright, um dos quais a espondilite. Cerca de 5% dos doentes com AP apresentam envolvimento axial exclusivo e em 25-70% o envolvimento axial coexiste com o periférico.

O objetivo primário é caracterizar a população com AP com envolvimento axial. Como objetivos secundários, identificar as características que a distinguem do envolvimento periférico e perceber como se estabelece o diagnóstico de AP axial na prática clínica.

Foram incluídos doentes adultos com o diagnóstico de AP em seguimento numa consulta de Reumatologia hospitalar. Definiu-se o envolvimento axial pela presença de lombalgia inflamatória e alterações imagiológicas sugestivas em radiografia axial, sacroileíte (SI) radiográfica de acordo com os critérios de Nova Iorque modificados ou SI em ressonância magnética (RM).

Foram incluídos 132 doentes, 48,5% do sexo feminino, com uma idade média de 58,9 (±14,1) anos. A poliartrite simétrica foi a apresentação mais comum (59,1%) seguida da oligoartrite assimétrica (26,5%). O envolvimento axial exclusivo correspondeu a 9,2% (N=12). No total, 17,4% (N=23) doentes apresentaram envolvimento axial (exclusivo ou concomitante com o periférico). O diagnóstico do envolvimento axial foi realizado por RM em 4 doentes (17,4%) e por radiografia em 17 (73,9%). Dois apresentavam apenas sindesmófitos em radiografia axial. Em 84,2% a SI foi simétrica e assi-

Tabela I.

	Axial Exclusivo (N=12)	Periférico (N=120)	Valor-p	Axial Exclusivo e Concomitante (N=23)	Periférico Exclusivo (N=109)	Valor-p
Sexo Feminino	3/12 (25%)	61/120 (50,8%)	0.088	6/23 (26%)	58/109 (53%)	0.015
HLA-B27	4/6 (67%)	11/36 (30,5%)	0.047	5/13 (38%)	10/40 (25%)	0.275
AP Psoríase *	4/12 (33%)	4/116 (29%)	0.671	23/23 (100%)	101/105 (96%)	0.448
AF Psoríase *	2/3 (67%)	13/38 (34,2%)	0.299	3/4 (75%)	12/34 (35,2%)	0.510
Dactilite	0/12 (0%)	38/114 (33,3%)	0.011	4/23 (17,4%)	34/103 (33%)	0.140
Entesite	1/12 (8,3%)	43/113 (38%)	0.034	6/23 (26%)	38/102 (37,2%)	0.311
Distrofia Ungueal	5/10 (50%)	43/102 (42,1%)	0.438	10/21 (47,6%)	38/91 (41,8%)	0.625
Uveíte	0/12 (0%)	2/113 (17,7%)	0.819	0/23 (0%)	2/104 (1,9%)	0.669
Anti-CCP *	1/9 (11,1%)	0/104 (0%)	0.08	1/21 (4,8%)	0/92 (0%)	0.186
FR *	1/9 (11,1%)	12/110 (10,9%)	0.660	2/21 (9,5%)	11/88 (12,5%)	0.589
FRCV * a	9/12 (75%)	69/120 (57,5%)	0,194	12/23 (52,1%)	66/109 (60,6%)	0.458
Hábitos Tabágicos b	4/8 (50%)	32/86 (37,2%)	0.363	27/78 (34,6%)	9/16 (56,3%)	0.105
Consumo alcoólico c	2/7 (28,6%)	24/80 (30%)	0.653	5/13 (38,5%)	21/74 (28,4%)	0.334
PCR ao diagnóstico *	1,64±1,71	1,84±3,64	0.870	1,31±1,72	1,98±3,82	0.507
VS ao diagnóstico *	32,70±25.08	33,47±25,65	0.930	29,35±24,1	34,64±25,87	0.459
Idade início sintomas	45,67±17,68	44,44±14,2	0,782	42,68±15,32	44,98±14,35	0.504
Idade ao diagnostico	50,08±17,2	49,95±13,64	0,461	47,83±15,81	47,11±13,60	0.824
IMC *	31,62±5,71	29,03±6,15	0,481	27,07±3,81	29,30±6,22	0,544

* AP – Antecedentes Pessoais; AF – Antecedentes Familiares; Anti-CCP – Anticorpos anti peptídeo citrulinado cíclico; FR – Fator Reumatoide; FRCV -Fatores de Risco Cardiovasculares; PCR – Proteína C Reativa; VS – Velocidade de Sedimentação; IMC – Índice de Massa Corporal; a Inclui hipertensão arterial, diabetes mellitus, dislipidemia e doença cardiovascular; b Refere-se a ex-fumadores e fumadores ativos; c Refere-se a consumo alcoólico prévio ou presente superior ao recomendado

métrica em 15,8%. Verificaram-se diferenças estatisticamente significativas na comparação entre o envolvimento axial e periférico. Identificou-se uma predominância do sexo masculino no envolvimento axial concomitante com o periférico. No envolvimento axial exclusivo verificou-se uma associação com a presença de HLA-B27 e uma menor prevalência de dactilite e entesite.
Na prática clínica o diagnóstico do envolvimento axial na AP foi realizado maioritariamente através de radiografia. É mais comum no sexo masculino e apresenta uma prevalência de 17,4%. O envolvimento axial exclusivo tem uma prevalência de 9,2% estando nestes casos associado à presença de HLA-B27. A dactilite e a entesite foram mais frequentes nos doentes com envolvimento periférico.

204 - LONG-TERM FOLLOW-UP OF STARTING AND SWITCHING FROM BIO-ORIGINATOR TO BIOSIMILAR: REAL-WORLD DATA IN AXIAL SPONDYLOARTHRITIS PATIENTS TREATED WITH ADALIMUMAB AND ETANERCEPT

Ana Lúcia Fernandes[1], Ana Sofia Pinto[2], Kalveer Flora[3], Dilpreet Matharu[3], Anthony Isaacs[3], Pedro M. Machado[3, 4]

[1]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal, [2]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, [3]Rheumatology, Northwick Park Hospital, London North West University Healthcare NHS Trust, London, United Kingdom, [4]Centre for Rheumatology & MRC Centre for Neuromuscular Diseases, University College London, London, UK., London, United Kingdom

Introduction: Biotherapeutics have revolutionized the treatment of axial spondyloarthritis (axSpA). The emergence of biosimilars allowed substantial savings and a wider access to treatment, and international recommendations now contemplate switching strategies from bio-originator (BO) to its biosimilars (BS).
Aim: To investigate treatment response to adalimumab (ADA) and etanercept (ETA) in BO- and BS-treated bDMARD-naïve patients with axSpA and in patients who switched from BO to BS respective drug; and to compare the effectiveness and safety of the BO and BS drugs by assessing their persistence rates (PR).
Material and methods: Retrospective tertiary care observational study that included bDMARD-naïve patients with a diagnosis of axSpA who initiated treatment with ADA/ETA BOs or BSs, and bDMARD-experienced patients who switched from ADA/ETA BO to a BS drug (Hyrimoz/Benepali). Response to treatment was assessed according to UK National Institute for Health and Care Excellence (NICE) guidelines (at least 2/10 units or 50% reduction in BASDAI and 2/10 units reduction in spinal pain) and rates compared at 3 months using the chi-square test. bDMARD PR were calculated using Kaplan-Meier "drug survival curves". Causes of drug discontinuation were summarized using descriptive statistics.
Results: A total of 228 patients were included (154 on ADA and 74 on ETA): regarding ADA, 83 patients were started on BO, 31 on BS, and 40 switched from BO to BS; regarding ETA, 25 patients were started on BO, 33 on BS, and 16 switched from BO to BS. Groups had similar demographic characteristics, apart from the ADA group that switched from BO to BS drug, which had longer disease duration. Regarding baseline clinical characteristics, lower acute phase reactants (CRP, ESR), spinal pain, BASDAI and BASFI scores at baseline were observed in the switch group compared to the bDMARD-naïve groups (except for CRP in the ETA bDMARD-naïve group). Response to treatment after 3 months was similar between originator and biosimilar drugs for both ADA (BO 81.0%; BS 78.3%, p=0.781)

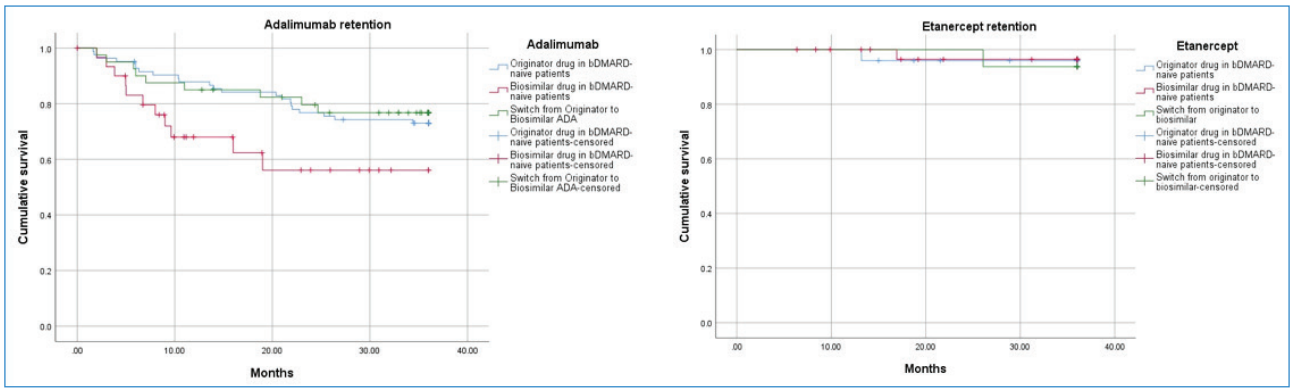


Figure 1. A – Drug survival in the three adalimumab groups (originator adalimumab – blue line, biosimilar adalimumab – red line and switch from originator to biosimilar adalimumab – green line). B – Drug survival in the three etanercept groups (originator etanercept – blue line, biosimilar etanercept – red line and switch from originator to biosimilar etanercept – green line).

and ETA patients (BO: 76.9%, BS: 76.5%, p=0.977). Three-year PR were similar across groups, both for ADA (figure 1A) and ETA (figure 1B): ADA PR were 73.5% for BO, 64.5% for BS, and 77.5% for the group that switched from BO to BS (p=0.959); ETA PR were 96.0% for BO, 97.0% for BS and 93.8% on the BO-to-BS switching group (p=0.449). Adverse events and inefficacy were the most frequent causes of discontinuation in ADA patients, and inefficacy and other reasons (e.g., fear of SARS-CoV-2 infection) were the most frequent causes of discontinuation in ETA patients.
Conclusion: First-line treatment with bio-originator or biosimilar ADA/ETA showed similar effectiveness and safety in our long-term cohort of patients with axSpA. Response to treatment was also similar between groups.

205 - CRYOGLOBULINEMIA CHARACTERIZATION FROM A TERTIARY CENTRE

Susana P. Silva[1, 2], Andrea Reboredo[3], Gisela Eugénio[1, 2], Anabela Barcelos[1, 2]
[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz Health Alliance, Portugal, Aveiro, Portugal, [3]Clinical Pathology, Centro Hospitalar Baixo Vouga, Aveiro, Portugal

Background: Cryoglobulinemia is defined as the persistent presence in serum of abnormal immunoglobulins (Ig) that precipitate at low temperatures and dissolve again upon warming. There are 3 types of cryoglobulins, accordingly to its composition: monoclonal Ig - type I; monoclonal and polyclonal Ig – mixed type II; polyclonal Ig – mixed type III. They can be associated to chronic infectious diseases, malignancies and rheumatic diseases and are responsible for rare systemic vasculitis.
Objectives: This study aims to analyze the clinical and analytical manifestations associated to the determination of cryoglobulins in our hospital.
Methods: We conducted a retrospective study on patients who performed a cryoglobulin test in our hospital, between January 2017 and December 2021. Clinical and analytical features of patients with positive cryoglobulins, were collected from the hospital database.
Results: A total of 183 tests were performed, 7 (3.8%) of which were positive. Six patients had cryoglobulinemia type II and one patient had type I. Eighty six percent of patients were male, with a mean age of 69.7±9.9 years. The most frequent presenting manifestation was palpable purpura (42.9%), followed by livedo reticularis (14.3%) and skin ulcers (14.3%). Biopsy specimens of skin lesions showed cutaneous vasculitis. Two patients were diagnosed after the beginning of a Raynaud's phenomenon. Regarding renal involvement, one patient had a nephrotic syndrome at clinical presentation and one developed nephrotic proteinuria over time. Renal biopsy revealed a membrane-proliferative and a mesangial proliferative glomerulonephritis in each patient. Arthritis (28.6%) and neurological (28.6%) involvement were also described. Two patients had low levels of complement and one had high levels of rheumatoid factor. Most cryoglobulins consisted of IgM and polyclonal IgG, but one also contained IgA. Hepatitis C was the main diagnosis in three patients with mixed cryoglobulinemia. Three patients were diagnosed with hematologic malignancy (two with multiple myeloma and one with Waldenstrom's macroglobulinemia). One patient with multiple myeloma was diagnosed with cryoglobulinemia type I and two with type II. One patient had rheumatoid arthritis and was diagnosed with cryoglobulinemia type II. In the follow-up, two patients died of hepatocellular carcinoma and one of metastatic multiple myeloma. Patients with Waldenstrom's macroglobulinemia and multiple myeloma were successfully treated with rituximab and corticosteroids.
Conclusions: Despite the small group of patients, our data show that cryoglobulinemia is a rare condition, associated with different diseases and different clinical presentations. Any of these clinical and analytical findings may be present in a rheumatologic patient, being important to consider cryoglobulinemia as a differential diagnosis. Thus, it is important to have a clinical suspicion about this entity and neoplasms should always be carefully searched in this group.

207 - AUTOLOGOUS HEMATOPOIETIC STEM CELL TRANSPLANTATION IN SYSTEMIC SCLEROSIS - DESCRIPTION OF A SINGLE CENTER EXPERIENCE

Augusto Silva[1], Vasco C Romão[1], Carlos Pinho Vaz[2], JE Fonseca[1], Catarina Resende[1]
[1]Rheumatology and Bone Metabolic Diseases Department, Centro Hospitalar Lisboa Norte, EPE - Hospital de Santa Maria, Lisbon Academic Medical Centre, Lisboa, Portugal, [2]Serviço de Transplantação de Medula Óssea, Instituto Português de Oncologia (IPO) Porto, Porto, Portugal

Introduction: Severe and progressive forms of systemic sclerosis (SSc) result in reduced quality of life and elevated mortality rates. Autologous hematopoietic stem cell transplantation (AHSCT) has been used in the treatment of patients with early diffuse cutaneous SSc (dcSSc) to promote immune depletion and to reprogramme the immune system, leading to the reestablishment of immune tolerance and, thereby, enhancing disease control and survival, even though this aggressive

approach is itself associated with a short-time increase in mortality.
Objective: To describe the clinical features and outcomes of patients with severe and refractory SSc treated with AHSCT in our department.
Methods: This study included adult patients who underwent AHSCT from 2017 to 2022. The procedure included harvesting of autologous hematopoietic progenitor stem cells with CYATG protocol (that uses cyclophosphamide and antithymocyte globulin) and subsequent infusion of 3.84x106 CD34+ cells/Kg after positive CD34+ selection.
Results: We treated 4 patients, all women, with a mean (range) age at diagnosis of 34 (11-43) years-old. The mean age at transplant was 38 (18-47) years and the mean time from diagnosis until transplant was 4.25 (2.5-7) years. Three patients had dcSSc and a fourth one had dcSSc and polymyositis overlap syndrome.
Before AHSCT, all patients had vasculocutaneous (4 patients with Raynaud's phenomenon, digital ulcers, sclerodactyly with a mean mRSS of 14/51 [4-22/51] and 2 had calcinosis), gastrointestinal (4 patients with dysphagia) and respiratory manifestations (dyspnea in 4 patients and dysphonia in 1). Two patients had arthritis. The patient with overlap syndrome showed marked tiredness and muscle and cardiac enzyme elevation (MMT8 pre-transplant: 104/150). All patients had positive antinuclear antibodies (titer: 1/160-1/640), anti-Ro52 in 2/4; anti-Scl70 in 2/4, PM75 and PM100 in 1/4. Before AHSCT, respiratory function tests showed a low forced vital capacity (FVC) in 3/4, low forced expiratory volume in the first second (FEV1) in 4/4, low total lung capacity (TLC) in 3/4 and a low single-breath diffusing capacity of the lungs for CO (DLCO) in 3/4. Computed tomography identified nonspecific interstitial pneumonia (NSIP) in only one patient.
All patients had previously received immunosuppressive therapy. Cyclophosphamide was the most commonly used (4/4), followed by methotrexate and mycophenolate mofetil (2/4) and azathioprine (1/4). All had progression of disease despite treatment.
All patients survived AHSCT but all had mild infectious and inflammatory complications. The mean follow-up duration was 2 years and 3 months (6 months - 5 years). After AHSCT, the patients had a favorable evolution with improvement of cutaneous manifestations and a mean mRSS of 8.75/51 (0-18/51). In the patient with NSIP, an improvement of respiratory function with an increase of FVC (22%), FEV1 (22%) and TLC (3%) were verified, while the single-breath DLCO was not changed. Muscle strength also improved in the overlap syndrome patient, with a MMT8 increasing to 123/150. None of the patients currently needs immunosuppressive therapy.
Conclusion: AHSCT is an option for the management of patients with severe and progressive SSc with an inadequate response to conventional immunosuppressive therapy. A multidisciplinary approach and good selection of patients are important for better outcomes.

210 - JUVENILE FIBROMYALGIA SYNDROME – AN UNDERDIAGNOSED DISORDER

Sofia Ferreira Azevedo[1, 2], Inês Genrinho[1, 2, 3], Inês Cunha[1, 2]
[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz Health Alliance, Portugal, Aveiro, Portugal, [3]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Introduction/Background: Juvenile fibromyalgia syndrome (JFS) is a chronic diffuse musculoskeletal pain condition affecting children and adolescents. Its worldwide prevalence is estimated of 1 to 6%. There is a strong female predominance. Pain is the hallmark of the disease although fatigue, headache, depression, anxiety and sleep disturbances are also common manifestations. Early diagnosis and intervention are of utmost importance in these patients. The standard of care is multidisciplinary, combining non-pharmacologic and pharmacologic approaches, with major importance of cognitive behavioral therapy, relaxing measures and physical activity. Prognosis depends on various factors and symptoms may persist into adulthood.
Materials and methods: Single center retrospective study of patients with JFS that fulfilled ACR 2016 fibromyalgia criteria, followed for a period of 2 years (2020-2022). Sociodemographic and clinical data were collected. Fibromyalgia Impact Questionnaire validated for portuguese population (FIQ-P), pain and fatigue visual analogic scales (VAS) values were registered.
Descriptive analysis was performed using means and medians, and standard deviation and interquartile range, with SPSS® software.
Results: Seven patients were enrolled. Mean age of patients at diagnosis was 14 ± 1.00 years old. All patients were female and Caucasian. There was no history of fibromyalgia in first degree relatives.
All patient presented with generalized widespread pain and fatigue, with median pain VAS of 5.00 [5.00-6.00] and mean fatigue VAS of 5.00 ± 2.16. Five patients (71.43%) had sleep disturbances, 4 (57.14%) had anxiety, 4 (57.14%) had chronic headache and 3 (42.86%) had depression. Mean FIQ-P value was 54.78 ± 17.25. Deficiency of vitamin D was found in all patient with a mean level of 19±8.89 ng/mL. Antinuclear antibodies (ANAs) were negative in 85.71% of patients.

Regarding pharmacologic treatment, all patients were treated with pregabalin, 5 (71.43%) with selective serotonin reuptake inhibitors (SSRI's), 5 (71.43%) with weak opioids, 3 (42.86 %) with muscular relaxants and 1 (14.29%) was taking benzodiazepines. Although all patient was advised to exercise, only 66.70% were compliant.
After a two year follow up 42.86% had symptomatic relief and improvement in well-being and functionality indicators.
Conclusion: Our study suggests that despite multimodal approach and combination of pharmacologic and non-pharmacologic treatments, an important percentage of patients will not improve. Compliance may represent an important aspect in disease course and prognosis.
Considering that, our study highlights the need of further investigation in this area, to better understand disease's physiopathology and identify effective treatment options. We also underline the importance of JFS syndrome awareness and early diagnosis, minimizing the negative impact on patients and their families.

212 - OSTEOGENESIS IMPERFECTA NO ADULTO: CASUÍSTICA DO CENTRO HOSPITALAR E UNIVERSITÁRIO DE COIMBRA

João Alexandre Oliveira[1], Maria João Cadório[1], Marlene Sousa[1], Ana Rita Prata[1], Mariana Luis[1], Armando Malcata[1]
[1]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

Introdução: A consulta de Doenças Ósseas Raras do Adulto (DORA) do CHUC foi criada em 2022 e tem como população-alvo os doentes adultos com displasias ósseas, incluindo com osteogenesis imperfecta (OI). A OI é a doença genética óssea caracterizada por fragilidade óssea e fraturas de repetição, com uma prevalência estimada de 1:20.000. Apesar de o diagnóstico ser na maioria dos casos realizado na infância, formas mais frustres ou incompletas podem passar despercebidas até à idade adulta. Para além do diagnóstico, outros desafios se impõem no adulto, nomeadamente a gestão de complicações, tais como deformidades osteoarticulares induzidas pelas fraturas, surdez, problemas na dentição e síndrome de apneia obstrutiva do sono.
Objetivo: Descrever as características clínicas e moleculares nos doentes adultos com OI atualmente em seguimento na consulta DORA do CHUC.
Resultados: Foi incluído um total de 13 doentes adultos com OI, sendo que 85% dos doentes apresentava uma OI do tipo 1. A maioria dos doentes era do sexo feminino (62%) e a idade atual mediana foi de 37.0 anos. A idade mediana no momento do diagnóstico foi de 4.0 anos, e a idade mediana da primeira fratura de 6.0 anos. Dos 10 doentes cuja estatura era conhecida, 3 apresentavam baixa estatura (mediana 130 cm).
Dos doentes submetidos a teste genético, em 90% dos casos foi identificada uma mutação do gene COL1A1. Em um doente não foi possível identificar uma mutação patogénica. Dos 10 doentes cuja história familiar era conhecida, metade apresentavam outros familiares (maioritariamente irmãos) com diagnóstico de OI (mediana de 3.0 familiares afetados por família). Para além das fraturas, as manifestações mais comuns na nossa coorte foram: escleras azuis (88%), problemas de dentição (88%), laxidez ligamentar (83%), escoliose (80%) e surdez (30%). Um total de 7 doentes realizou terapêutica com bifosfonatos e 1 doente recusou terapêutica com qualquer um destes fármacos. Dos 7 doentes tratados, 4 mantêm a terapêutica até à atualidade. O ácido zoledrônico foi o bifosfonato mais frequentemente prescrito (3 doentes), seguido do alendronato e pamidronato, risedronato e ibandronato (1 doente, cada).
Discussão: A OI é uma doença óssea genética habitualmente diagnosticada na infância, mas que se mantém na idade adulta e para o resto da vida do doente sendo essencial garantir o seu acompanhamento ao longo das diversas etapas da vida, identificando e gerindo da melhor forma os desafios impostos em cada uma destas etapas.

213 - CLINICAL AND IMMUNOLOGICAL FEATURES OF SYSTEMIC SCLEROSIS WITH INTERSTITIAL LUNG DISEASE: RESULTS FROM A PORTUGUESE TERTIARY CENTER

Ana Isabel Maduro[1], André Saraiva[1], Helena Assunção[1], Tânia Santiago[1, 2], Maria João Salvador[1, 2]
[1]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [2]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal

Background: Systemic sclerosis (SSc) is frequently complicated by interstitial lung disease (ILD), and is associated with worse outcomes and mortality. The proportion of patients who develop SSc-ILD versus SSc-non ILD is incompletely understood.
Objective: To describe and compare the demographic features, SSc subsets, and main clinical and immunological features in SSc patients with and without ILD.
Materials and methods: Cross-sectional single-center study including SSc patients fulfilling the ACR'1980, ACR/EULAR'2013 or the LeRoy's classification criteria, followed up at the Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, between 2010 and 2021. The patient characteristics were collected

and assessed at baseline, including gender, age, age at disease onset, SSc subtype, positivity to antinuclear antibodies (ANA) and specific autoantibodies [anti-centromere (ACA), anti-topoisomerase I (anti-Scl70), anti-ribonucleic acid polymerase III (anti-RNA-pol III)], cumulative disease manifestations [Raynaud phenomenon, telangiectasia, skin thickening, digital ulcers or pitting scars, articular involvement, myositis, gastrointestinal and renal involvement, pulmonary artery hypertension (PAH)], respiratory function tests [diffusing lung capacity for carbon monoxide (DLCO) and forced vital capacity (FVC)] and nailfold capillaroscopy pattern. ILD was confirmed by chest high-resolution computed tomography. Chi-square test, two-sample t-test and Mann Whitney U test were used to compare the groups with and without DPI. The statistical analysis was carried out using SPSS 26.0. P-value <0.05 was considered statistically significant.

Results: 170 patients were included (85.9% women, mean age 54.5±14.2 years). ILD was found in 41 (24.1%) patients. Male sex (24.4% vs 10.9%, p=0.03) and diffuse cutaneous SS subtype (43.9 vs 10.1%, p<0.01) were more prevalent in the ILD group, with a statistically significant difference. Regarding clinical manifestations, it was found a statistically significant greater percentage of skin thickening (92.7 vs 51.2%, p<0.01), digital ulcers or pitting scars (51.2% vs 22.5%, p<0.01), gastrointestinal involvement (48.8 vs 18.6%, p<0.01) and PAH (34.1 vs 6.9%, p<0.01). Concerning specific antibodies, a positive titer of the anti-Scl70 was more frequent (46.3 vs 13.2%, p<0.01) and a positive titer of the ACA was less frequent in SS patients with

Table I. Frequency of SSc-associated clinical manifestations and laboratory features in SS patients with and without interstitial lung disease

	Whole cohort n = 170 (100%)	With ILD n = 41 (24.1%)	Without ILD n = 129 (75.9%)	p-value
Age at diagnosis (mean DP)	51.87 14.045	51.46 11.792	52.00 14.728	0.110
Sex (n [%])				
- Female	146 (85.9)	31 (75.6)	115 (89.1)	0.03
- Male	24 (14.1)	10 (24.4)	14 (10.9)	
Subtype (n [%])				
- Pre-SSc	48 (28.4)	4 (2.4)	72 (55.8)	<0.01
- Limited cutaneous	90 (52.7)	17 (41.5)	73 (56.6)	0.214
- Diffuse cutaneous	31 (18.3)	18 (43.9)	13 (10.1)	<0.01
Clinical features (n [%])				
- Raynaud´s Phenomenon	160 (94.1)	38 (92.7)	122 (94.6)	0.705
- Skin thickening	104 (61.2)	38 (92.7)	66 (51.2)	<0.01
- Telangiectasis	51 (30)	14 (34.1)	37 (28.7)	0.506
- Digital ulcers or pitting scars	50 (29.4)	21 (51.2)	29 (22.5)	<0.01
- Articular involvement	43 (25.3)	12 (29.3)	31 (24.0)	0.502
- Myositis	6 (3.5)	3 (7.3)	2 (1.5)	0.152
- Gastrointestinal involvement	45 (26.5)	20 (48.8)	25 (18.6)	<0.01
- Renal involvement	1 (0.6)	1 (2.4)	0 (0.0)	0.241
- Pulmonar arterial hypertension	23 (13.5)	14 (34.1)	9 (6.9)	<0.01
Laboratory features				
- Antinuclear antibodies (n [%])	167 (98.2)	41 (100)	126 (97.7)	0.579
- Anti-centromere (n [%])	100 (58.8)	11 (26.8)	89 (69.0)	<0.01
- Anti-topoisomerase I (n [%])	36 (21.2)	19 (46.3)	17 (13.2)	<0.01
- Anti-RNA polymerase III (n [%])	4 (2.4)	1 (2.4)	3 (2.3)	1.00
- C Reactive Protein (median IQR)	0.25 (0.58)	0.7 (1.29)	0.21 (0.48)	<001
- Erythrocyte sedimentation rate (median IQR)	14 (16)	19 (18.25)	12 (16)	0.033
Respiratory Function Tests (median IQR)				
- DLCO (mmol/min/kpa)				
- FVC (%)	91 (39.75)	62 (27)	95 (24.5)	0.01
- FEV1/FVC (%)	101 (22.5)	94.5 (21)	108 (24)	0.002
	1.01 (0.11)	0.99 (0.19)	1.01 (0.05)	0.382
Nailfold capillaroscopy pattern (n [%])*				
- Early	34 (21.4)	6 (15.8)	28 (23.1)	0.160
- Active	34 (21.4)	17 (44.7)	17 (14.0)	<0.01
- Late	24 (14.1)	5 (13.2)	15 (12.4)	0.567
- Unespecific CTD	61 (38.4)	10 (26.3)	51 (42.1)	0.073
- Normal	10 (5.9)	0 (0)	10 (8.3)	0.201

ILD: interstitial lung disease; IQR: Interquartile range; DLCO: Diffusing lung capacity for carbon monoxide; FVC: Forced vital capacity; FEV1/FVC: forced expiratory volume in the first second/forced vital capacity ratio; CTD: connective tissue disease. *missings

ILD (26.8 vs 69.0%, p<0.01), both with statistical significance. The median C Reactive Protein and erythrocyte sedimentation rate were also higher in the group of patients with ILD. Considering respiratory function tests, there was a statistically significant decrease in the median of DLCO [62 (IQR 27) vs 95 (IQR 24.5) mmol/min/kPa, p=0.01] and FVC [101 (IQR 22.5) vs 108% (IQR 24), p=0.002] in this group. The active pattern was the most prevalent nailfold capillaroscopy pattern in SSc patients with ILD. No differences in age at diagnosis, Raynaud's phenomenon, telangiectasis, articular and renal involvement, myositis and anti-RNA polymerase III antibodies were found between the groups.

Conclusion: In this cohort, it found a difference statistically significant between SSc patients with ILD and male sex, diffuse cutaneous SSc subtype and the presence of skin thickening, digital ulcers or pitting scars, gastrointestinal involvement, PAH, a positive titer of the anti-Scl70, active nailfold capillaroscopy pattern and decrease in the median of DLCO and FVC. The identification of these features can be useful to suspect ILD in SSc.

215 - RADIOGRAPHIC PROGRESSION OF PATIENTS FOLLOWING A BIOLOGIC TAPERING PROTOCOL FOR RHEUMATOID ARTHRITIS, PSORIATIC ARTHRITIS AND AXIAL SPONDYLOARTHRITIS

Pedro Ávila Ribeiro[1, 2], Tomás Marques-Ramalho[3], Fernando Saraiva[1, 2], Patrícia Martins[1, 2], Ana Rita Cruz-Machado[1, 2], Maria João Saavedra[1], JE Fonseca[1, 2], Elsa Vieira-Sousa[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, Lisboa, Portugal, [3]Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal

Aim: To determine the rate of radiographic progression in rheumatoid arthritis (RA), psoriatic arthritis (PsA) and axial spondyloarthritis (axSpA) patients after 1 to 2 years of progressive biologic tapering according to a standardized protocol. Secondary aims: to investigate the association of clinical variables with radiographic progression.

Materials and **Methods:** Patients diagnosed with RA, PsA (with predominant peripheral (pPsA) or axial (axPsA) involvement) and axSpA, treated with tumor necrosis factor inhibitors (TNFi), in sustained Disease Activity Score (DAS) 28 remission (DAS28<2.6, if RA or pPsA) or Ankylosing Spondylitis Disease Activity Score (ASDAS) low disease activity (LDA ASDAS<2.1, if axSpA or axPsA), 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). In case of disease flare (defined as DAS28>2.6/ ASDAS>2.1), the previous treatment frequency was reintroduced by protocol.

Radiographs were taken at baseline and after 1-2 years. Two blinded readers rated the hands and feet (for patients with RA and pPsA) using the modified Sharp-van der Heijde (SvdH) score, and cervical and lumbar spine (for axSpA and axPsA) using the modified Stoke Ankylosing Spondylitis Spine Score (mSASSS). Inter-rater reliability was assessed with intraclass correlation (ICC) and changes in radiographic scores were quantified, tested for statistical significance (t-test) and compared to the minimal clinically important difference (MCID) and smallest detectable change (SDC, calculated using the Bland & Altman method).

Results: 7 RA, 10 PsA and 14 axSpA patients initiated tapering and had radiographs available at baseline and after 17.6 (±4.9) months. Inter-rater reliability was high across all diseases (ICC 0.996, 0.997 and >0.999 for RA, peripheral PsA and axSpA/axial PsA, respectively). There was no statistically significant change in structural damage at baseline vs. after 1-2 years of tapering protocol: SvdH=10.6 vs. 12.1 in RA (p=0.13); SvdH=7.8 vs. 8.1 in pPsA (p=0.40); mSASSS=17.5 vs. 18.0 in axSpA/axSpA (p=0.07). We found, however, a small trend for damage progression, which was larger than the SDC in 3 RA patients (42.3%; SDC=1.7), 2 pPsA patients (22.2%; SDC=1.0) and 5 axSpA/axPsA patients (33.3%; SDC=0.9). No RA or pPsA patients exceeded the MCID (SvdH=5; MCID not established for the mSASSS). pPsA patients whose change was larger than the SDC had significantly more damage at baseline (SvdH= 20.5 vs. 4.1, p<0.01); similar trends were found in RA (SvdH=19.5 vs. 4.0, p=0.26) and axSpA/axSpA (mSASSS=20.4 vs. 16.0, p=0.73). No difference in disease flare rate was found between patients who had radiographic progression larger than the SDC vs. no progression.

Conclusion. After 1-2 years of biologic tapering, small and not statistically significant increases in radiographic damage were found in a minority of RA, PsA and axSpA patients. SDC were lower than the MCID. The occurrence of short-lived flares did not accelerate radiographic progression.

Acknowledgments: TaPRA and TaSpA teams: Inês Cordeiro, Joaquim Pereira, Nikita Khmeliskii, Rita Barros, 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.

219 - CARACTERÍSTICAS CLÍNICAS E FATORES PREDITORES DE RECIDIVA EM DOENTES COM POLIMIALGIA REUMÁTICA

Carla Campinho Ferreira[1], Ana Margarida Correia[1], Emanuel Costa[1], Diogo Esperança Almeida[1], Joana Sousa-Neves[1], Marcos Cerqueira[1], José Redondo[1], Ana Ribeiro[1], Joana Leite Silva[1]

[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

Introdução: A Polimialgia Reumática (PMR) é uma doença inflamatória caracterizada por dor e rigidez articular das cinturas escapular e pélvica e aumento dos parâmetros inflamatórios, que afeta indivíduos a partir da 5ª década de vida. O tratamento inicial recomendado é a corticoterapia (CCT), havendo uma porção significativa de doentes que apresenta recidiva da doença durante o desmame ou após suspensão do tratamento, não sendo ainda claros quais os fatores responsáveis por estas diferenças na resposta ao tratamento.
Objetivos: Caracterizar clínica e analiticamente uma coorte de doentes com PMR e avaliar possíveis fatores preditores do risco de recidiva.
Métodos: Estudo retrospetivo e unicêntrico. Incluídos todos os doentes com primeira consulta de Reumatologia entre 1/01/2017 e 31/12/2019, com diagnóstico de PMR. A análise estatística foi efetuada com recurso ao SPSS, tendo sido utilizado, para a comparação entre os grupos, os testes de Qui-quadrado, Exato de Fisher ou Mann-Whitney, conforme as variáveis em análise. A significância estatística foi definida como valor de $p \leq 0.05$.
Resultados: Foram incluídos um total de 29 doentes com PMR, com idade média de 70,8±9,91 anos, sendo 15 (51,7%) do sexo feminino. Relativamente às características ao diagnóstico, observou-se que 12 (41,4%) eram diabéticos, 4 (13,8%) reportavam perda ponderal desde o início do quadro e em 12 (41,4%) foi objetivada artrite periférica. Analiticamente verificou-se que a média da velocidade de sedimentação (VS) era de 80,2±29,07 mm/h e a média da proteína C reativa (PCR) de 59,3±48,52 mg/L. A média de tempo decorrido desde o início dos sintomas até ao diagnóstico era de 4,3±3,10 meses. Todos os doentes tinham iniciado tratamento com prednisolona (PDN) à data do diagnóstico, com uma dose média inicial de 18,6±10,44 mg/d. Nos 24 meses de follow-up analisados, foi observada recidiva da doença em 10 (34,5%) doentes, sendo que esta ocorreu em média 8,4±5,74 meses após o início do tratamento, encontrando-se os doentes à data da recidiva com uma dose de PDN em média de 3,9±4,1 mg/d. Verificou-se ainda que 20,7% (n=6) iniciou tratamento com metotrexato (MTX) durante o período de seguimento. Na análise estatística, verificou-se que o grupo de doentes que apresentou pelo menos 1 recidiva nos primeiros 24 meses após o diagnóstico de PMR, apresentava valores de VS e PCR mais elevadas ao diagnóstico, iniciaram doses de PDN mais elevadas e foi efetuado um desmame mais rápido da CCT nos primeiros 3 meses. No entanto, nenhuma destas diferenças apresentou significância estatística (tabela 1), não tendo sido por isso possível proceder à análise de possíveis fatores preditores de recidiva.
Conclusão: As recidivas são eventos comuns nos doentes com PMR, tendo-se observado neste estudo uma taxa de recidiva semelhante à descrita na literatura. Nesta coorte não foi possível avaliar possíveis fatores preditores, dado não se ter encontrado diferenças estatisticamente significativas entre o grupo de doentes com e sem recidiva. Este resultado pode dever-se a vários fatores como sejam o reduzido número de doentes e o facto de se tratar de um estudo retrospetivo, o que condiciona a qualidade dos dados. Os resultados publicados nesta área têm apresentado resultados contraditórios, sendo que os resultados deste estudo vão de encontro ao estudo de Mackie SL et al., numa coorte de 176 doentes com PMR, no qual também não foi possível encontrar fatores preditores de recidiva.

Tabela I. Comparação das características demográficas, clínicas e analíticas dos doentes com PMR que desenvolveram ou não recidiva*.

	Grupo sem recidiva (n=19)	Grupo com recidiva (n=10)	valor de p
Idade	70,5±9,45	71,4±11,23	0,765
Sexo feminino, n (%)	11 (57,9%)	4 (40,0%)	0,450
Perda Ponderal, n (%)	3 (15,8)	1 (10,0%)	1,000
Artrite periférica, n (%)	8 (42,1%)	4 (40,0%)	1,000
Diabetes mellitus ao diagnóstico, n (%)	7 (36,8%)	5 (50,0%)	0,694
Tempo, em meses, desde o início dos sintomas até ao diagnóstico	4,9±3,43	2,7±0,76	0,218
VS ao diagnóstico	77,2±30,79	87,4±25,00	0,546
PCR ao diagnóstico	51,2±35,91	79,4±71,60	0,484
Dose de PDN inicial	16,0±4,30	23,5±15,86	0,372
Taxa de desmame de PDN, em mg/mês, nos primeiros 3 meses	2,4±1,61	3,9±3,21	0,496

*valores apresentados como média±desvio padrão, se não especificado o contrário.

220 - PERFORMANCE DOS NOVOS CRITÉRIOS DE CLASSIFICAÇÃO ACR/EULAR 2022 PARA GRANULOMATOSE COM POLIANGEÍTE E GRANULOMATOSE EOSINOFÍLICA COM POLIANGEÍTE

Adriana Carones[1], Beatriz Mendes[1], Mariana Luis[1, 2], Helena Assunção[1]

[1]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [2]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal

Introdução: As vasculites ANCA são doenças raras e que comportam um prognóstico reservado na ausência de tratamento, pelo que o diagnóstico precoce é essencial ao sucesso terapêutico. Recentemente o American

Tabela I. Caraterização demográfica e manifestações clínicas e laboratoriais à data de diagnóstico dos doentes com GPA

Doente nº			1	2	3	4	5	6	7	8
Sexo			M	M	F	M	M	M	M	M
Idade (anos)			59	41	40	41	43	43	59	30
Critérios ACR 1990										
Inflamação oral ou nasal (úlceras orais, epistáxis ou crusting)			1	1	1	1	1	1	1	1
Alterações no sedimento urinário (hematúria microscópica ou cilindros hemáticos na urina)			1	0	1	0	0	1	0	0
Alterações radiografia tórax (nódulos, infiltrados fixos ou cavitações)			1	1	0	0	0	0	1	1
Biópsia com inflamação granulomatosa			0	1	0	1	1	0	1	1
Total – classificação se ≥2/4			3/4	3/4	2/4	2/4	2/4	2/4	3/4	3/4
Critérios ACR/EULAR 2022										
Clínicos	Nasal (epistáxis, crusting ou perfuração septo)	+3	+3	+3	+3	+3	+3	+3	+3	+3
	Cartilagíneo (ouvido/nasal/VA)	+2	0	+2	+2	+2	+2	0	0	0
	Perda auditiva de condução ou neurossensorial	+1	+1	0	0	0	0	0	0	0
Laboratoriais, Imagiológicos e Histológicos	c- ANCA+/PR3+	+5	+5	+5	+5	0	0	+5	0	+5
	Pulmonar (nódulos/cavitações/massas)	+2	+2	+2	0	0	0	0	+2	+2
	Inflamação granulomatosa biópsia	+2	0	+2	0	+2	+2	0	+2	+2
	Inflamação SPN/mastóide imagem	+1	+1	0	+1	+1	+1	+1	+1	+1
	GNF pauci-imune biópsia	+1	+1	0	+1	0	0	+1	0	0
	p-ANCA/MPO	-1	0	0	0	-1	0	0	0	0
	Eosinofilia periférica	-4	0	0	0	0	0	0	0	0
Total - classificação se ≥ 5			13	14	12	7	8	10	8	13

Legenda: F – feminino; GNF – glomerulonefrite; M – masculino; SPN - Seios perinasais

Tabela II. Caraterização demográfica e manifestações clínicas e laboratoriais à data de diagnóstico dos doentes com GEPA

Doente nº			1	2	3	4	5	6
Sexo			F	M	M	M	M	F
Idade (anos)			50	58	68	58	70	16
Critérios ACR 1990								
Asma			1	1	1	1	1	1
Eosinofilia periférica > 10%			1	1	1	1	0	1
Mononeuropatia ou polineuropatia			1	1	1	0	0	0
Infiltrados pulmonares migratórios ou transitórios na radiografia de tórax			1	1	1	1	0	0
Alteração SPN (dor aguda ou crónica ou opacificação radiológica)			1	0	1	1	1	1
Biópsia com eosinófilos extra-vasculares			1	1	1	0	1	1
Total – classificação se ≥ 4/6			6/6	5/6	5/6	4/6	3/6	4/6
Critérios ACR/EULAR 20								
Clínicos	Patologia obstrutiva da via aérea	+3	+3	+3	+3	+3	+3	+3
	Pólipos nasais	+3	+3	+3	+3	+3	+3	+3
	Mononeurite multiplex	+1	+1	0	0	0	0	0
Laboratoriais, Imagiológicos e Histológicos	Eosinofilia periférica	+5	+5	+5	+5	+5	0	+5
	Biópsia com eosinofilia extra-vascular	+2	+2	+2	+2	0	+2	+2
	Hematúria	0	0	0	-1	0	0	0
	c-ANCA/PR3	-3	0	0	0	0	0	0
Total - classificação se ≥ 6			14	13	12	11	8	13

Legenda: F – feminino; M – masculino; SPN – Seios perinasais

College of Rheumatology (ACR) e a European League Against Rheumatism (EULAR) colaboraram no desenvolvimento dos novos critérios de classificação ACR/ EULAR 2022 para vasculites ANCA. Estes critérios vieram substituir os prévios critérios de classificação ACR 1990. De entre as novidades introduzidas nos novos critérios incluem-se a pesquisa de anticorpos citoplasmáticos anti-neutrófilo (ANCA) e a atribuição de uma ponderação específica a cada critério em função do seu potencial contributo para o diagnóstico.
Objetivo: Avaliar a performance dos novos critérios de classificação ACR/EULAR 2022 em doentes com diagnóstico de granulomatose com poliangeíte (GPA) e granulomatose eosinofílica com poliangeíte (GEPA) seguidos num centro terciário em Portugal.
Métodos: Foram identificados todos os doentes com diagnóstico clínico inequívoco de GPA e GEPA seguidos em consulta de reumatologia no serviço do Centro Hospitalar e Universitário de Coimbra. Dados clínicos, laboratoriais, imagiológicos e histológicos relevantes relativos ao momento do diagnóstico foram colhidos retrospetivamente e através da consulta do processo clínico e da base de dados Reuma.pt/vasculites. Foi averiguado o cumprimento dos critérios de classificação ACR 1990 e ACR/EULAR 2022 ao momento do diagnóstico.
Resultados: Foram incluídos 15 doentes com diagnóstico inaugural de GPA (n=8) ou GEPA (n=6). A caracterização demográfica, manifestações clínicas e laboratoriais inaugurais relevantes e aplicação dos critérios de classificação ACR 1990 e ACR/EULAR 2022 estão descritos nas Tabelas 1 (GPA) e 2 (GEPA). A totalidade dos doentes com GPA cumpriam ambos os critérios. Dos doentes com diagnóstico de GEPA, à exceção de um doente (Doente 5) que cumpria apenas critérios de classificação ACR/EULAR 2022, todos os outros cumpriam ambos os critérios. Relativamente ao doente com GEPA que cumpre apenas critérios de classificação ACR/EULAR 2022 trata-se de um doente com antecedentes de asma que se apresentou com colite eosinofílica confirmada histologicamente, sem que tivessem sido objetivadas eosinofilia periférica ou envolvimento do sistema nervoso periférico. A presença de pólipos nasais neste doente (característica não contemplada nos critérios ACR 1990) permitiu a classificação deste doente após aplicação dos novos critérios. Contudo, a elevada ponderação atribuída à patologia obstrutiva das vias aéreas e à presença de pólipos nasais nos critérios mais recentes permitiu a sua classificação como GEPA.
Conclusões: Todos os doentes com diagnóstico clínico de GPA/ GEPA cumpriram os novos critérios de classificação ACR/ EULAR 2022. No único doente não classificado como GPA/GEPA de acordo com os critérios ACR 1990, verificou-se o cumprimento dos novos critérios de classificação ACR/ EULAR 2022. A inclusão de critérios clínicos de baixa gravidade mas de elevada especificidade, como é o caso dos pólipos nasais, permite a classificação do doente para GEPA numa fase mais precoce da doença, comparativamente aos critérios prévios. Tal permite a inclusão de formas progressivamente mais precoces da doença em ensaios clínicos o que, em última análise, resultará em oportunidades de intervenção mais precoces com melhoria do prognóstico.

225 - JUVENILE PSORIATIC ARTHRITIS - CLINICAL CHARACTERIZATION AND DIFFERENCES FROM ADULT-ONSET DISEASE

Vanessa Fraga[1], Catarina Abreu[1], Sandra Sousa[1], Maria José Santos[1, 2]

[1]Reumatology Department, Hospital Garcia de Orta, EPE, 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: Juvenile psoriatic arthritis (jPsA) is a category of juvenile idiopathic arthritis (JIA), clinically heterogenous.
Objectives: Characterize clinical features of jPsA and compare with adult-onset psoriatic arthritis (aPsA).
Methods: Children fulfilling the ILAR criteria for jPsA and adults fulfilling CASPAR criteria for PsA were included. Demographic and clinical data was retrieved from the Portuguese registry Reuma.pt. A cross-sectional descriptive analysis of jPsA followed by a comparison with aPsA was performed.
Results: A total of 11 jPsA and 132 aPsA were included. jPsA represented 8.9% of JIA patients: 8 girls (72.7%), mean age at disease onset 10.4 ± 3.7 years and at diagnosis 11.9 ± 3.9 years, all Caucasian. Five (45.5%) developed psoriasis before arthritis and in 2 patients, psoriasis followed the onset of arthritis. Family history of psoriasis in 1st degree relatives was positive in 63.6%. The most frequent articular pattern was oligoarthritis (63.6%, n=7) and the most common musculoskeletal features were dactylitis (27.3%, n=3) and enthesitis (27.3%, n=3). Axial involvement was present in only one case. All jPsA patients were rheumatoid factor negative, six (54.5%) were ANA positive and none was HLA-B27 positive. Seven were treated with methotrexate, in combination with a biologic in four of them. The mean age of aPsA onset was 58.9 ± 14.1 years, 51.5% (n=68) were males, and the most common joint pattern was polyarthritis (59.1%, n=78). Presence of psoriasis at diagnosis was significantly more frequent in aPsA than in jPsA (84,8% vs 45.5%; p-value= 0.01). A total of 77 patients were treated with a biologic, most

Table I. Extra-articular manifestations in jPsA and aPsA

	jPsA %(n)	aPsA% (n)	p-value
Psoriasis at diagnosis	45.5 (5)	84.8 (112)	p=0.01
Dactylitis	27.3 (3)	30.2 (38)	p=0.572
Enthesitis	27.3 (3)	35.3 (44)	p=0.748
Nail involvement	18.2 (2)	42.9 (48)	p=0.197
Axial involvement	9.1 (1)	17.4 (23)	p=0.418
Inflammatory bowel disease	9.1 (1)	0 (0)	p=0.080
Uveitis	0 (0)	1.6 (2)	p=0.846

frequently adalimumab (n=27), etanercept (n=21) and infliximab (n=15). Seventy-two patients were treated with csDMARDs alone or in combination with a biologic.

Conclusion: At disease onset, less than half of the jPsA had psoriasis, and the most common joint pattern was oligoarthritis. The presence of dactylitis, enthesitis, and a positive family history of psoriasis are helpful in establishing the diagnosis. Conversely, aPsA patients were more often male, with psoriasis at the time of diagnosis and with polyarticular disease. No differences were found concerning extra-articular manifestations.

226 - POLIANGEITE MICROSCOPICA E OS NOVOS CRITÉRIOS DE CLASSIFICAÇÃO ACR/EULAR 2022

Adriana Carones[1], Beatriz Mendes[1], Helena Assunção[1]
[1]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

Introdução: A poliangeite microscópica (MPA) é uma vasculite necrotizante de pequenos vasos que afeta predominantemente os capilares renais e pulmonares, com frequente positividade para p-ANCA (imunofluorescência) ou anti-MPO (ELISA), não incluindo o envolvimento do trato respiratório superior habitual nas outras vasculites ANCA. Adicionalmente, a histopatologia demonstra vasculite necrotizante de pequenos vasos mas não inclui as características distintivas de inflamação granulomatosa e eosinofilia extra-vascular presentes nas outras vasculites ANCA. O diagnóstico é baseado na combinação dos achados clínicos, serológicos e histológicos. Apenas estabelecida como entidade independente e separada da poliarterite nodosa na 1ª conferência de consenso de Chapel Hill 1994, não existiam critérios de classificação amplamente aceites para esta patologia até ao desenvolvimento dos critérios de classificação ACR/EULAR 2022.

Objetivos: Demonstrar a aplicação dos novos critérios de classificação ACR/EULAR 2022 em doentes com diagnóstico inaugural de MPA seguidos num centro terciário em Portugal.

Métodos: Foram identificados todos os doentes com diagnóstico clínico inequívoco de MPA seguidos em consulta de Reumatologia no serviço do CHUC. Dados clínicos, laboratoriais, imagiológicos e histológicos relevantes relativos ao momento do diagnóstico foram colhidos retrospetivamente através da consulta do processo clínico e do Reuma.pt e foi averiguado o cumprimento dos itens incluídos nos critérios de classificação ACR 1990 e ACR/EULAR 2022 ao momento do diagnóstico.

Resultados: Foram identificados 2 doentes com MPA, cuja caracterização demográfica manifestações clínicas e laboratoriais inaugurais relevantes para a classificação segundo os critérios ACR/EULAR 2022 estão descritos na Tabela 1.

Discussão/Conclusões: A MPA é uma doença rara, até recentemente pouco reconhecida e que comporta um prognóstico reservado na ausência de reconhecimento e tratamento atempados. Assim, é importante uma elevada suspeição clínica, radicada no conhecimento das características clínicas e laboratoriais associadas a estas condições, bem como das características que nos devem por na pista de outras vasculites de pequenos vasos com as quais partilha semelhanças. Os novos critérios de classificação EULAR/ACR 2022 para a MPA visam a

Tabela I. Características demográficas, clínicas e laboratoriais inaugurais e cumprimento dos critérios de classificação ACR/EULAR 2022

MPA				
ID			1	2
Sexo			M	M
Idade (anos)			46	48
Clínicos	Nasal (epistáxis, crusting ou perfuração septo)	-3	0	0
Laboratoriais, Imagem ou Biópsia	p-ANCA/MPO	+6	+6	+6
	Fibrose ou DPI na radiografia de tórax	+3	0	0
	GNF pauci-imune biópsia	+3	+3	+3
	cANCA ou anti-PR-3	-1	0	0
	Eosinofilia periférica	-4	0	0
Total - classificação se ≥ 5			6	9

identificação de grupos homogéneos de doentes com MPA recém-diagnosticada mas o sistema de ponderação utilizado pode auxiliar o raciocínio diagnóstico, na medida em que atribui ponderação elevada às manifestações mais paradigmáticas da doença, nomeadamente envolvimento renal e positividade para anti-MPO, permitindo a pontuação negativa de manifestações clínicas e laboratoriais comuns nas outras vasculites ANCA.

230 - PREDICTORS OF MORTALITY IN CONNECTIVE TISSUE DISEASES – INTERSTITIAL LUNG DISEASE

Francisca Guimarães[1], Mariana Serino[2], Catarina Soares[1], Hugo Parente[1], Miguel Bernardes[3,4], Lúcia Costa[3], André Terras Alexandre[2], Natália Melo[2], Patrícia Mota[2,4,5], Hélder Bastos[2,4,5], António Morais[2,4,5]
[1] Rheumatology department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal; [2] Interstitial Lung Diseases Unit, Pneumology department, Centro Hospitalar Universitário do São João, Porto, Portugal; [3] Rheumatology department, Centro Hospitalar Universitário do São João, Porto, Portugal; [4] Faculdade de Medicina da Universidade do Porto, Porto, Portugal; [5] Instituto de Investigação e Inovação em Saúde (i3S).

Background: Connective tissue diseases (CTD) are an heterogenous group of systemic diseases that are often associated with lung involvement, usually with interstitial lung disease (ILD) presentation. Additionally, ILD is considered a main cause of mortality in CTD, therefore identifying high-risk patients is crucial to proper management. This study aimed to evaluate predictors of mortality in CTD-ILD.
Methods: Retrospective and longitudinal study including patients diagnosed with CTD-ILD followed between January 2019 until February 2022, in a tertiary pneumology centre. Sociodemographic data, CTD manifestations and immunology were collected. Baseline pulmonary functional tests (PFT) and radiologic pattern on high-resolution computer tomography (HRCT), and after 6 (ΔPFT-6), 12 (ΔPFT-12) months of the diagnosis and on last appointment were recorded. Mortality rate and causes of death were also recorded. Survival analysis was performed using Kaplan-Meier analysis and Cox regression. SPSS v25 was used for statistical analysis and significance level was defined as 2-sided $p< 0.05$.
Results: In total, 135 patients were included, 68.9% were female, with a mean age at diagnosis of 60.05±12.77 years. The most common CTD were systemic sclerosis - SSc - (28.9%) and rheumatoid arthritis - RA - (28.1%). Non-specific interstitial pneumonia (NSIP) and usual interstitial pneumonia (UIP) were the most common radiological pattern (40.7% and 37.0%, respectively). Most of the patients had a restrictive pat-

Table I. Patient and disease characteristics at baseline

	All patients n=135
Sociodemographic characteristics	
Age (years)	66.05±12.77
Gender (Female)	93/135 (68.9%)
Race (White European origin)	129/135 (95.6%)
Smoking status (Never smoked)	85/132 (64.4%)
Comorbidities	
Hypertension (yes)	79/135 (51.1%)
Dyslipidemia (yes)	64/135 (47.4%)
Diabetes mellitus (yes)	17/134 (12.7%)
Sleep apnea (yes)	24/135 (17.8%)
Chronic obstructive pulmonary disease (yes)	7/134 (5.2%)
Cardiac failure (yes)	16/135 (11.9%)
CTD- Diagnosis	
Rheumatoid arthritis	38/135 (28.1%)
Systemic sclerosis	39/135 (28.9%)
Inflammatory immune mediated myopathy	18/135 (13.3 %)
Antisyntetase syndrome	11/18 (61.1%)
Dermatomyositis	6/18 (33.3 %)
Polymyositis	1/18 (5.6 %)
Sjögren Syndrome	14/135 (10.4%)
Systemic lupus erythematosus	11/135 (8.1%)
Overlap syndrome	9/135 (6.7%)
Rheumatoid arthritis/Systemic sclerosis	5/9 (55.6%)
Systemic lupus erythematosus/ Systemic sclerosis	3/9 (33.3%)
Systemic sclerosis/Polymyositis	1/9 (11.1%)
Mixed connective tissue disease	4/135 (3.0%)
Antiphospholipid syndrome	1/135 (0.7%)
Undifferentiated Connective Tissue disease	1/135 (0.7%)
Secondary Sjögren syndrome	18/132 (13.6%)
ILD-disease characteristics	
Age at diagnosis (years)	60.51±13.23
Previous CTD diagnosis (yes)	106/135 (78.5%)
PFT pattern	
Normal	18/135 (13.3%)
Restrictive	44/135 (32.6%)
Obstructive	21/135 (15.6%)
Isolated decreased DLCO	52/135 (38.5%)
ILD classification	
Non-specific interstitial pneumonia	55/135 (40.7%)
Usual interstitial pneumonia	50/135 (37.0%)
Lymphocytic interstitial pneumonia	5/135 (3.7%)
Organizing pneumonia	10/135 (7.4%)

continues on the next page

Table I. Continuation

	All patients n=135
Organizing pneumonia/non-specific interstitial pneumonia	7/135 (5.2%)
Desquamative interstitial pneumonia	1/135 (0.7%)
Diffuse alveolar hemorrhage	1/135 (0.7%)
Fibroelastosis	2/135 (1.4%)
Interstitial lung abnormalities	4/135 (3.0%)
Baseline PFT	
FVC < 70%	26/115 (22.6%)
DLCO < 70%	74/93 (79.6%)
Baseline changes in HRCT	
Reticulations (yes)	104/135 (77.7%)
Ground Glass (yes)	84/135 (62.2%)
Traction bronchiectasis (yes)	66/135 (48.9%)
Honeycombing (yes)	25/135 (18.5%)
Fibrosis (yes)	73/135 (54.1%)
Lung extension > 20% (yes)	42/135 (31.1%)
Treatment	
Glucocorticoid (ever)	109/134 (81.3%)
Hydroxychloroquine (ever)	41/135 (30.4%)
Methotrexate association (ever)	51/106 (48.1%)
Mycophenolate mofetil association (ever)	49/135 (36.3%)
Cyclophosphamide (ever)	27/135 (20.0%)
Rituximab (ever)	44/135 (32.6%)
Tocilizumab (ever)	1/135 (0.7%)
Nintedanib (ever)	21/135 (15.7%)
Pirfenidone (ever)	2/135 (1.4%)
Mortality	
Deaths (yes)	12/135 (8.9%)
Causes of dead	
Infection	10/12 (83.3%)
Malignancy	1/12 (8.3%)
Follow-up time (months), median (IQR)	58.80 (79.98)

Legend: CTD: connective tissue disease; DLCO: diffusion lung carbon oxide; FVC: functional vital capacity; HRCT: high resolution computerized tomography; ILD: Interstitial lung disease; IQR: Interquartile range, PFT: Pulmonary functional tests. Continuous variables are presented in mean ± standard deviation and discrete variables number/total population available (percentage).

tern on PFT (32.6%). Baseline characteristics are described in Table 1. After a median follow-up time of 58.1 (IQR 79.98) months, 8.9% of the patients died, mainly due to respiratory infection (83.3%).

In univariate analysis, male gender ($\chi2$=13.26, p<.0001), smoking history ($\chi2$=5.56, p=.018), chronic obstructive lung disease ($\chi2$=20.38, p<.0001), lack of a previous CTD-diagnosis ($\chi2$=4.52, p=.034), UIP pattern ($\chi2$=9.95, p=.002), baseline evidence of honeycombing ($\chi2$=4.37, p=.037), baseline lung extension>20% on HRCT ($\chi2$=4.524, p=.033), lung emphysema ($\chi2$=12.97, p<.0001) and RA ($\chi2$=3.88, p=0.049) were associated with a worse prognosis. Even though Sjögren Syndrome (SSj) diagnosis didn't reach statistical significance, the mean survival time was numerically inferior comparing to other diseases (149 VS 206 months, p=0.08). In this study, neither baseline PFTs nor ΔPFT-6 and ΔPFT-12 were associated with mortality. In multivariate analysis, using Cox regression, age at diagnosis (HR 1.13, p=0.002), emphysema (HR 10.98, p=0.015), honeycombing at baseline (HR 5.62, p=0.024) and RA (HR=6.045, p=0.048) were independents predictors of mortality.

Conclusions: Age, concomitant emphysema, baseline honeycomb on HRCT and RA were independent predictors of worse prognosis in this CTD-ILD cohort which are in line with the literature. The honeycombing pattern, an advance disease manifestation, suggests that an early diagnosis and treatment could be determinant to improve CTD-ILD outcome. Also, RA patients require a tighter follow up and probably earlier therapy.

234 - NINTEDANIB IN RHEUMATIC DISEASE-ASSOCIATED INTERSTITIAL LUNG DISEASES – DESCRIPTION OF A SINGLE CENTER EXPERIENCE

Augusto Silva[1], Nikita Khmelinskii[2], Vasco C Romão[2], Cristina Ponte[2], JE Fonseca[2], Catarina Resende[2]

[1]Rheumatology and Bone Metabolic Diseases Department, Centro Hospitalar Lisboa Norte, EPE - Hospital de Santa Maria, Lisbon Academic Medical Centre, Lisboa, Portugal, [2]Serviço de Reumatologia e Doenças Ósseas Metabólicas do Hospital de Santa Maria, Centro Hospitalar Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introduction: Interstitial lung disease (ILD) may be a severe manifestation of inflammatory rheumatic diseases (IRD). Conventional treatments have shown modest and short-lived success rates in the control of ILD. Nintedanib, an oral tyrosine kinase inhibitor with antifibrotic properties, was one of the first drugs approved for treating idiopathic pulmonary fibrosis (IPF) and has recently been approved for systemic sclerosis-associated ILD. However, experience with the use of nintedanib in other rheumatic disorders with associated ILD is still scarce.

Objective: To describe the clinical features and outcomes of patients with IRD-ILD treated with nintedanib in our department.

Methods: Chart review, from 2018 to 2022, of the

Rheumatology and Pulmonology medical records for inclusion of patients with rheumatic disorders and ILD treated with nintedanib.

Results: We identified 11 patients (90.9% women) with IRD-ILD treated with nintedanib for a mean of 20.2±12.8 (11-52) months (Table 1). The mean age at diagnosis was 58.9±16.2 (33-78) years and the mean time for nintedanib initiation after diagnosis was 6.36±3.4 (1-15) years. The most frequently underlying IRD was systemic sclerosis (8/11; 72.7%). ILD was diagnosed prior to the IRD only in two patients (18.2%). Computed tomography identified nonspecific interstitial pneumonia in 6 (54.5%) cases and usual interstitial pneumonia in 5 (45.5%). All patients had positive antinuclear antibodies (titer: 1/160-1/1280): anti-Scl70 in 8 (72.7%) cases, anti-Ro60 in 3 (27.3%) and anti-Ro52 in 1 (9.1%). Rheumatoid factor was present in 3 (27.3%) patients, anti-citrullinated peptide antibody in 2 (18.2%), and antineutrophil cytoplasmic antibody against myeloperoxidase in 1 (9.1%). Before the initiation of nintedanib, respiratory function tests showed a low forced vital capacity (FVC) in all patients, low forced expiratory volume in the first second (FEV1) in 9 (81.8%), low total lung capacity (TLC) in 10 (90.9%) and a low single-breath diffusing capacity of the lungs for CO (DLCO) in 7 (63.6%). Pulmonary hypertension was present in 6 (54.5%) patients, confirmed by right heart catheterization, and oxygen therapy was necessary for 10 (90.9%) patients.

All patients had previously received immunosuppressive therapy. Glucocorticoids and mycophenolate mofetil were the most commonly used (10/11; 90.9%), followed by azathioprine and cyclophosphamide (3/11, 27.3%). After a mean exposure of 20.2±12.8 (11-52) months to nintedanib, mild improvement of the FVC (8.75%) and FEV1 (7.25%) was observed in 4 (36.6%) patients and of the TLC (3.25%) in 3 (27.3%), while the single-breath DLCO improved in only one (13%) patient. The best outcomes were observed in the patient with microscopic polyangiitis who started nintedanib one year after the ILD diagnosis. During follow-up, 5 (45%) patients died as a result of ILD, 3 (27%) had ILD

Table I. Clinical features of patients with IRD-ILDs

Sex	Diagnosis	Immune profile	Age at diagnosis	Start of NTD	Illness duration before NTD initiation	ILD pattern on CT	Previous treatments	NTD duration of treatment	Outcome
Male	PM	anti-Ro60 + anti-Ro52 + ACPA + RF +	33	02.2021	6 years	UIP	PDN, MPM, CYC, MPM + tacrolimus	16 months	ILD progression
Female	dcSSc	anti-Scl70 +	49	02.2018	15 years	NSIP	PDN, CYC, MPM, MA, bosentan	52 months	ILD progression
Female	dcSSc	anti-Scl70 + anti-Ro60 +	52	05.2019	8 years	NSIP	PDN, AZA, CYC, MPM	13 months	Stopped NTD ILD progression
Female	dcSSc	anti-Scl70 +	79	08.2020	6 years	UIP	DFZ, MPM	14 months	Stopped NTD Remains stable
Female	RA	ACPA + RF +	66	07.2019	8 years	UIP	PDN, MTX, SLZ, HCQ, ETN, IFX, RTX, ABA	26 months	Died
Female	dcSSc	anti-Scl70 +	58	03.2018	5 years	NSIP	PDN, CYC, MPM, riociguat	27 months	Died
Female	dcSSc	anti-Scl70 +	74	12.2018	6 years	UIP	PDN, MPM	12 months	Died
Female	dcSSc	anti-Scl70 + anti-Ro60 +	78	07.2021	5 years	UIP	MPM	11 months	Remains stable
Female	dcSSc	anti-Scl70 +	38	12.2019	5 years	NSIP	PDN, AZA, MPM	11 months	Died
Female	dcSSc	anti-Scl70 +	48	12.2020	5 years	NSIP	MP, MTX, AZA, MPM	10 months	Died
Female	MPA	ANCA-MPO + RF+	73	12.2019	1 year	NSIP	PDN, MPM, MA	30 months	Remains stable

Abbreviations: ABA - abatacept; ACPA - anti-citrullinated peptide antibody; ANCA-MPO - antineutrophil cytoplasmic antibody against myeloperoxidase; AZA - azathioprine; CYC - cyclophosphamide; dcSSc - diffuse cutaneous systemic sclerosis; DFZ - deflazacort; ETN - etanercept; HCQ - hydroxychloroquine; IFX - infliximab; ILD - interstitial lung disease; MA - mycophenolic acid; MP - methylprednisolone; MPA - microscopic polyangiitis; MPM - mycophenolate mofetil; MTX - methotrexate; NSIP - nonspecific interstitial pneumonia; NTD - nintedanib; PDN - prednisolone; PM - polymyositis; RA - rheumatoid arthritis; RF - rheumatoid factor; RTX - rituximab; SLZ - sulfasalazine; UIP - usual interstitial pneumonia.

progression and 3 (27%) remained stable. Regarding tolerance: 5/11(45.5%) patients were able to maintain a daily dose of 150 mg bd, 4/11 (36.4%) required dose reduction to 100 mg bd due to gastrointestinal symptoms, and 2/11 (18.2%) discontinued treatment due to gastrointestinal intolerance or hepatotoxicity.
Conclusion: ILD is a potential complication of IRDs with a poor prognosis, having a dramatic impact and burden on these patients. Nintedanib may improve lung function and halt ILD progression in a subset of patients. A multidisciplinary approach and early initiation of nintedanib seem to be associated with better outcomes.

235 - SÍNDROME DE IGG4 – A COORTE PORTUGUESA

Hugo Parente[1], Adriana Carones[2], Augusto Silva[3], Carlos Costa[4], Catarina Soares[1], Inês Santos[5], Manuel Silvério-António[3, 6], Filipa Teixeira[1]
[1]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [2]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [3]Serviço de Reumatologia, Centro Hospitalar Lisboa Norte - Hospital de Santa Maria, Lisboa, Portugal, [4]Serviço de Reumatologia, Centro Hospitalar Trás-os-Montes e Alto Douro, Vila Real, Portugal, [5]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [6]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Lisboa, Portugal

Introdução: A doença relacionada a IgG4 (DRIgG4) é recente, de base pseudotumoral fibro-inflamatória, carácter indolente e atingimento difuso - pancreático, glandular salivar e lacrimal, mas também retroperitoneal, vascular, etc. Afecta sobretudo homens a partir da meia-idade1. Pode mimetizar neoplasias, sendo o seu diagnóstico complexo. Tem resposta excelente a corticoterapia2, mas recidiva apreciável3.
Objectivos: Caracterizar a coorte portuguesa de doentes com DRIgG4.
Métodos: Estudo observacional multicêntrico nacional, incluindo doentes com DRIgG4 seguidos em Reumatologia.
Resultados: Incluíram-se 15 doentes, com idade média de 61,21 anos (DP=10,79), mediana de idade ao diagnóstico de 58 (AIQ 36) e ao início de sintomas de 53,5 (AIQ 22). Sete (46,7%) eram do sexo masculino. Apenas 1 doente (6,7%) revelou envolvimento pancreático, 2 (13,3%) pulmonar, 2 (13,3%) retroperitoneal, 2 (13,3%) renal, 6 (40%) orbital, 3 (20%) de vias biliares, 5 (33,3%) de glândulas lacrimais (4 deles, bilateralmente), 3 (20%) da aorta, 4 (26,7%) de glândulas salivares (bilateralmente) e 1 (6,7%) de tiróide.

Tabela I.

Dados demográficos	
Sexo masculino, n/N (%)	7/15 (46,7)
Idade ao início de sintomas, mediana (AIQ)	53,5 (22)
Idade ao diagnóstico, mediana (AIQ)	58 (36)
Idade actual, média (DP)	61,21 (10,79)
Óbitos, n/N (%)	1/15 (6,7)
Estilo de vida	
Tabagismo	
Prévio, n/N (%)	5/15 (33,3)
Actual, n/N (%)	1/15 (6,7)
Dados clínicos	
Envolvimento pancreático, n/N (%)	1/15 (6,7)
Envolvimento pulmonar, n/N (%)	2/15 (13,3)
Envolvimento retroperitoneal, n/N (%)	2/15 (13,3)
Obstrução renal, n/N (%)	1/15 (6,7)
Envolvimento renal, n/N (%)	2/15 (13,3)
Nefrite tubulointersticial, n/N (%)	1/15 (6,7)
Envolvimento orbital, n/N (%)	6/15 (40)
Dacrioadenite, n/N (%)	3/15 (20)
Dacriocistite, n/N (%)	1/15 (6,7)
Pseudotumor orbital, n/N (%)	1/15 (6,7)
Envolvimento de vias biliares, n/N (%)	3/15 (20)
Espessamento e fibrose ductal, n/N (%)	3/15 (20)
Envolvimento de glândulas lacrimais, n/N (%)	5/15 (33,3)
Bilateral, n/N (%)	4/15 (26,7)
Envolvimento de meninges, n/N (%)	0/15 (0)
Envolvimento aórtico, n/N (%)	3/15 (20)
Aorta infra-renal, n/N (%)	2/15 (13,3)
Artérias ilíacas, n/N (%)	2/15 (13,3)
Outro envolvimento vascular, n/N (%)	2/15 (13,3)
Aneurisma cerebral, n/N (%)	1/15 (6,7)
Encapsulamento de artéria pulmonar por pseudotumor, n/N (%)	1/15 (6,7)
Envolvimento de glândulas salivares, n/N (%)	4/15 (26,7)
Bilateral, n/N (%)	4/15 (26,7)
Envolvimento tiroideu, n/N (%)	1/15 (6,7)
Hepatite autoimune, n/N (%)	0/15 (0)

continua na próxima página

Tabela I. continuação

Pericardite constrictiva, n/N (%)	0/15 (0)
Mastite esclerosante, n/N (%)	0/15 (0)
Gastrite, n/N (%)	0/15 (0)
Neoplasia, n/N (%)	0/15 (0)
Hidraadenite supurativa, n/N (%)	1/15 (6,7)
Linfadenopatias, n/N (%)	2/15 (13,3)
Polisserosite, n/N (%)	1/15 (6,7)
Pseudotumor mediastínico, n/N (%)	2/15 (13,3)
Pseudotumor epidural com compressão medular, n/N (%)	1/15 (6,7)
Imagiologia	
Tomografia - Sausage pancreas, n/N (%)	0/15 (0)
Tomografia - Nodulose pulmonar, n/N (%)	2/15 (13,3)
Tomografia - Padrão broncovascular pulmonar, n/N (%)	0/15 (0)
Tomografia - Padrão intersticial pulmonar, n/N (%)	1/15 (6,7)
Tomografia - Vidro despolido pulmonar, n/N (%)	4/15 (26,7)
PET – achados inflamatórios, n/N (%)	8/15 (53,3)
Histologia	
Fibrose storiform, n/N (%)	4/15 (26,7)
Infiltrado linfoplasmocítico, n/N (%)	12/15 (80)
Flebite obliterante, n/N (%)	0/15 (0)
Rácio IgG4/IgG >40% em campo de grande ampliação, n/N (%)	4/15 (26,7)
Análises laboratorais	
Factor reumatóide positivo, n/N (%)	0/15 (0)
Anticorpos anti-nucleares positivos, n/N (%)	1/15 (6,7)
Eosinofilia periférica, n/N (%)	3/15 (20)
Proteina C-reactiva elevada, n/N (%)	8/15 (53,3)
Velocidade de sedimentação elevada, n/N (%)	8/15 (53,3)
Níveis séricos de IgG4 elevados, n/N (%)	10/15 (66,7)
Níveis séricos de IgG1 elevados, n/N (%)	1/15 (6,7)
Níveis séricos de IgE elevados, n/N (%)	1/15 (6,7)
Tratamento inicial	
Corticosteróides - pulsos, n/N (%)	0/15 (0)
Corticosteróides - oral, n/N (%)	11/15 (73,3)
Dose em miligramas, média (DP)	23,8 (23,76)
Ciclofosfamida, n/N (%)	0/15 (0)
Micofenolato de mofetil, n/N (%)	2/15 (13,3)

continua na próxima coluna

Tabela I. continuação

Azatioprina, n/N (%)	0/15 (0)
Metotrexato, n/N (%)	2/15 (13,3)
Leflunomida, n/N (%)	0/15 (0)
Hidroxicloroquina, n/N (%)	0/15 (0)
Tacrolimus, n/N (%)	0/15 (0)
Ciclosporina, n/N (%)	0/15 (0)
Imunoglobulinas intravenosas, n/N (%)	0/15 (0)
Rituximab, n/N (%)	0/15 (0)
Anti-TNF, n/N (%)	0/15 (0)
Outro - antibiótico, n/N (%)	1/15 (6,7)
Tratamento actual	
Corticosteróides - pulsos, n/N (%)	1/15 (6,7)
Corticosteróides - oral, n/N (%)	9/15 (60)
Dose em miligramas, média (DP)	5,33 (4,76)
Ciclofosfamida, n/N (%)	0/15 (0)
Micofenolato de mofetil, n/N (%)	2/15 (13,3)
Azatioprina, n/N (%)	1/15 (6,7)
Metotrexato, n/N (%)	0/15 (0)
Leflunomida, n/N (%)	0/15 (0)
Hidroxicloroquina, n/N (%)	1/15 (6,7)
Tacrolimus, n/N (%)	0/15 (0)
Ciclosporina, n/N (%)	0/15 (0)
Imunoglobulinas intravenosas, n/N (%)	0/15 (0)
Rituximab, n/N (%)	5/15 (33,3)
Anti-TNF, n/N (%)	0/15 (0)
Outro - cirurgia, n/N (%)	1/15 (6,7)
Resultado	
Resposta a corticosteróides, n/N (%)	13/15 (86,7)
Redução dos níveis séricos de IgG4, n/N (%)	9/15 (60)
Normalização dos níveis séricos de IgG4, n/N (%)	5/15 (33,3)

DP: desvio padrão; AIQ: amplitude interquartil

Nenhum exibiu acometimento meníngeo. Não se documentaram neoplasias. Tomograficamente, nenhum revelou sausage pancreas, 2 (13,3%) demonstraram nodulose pulmonar, 1 (6,7%) padrão intersticial pulmonar e 4 (26,7%) vidro despolido. Histologicamente, 4 (26,7%) apresentavam fibrose storiform, 12 (80%) infiltrado linfoplasmocítico e 4 (26,7%) rácio IgG4/IgG

> 40%. Em 8 (53,3%) dos casos, a PET evidenciou inflamação. Analiticamente, 3 (20%) cursaram com eosinofilia periférica, 8 (53,3%) com elevação de VS e PCR, 10 (66,7%) com IgG4 aumentada, e 1 (6,7%) com ANA positivos, IgG1 e IgE elevados. Registou-se 1 (6,7%) morte, de causa desconhecida, aos 65 anos. Em primeira linha, 11 (73,3%) dos doentes iniciaram corticóide (CCT) oral, de dose média 23,8mg (DP=23,76) (máx=80mg/d), 2 (13,3%) micofenolato de mofetil (MMF), e 2 (13,3%) metotrexato (MTX). Actualmente, 1 (6,7%) doente encontra-se sob pulsos de CCT, pelo envolvimento pulmonar e aórtico, 9 (20%) com CCT oral, de dose média 5,33mg (DP=4,26), 2 (13,3%) com MMF, 1 (6,7%) com azatioprina, 1 (6,7%) com hidroxicloroquina e 5 (33,3%) com Rituximab. Treze (86,7%) responderam bem ao CCT inicial, 9 (60%) reduziram os níveis séricos de IgG4, tendo 5 (33,3%) normalizado.

Conclusão: A coorte portuguesa é heterogénea, com um discreto predomínio no sexo feminino, a partir da meia idade, e envolvimento mais comum orbital e glandular. Verificou-se boa resposta a CCT, com redução dos níveis circulantes de IgG4, mas necessidade de progressão terapêutica para outros imunomoduladores. A sua escolha depende dos acometimentos orgânicos – da terapêutica inicial, verificou-se falência ao MTX, boa resposta ao MMF, e necessidade moderada de RTX.

Referências: [1] Uchida K, Masamune A, Shimosegawa T, Okazaki K. Prevalence of IgG4-Related Disease in Japan Based on Nationwide Survey in 2009. Int J Rheumatol. 2012; 2012:358371. [PubMed: 22899936] [2] Tzaribachev et al. Arthritis Care Res 2009; [2] Abraham M, Khosroshahi A. Diagnostic and treatment workup for IgG4-related disease. Expert Rev Clin Immunol. 2017;13(9):867-875. doi:10.1080/1744666X.2017.135469; [3] Hart PA, Topazian MD, Witzig TE, Clain JE, Gleeson FC, Klebig RR, et al. Treatment of relapsing autoimmune pancreatitis with immunomodulators and rituximab: the Mayo Clinic experience. Gut. 2013; 62(11):1607–15. A thougtful description of experince with treatment of relapses and maintenance of remission in IgG4-RD using different steroid-sparing agents. [PubMed: 22936672].

237 - CUMULATIVE DOSE AND EXPOSURE TIME TO METHOTREXATE WERE NOT SHOWN TO BE PREDICTORS OF HEPATIC FIBROSIS BY ELASTOGRAPHY - A MONOCENTRIC COHORT STUDY

Filipe Oliveira Pinheiro[1, 2], Rui Gaspar[2, 3], Bruno Miguel Fernandes[1], Armando Peixoto[2, 3], Guilherme Macedo[2, 3], Iva Brito[2, 4]

[1]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal, [3]Gastroenterology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [4]Paediatric and Young Adult Rheumatology Unit, Centro Hospitalar Universitário de São João, Porto, Portugal

Background: Methotrexate is an immunomodulatory agentu sed in several inflammatory diseases, such as rheumatoid arthritis (RA), spondyloarthritis (SpA) or inflammatory bowel disease (IBD). Despite being previously associated with the development of liver damage, recent studies have showed significant less hepatotoxicity. The aim of our study was to evaluate the prevalence and development of liver injury in methotrexate-treated patients with inflammatory diseases.

Methods: We performed a cross-sectional study where consecutive patients followed at the Rheumatology and Gastroenterology Departments of a tertiary center, diagnosed with RA, SpA or IBD, treated with methotrexate, were submitted to liver elastography and liver function tests. Clinical and laboratory data were collected, including duration of treatment and cumulative dose of methotrexate. The cutoff for significant fibrosis was ≥7.1 kPa. Comparisons between groups (significant versus non-significant fibrosis) was evaluated using chi-square, t test and Mann-Whitney U test. Spearman's correlation was used to assess associations between two or more continuous variables. Logistic regression was performed to determine predictors of significant fibrosis.

Results: A total of 101 patients were included, 60 (59.4%) females, aged 46.2±12.6 years at the start of methotrexate. Forty-nine patients (48.5%) had RA, 17 (16.8%) SpA and 35 (34.7%) IBD, and 35 (34.7%) consumed alcohol daily. Patients were exposed to methotrexate for a median of 204 (72-146) weeks, with a median cumulative dose of 2385 (940-5200) mg. Eleven patients (10.9%) had significant fibrosis, with a median score of 4.8 (4.1-5.9) kPa.

Patients with significant fibrosis had higher rates of daily alcohol consumption (63.6% vs 31.1%, p=0.045), but there were no differences in methotrexate's exposure time or cumulative dose. There was a very significant correlation between exposure time to methotrexate and cumulative dose (Spearman's rho 0.904, p<0.001), but no correlations were found between cumulative dose and elastography scores. Methotrexate exposure time (OR 1.001, 95% CI 0.999-1.003, p=0.549) and cumulative dose (OR 1.000, 95% CI 1000-1000, p=0.629) were shown not to be predictors of significant fibrosis, unlike alcohol (OR 3.875, 95%CI 1.049-014319, p=0.042). In multivariate logistic regression analysis, methotrexate cumulative and exposure times were not predictors of significant fibrosis, even when adjusted for alcohol consumption.

Another analysis was performed with the division of

groups at the median – ≤4.8kPa (n=55) and >4.8 kPa (n=46). Methotrexate exposure time (OR 1.000, 95% CI 0.999-1.002, p=0.607) and cumulative dose (OR 1.000, 95% CI 1.000-1.000, p=0.376) were also shown not to be predictors of fibrosis in patients on methotrexate.

Conclusion: In this study, we found that fibrosis detected on hepatic elastography was not associated with the cumulative dose or time of exposure to methotrexate, unlike alcohol. Therefore, it is of paramount importance to redefine risk factors for liver toxicity In patients with inflammatory diseases under treatment with methotrexate.

238 - CLINICAL OUTCOMES OF COVID-19 PATIENTS WITH RHEUMATIC AND MUSCULOSKELETAL DISEASES: A SINGLE CENTRE COHORT STUDY

Carolina Mazeda[1, 2], Barcelos A.[1, 2]

[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz Health Alliance, Portugal, Aveiro, Portugal

Introduction: The coronavirus disease 2019 (COVID-19) pandemic, caused by the novel acute respiratory syndrome coronavirus 2 (SARS-CoV-2), has triggered a global health crisis. In the majority of the population, COVID-19 is a self-limiting viral disease with a good prognosis. Nevertheless, proportionately, severe disease occurs in patient with risk factors such as advanced disease, male gender and/or underlying diseases. Many of the large population-based or healthcare-based studies performed have suggested an increased risk of hospitalization or death from COVID-19 in people with rheumatic and musculoskeletal diseases (RMDs). European Alliance of Associations for Rheumatology (EULAR) recently published its recommendations for the management and vaccination of people with RMDs in the context of SARS-CoV-2. These reinforce that in general, patients with RMDs do not face more risk of contracting SARS-CoV-2 but may generally face worse outcomes and increased mortality.3 The aim of this study is describe our experience with a COVID-19 outbreak by identifying the clinical outcomes of patients with COVID-19 and RMDs.

Methods: Retrospective monocentric study that included patients with COVID-19 observed at the emergency department (ED) in a secondary hospital, between 2nd March 2020 and 31st December 2021. COVID-19 diagnosis was identified through the 10th International Classification of Diseases and RMDs were identified after revision of the clinical record. A COVID-19 diagnosis was based on polymerase chain reaction (PCR) test. Data collected included demographic and clinical data,

Table I. Patient demographic and clinical characteristics

	RMDs
Age, years (mean±SD)	76.93±27.90
Sex (M/F), n	126/87
Rheumatic disease, n (%)	
Osteoarthritis	94 (44.1)
Gout	36 (16.9)
Osteoporosis	30 (14.1)
Rheumatoid arthritis	27 (12.7)
Polymyalgia rheumatica	8 (3.8)
Spondyloarthritis	4 (1.9)
Systemic sclerosis	3 (1.4)
Vasculitis	3 (1.4)
Myositis	2 (0.9)
Systemic lupus erythematosus	2 (0.9)
Undifferentiated connective tissue disease	2 (0.9)
Sjögren's syndrome	1 (0.5)
Psoriatic arthritis	1 (0.5)
Disease duration, years (mean±SD)	6.3±3.2
Disease activity, n (%)	
Remission	
Low	20 (9.4)
Moderate	18 (8.5)
High	1 (0.5)
Not applicable	174 (81.7)
Immunosuppressive treatment, n (%)	
None	174 (81.7)
Glucocorticoids	44 (20.7)
csDMARDs	30 (14.1)
TNF inhibitors	1 (0.5)
Rituximab	2 (0.9)
Other b/tsDMARDs	2 (0.9)
Smoking Status (%)	
Smoker	4.7
Ex-smoker	1.4
Comorbidities	
Arterial hypertension	146 (68.5)
Dyslipidemia	102(47.9)
Diabetes mellitus	58 (27.2)
Obesity	52 (24.4)
Atrial fibrillation	41 (19.2)
Obstructive lung disease	14 (6.6)
Interstitial lung disease	5 (2.3)
Vaccination Status (%)	
Yes	64.8
No	35.2

RMDs - rheumatic and musculoskeletal diseases; cs - conventional synthetic; DMARDs - disease-modifying anti-rheumatic drugs; TNFi – tumour necrosis factor inhibitors; ts - targeted synthetic; b - biological

as well the complete course of the disease in the hospital. A descriptive analysis was performed using SPSS ® version 25, p-value≤0.05 was statistically significant. In multivariate analysis we included variables with a significant association in univariate analysis and those with clinical relevance (reported in other studies).
Results: Among 2169 patients with COVID-19 diagnosis, 213 (9.8%) had RMDs. Most of the patients included were women (59.2%) with a mean age of 76.93±27.90. Patient demographic and clinical characteristics are listed in table 1. One hundred and sixty (75.1%) patients with RMDs required hospitalization with one hundred and forty-nine (70%) requiring oxygen supply, six (2.8%) non-invasive ventilation and nine (4.2%) mechanical ventilation. A total of fifty-three (24.9%) patients died. Regarding COVID-9 management, glucocorticoids were administered to one hundred and five patients (49.3%) and nonspecific antivirals to four patients (1.9%). We found an association between the patients with COVID-19 requiring hospitalization (moderate/severe disease) and age (p=0.013), arterial hypertension (p=0.01), gout (p=0.01) and vaccination status (n=0.05). We found an increased risk of hospitalization in older patients [OR 1.07 (95% CI 1.04, 1.10)], gout [OR 1.16 (95% CI 1.01, 1.34)] and no vaccination [OR 2.63 (95% CI 1.23, 5.62)].
Conclusion: With our work, we were able to analyze how sociodemographic characteristics, comorbidities and therapeutic can affect the prognosis of patients with RMDs and COVID-19, as well as the identification of possible risk factors associated with hospitalization. Our results are not fully consistent with published studies, eg male and certain immunosuppressive treatments are no longer a significant risk factor for hospitalization. Larger studies are needed to identify groups with greater vulnerability as well as to understand the impact of vaccination.

239 - ULTRASOUND AND ELASTOGRAPHY IN THE ASSESSMENT OF SKIN INVOLVEMENT IN SYSTEMIC SCLEROSIS: A SYSTEMATIC LITERATURE REVIEW FOCUSING ON VALIDATION AND STANDARDIZATION

Tânia Santiago[1, 2], Eduardo Santos[3], Barbara Ruaro[4], Gemma Lepri[5], L Green[6], M Wildt[7], S Watanabe[8], Alain Lescoat[9], R Hesselstrand[7], Francesco Del Galdo[6], J Pauling[10], Annamaria Iagnocco[4], JAP da Silva[1, 2]
[1]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal, [2]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [3]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [4]Department of Pulmonology, University Hospital of Cattinara, University of Trieste, Italy., Trieste, Italy, [5]Department of Experimental and Clinical Medicine, University of Florence, and Division of Rheumatology AOUC, Firenze, Italy, [6]Rheumatology Department, Chapel Allerton Hospital, Leeds, United Kingdom, [7]Department of Rheumatology, Skåne University Hospital, Lund University, Lund, Stockholm, Sweden, [8]Department of Allergy and Rheumatology, Nippon Medical School Graduate School of Medicine, Tokyo, Japan., Tokyo, Japan, [9]Univ Rennes, CHU Rennes, Inserm, EHESP, Irset (Institut de Recherche en Santé, Environnement et Travail) - UMR_S 1085, Rennes, France, Rennes, France, [10]Royal National Hospital for Rheumatic Diseases (at Royal United Hospitals NHS FT), Bath, UK., Bath & North East Somerset, United Kingdom

Objective: Ultrasound and elastography are promising tools to foster much-needed improvement of skin assessment in systemic sclerosis (SSc). To summarize the published evidence in the literature on the role of ultrasound and elastography to assess skin involvement in SSc.
Methods: A systematic literature review (SLR) was performed within the "Skin Ultrasound Working Group" of the World Scleroderma Foundation, according to the Cochrane Handbook. A search was conducted in Pubmed, Cochrane Library and Embase databases from 1/1/1979 to 31/5/2021, using the participants, intervention, comparator and outcomes (PICO) framework (fig. 1). Only full-text articles involving adults, reported in any language, assessing ultrasound to quantify skin pathology in SSc patients. Two reviewers performed the assessment of risk of bias, data extraction and synthesis, independently.
Results: Forty-six studies out of 3248 references evaluating skin ultrasound and elastography domains were included. B-mode ultrasound was used in 30 studies (65.2%), elastography in nine (19.6%), and both methods in seven (15.2%). The ultrasound outcome measure domains reported were thickness (57.8%) and echogenicity (17.2%); the elastography domain was stiffness (25%). Methods used for image acquisition and analysis were remarkably heterogeneous and frequently under-reported, precluding data synthesis across studies. The same applies to contextual factors and feasibility. Our data syntheses indicated evidence of good reliability and convergent validity for ultrasound thickness evaluation against mRSS and skin histological findings (table 1). Stiffness and echogenicity have limited evidence for validity against histological findings. Evidence for sensitivity to change, test-retest reliability, clinical trial discrimination or thresholds of meaning is limited or absent for reported ultrasound domains.
Conclusion: Ultrasound is a valid and reliable tool for skin thickness measurement in SSc but there are sig-

nificant knowledge gaps regarding skin echogenicity assessment by ultrasound and skin stiffness evaluation by elastography in terms of feasibility, validity and discrimination. Standardization of image acquisition and analysis is needed to foster progress.

240 - WHY DON'T WE HAVE A INTERSTITIAL LUNG DISEASE SCREENING PROTOCOL FOR RHEUMATOID ARTHRITIS AS FOR SYSTEMIC SCLEROSIS?

Francisca Guimarães[1], Mariana Serino[2], Catarina Soares[1], Hugo Parente[1], Miguel Bernardes [3,4], Lúcia Costa [3], André Terras Alexandre[2], Natália Melo[2], Patrícia Mota[2,4,5], Hélder Bastos[2,4,5], António Morais[2,4,5]
[1] Rheumatology department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal; [2] Interstitial Lung Diseases Unit, Pneumology department, Centro Hospitalar Universitário do São João, Porto, Portugal; [3] Rheumatology department, Centro Hospitalar Universitário do São João, Porto, Portugal; [4] Faculdade de Medicina da Universidade do Porto, Porto, Portugal; [5] Instituto de Investigação e Inovação em Saúde (i3S).

Background: Interstitial lung disease (ILD) affects around 10% of Rheumatoid Arthritis (RA) patients, rising to 60% when considering subclinical lung involve-

Table I. Patient and disease characteristics at baseline

		RA patients N=38	SSc patients N=39	P value
Sociodemographic characteristics	Age (years)	70.38±7.70	65.87±13.54	0.077
	Gender (Female)	23/38 (60.5%)	31/39 (79.5%)	0.07
	Race (White European origin)	38/38 (100%)	37/39 (94.9%)	0.494
	Smoking status (Never smoked)	20/37 (54.1%)	26/39 (66.7%)	0.261
ILD-disease characteristics	Age at diagnosis (years)	65.13±9.23	60.55±14.52	0.111
	Previous CTD diagnosis (yes)	34/40 (85.0%)	30/39 (76.9%)	0.360
PFT pattern	Normal	8/38 (21.1%)	3/39 (7.7%)	-
	Restrictive	12/38 (31.6%)	11/39 (28.2%)	-
	Obstructive	7/38 (18.4%)	5/39 (12.8%)	-
	Isolated decreased DLCO	11/38 (30.0%)	20/39 (51.3%)	-
ILD classification	Non-specific interstitial pneumonia	9/38(23.7%)	22/39 (56.4%)	-
	Cellular	3/9(33.3%)	6/22 (27.3%)	-
	Fibrotic	6/9 (66.7%)	16/22 (72.7%)	-
	Usual interstitial pneumonia	22/38 (45.0%)	14/39 (35.9%)	0.053
	Lymphocytic interstitial pneumonia	0/38 (0.0%)	1/39 (2.6%)	-
	Organizing pneumonia	2/38 (5.3%)	1/39 (2.6%)	-
	Organizing pneumonia/non-specific interstitial pneumonia	3/38 (7.9%)	0/39 (0.0%)	-
	Fibroelastosis	1/38 (2.6%)	0/39 (0.0%)	-
	Interstitial lung abnormalities	1/38 (2.6%)	1/39 (2.6%)	-
Baseline PFT	FVC < 70%	6/31 (19.4%)	6/36 (16.7%)	0.775
	DLCO < 70%	15/22 (68.2%)	23/31 (74.2%)	0.632
Baseline changes in HRCT	Reticulations (yes)	33/38(86.8%)	35/39 (89.7%)	0.692
	Ground Glass (yes)	22/38 (57.9%)	22/39 (56.4%)	0.895
	Traction bronchiectasis (yes)	18/38 (47.4%)	22/39 (56.4%)	0.427
	Honeycomb (yes)	10/38(26.3%)	7/39 (17.9%)	0.376
	Fibrosis (yes)	23/38(60.5%)	25/39 (64.1%)	0.746
	Lung extension > 20% (yes)	9/38 (23.7%)	12/39 (30.8%)	0.485
Mortality	Deaths (yes)	6/40 (8.6%)	1/39 (2.56%)	0.042
	Follow-up (months), median (range)	62.18 (76.40)	49.7 (90.07)	0.427

Legend: DLCO: diffusion lung carbon oxide; FVC: functional vital capacity; HRCT: high resolution computerized tomography; ILD: Interstitial lung disease; PFT: Pulmonary functional tests. Continuous variables are presented as mean ± standard deviation and discrete variables number/total population available (percentage).

ment. Older age at diagnosis, male gender, smoking history and seropositive disease are well known risk factors for ILD. Usual interstitial pneumonia (UIP) pattern is the most common and associated with a worse prognosis. Delays in the diagnosis may compromise response to therapy, with recent studies showing similar prognosis compared with idiopathic pulmonary fibrosis (IPF). Nevertheless, there are no guidelines or validated standardized ILD-screening in RA as for Systemic Sclerosis, where a high-resolution computer tomography (HRCT) and pulmonary functional tests (PFT) are recommended on baseline, and PFT repeated every year. This study aims to compare mortality rates of RA-ILD with SSc-ILD.

Methods: Retrospective and longitudinal study including patients diagnosed with RA-ILD and SSc-ILD followed between January 2019 until February 2022, in a tertiary pneumology centre. Overlap syndromes were excluded. Sociodemographic data, comorbidities, baseline PFTs and radiologic pattern on HRCT and mortality rates were recorded. Categoric variables were compared using Chi-Square analysis and continuous variables through T-test for independent samples or Mann-Whitney test depending on whether data was normally distributed or not. Kaplan-Meier analysis was performed to compare mortality rates of RA-ILD and SSc-ILD. Primary endpoint was death (yes/no). SPSS v25 was used for statistical analysis and significance level was defined as 2-sided $p< 0.05$.

Results: In total, 77 patients were included, 70.15% were female, with a mean age at diagnosis of 62.75±12.41. Of those, 39 patients had SSc and 38 were diagnosed with RA. There were no significant differences between groups regarding sex, age at diagnosis, smoking history, comorbidities, baseline FVC or DLCO and UIP pattern (Table 1). After a median follow-up time of 59.7 months, 7 patients have died (9.1%), 6 with RA and 1 with SSc. RA-ILD showed higher mortality rates than SSc-ILD (log rank = 4.125, p=0.042), especially 5 years after diagnosis.

Conclusions: Screening for CTD-ILD in SSc is a well-known routine in most rheumatologists practice, due to the existence of a well-defined screening protocol on international guidelines. This allows an earlier diagnosis and proper follow up, in order to define which patients, benefit the most with treatment (immunosuppressive/antifibrotic drugs). However, for RA ILD-screening is not well defined and depends mostly on symptoms and changes on physical examination. Yet, a great proportion of patients became symptomatic only when presenting a more extensive lung involvement, which leads to a delayed diagnosis and treatment. This study shows that RA-ILD has a higher mortality rate than SSc-ILD, even when there were no significant differences in sociodemographic and baseline characteristics. A larger sample is needed to allow a more complex survival analysis.

This study aims to alert pneumologists and rheumatologists the need to define a standardized screening protocol, that would allow an earlier diagnosis and treatment in RA-ILD, which might reduce mortality in those patients.

241 - OUTCOME OF UVEITIS IN JUVENILE IDIOPATHIC ARTHRITIS AND SPONDYLOARTHRITIS PATIENTS – A 5-YEAR FOLLOW UP STUDY

Filipe Oliveira Pinheiro[1, 2], Mariana Leuzinger-Dias[3], Bruno Miguel Fernandes[1], Diogo Guimarães da Fonseca[4], Joana Vilaça[5], Luís Figueira[3, 6], Iva Brito[2, 7]

[1]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal, [3]Ophthalmology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [4]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [5]Paediatrics Department, Hospital de Braga, Braga, Portugal, [6]Pharmacology and Therapeutics Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [7]Paediatric and Young Adult Rheumatology Unit, Centro Hospitalar Universitário de São João, Porto, Portugal

Background: Uveitis is a frequent complication of juvenile idiopathic arthritis (JIA) and juvenile spondyloarthritis (jSpA), and diagnosis is often challenging. The importance of uveitis relates to potential complications, and prognosis depends on early recognition and treatment. The aim of this study is to evaluate the prevalence and risk factors for complications associated with uveitis in patients with JIA and jSpA.

Methods: A longitudinal, monocentric cohort study that included patients diagnosed with JIA and jSpA, who developed uveitis. Demographic, laboratory, and clinical data were collected including complications of uveitis, HLA-B27, antinuclear antibodies, erythrocyte sedimentation rate, C-reactive protein, visual acuity and DMARD treatment. Follow-up was 5 years after uveitis' diagnosis. Comparison between groups (complicated versus uncomplicated uveitis) was evaluated using chi-square, t test and Mann-Whitney test. Logistic regression was performed to determine predictors of complications.

Results: A total of 270 patients were evaluated, of which 37 patients (13.7%) had uveitis and were included in this study. Twenty patients were female (54.1%), age 11.9±8.7 years at diagnosis of jSpA/JIA and 15.3±9.9 years at diagnosis of uveitis. Twenty-seven patients

(73.0%) had a diagnosis of JIA (23 with oligoarticular disease) and in 12 patients (32.4%) uveitis was the first manifestation of the rheumatic disease. Fifteen (40.5%) patients exhibited complications during follow-up period, namely cataract (n=11), synechiae and ocular hypertension (n=7), keratopathy (n=6), vitritis (n=3), retinal detachment (n=2), hemovitreous and glaucoma (n=1). Eleven patients (29.7%) underwent ophthalmologic surgery.

Comparisons between complicated versus uncomplicated uveitis patients showed that complications were significantly more frequent in those who had uveitis as the initial presentation (53.3% vs 18.2%, p=0.04), diagnosis of JIA (93.3% vs 59.1%, p=0.03), a younger age at diagnosis of uveitis (8.9 ± 5.7 vs 17.7 ± 10.1, p=0.01) and age at the diagnosis of rheumatic disease (5.4 ± 4.6 vs 14.3 ± 8.7, p=0.01); no significant differences were found between the groups in the other variables studied.

Univariate logistic regression analysis showed that JIA (OR 9.69, CI 1.07-87.44, p=0.04), presentation as uveitis (OR 5.14, CI 1.17-22.69, p=0.03), age at diagnosis of jSpA/JIA (OR 0.88, CI 0.79-0.98, p= 0.02) and age of uveitis (OR 0.90, CI 0.82-0.99, p=0.02) were predictors of complications. When adjusting for age of uveitis, age at diagnosis and JIA, uveitis as first manifestation of rheumatic disease was found to be an independent predictor of complications (OR 101.77, CI 3.08-3358.7, p=0.01).

Conclusion: Ophthalmologic complications of uveitis occur in a significant percentage of patients with JIA and jSpA. The initial presentation of rheumatic disease as uveitis is significantly associated with the occurrence of uveitis complications, so it is essential that there is a collaboration between ophthalmologist and rheumatologist in the diagnosis and treatment of these patients.

242 - ESTIMATION OF THE 10-YEAR RISK OF FATAL CARDIOVASCULAR DISEASE IN A PRIMARY SJÖGREN'S SYNDROME POPULATION

Sara Paiva Dinis [1], Ana Sofia Pinto[1], Filipe Cunha Santos[1], Nathalie Madeira[1], Cláudia Vaz[1, 2], 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

Background: Higher prevalence of subclinical atherosclerosis has been observed in primary Sjögren's syndrome (pSS) patients and recent studies demonstrated an increased risk ofcardiovascular (CV) events in these patients in comparison to general population. EULAR guidelines recommended thorough assessment of traditional CV risk factors using of CV prediction tools for the general population. In Portugal Systematic Coronary Risk Evaluation (SCORE) is recommended by Directorate General of Health. The objective of this study was to estimate the cardiovascular risk in a cohort of patients with pSS.

Methods: A retrospective analysis of pSS patients was done. CV risk factors were collected, and the 10-year CV risk was assessed by SCORE for low-risk countries. Very high CV risk was considered when the SCORE was ≥ 10%, the patient had an established CV disease or diabetes (DM) with target organ damage, high risk was considered when the SCORE was ≥ 5% and < 10% or the patient had DM and additional CV risk factors, moderate risk was considered when the SCORE was ≥ 1% and < 5% or the patient had well controlled DM and low risk was considered when the SCORE was < 1%.

Results: A consecutive cohort of 23 pSS patients with and without past CV events was included, 8.7% male (2), with a mean age of 59.6 ± 13.5 years. Globally, the prevalence of type 2 DM in the population was 13% and 26.1% of patients were hypertensive. Overall, the mean body mass index (BMI) was 27.1± 6.1 Kg/m2 and 17.4% (4) of the patients were smokers in their lifetime. Anti-hypertensive medication was reported in 21.7% of the patients, cholesterol medication in 47.8% and antidiabetic medications in 13%. The mean of systolic blood pressure was 129± 16.7 mmHg, diastolic blood pressure of 68.9 ± 9 mmHg, total cholesterol of 195.7± 42.5 mg/ dL and high-density lipoprotein cholesterol of 57.8 ± 12.9 mg/dL. Four (17.4%) patients had acardiovascular event. According to SCORE, 13% (3) of population was classified at very highrisk, 8.7% (2) at high risk, 56.5% (13) at moderate risk and 21.7% (5) at low risk of having a fatal

CV event in the following 10 years.

Conclusions: Our findings showed that most of the patients included were at moderate risk of suffering a CV event in the following 10 years and that circa one fifth had high or very high CV risk. Traditional CV risk factors, namely, hypertension and dyslipidemia are present and should be routinely access in Rheumatology appointments.

244 - BARREIRAS E ALTERNATIVAS NA REFERENCIAÇÃO À REUMATOLOGIA PEDIÁTRICA

Hugo Parente[1], Maria Pontes Ferreira[1], Catarina Soares[1], Emanuel Costa[2], Francisca Guimarães[1], Soraia Azevedo[1], Daniela Santos-Faria[1], José Tavares-Costa[1], Filipa Teixeira[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: A Reumatologia Pediátrica (RP) é a área médica especializada na patologia reumatismal em idade infantil. Nos EUA[1], a referenciação à RP revelou atrasos relacionados com tempo de espera, distância, falta de formação, com implicações óbvias no outcome destas patologias. Na Europa, há dados igualmente preocupantes[2,3,4]. Em Portugal, esta realidade é desconhecida.
Objetivos: Identificar as barreiras à referenciação para a RP, quer pela Pediatria quer pela Medicina Geral e Familiar (MGF) e caracterizar os padrões de referenciação atuais, assim como determinar medidas consensuais de melhoria.
Métodos: Questionário online, enviado por email a uma amostra representativa dos Pediatras e Médicos de Família a nível nacional, com posterior análise descritiva e comparativa. A significância estatística foi definida com um valor p<0.05.
Resultados: Obteve-se um total de 292 respostas, 24,7% da Pediatria e 75,3% de MGF. No grupo da Pediatria, 39% dos respondedores desempenham funções num hospital central e 61,1% num distrital. Apenas 11,3% afirmaram ter tido formação específica em RP. O número de formações nesta área nos últimos 5 anos foi inferior na MGF, sendo que não assistir a nenhuma formação demonstrou estar associado a uma menor referenciação. A maioria dos inquiridos referiu existir um centro de RP a menos de 60 minutos de distância. Ser Pediatra e ser especialista associou-se a uma maior taxa de referenciação.

Tabela I.

	Total (n=292)	MGF (n=220)	Pediatria (n=72)	p-value
Idade (anos), Mdn (AIQ)	32,5 (10)	32 (9)	36 (13)	NS
Sexo, feminino % (n/N)	82,5% (241/292)	81,4% (179/220)	86,1% (62/72)	NS
Categoria profissional, especialista % (n/N)	55,8% (163/292)	51,8% (114/220)	68,1% (49/72)	0,016
Número de anos de trabalho % (n/N)				
0-2	18,5% (54/292)	21,8% (48/220)	8,3% (6/72)	<0,001
3-5	34,9% (102/292)	37,3% (82/220)	27,8% (20/72)	NS
6-10	16,8% (49/292)	17,7% (39/220)	13,9% (10/72)	NS
10-20	19,5% (57/292)	14,5% (32/220)	34,7% (25/72)	<0,001
>20	10,3% (30/292)	8,6% (19/220)	15,3% (11/72)	NS
Distrito % (n/N)				
Aveiro	5,8% (17/292)	3,6% (8/220)	12,5% (9/72)	NS
Braga	16,4% (48/292)	16,8% (37/220)	15,3% (11/72)	NS
Bragança	1,0% (3/292)	0,5% (1/220)	2,8% (2/72)	NS
Castelo Branco	1,0% (3/292)	0,9% (2/220)	1,4% (1/72)	NS
Coimbra	4,4% (13/292)	5,0% (11/220)	2,8% (2/72)	NS
Faro	3,1% (9/292)	2,7% (6/220)	4,2% (3/72)	NS
Guarda	1,0% (3/292)	1,4% (3/220)	0% (0/72)	NS
Leiria	2,7% (8/292)	3,2% (7/220)	1,4% (18/72)	NS
Lisboa	7,4% (22/292)	7,4% (16/220)	8,3% (6/72)	NS
Porto	35,9% (105/292)	35,5% (78/220)	37,5% (27/72)	NS
Madeira	0,3% (1/292)	0% (0/220)	1,4% (1/72)	NS
Santarém	3,4% (10/292)	3,6% (8/220)	2,8% (2/72)	NS
Setúbal	0,3% (1/292)	0,5% (1/220)	0% (0/72)	NS
Viana do Castelo	10,9% (32/292)	11,5% (25/220)	9,7% (7/72)	NS
Vila Real	2,1% (6/292)	2,7% (6/220)	0% (0/72)	NS
Viseu	3,8% (11/292)	5,0% (11/220)	0% (0/72)	NS
Hospital, central % (n/N)	9,6% (28/292)	-	38,9% (28/72)	-

Mdn: Mediana; NS: não significativo; AIQ: amplitude interquartil

Nove por centro dos respondedores, afirmou não referenciar apesar de o considerar necessário, existindo uma associação significativa com ser MGF. Nestas situações, a referenciação foi para outra especialidade/subespecialidade, nomeadamente Pediatria Geral, Reumatologia Geral (RG) e Ortopedia Pediátrica. Vinte e quatro porcento consideraram o apoio da RP suficiente, 21,2% insuficiente e 54,8% desconheciam esta subespecialidade.
Como sugestões de melhoria, valorizaram a necessidade de aumentar o número de reumatologistas pediátricos, alargar a sua distribuição nacional, constituir algoritmos de fast track em zonas carenciadas, e aumentar a formação nesta área.
Porto, Santa Maria da Feira, Cascais, Santarém, Viana do Castelo e Vila Franca de Xira foram os concelhos com maior taxa de referenciação. Em Braga, Faro e Setúbal, identificou-se haver necessidade de aumentar a cobertura regional de RP, enquanto no Porto e em Guimarães considerou-se que a cobertura atual é suficiente.
Conclusões: A falta de formação, a maior distância a centros de RP e a desigual cobertura nacional são as principais barreiras na referenciação à RP, em Portugal. A especialidade de Pediatria parece ter maior formação nesta área, com maior contacto com diversas patologias deste foro assim como um maior historial de referenciação a RP. A Pediatria e a RG parecem constituir as principais alternativas à RP. A medida considerada mais importante para melhorar esta lacuna foi o aumento da formação.

REFERENCES

1. Correll, C.K., Spector, L.G., Zhang, L. et al. Barriers and alternatives to pediatric rheumatology referrals: survey of general pediatricians in the United States. Pediatr Rheumatol 13, 32 (2015). https://doi.org/10.1186/s12969-015-0028-6;
2. Tzaribachev et al. Arthritis Care Res 2009;
3. Freychet et al. Joint bone spine 2019; [4] Mcerlane et al. Ped Rheumatol 2014.

245 - CARDIAC TOXICITY TO HYDROXYCHLOROQUINE IN INFLAMMATORY RHEUMATIC DISEASES

Filipe Oliveira Pinheiro[1, 2], Miguel Carvalho[2, 3], Pedro Madureira[1], Maria Seabra Rato[1, 2], Filipe Macedo[3], Lúcia Costa[1]

[1]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal, [3]Cardiology Department, Centro Hospitalar Universitário de São João, Porto, Portugal

Background: Hydroxychloroquine (HCQ) is used in the treatment of inflammatory rheumatic diseases and is considered a safe drug. The role of HCQ in the

COVID-19 pandemic highlighted some deleterious effects of HCQ, especially cardiac. Cardiac adverse effects are potentially fatal but, although previously described in rheumatic patients, are very rare in this population. The aim of our study is to evaluate the prevalence and development of cardiac adverse events in HCQ-treated patients with inflammatory rheumatic diseases.

Methods: We performed a cross-sectional study where patients aged ≥18 years with a diagnosis of inflammatory rheumatic disease currently exposed or not to hydroxychloroquine underwent analytical evaluation, electrocardiogram (ECG) and echocardiogram. Clinical data were collected, namely HCQ exposure time, cumulative dose, and weight-adjusted dose. Comparisons between groups were evaluated using chi-square, t test and Mann-Whitney U test. Logistic regression was performed to determine predictors of changes in ECG and echocardiography.

Results: A total of 80 patients were included, 75 (93.8%) female, aged 52±13 years. Fifty patients (62.5%) had systemic lupus erythematosus (SLE), 20 (25.0%) Sjögren's syndrome, 8 (10.0%) undifferentiated connective tissue disease, and 2 (2.5%) SLE/systemic sclerosis overlap. Forty-one (51.2%) patients were currently under HCQ, with a median cumulative dose 584 (438-1606) g and a median time of exposure to HCQ of 4 (3-11) years. Thirty-two patients (40.0%) had changes in ECG and 24 (30.0%) changes in echocardiogram. Patients under HCQ had a shorter duration of disease (7±6 versus 16±10.3 years, p<0.001) and a higher PR interval (157±16 versus 149±22 ms, p=0.046). There were no differences regarding weight-adjusted doses of <5 or ≥5mg/kg. ECG changes were seen in higher proportion in patients with hypertension (40.6% versus 12.5%, p=0.004) and with higher median potassium levels – 4.5 (4.1-4.8) versus 4.2 (4.0-4.4), p=0.023. Echocardiography changes were seen in older patients (59±11 versus 50±13 years, p=0.003) and in patients with higher cumulative dose – 1752 (785-2190) versus 438 (328-1022) g, p=0.008 – and time of exposure to HCQ – 12 (6-15) versus 4 (2-9) years, p=0.028.

In univariate logistic regression analysis, hypertension (OR 4.789, CI95% 1.580-14.515, p=0.006) and potassium levels (OR 4.461, CI95% 1.225-16.241, p=0.023) were shown to be predictors of ECG changes; age (OR 1.065, CI95% 1.018-1.113, p=0.006), HCQ cumulative dose (OR 1.001, CI95% 1.000-1.002, p=0.033), and exposure time (OR 1.136, CI95% 1.000-1.289, p=0.049) were predictors of echocardiography changes. In multivariate logistic regression analysis, when adjusted for age, neither HCQ cumulative dose nor exposure time were predictors of echocardiography changes.

Conclusion: In this study, no association was found between changes in ECG and echocardiogram in patients under HCQ, which remains to be a safe drug in patients with inflammatory rheumatic diseases.

246 - WHAT ARE THE DIAGNOSES ASSOCIATED WITH UVEITIS?

Carolina Mazeda[1, 2], Gisela Eugénio[1, 2], Barcelos A.[1, 2]
[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz, Aveiro, Portugal

Introduction: Uveitis is a group of ocular disorders characterized by intraocular inflammation which can cause significant visual impairment. There are multiple differential diagnoses for this pathology, changes with time, and it is highly variable, being influenced by numerous factors, including genetic, ethnic, geographic, and environmental factors, diagnostic criteria, and referral patterns. In many instances, uveitis is not a primary ocular disease but reflects ocular involvement related to a systemic disorder. The main objective of our study was to describe the frequency of uveitis seen in emergency department (ED) and to describe their clinical characteristics.

Methods: This retrospective study included all patients with diagnosis of uveitis observed at ED in a secondary hospital, between January 2018 and December 2021. Uveitis diagnosis was identified through the 9th and 10th International Classification of Diseases. Data collected for each patient included sociodemographic and clinical data, type of uveitis, number of recurrences of uveitis per patient and type of treatment. A descriptive analysis was performed using SPSS® version 25.

Results: A total of 77 patients with diagnosis of uveitis were included. Forty-one were females and 36 men, with a median age at first referral of 59.9 years (interquartile range 13–87 years) and had a median number of consultations per patient of 3.3. Seventy-two (93.5%) patients were diagnosed with anterior uveitis, three (3.9%) patients had posterior uveitis and one patient had intermediate uveitis and panuveitis. The uveitis was unilateral in the majority of patients, with only one case documented as bilateral; recurrence occurred in 18.2% of these patients. HLA-B27 was positive in 11.8% of patients. Regarding its etiology, 18.2% of the patients with uveitis were associated with systemic autoimmune conditions and 14.3% patients were classified as infectious. In patients with systemic autoimmune conditions, 5.2% cases were diagnosed with sarcoidosis (2 anterior, 1 intermediate and 1 panuveitis), 1.3% with ulcerative colitis (anterior) and the remain (11.7%) with spondyloarthritis (all anterior uveitis). In two cases, the uveitis was the first manifestation. In the case of infectious etiology, the most frequent was herpes simplex (10.4%), syphilis (2.6%) and toxoplasmosis (1.3%). As

for the treatment regimen, most patients started topical ocular corticosteroids (81.8%), antivirals (10.4%), azathioprine (2.6%) and antibiotic therapy (3.9%).
Conclusion: Unilateral, non-idiopathic, non-infectious anterior uveitis was the most frequent presentation. Despite the evolution of diagnostic investigations, etiology remained unknown in many cases of uveitis. It is important for physicians to be familiar with the different etiologies for early diagnosis and therapy.

247 - THE EFFECT OF BMI IN PSORIATIC ARTHRITIS

Vanessa Fraga[1], Catarina Abreu[1], Sandra Sousa[1], Maria José Santos[1, 2]
[1]Reumatology Department, Hospital Garcia de Orta, EPE, 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: Psoriatic arthritis (PsA) is a chronic inflammatory disease. Obesity is among the most prevalent comorbidities in PsA.1This may be particularly important as adipose tissue plays a role not only in metabolism but also in immune and inflammatory processes.1 On the other hand, obesity may contribute to mechanical stress and inflammatory response on the entheseal structures.2
Objectives: The aim of this study was to characterize the body mass index (BMI) in PsA patients and to investigate if increased BMI is associated with PsA clinical features, particularly enthesitis.
Methods: Patients who met the CASPAR criteria for PsA and with information on BMI available were included. Demographic and clinical features were retrieved from the Portuguese registry Reuma.pt and completed with information from clinical records.
BMI was classified according to the WHO in normal weight, overweight and obesity. Clinical manifestations were compared between normal weight and overweight/obese patients, using Fisher's exact test.
Results: A total of 80 patients were included, 96.3% Caucasian, 41 (51.2%) males. The mean age of patients was 58.51 ± 13.39 years. The mean BMI was 29.13 ± 5.79 kg/m2, sixty-four patients (80%) had increased BMI, being 37 overweight (46.3%) and 27 obese (33.8%).Three patients had obesity class 3 (BMI > 40 kg/m2), 7 had obesity class 2 (BMI 35 – 39.9kg/m2) and 17 had obesity class 1 (BMI 30-34.9 kg/m2). Most of obese patients were female (66.7% vs 33.3%, p-value 0.02). Patients with elevated BMI had other comorbidities more frequently than patients with normal BMI, such as hypertension (66.7% vs 18.8%; p=0.001), diabetes mellitus (27% vs 0%; p=0.013), and dyslipidemia (46% vs 18.8%; p=0.041). Regarding PsA clinical features, there were no significant differences in the presence of enthesitis, dactylitis, or uveitis (table I).

Table I. Clinical manifestation in PsA patients with normal BMI and elevated BMI.

	BMI[3] 25 kg/m² N=64	BMI < 25 kg/m² N=16	p-value
Enthesitis	31.1% (19)	31.3% (5)	0.607
Dactylitis	30.6% (19)	25% (4)	0.457
Psoriasis	100% (62)	93.8% (15)	0.205
Uveitis	1,6% (1)	0	0.792
Axial involvement	9.4% (6)	18.8% (3)	0.254
Nail involvement	42.3% (22)	57.1 % (8)	0.239

Conclusion: Overweight obesity and are present in the vast majority of PsA patients and are associated with other comorbidities. We did not fount differences in the prevalence of extra-musculoskeletal manifestations associated with overweight/obesity, although enthesitis may be more difficult to access clinically in patients with increased BMI. Besides, the small sample size prevents a definitive conclusion. Therefore, in the future we intend to address this question in a larger cohort.

REFERENCES:

1. Kumthekar, A., Ogdie, A. Obesity and Psoriatic Arthritis: A Narrative Review. Rheumatol Ther 7, 447–456 (2020). https://doi.org/10.1007/s40744-020-00215-6; 2. Schett G, Lories RJ, D'Agostino MA, Elewaut D, Kirkham B, Soriano ER, McGonagle D. Enthesitis: from pathophysiology to treatment. Nat Rev Rheumatol. 2017 Nov 21;13(12):731-741. doi: 10.1038/nrrheum.2017.188. PMID: 29158573.)

Casos Clínicos

014 - A RARE DISEASE WITH AN EVEN RARER CAUSE

Beatriz Samões[1], Bruna Silva[2], Ana Martins[2], Daniela Santos Oliveira[2, 3], Frederico Rajão Martins[4], Diogo Guimarães da Fonseca[1], Lúcia Costa[2], Miguel Bernardes[2, 5]

[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [2]Rheumatology Department, Centro Hospitalar 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]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal

Introduction: Osteomalacia is an uncommon disease characterized by an impaired mineralization with accumulation of non-mineralized bone matrix that may have several etiologies.

Case report: A 70-year-old male was admitted to our Rheumatology department for severe lower limb pain associated with phospho-calcium metabolism abnormalities. He had Rendue-Osler-Weber disease requiring biweekly infusions of ferric carboxymaltose, osteoporosis treated with annual zoledronic acid 5 mg and calcium and vitamin D supplementation for about 1 year, dyslipidaemia treated with pitavastatin 2 mg and allergic rhinosinusitis. He reported an onset of disabling pain and swelling of the left ankle and midfoot with additive involvement of the contralateral about 18 months ago, associated with temperature variations, allodynia and paresthesia. He was hemodynamically stable, with discoloured mucous membranes, telangiectasias in the face and fingers, with limited passive mobilization of the tibiotarsal and subtalar joints bilaterally and with pain on passive mobilization of the tarsus, without arthritis but highly dependent on crutches for walking. He had already performed several radiological exams: feet radiography and CT scan that showed bilateral severe diffuse bone demineralization; feet MRI that displayed bone marrow edema of the navicular and cuneiform, subchondral fracture of the cuneiform-navicular joints, synovitis of the intertarsal and tarsometatarsal joints and periarticular soft tissue edema; bone scintigraphy that revealed hyperuptake of the tibiotarsal, tarsal and metatarsophalangeal joints in the vascular and bone phases. He had been diagnosed with complex regional pain syndrome about a year ago and was started on therapy with tapentadol 200 mg/day, amitriptyline 25 mg/day and pregabalin 400 mg/day with little improvement. Additional investigation showed persistent hypophosphatemia (1.6 mg/dL), increased PTH (110 pg/mL) and bone alkaline phosphatase (40.1 mcg/L), in addition to increased fractional excretion of phosphate (43%, UP04 of 118.3 mg/dL) and hypocalciuria (49 mg/24h), with normal serum calcium and 25(OH) vitamin D3 (61.4 ng/mL). Serum FGF-23 levels were raised (324 UA/mL [< 180]). The radiographic study revealed dorsolumbar vertebral flattening and changes in bone trabeculation, without Looser's zones. Somatostatin receptor PET imaging showed several areas of anomalous Ga68-DOTA-TOC uptake throughout the skeleton suggestive of stress fractures, with no evident lesions to support a diagnosis of oncogenic osteomalacia. Bone biopsy with histomorphometry showed osteoid accumulation and confirmed the presence of osteomalacia. After excluding other causes, we assumed that it was related to the frequent ferric carboxymaltose infusions. The later were replaced by saccharated ferric oxide infusions. Treatment was also started with progressive dose titration of calcitriol up to 1.5mg/day and oral disodium phosphate up to 5740mg/day. After 6 months, there was a clear clinical and analytical improvement, with normalization of phosphatemia (3.7 mg/dL), serum FGF-23 (43 UA/mL), PTH (60.4 pg/mL) and alkaline phosphatase (90 U/L).

Conclusions: Intravenous iron supplementation with ferric carboxymaltose is a rare cause of osteomalacia with only 18 cases reported. Repeated infusion can lead to increased FGF-23 and subsequent persistent hypophosphatemia. Apart from treatment with phosphate and active forms of vitamin D, discontinuing or switching iron preparation seems to be the most effective intervention.

036 - A SEVERE CASE OF IMMUNE RECONSTITUTION INFLAMMATORY SYNDROME IN AN HIV-INFECTED PATIENT WITH PSORIATIC ARTHRITIS

Margarida Lucas Rocha[1], Laura Gago[2], Maria Helena Lourenço[2, 3], Rita Pinheiro Torres [2, 3, 4], Manuela Costa[2], Jaime C. Branco[2, 3, 4], Alexandre Sepriano[2, 3]

[1]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, [2]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [3]CEDOC, NOVA Medical School, Lisboa, Portugal, [4]NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

Introduction: The long-term prognosis of the infection by the human insufficiency virus (HIV) has dramatically changed with the use of combination anti-retroviral therapy (cART). The immune reconstitution inflammatory syndrome (IRIS) describes the phenomenon in which a clinically silent immune-mediated disease appears or is exacerbated by the initiation of cART. We report a case of a severe flare of psoriatic arthritis in the context of IRIS.
Clinical Case: A 53-year-old man with a 10-years history of psoriatic arthritis as well as infection by HIV, C and B hepatitis for the same amount of time, was seen in our Rheumatology department. The patient had stopped all medication and was lost to medical follow up for more than 7 years. The patient however did not report any symptoms pertaining to PsA up until he had resumed cART therapy (efavirenz, emtricitabine and tenofovir) only a few days before being admitted to our department. The patient referred acute onset of extensive psoriatic lesions as well as sudden onset of polyarthralgia and joint swelling, prolonged morning stiffness, severe disability and weight loss. Joint symptoms started at the left and right elbows, then shoulders, small joints of the hands and feet, knees and ankles. On the physical examination he had arthritis of the elbows and shoulders and dactylitis in all fingers of the right hand, first and second finger of the left hand (image 1A and 1B), all toes of the right foot and all, except the fourth, toes of the right foot (image 1E). He also had extensive psoriatic lesions at the hands, feet, dorsal region and abdomen, elbow, scalp and nails. Inflammatory parameters were elevated (C-reactive protein: 13.3mg/dL and erythrocyte sedimentation rate: 81 mm/h) (DAS28-VS: 6,64). After confirming that the HIV and C and B hepatitis viral loads were negative, the patient was treated with 500mg of methylprednisolone intravenous, followed by 25mg/day of oral prednisolone, subcutaneous methotrexate (up to 25mg/week) and oral leflunomide 20mg/day. Viral loads remained negative, there were no side effects and the patient was in remission (DAS28-VS: 2,58) with no arthritis and no skin lesions 8 months after starting the treatment for PsA (Figure 1C, 1D and 1F).
Discussion: The incidence of IRIS has been increasing in parallel with the widespread use of cART around the world. On average, symptoms occur within 9 months after the start of cART, but can also occur within days as in our case. The use of immunosuppressive drugs to treat inflammatory rheumatic diseases in patients infected by HIV can be challenging. This case shows that immunosupressive treatment can however be safe, well tolerated and highly effective under the proviso of complete suppression of viral activity.

Figure 1. Physical examination

042 - THE SKIN AS THE FIRST SIGN OF A RARE COMPLICATION OF RHEUMATOID ARTHRITIS

Maria Seabra Rato[1], Alexandra Bernardo[1], Filipe Oliveira Pinheiro[1], Diogo Guimarães da Fonseca[2], Ana Martins[1], Daniela Santos Oliveira[1], Lúcia Costa[1]
[1]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Introduction: Felty syndrome (FS) is defined as the coexistence of rheumatoid arthritis (RA), neutropenia and splenomegaly and it occurs in less than 1% of RA patients. Neutropenia is characteristically associated with recurrent bacterial infections which may lead to increased mortality. Ecthyma gangrenosum (EG) is an uncommon cutaneous infection usually caused by Pseudomonas aeruginosa, an opportunistic bacterium affecting typically immunocompromised patients. The anogenital and axillary areas are the most commonly affected (57%), followed by the extremities (30%), making the trunk (6%) and face (6%) rarer anatomic locations. Multiple skin lesions of EG are usually secondary to haematogenous dissemination.
Presentation: A 45-year-old caucasian female was admitted for non-painful ulcers involving face, trunk and limbs. Lesions started 3 days before as red to violaceous macules evolving rapidly (within 12-24h) into ulcers with a black eschar. The patient also reported fever, rhinorrhea, nasal obstruction and cough present for 5 days. Her past medical history includes 13-year of seropositive and erosive rheumatoid arthritis (RA) treated

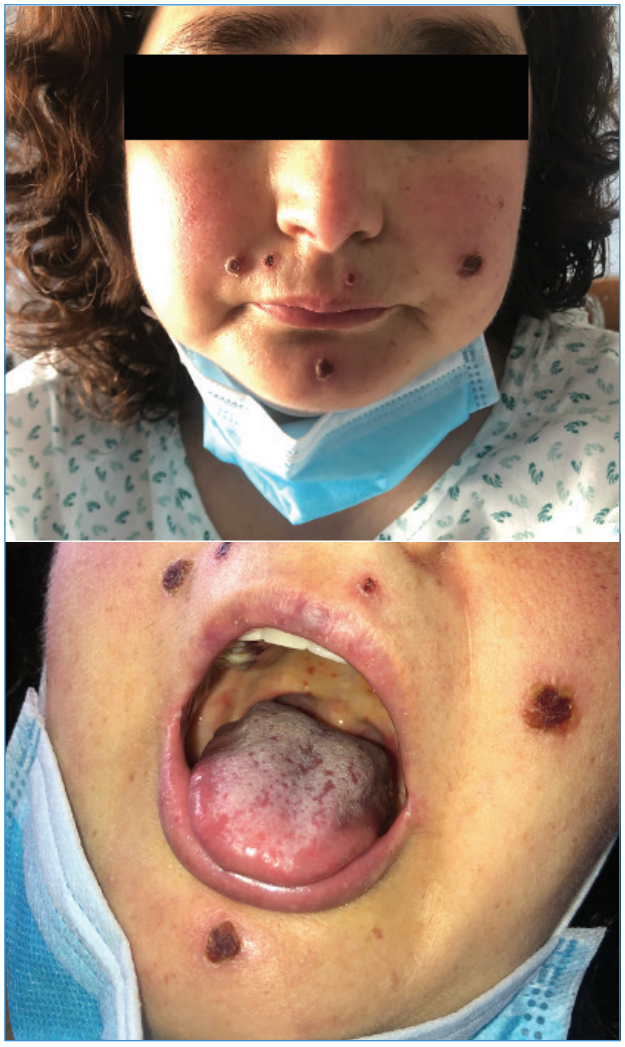


Figure 1. Necrotic ulcers on the face, surrounded by erythema, suggestive of ecthyma gangrenosum.

with a low dose of prednisolone and non-steroidal anti-inflammatory drugs. She had an irregular follow-up at the Rheumatology consultation and had never used any disease-modifying antirheumatic drug. On physical examination several ulcerations were observed, surrounded by erythema and covered by a necro-hemorrhagic crust (Fig. 1). Examination of the nose and paranasal sinuses showed crusting and mucous rhinorrhea. Laboratory results revealed pancytopenia with a white blood cell count of 2200/mm3 (4000–11 000/mm3) with an absolute neutrophil count of 230/mm3 (1500–8000/mm3), hemoglobin of 10.0 g/dL (10.5–13.5 g/dL) and platelet count of 139 000/mm3 (150 000–400 000/mm3). Peripheral blood smear showed no significant abnormalities with normal reticulocytes. C-reactive protein was 230 mg/L (<3 mg/L). Serology for B19 parvovirus, HIV, viral hepatitis and Epstein-Barr virus infection were negative. On abdominal ultrasonography splenomegaly of 17.9 cm was identified. Chest radiography and echocardiography were unremarkable. Bone marrow biopsy and flow cytometry showed no evidence of large granular lymphocytic leukemia. Skin and sputum culture showed the growth of multisensitive Pseudomonas aeruginosa, although blood cultures were negative. Based on these findings, the diagnosis of FS complicated with ecthyma gangrenous due to Pseudomonas aeruginosa was established. Piperacillin/tazobactam was started with complete healing of the lesions within 7 days. Prednisolone was increased to 0.5mg/kg/day with normalization of the absolute neutrophil count. One month later, methotrexate was started (up to 20mg/week) and prednisolone was progressively tapered.

Discussion: Our patient had positive sputum and wound cultures to Pseudomonas Aeruginosa, although blood cultures were sterile. This could be explain by a transient bacteremia with the source of infection being the respiratory tract. At our knowledge, the association between FS and EG was not previously described. The diagnosis of EG should be suspected in patients with classic skin findings and neutropenia, even in the absence of bacteremia.

043 - GASTROINTESTINAL BLEEDING – AN ATYPICAL PRESENTATION OF GRANULOMATOSIS WITH POLYANGIITIS

Ana Catarina Moniz[1], Marta Teixeira de Almeida[2], Cristina Ponte[3, 4], Ana Valido[5]

[1]Serviço de Reumatologia, Centro Hospitalar Lisboa Ocidental | Hospital Egas Moniz, Lisboa, Portugal, [2]Unidade de Cuidados Intensivos, Hospital de Vila Franca de Xira, Vila Franca de Xira, Portugal, [3]Rheumatology and Bone Metabolic Diseases Department, Centro Hospitalar Lisboa Norte, EPE - Hospital de Santa Maria, Lisbon Academic Medical Centre, Lisboa, Portugal, [4]Rheumatology Research Unit, Instituto de Medicina Molecular, Faculdade de Medicina da Universidade de Lisboa, Lisboa, Portugal, [5]Serviço de Reumatologia, Unidade Local de Saúde do Litoral Alentejano, Santiago do Cacém, Portugal

Granulomatosis with polyangiitis (GPA) is an anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis in which there is necrotizing granulomatous inflammation involving the small but also the medium vessels, leading to vascular destruction and potential organ- and life-threatening consequences. It mostly affects the ear, nose, throat (ENT), lower respiratory tract and kidneys, and it is typically associated with proteinase 3 (PR3)-ANCA.

We describe a case of a 60-year-old man presenting at the emergency department complaining of bloody diarrhoea for the past week. Alongside, he referred

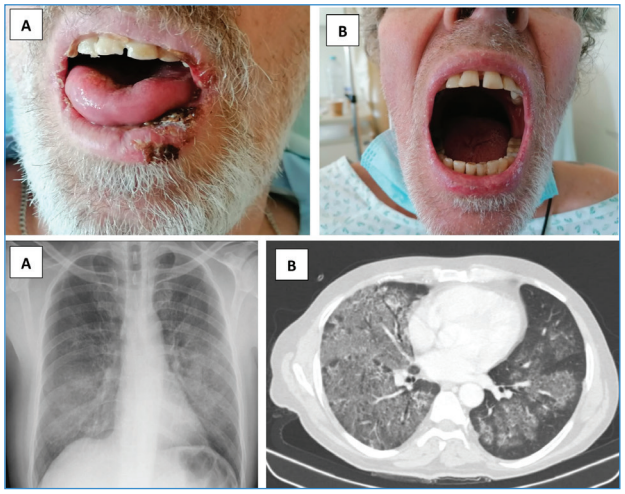


Figure 1. Oral ulcers prior (A) and after glucocorticoids (B). Chest PA radiography (C) and CT (D) show diffuse alveolar haemorrhage.

otalgia and ear fullness, for which he had already started amoxicillin/clavulanate. On physical exam he was sweaty, hypertensive and slightly tachycardic (arterial pressure of 152/100 mmHg and heart rate of 101 bpm). The abdominal exam showed hyperactive bowel sounds. Laboratory studies showed haemoglobin of 12.8 g/dL, leucocytes of 7100 cells/mm3 without eosinophilia, erythrocyte sedimentation rate (ESR) of 83 mm/h and C-reactive protein (CRP) of 12.89 mg/dL. Head computerized tomography (CT) revealed otitis with mastoiditis and the patient was admitted to the ward for intravenous (IV) antibiotic therapy with piperacillin/tazobactam. After five days of hospital admission, bloody diarrhoea progressed to heavy haematochezia, resulting in severe anaemia requiring multiple blood transfusions. Colonoscopy showed ulcers in the right colon and colon biopsies revealed inflammatory cell infiltrates compatible with colitis, fibrosis and a single granuloma. The patient developed progressively worsening acute kidney injury (serum creatinine of 2.3 mg/dL, microscopic haematuria and 24h-protein of 1 g), increasing inflammatory parameters (ESR of 103 mm/h and CRP of 20.14 mg/dL) and oral ulcerations (Figure 1). At day 26 of admission, the patient presented with massive alveolar haemorrhage (Figure 2), for which he was treated with IV methylprednisolone (1 g/day for three days), followed by oral prednisolone (1 mg/kg/day), and IV immunoglobulin (50 g/day for three days). The immunology results confirmed a positive PR3-ANCA (167 UA/mL), a negative myeloperoxidase (MPO)-ANCA and negative antinuclear antibodies (ANA). The diagnosis of GPA was definitively established and the patient was started on IV cyclophosphamide (six pulses of 15 mg/kg). Disease remission was attained with improvement of anaemia, renal function, and respiratory and gastrointestinal (GI) symptoms. The patient was discharged after 34 days of hospitalization and is under remission maintenance therapy with methotrexate 15 mg/week and prednisolone 7.5 mg/day in a taper schedule, without registered relapses after one year of follow-up.

GI involvement is uncommon in GPA, particularly at initial presentation, and it has recently been estimated to occur in 12.38% of patients, with GI bleeding representing approximately 6% of cases. To our knowledge, there are only a few case reports describing GI bleeding at the onset of GPA worldwide. Therefore, our case embodies a unique presentation of a rare disease, in which timely diagnosis contributes largely to preventing catastrophic outcomes. Nonetheless, the patient eventually developed the typical picture of ENT, pulmonary and renal involvement, having responded well to immunosuppressive therapy with glucocorticoids and cyclophosphamide.

045 - ELDERLY-ONSET RHEUMATOID ARTHRITIS: A CASE REPORT

Ana Martins[1, 2], Daniela Santos Oliveira[1, 2, 3], Frederico Rajão Martins[4], 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, [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: Elderly-onset Rheumatoid Arthritis(EORA) is defined as rheumatoid arthritis(RA) with onset at age 60 or over and differs slightly at presentation from younger-onset RA(YORA). The atypical manifestations of EORA, such as fever, myalgia, anemia, and polymyalgia rheumatica-like syndrome increase the difficulties in the diagnosis. EORA patients develop an upgraded systemic inflammatory status, more declined physical and functional assessment and worse prognosis than the elderly patients with YORA.

Case report: A 79-year-old female with arterial hypertension and osteoporosis presented to the emergency department with fever and bilateral shoulder pain with functional limitation in the last two weeks. She described morning stiffness lasting for more than an hour. She had no symptoms suggestive of infection nor trauma history. On physical examination her vital signs were stable. Examination of both shoulders showed a significant limitation in active shoulder movements with a slightly improve in passive movement. No inflammatory signs were found. The initial laboratory

workup showed normochromic normocytic anemia (11.1g/dL), thrombocytosis (453000/µL), high erythrocyte sedimentation rate (ESR, 96mm/h) and C-reactive protein (CRP, 190 mg/L), high serum ferritin (1080ng/mL) and low serum albumin (22.4g/L). Results of microbiological workup were all negative. Bilateral shoulder ultrasound showed bilateral joint effusion with significant synovial thickening on the left shoulder. In magnetic resonance imaging no signs of osteomyelitis or septic arthritis were found.

During the period of hospitalization, she developed polyarthralgia involving the wrists, proximal interphalangeal joints, knees and feet. She has also developed exuberant peripheral edema of the upper and lower limbs. An immunological study was conducted. Rheumatoid factor and anti-cyclic citrullinated peptide were positive (30.3 IU/mL and 401 U/mL, respectively). The remaining immunological study was normal.

The diagnosis of EORA was established and she started on a low dose prednisolone and methotrexate. She was discharged and after 3 months of follow-up, her shoulder movements were practically normal, no arthritis was found on physical examination and laboratory analysis showed a significantly improvement with almost normal ESR and CRP and resolution of anemia, thrombocytosis, hypoalbuminemia and hyperferritinemia.

Conclusion: As our case describes, EORA seems to have a characteristic pattern with more acute onset and systemic involvement with a higher level of nonspecific inflammatory parameters. Treatment of EORA patients is crucial to improve the prognosis. However, the therapeutic approach must be balanced against the risk profile of elderly patients with a careful drug selection, dose adjustment and disease activity monitoring.

046 - PROGRESSIVE PROXIMAL MUSCLE WEAKNESS WITH SUBACUTE ONSET IN AN ELDERLY PATIENT: A CASE REPORT

Ana Martins[1, 2], Sofia Pimenta[1, 2], Goreti Nadais[3], Miguel Pinto[4], Ricardo Taipa[4], 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, [3]Serviço de Neurologia, Centro Hospitalar Universitário de São João, Porto, Portugal, [4]Neuropathology Unit, Department of Neurosciences, Centro Hospitalar Universitário do Porto, Porto, Portugal

Background: Statins are some of the most widely prescribed medications. Although generally well tolerated, statins can lead to musculoskeletal side effects. Statin-induced necrotizing autoimmune myositis (SINAM) is a rare condition and the prevalence is only 1/100.000 people. This disorder is characterized by progressive and severe symmetric muscle weakness, marked elevation of creatine kinase (CK), nonspecific irritable myopathy on electromyography, necrotic fibers and regenerating fibers without significant inflammatory cells on muscle biopsy, positive anti-3-hydroxy-3-methylglutaryl-coenzyme A reductase (anti-HMGCR) antibodies and persistent symptoms despite statin discontinuation.

Case report: A 70-year-old female with a past medical history of hypertension, diabetes and hyperlipidemia (treated with simvastatin 20 mg/day for the last 7 years) presented with several months of progressive proximal muscle weakness, myalgia, anorexia and weight loss. She denied fever, chills, rashes, oral ulcers, shortness of breath, chest pain, dysphagia or visual disturbances. There was no family history of neuromuscular disease.

Physical examination revealed significant proximal upper and lower extremity muscle weakness (shoulder abduction 3/5, elbow flexion 4/5, hip flexion 3/5, knee flexion 4/5 bilaterally) and inability to rise from a chair. Deep tendon reflexes, sensation and coordination were intact. Heart and lung auscultation and abdominal examination didn't reveal abnormalities. No cutaneous manifestations were found.

Laboratory workup showed an elevated CK (2954 U/L), aldolase (45 U/L), aspartate transaminase (124 U/L), and alanine transaminase (95 U/L). Serologic tests for herpes simplex, Epstein-Barr, cytomegalovirus, varicella-zoster, human immunodeficiency virus and hepatitis C were negative. Serology test for hepatitis B revealed a past infection (Negative HBV-DNA). Thyroid function was normal. Electromyography showed abnormal spontaneous muscle activity in proximal muscles of upper and lower extremities suggestive of an irritable myopathy. Right deltoid muscle biopsy showed profound myopathic features with numerous necrotic fibers, some regenerating fibers and perimysial inflammatory cell infiltrates (predominantly composed of macrophages and T cells), combined with a diffuse overexpression of MHC class I products (figure 1). Results for myositis specific and associated autoantibodies showed anti-HMGCR antibodies positivity (>200, N<20). Given the association between inflammatory myopathies and malignancy, an investigation was performed and ruled out malignancy.

She was diagnosed with SINAM and statin was suspended. Methylprednisolone 1g/day, for 3days followed by prednisolone 60mg/day (0.75mg/kg/day) was started. Due to minimal improvement, concomitant intravenous immunoglobulin was prescribed for 5 consecutive days (0.4g/kg/day). A significant improvement in muscle strength, myalgia and substantial reduction in the CK level were observed. After 21 days, she was discharged on a tapering dose of steroids and metho-

trexate. After three months of follow up, neurological examination was normal and CK returned to normal (15 U/L).

Discussion: SINAM is an extremely rare and severe form of statin myopathy, which can lead to debilitating weakness. Exclusion of inflammatory myopathies, metabolic and genetic muscle disorders and toxic statin myopathy is necessary for establishing a reliable diagnosis. In SINAM, simply discontinuing statin is often insufficient and aggressive immunosuppression therapy is needed to achieve the disease remission.

049 - PSEUDO-PSEUDO MEIGS SYNDROME: AN UNCOMMON ONSET OF SYSTEMIC LUPUS ERYTHEMATOSUS

Ana Martins[1, 2], Maria Seabra Rato[1, 2], Daniela Santos Oliveira[1, 2, 3], Frederico Rajão Martins[4], Filipe Oliveira Pinheiro[1, 2], Diogo Guimarães da Fonseca[5], 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, [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]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Introduction: Serositis is seen in approximately 12% of patients with systemic lupus erythematosus (SLE), usually in the form of pleuritis and/or pericarditis(1). Peritoneal serositis with ascites is an extremely rare manifestation and ascites as initial manifestation of SLE is even rarer(1).

Case report: A 48-year-old female with no past medical history presented to the emergency department with periumbilical abdominal pain in the last month, history of increasing abdominal girth, fatigue, weight loss and anorexia. She, also, complained of polyarthralgia with morning stiffness for the past 4 months involving the wrists, proximal interphalangeal joints (PIP) and knees.

On physical examination, she looked older than she was, underweight and pale. Abdominal examination revealed distension with diffuse tenderness and dull note on percussion of the flanks, no abdominal masses or organomegaly. Mild tenderness at the PIP joints without swelling or deformities was found. No peripheral edema, cutaneous lesions or stigmata of chronic liver disease were found.

Laboratory workup showed pancytopenia and elevation of inflammatory markers. Renal, liver and thyroid functions were normal. Urine analysis showed leukocyturia, proteinuria and hematuria. Spot urine protein-to-creatinine ratio was 765 mg/g. Urine culture identified an Escherichia coli and suitable antibiotic was started.

Since the patient had a history of weight loss, abdominal pain and distension, a study was carried out to look for underlying malignancy. CT of chest, abdomen and pelvis showed moderate ascites, mild bilateral pleural effusion and very small pericardial effusion. No other alterations were found. Cancer antigen 125 level was 60 U/mL (N 0-35). Ascitic fluid workup showed characteristics of exudate with a low serum-ascites albumin gradient. Culture and mycobacterium tuberculosis polymerase chain reaction were negative and no malignant cells were visualized. Autoimmune workup showed positive ANA (1:1280, homogenous pattern), anti-dsDNA (>800 IU/mL) and anti-nucleosome antibodies and low complement levels (C3 54mg/dL, C4 17mg/dL). SLE associated with pseudo-pseudo Meigs syndrome (PPMS) has been diagnosed and oral prednisolone and hydroxychloroquine were started.

During hospitalization, the patient had a progressive worsening of the pancytopenia and a blood transfusion was required. Also, an increase in spot urine protein-to-creatinine ratio was noticed. A 24-hour urine protein test revealed 3080 mg/day and renal biopsy showed class III lupus nephritis. Intravenous methylprednisolone pulse (1g daily, 3days), followed by 0.5mg/kg daily of oral prednisolone and bolus intravenous cyclophosphamide (1 g/month, 6months), angiotensin-converting-enzyme inhibitor, calcium and vitamin D supplementation were also started.

The patient was discharged after 3 weeks. Steroid was tapered and mycophenolate mofetil was initiated after the 6 monthly cycles of cyclophosphamide. At 3 months follow-up, a significant clinical and laboratory improvement was noticed with recovery of her usual weight and complete resolution of pancytopenia, proteinuria and polyserositis.

Discussion: PPMS is a newly emerging manifestation of SLE, characterized by the presence of ascites, pleural effusion and raised CA-125 level. Only few cases have been published before. Awareness of this entity is crucial to warranty an early recognition, effective treatment and consequently a better prognosis.

REFERENCES

1. Man BL, Mok CC. Serositis related to systemic lupus erythematosus: prevalence and outcome. Lupus. 2005;14(10):822-6.

053 - PRIMARY SJÖGREN'S SYNDROME PRESENTING AS THROMBOTIC THROMBOCYTOPENIC PURPURA IN A MALE PATIENT WITH PREVIOUS KIKUCHI-FUJIMOTO DISEASE

Manuel Silvério-António[1, 2], Lídia Alves Ribeiro[3], Júlia Medeiros[3], Vasco C Romão[1, 2], JE Fonseca[1, 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]Serviço de Hematologia e Transplantação de Medula, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

A 28-year-old Caucasian man, with a past medical history of isolated self-limited cervical Kikuchi-Fujimoto Disease (KFD) 9 years before, presented with a 5-day history of fatigue, fever and mucocutaneous pallor. Two days later, he developed headache, progressive weakness of right upper limb, confusion and slurred speech. The patient had mild xerostomia and extensive teeth lack. Laboratorial investigation showed intravascular haemolytic anaemia (haemoglobin [Hb] 7.0 g/dL, total bilirubin 1.7 mg/dL, lactate dehydrogenase [LDH] 1400 IU/L, haptoglobin <10 mg/dL) and thrombocytopenia (platelets 11,000/mm3). Blood smear revealed anisocytosis and poikilocytosis with schistocytes and polychromatic erythrocytes. Erythrocyte sedimentation rate was 94 mm/1sth and C-reactive protein was normal range. Creatinine was mildly raised at 1.14 mg/dL and general biochemistry was otherwise unremarkable. Prothrombin time, partial thromboplastin time, fibrinogen and D-dimers were normal. Cultures and serology did not reveal any acute infection. Cranial and thoraco-abdominal CT scans were unremarkable. A diagnosis of thrombotic thrombocytopenic purpura (TTP) was suspected. The patient started daily plasma exchange therapy (PET) with resolution of neurologic defects and laboratorial improvement. The TTP diagnosis was subsequently confirmed by a severe deficiency of ADAMTS13 activity (<1%) and anti-ADMATS13 IgG autoantibodies (>95 UI). At 7th day of PET there was a laboratorial disease relapse treated with high-dose glucocorticoid therapy and rituximab. The patient had also positive ANA (1:320 speckled), anti-Ro-52 and anti-Ro-60 antibodies. He had a high clinical oral dryness score (CODS=7) and diminished unstimulated whole salivary flow rate at 0.1 mL/min. Schirmer's test (15mm/5'), ocular staining score (0) and tear break-up time were unremarkable. Salivary gland ultrasound revealed grade 2 changes in parotid glands, according to OMERACT/EULAR definition. Salivary gland biopsy showed focal lymphocytic sialadenitis with a focus score <1/4 mm2. According to the 2016 American College of Rheumatology/European League Against Rheumatism classification criteria, a diagnosis of pSS was confirmed. Additional extra-glandular involvement was ruled out and there was no systemic disease activity (ESSDAI=0).

Steroids were tapered and discontinued after 6 months. At 2 months normalization of Hb, platelets, LDH and ADAMTS13 activity were observed and anti-ADAMTS13 auto-antibodies were negative. There were no signs of TTP relapse after one year of follow-up.

There are 16 reports of TTP associated with pSS: mostly female patients (14/16) and with SS diagnosis prior to TTP in less than 50% (7/16). Studies in systemic lupus erythematosus (SLE) and other autoimmune diseases suggest that older age and female sex are risk factors for TTP. Our case presents uncommon features, such as young age, male sex and presentation with the 5 typical clinical manifestations.

Only 10 case reports linked KFD with SS, mostly in women (9/10), with the KFD diagnosis generally preceding SS onset (7/10). As for SLE, patients with KFD and SS have a higher recurrence rate than reported for idiopathic forms (7/10 patients vs 3-4%). Our patient had some atypical features such as male gender and lack of recurrences.

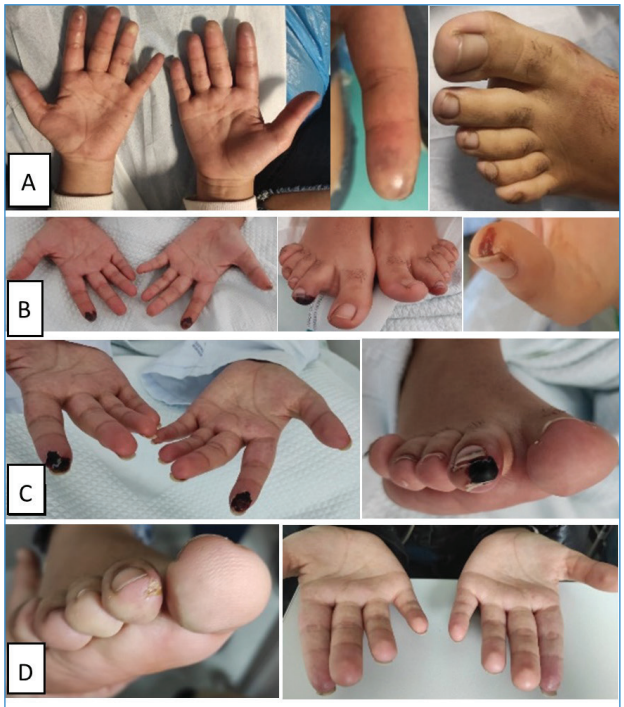
This report presents the first association between these three clinical entities and highlights the possibility of atypical characteristics of TTP and KFD in patients with SS.

062 - SEVERE DIGITAL ISCHEMIA OF UNKNOWN ETIOLOGY

Mariana Santos[1], Ana Teodósio Chícharo[2], Laura Gago[1], Maria João Gonçalves[1], Maria Manuela Costa[1], Jaime C. Branco[1, 3], Alexandre Sepriano[1, 3]
[1]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [2]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, [3]CEDOC, NOVA Medical School, Lisboa, Portugal

Introduction: Digital ischemia is caused by a reduction of the arterial blood flow and may have a wide variety of underlying etiologies, including embolism, local thrombosis, systemic connective tissue diseases, vasculitis or paraneoplastic syndromes. However, it can be also idiopathic. The rheumatologists are frequently the first health professionals to manage patients with digital ischemia.

Cases report: We report a case of a 19-year-old girl of African descendent, who presented in the emergency department of a tertiary hospital in January 2022 complaining of pain and changes in color of the fingers and toes while expose to cold. She also had inflammatory, additive and symmetric polyarthralgia, with 1 month duration, as well as anorexia and unintentional weigh



A – clinical status at emergency department; B – ischemia progression to necrosis 1 week after initial treatment; C – stopped progression after intensive glucocorticoid treatment; D – Re-evaluation after discharge and rituximab initiation

Figure 1. Ischemic lesions progression

loss. There was no previous relevant medical history or medications. In the 2 years before, she started to smoke (1.20 pack-year), and reported drug abuse (MDMA) and binge-drinking episodes.

At physical observation, she had cold extremities, digital cyanosis of both feet and hand with rare petechiae. One digital pulp was pale and 3 were erythematous (Figure 1A). All pulses were present. Laboratory results revealed leukocytosis and a slight increase of the erythrocyte sedimentation rate and C-Protein Reactive (27 mm/h and 3.45 mg/dL, respectively). The patient was admitted into our department for diagnosis and treatment.

The immunologic study showed only weak positive values of anti-neutrophils antibodies, anti-nRNP and cryoglobulins (<1%). The capillaroscopy revealed unspecific abnormalities and the remaining diagnostic study was negative (e.g. no underlying malignance and no vascular abnormalities).

The patient was (empirically) treated with iloprost, prednisolone 30 mg/day and acetylsalicylic acid (AAS) 100 mg/day with little success. The digital cyanosis aggravated, progressing to ischemia and necrosis of the digital pulp of the second finger of both hands and of the second left toe (Figure 1B). Because of this unfavorable progression, three pulses of methylprednisolone (1000 mg/day during 3 days) with a subsequent switch to 1 mg/Kg/day of prednisolone, was added to iloprost and AAS. The patient also initiated hyperbaric treatment. One week after the new treatment the lesions stopped to progress (Figure 1C), and then eventually reduced their size. the improvement observed, the patient not initiated rituximab, assuming an wait and see attitude. Initially, it was considered the possibility of starting rituximab before discharge. However, due to the significant clinical improvement observed, it was chosen a wait and see attitude. The lesions continued to progress well after the suspension of the iloprost and hyperbaric treatment. In the most recent visit, the patient was asymptomatic and free of lesions (Figure 1D).
Discussion: Since the patient responded well to immunosuppressive therapy (in combination with iloprost and hyperbaric treatment), an early manifestation of a systemic rheumatic disease, cannot be completely ruled out. Tromboangiitis obliterans is also an option given the history of smoking, even though it occurs more frequently in males with heavy tobacco load, and it does not explain the constitutional symptoms of our patient. Irrespective of the underlying cause, this case shows the benefits of quick and aggressive treatment in cases of severe digital ischemia.

067 - THE MANY FACES OF PULMONARY INVOLVEMENT IN A PATIENT WITH A SYSTEMIC SCLEROSIS-OVERLAP SYNDROME

Rafaela Nicolau[1], Ana Martins[2], Frederico Rajão Martins[3], Beatriz Samões[4], Diogo Guimarães da Fonseca[4], Miguel Bernardes[2, 5], Lúcia Costa[2]
[1]Rheumatology Unit, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [2]Rheumatology Department, Centro Hospitalar São João, 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

Systemic Sclerosis (SSc) can present alone or as part of an overlap syndrome with other autoimmune diseases like Sjögren's syndrome (SS). Pulmonary manifestations of SSc, namely interstitial lung disease (ILD) and pulmonary arterial hypertension (PAH), are the leading cause of death. Undiagnosed contributors to PH, such as chronic thromboembolic disease, should be properly investigated.

We report a case of thromboembolic complication in a patient with SSc sine scleroderma/SS overlap syndrome who showed worsening of her usual dyspnea.
Case report: We report the case of a 69-year-old female

with the diagnosis of a SSc sine scleroderma/SS overlap syndrome at the age of 55, when she presented with anti-nuclear antibodies (1/1000, speckled pattern), anti-SSA and anti-SSB antibodies, polyarthralgias, sicca symptoms and minor salivary glands histology compatible with SS. In addition, there were features of SSc such as nailfold capillaroscopy with active scleroderma pattern, nonspecific Interstitial pneumonia (NSIP), PAH, Raynaud's phenomenon, digital ulcers, pitting scars, telangiectasias and dysphagia, meeting the revised classification criteria of Ssc. Currently, she was taking Rituximab every 6 months, sildenafil 150 mg/day, ambrisentan 10 mg/day pentoxifylline 400 mg/day, prednisolone 5 mg daily and home oxygen therapy (4 L/min).

She was admitted to our hospital complaining of a 3-month history of progressive worsening of dyspnea on minor exertion and rest, with worsening lower limb edema in the afternoon. Moreover, pulmonary function tests performed 4 months ago, showed an isolated decline DLCO (11%) with normal total lung capacity and forced vital capacity.

On admission to the rheumatology department, she was apyretic, hemodynamically stable and with an oxygen saturation of 95%. Cardiopulmonary auscultation exhibited rhythmic beats, without murmurs, diffused wheezing and inspiratory basal crackles.

Laboratory testing revealed normal blood count and normal inflammatory parameters, highlighting the high serum levels of brain natriuretic peptide (1095.8 pg/mL). A high-resolution chest CT was performed, confirming a stable NSIP pattern when compared to previous exams. The echocardiogram detected worsening of estimated pulmonary artery systolic pressure (100 mmHg) and of right ventricular dysfunction. Additionally, ventilation-perfusion lung scintigraphy of the lung revealed an acute and chronic thromboembolism. Thus, anticoagulation was started and sildenafil was replaced by talafafil 40 mg/day.

Discussion/Conclusion: In conclusion, we report a rare case of a patient with SSc/SS overlap syndrome who developed acute and chronic thromboembolic PAH. PAH in SSc may comprise a variety and overlap of phenotypes. Most of them due to impairment of pre-capillary arterioles, but also due to ILD and/ or pulmonary thrombosis. SSc thromboembolic complications among patients with PAH should be promptly identified to improve treatment decision and prognosis.

068 - EXTENSIVE SPONTANEOUS PNEUMOMEDIASTINUM, A RARE MANIFESTATION OF AMYOPATHIC DERMATOMYOSITIS

Margarida Lucas Rocha[1], Laura Gago[2, 3], Alexandre Sepriano[2, 3], Tiago Saldanha[4], Ana Filipa Mourão[2, 3], Manuela Costa[2], MD[5], Jaime C. Branco[1, 3]
[1]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, [2]Rheumatology department, Hospital de Egas Moniz, Centro Hospitalar de Lisboa Ocidental, Lisboa, Portugal, [3]CEDOC, NOVA Medical School, Nova University, Lisboa, Portugal, [4]Serviço de Radiologia, Hospital de Egas Moniz, Centro Hospitalar de Lisboa Ocidental, Lisboa, Portugal, [5]Serviço de Pneumologia, Hospital de Egas Moniz, Centro Hospitalar de Lisboa Ocidental, Lisboa, Portugal

Introduction: Amyopathic dermatomyositis (ADM) is a rare rheumatic and musculoskeletal disease (RMD) characterized by skin lesions typical of dermatomyositis but without muscle involvement. Respiratory involvement occurs in about half of cases and spontaneous pneumomediastinum (PnM) is a rare and often fatal complication. So far, there is no consensus for the management of PnM associated with RMDs. We report the case of a patient diagnosed with ADM, anti-melanoma differentiation-associated gene 5 (anti-MDA5) positivity and interstitial lung disease (ILD) complicated by PnM and subcutaneous emphysema.

Clinical Case: A 61-year-old non-smoker female patient, with asymmetric polyarthralgia, Gottron papules and a 6-month history of non-productive cough, fatigue and dyspnoea for moderate efforts was evaluated in our department. On physical examination, the patient had bilateral velcro crackles in the thorax, normal muscle strength and normal muscle enzymes. She tested positive for anti-MDA5 and had a nonspecific interstitial pneumonia pattern in the high-resolution computed tomography (HRCT). Respiratory function tests showed moderate restrictive respiratory failure and moderate reduction in alveolar-capillary transference capacity for carbon monoxide causing hypox-

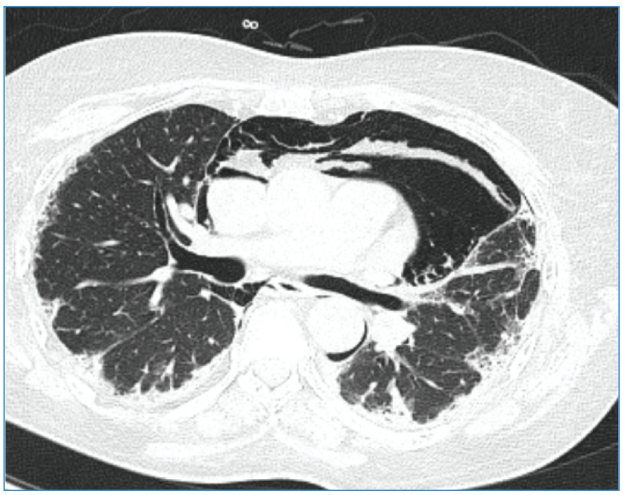


Figure 1. Extensive pneumomediastinum

emia at rest. The diagnosis of ADM with ILD was made and the patient started mycophenolate mofetil (MMF) 1500mg/day, prednisolone (PDN) 1mg/kg/day and long-term oxygen (2 L/min for 16h/day). The patient responded well to therapy, but soon after PDN tapering was started, she complained of severe dysphagia and was therefore readmitted. An upper digestive endoscopy revealed esophageal candidiasis and six days after, the patient complained of chest pain with accompanying cervical and upper thoracic subcutaneous emphysema. An urgent thoracic HRCT revealed an extensive PnM. A ventilation/perfusion scintigraphy identified a possible leak at the level of the trachea, which was not confirmed by bronchoscopy. No evidence supporting paraesophageal abscesses or an esophagus perforation was found. Despite the extensive PnM, the patient remained asymptomatic under treatment with continuous oxygen and bronchodilators. The dose of MMF was increase to 2000mg/day, with a deescalating dose of PDN up to 25mg/day. Upon discharge, the patient was under 4L/min of continuous oxygen therapy and was already able to walk short distances.

The chest HRCT performed 4 months after the onset of the PnM showed a marked reduction of its dimensions. Three months later she was hospitalized due to emphysematous cystitis and developed hypoxemic nosocomial pneumonia. Despite invasive ventilation the patient showed progressive worsening of respiratory failure as a consequence of an increase of the PnM, which resulted in her death 6 months after the initial diagnosis.

Discussion: PnM can occur with different forms of ILD associated RMDs. Although this is a rare complication, patients with dermatomyositis are reported to be at particular high risk of PnM. By definition, spontaneous PnM is not caused by trauma, surgery or other medical procedure. The evidence supporting the efficacy of interventions for PnM associated with ADM is yet scarce. We opted for non-invasive approach with close monitoring which was followed by an initial reduction of the PnM. However, the follow-up HRCT did show an increase of pulmonary lesions and a major increase in the dimensions of the PnM. Evidence clarifying the optimal management of PnM associated with RMDs is greatly needed.

072 - LYTIC SKULL LESIONS - FINDING THE CAUSE

Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Tiago Beirão[1], Catarina Rua[1], Flávio Campos Costa[1], Romana Vieira[1], Taciana Videira[1], Margarida Badior[2], Joana Aleixo[1], P Pinto[1]

[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [2]Haematology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Introduction: Lytic skull lesions can be attributable to a wide range of diagnoses, like bone metastases, multiple myeloma, or metabolic bone disease. This case report describes an atypical cause for these lesions.

Case report: 67-year-old women referred to the Rheumatology consultation for an incidental finding of lytic lesions in the skull on a cranial CT scan during a recent-onset headache study. She had a history of arterial hypertension, dyslipidaemia, chronic gastritis, depression and osteoporosis with previous fragility fracture. She was chronically medicated with lansoprazole 30 mg, bisoprolol 1.25 mg, losartan-hydrochlorothiazide 50/12.5 mg, atorvastatin 20 mg, annual zoledronate 5 mg, cholecalciferol 600 UI and sertraline 50 mg. She was initially observed on the Haemato-Oncology consultation to exclude multiple myeloma. She only had asthenia and the physical examination was unremarkable. The blood tests showed normal blood count, protein electrophoresis, immunofixation, immunoglobulin assay, beta-2-microglobulin and light chain ratio as well as proteinuria, light chains, immunofixation and immunoglobulin on 24-hour urine. Vitamin D and serum and urinary calcium and phosphorus were normal. Parathyroid hormone was slightly elevated (76,3 pg/mL) but primary hyperparathyroidism was excluded. Radiographs of the skeleton were performed showing no other lytic lesions (figure 1). FDG-PET scan excluded foci of hypermetabolism suggestive of plasmacytoma. The bone marrow biopsy ruled out non-secretory multiple myeloma. The paraneoplastic study was also negative. The patient's clinical history was reviewed, and it came to our attention that she was exposed to irradiation therapy for the treatment of Tinea capitis in childhood, exactly in the same area of the skull. Years later, the patient's sister was referred to the consultation

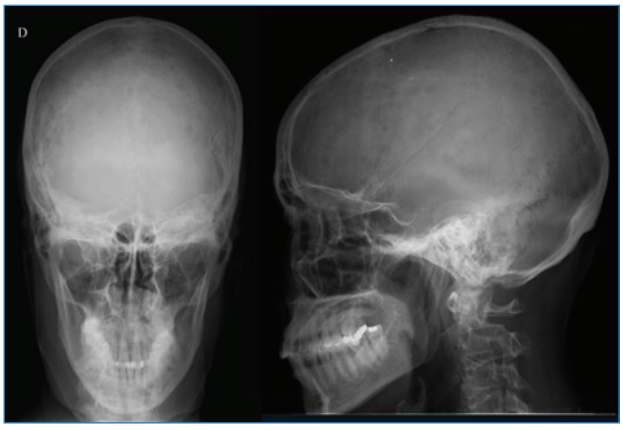


Figure 1. Lytic skull lesions

for the same incidental finding and she too had been exposed to the same treatment.

Conclusion: Radiation therapy was a treatment used up to 1960s for Tinea capitis that has been linked to the later development of cancer in the exposed areas.1 To our knowledge, this is the first report to suggest an association between this ancient therapy and benign lytic skull lesions, mimicking other clinical situations.

REFERENCES

1. Antunes L, Bento MJ, Sobrinho-Simões M, Soares P, Boaventura P. Cancer incidence after childhood irradiation for tinea capitis in a Portuguese cohort. Br J Radiol 2020; 93: 20180677

076 - A NEW GENETIC ASSOCIATION WITH OSTEOGENESIS IMPERFECTA

Frederico Rajão Martins[1], Francisca Aguiar[2], Sara Ganhão[3], Mariana Rodrigues[2, 4], Esmeralda Rodrigues[5], Iva Brito[2, 4]

[1]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [2]Paediatric and Young Adult Rheumatology Unit, Centro Hospitalar Universitário de São João, Porto, Portugal, [3]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [4]Faculdade de Medicina da Universidade do Porto, Porto, Portugal, [5]Pediatrics Department, Centro Hospitalar Universitário de São João, Porto, Portugal

Introduction: Osteogenesis imperfecta (OI) is a rare skeletal dysplasia, affecting approximately 1 in every 15000 to 20000 children. Characteristic features include bone fragility and high frequency of fractures, growth retardation, blue sclerae and dentinogenesis imperfecta. OI is classified in several subtypes according to genetic, radiographic and clinical features. New mutations associated with this condition continue to be identified.

Clinical Case: A five-year-old girl with dorsolumbar scoliosis was evaluated for bilateral diffuse lower limb pain after multiple spontaneous fractures: right cuboidal bone, left talus, left calcaneus and 2nd left metatarsal bone. Light blue sclerae, translucid skin of the feet, lower limb length discrepancy and a positive bending test were observed. Body stature was appropriate for age and there were no dental abnormalities. There was no family history of metabolic bone disease. Successive new fractures of the right radius after minor trauma and right spontaneous patellar fracture occurred in the following months. She was evaluated in ENT for hypoacusis, without any objective findings. Stomatological evaluation identified several decidual teeth fractures derived from multiple cavities, with no signs of dentinogenesis imperfecta. Besides low 25(PH)D3 levels (16 ng/mL) there were no abnormal findings regarding phosphocalcic metabolism, osteocalcin, beta-crosslaps or bone density (Z score = - 0.2). Radiographic imaging showed consolidation of previously described fractures, periarticular osteopenia, cranial wormian bones and patchy skull density. Genetic testing revealed a new likely pathogenic variant of the CREB3L1 gene - c.1187A>G (p.(Glu396Gly)).

Diagnosis of type XVI OI was established and cholecalciferol supplementation (22400 UI) initiated. In the absence of new fractures, and with optimal pain relief, no additional treatment was required.

Conclusion: We report a case of OI in a child with recurrent bone fractures, blue sclerae and radiological abnormalities, in the absence of skeletal deformities, hearing loss or family history.

This case highlights the clinical heterogeneity of OI. Type XVI OI is classically associated with an autosomal recessive inheritance pattern, contrasting with the classical subtypes of OI, delaying the diagnosis. This is the mildest form of CREB3L1 associated OI reported, further expanding the clinical spectrum of this disease.

078 - AN ATYPICAL CASE OF ERYTHEMA NODOSUM

Frederico Rajão Martins[1], Beatriz Samões[2], Ana Martins[3], Rafaela Nicolau[4], Lúcia Costa[3]

[1]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [2]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [3]Rheumatology Department, Centro Hospitalar de São João, Porto, Portugal, [4]Rheumatology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Introduction: Erythema nodosum (EN) is a delayed-type hypersensitivity reaction mostly presenting as painful erythematous nodules on the lower limbs. Mild musculoskeletal symptoms may occur, such as arthralgia and articular swelling. Common triggers include infection, drugs, malignancy and inflammatory conditions, yet a significant proportion of cases are idiopathic.

Clinical Case: A 33-year-old healthy woman with no contraceptive intake presented in the emergency department for chronic recurring erythematosus painful nodular lesions in the lower limbs.

There was history of vaginal discharge associated with cutaneous erythematous macules in the lower limbs three months' prior, resolved with topical antifungal, amoxicillin/clavulanate and etodolac.

Few weeks before evaluation, new nodules appeared in association with polyarthralgia involving elbows, right knee and ankle, malaise and anorexia. Prednisolone (PDN) was administered in rapid taper (40mg – 20 mg – 10mg, three days each), with early improvement but relapse at the lowest dosage.

Physical examination in the emergency department showed multiple areas of residual skin hyperpigmentation and discrete painless nodules in the anterior lower limbs, the biggest measuring 5 x 3 cm. Right elbow palpation was painful, with preserved range of motion (ROM). The patient was admitted for further study and PDN was suspended. She became subfebrile (37,1 to 37,4ºC), and there was recurrence of erythematosus non-ulcerative painful nodules in the anterior and posterior lower limbs and forearms, and polyarthralgia involving right elbow, right knee, ankle and talocalcaneal areas. New contracture (~40º) of the right elbow was evident, with ultrasound revealing mild synovial effusion.

Laboratorial data showed mild leucocytosis (10,68 x109) with discrete neutrophilia (8,13x109), an elevated CRP (36,3 mg/L), ESR (60 mm/hr), serum amyloid A protein (37,9 mg/L) and antistreptolysin O titters (ASO, 954 U/mL). Angiotensin converting enzyme was normal. Immunological studies revealed positive ANA 1/320 in a nucleolar pattern and IgM anti-beta2 glycoprotein I (58 SMU U/ml), and negative ANCA, ACPA, anti-dsDNA and rheumatoid factor. C3, C4, C1 q and its inactivator were in the normal range. Serological studies only revealed a positive IgM for Chlamydia Pneumoniae; Streptococcus group B was negative in vaginal exudate. Chest radiograph was normal. Skin biopsy showed septal and lobular panniculitis with predominantly lymphocytic infiltrates, associated with small and medium vein vasculitic lesions, in the absence of fibrinoid necrosis. No microorganisms were isolated in histochemistry.

An EN of probable streptococcal origin was diagnosed and the patient was initiated on PDN 10mg id and indomethacin 50mg bid. There was resolution of skin lesions and skeletal complaints, with restoration of normal ROM. Inflammatory markers subsided and ASO titters halved 3 months later.

At six months of follow-up, the patient is asymptomatic without treatment.

Conclusion: This report evidences EN with exuberant cutaneous and musculoskeletal manifestations in the setting of high ASO titters. Arthralgia and limited ROM were more evident in this case than reported in literature, dramatically involving less affected joints such as the elbow.

Histologic findings weren't typical for the disease, as vasculitis lesions are characteristically absent or only involving small vessels on early stages.

079 - AN INNOCENT RAYNAUD PHENOMENON SPELLING TROUBLE

Frederico Rajão Martins[1], Beatriz Samões[2], Ana Martins[3], Daniela Santos Oliveira[3, 4], Lúcia Costa[3]

[1]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [2]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [3]Rheumatology Department, Centro Hospitalar de São João, Porto, Portugal, [4]Center for Health Technology and Services Research (CINTESIS), Faculdade de Medicina da Universidade do Porto, Porto, Portugal, Porto, Portugal

Introduction: Anti-synthetase syndrome (ASS) is an inflammatory myopathy with diverse clinical subsets, and neoplastic disease is associated with myositis specific antibodies. Venous thrombosis has been documented in patients with ASS and positive antiphospholipid antibodies, with no arterial events reported.

Clinical Case: A 41-year-old woman with smoking habits and no contraceptive pill use was admitted in the emergency department for sudden dysarthria, right hemiplegia and prolonged chest pain. Severe motor aphasia, central facial palsy and right hypoesthesia were identified, and angio-CT revealed endoluminal thrombi in the medial cerebral and right profound femoral arteries. Electrocardiogram revealed inverted T wave and prolonged QT segment in anterolateral chest derivations, in association with troponin I levels of 1884 ng/mL and echocardiographic evidence of septal akinesia, diagnostic of myocardial infarction. Thrombolysis and thrombectomy were performed with good clinical response and femoral reperfusion was observed in Doppler ultrasound (US). Coronary CT calcium scoring and cerebral MRI excluded atherosclerotic coronary disease or carotid dissection. Echocardiogram excluded right-left atrial shunt. Neoplasia was excluded through thoracic-abdominal-pelvic CT, mammography, mammary US and gynaecological evaluation. Coagulation studies evidenced a discreet self-limited acute antithrombin III deficit. The patient started warfarin, with no new events.

Rheumatology consultation was requested due to chronic mechanical polyarthralgia of small hand joints and long-standing biphasic Raynaud phenomenon involving the distal digital region in both hands. No other symptoms were present. For the exception of puffy fingers, physical examination was unremarkable and skin ulcerations were absent. Immunological studies revealed positive ANA 1/320 in a speckled pattern and a borderline C1q level (19 mg/dl) without additional immunological or serological findings. Videocapillaroscopy revealed unspecific changes in capillary morphology.

After 6 months of follow-up the patient developed proximal painful muscular weakness of the scapular girdle appear (grade 3/5), associated with focal leg induration, persistent xerostomia and progressive dyspnoea on exertion. Minor salivary gland biopsy showed unspecific mild chronic sialadenitis. Left leg ultrasound

revealed soft tissue calcifications. Upper limb electromyography was normal. Laboratorial data revealed a slightly increased aldolase (10.8 U/l), low C1q (16 mg/dl), C3 (74 mg/dl) and CH50 (32.8 U/ml) levels. Despite initial negative antiphospholipid antibodies, revaluation showed increased IgG anticardiolipin titres in two separate assessments (155 and 36.8 GPL) and IgG anti-beta2-glycoprotein I antibody elevated in one measurement (34.4 SGU). Myositis specific antibodies anti-PL7 and anti-PL12 were identified.

Probable catastrophic antiphospholipid syndrome diagnosis was established, in possible association with an idiopathic inflammatory myopathy. Pulmonary function tests, high resolution lung CT scan and proximal limb muscle MRI are pending.

Conclusion: To the best of our knowledge this is the first case of antiphospholipid syndrome with multiple arterial thrombi at presentation associated with a probable ASS. Anti-PL7 and anti-PL12 positivity is rare and clinical characteristics are poorly documented. Evidence in the literature indicates mild to none muscular involvement and significant early interstitial lung disease in these patients, warranting further studies in this situation.

090 - THE ROLE OF TERIPARATIDE ON ATYPICAL FEMORAL FRACTURE AFTER PROLONGED USE OF BISPHOSPHONATES

Daniela Santos Oliveira[1, 2, 3], Salomé Garcia[3], Georgina Terroso[3], Luciano Pereira[4], Lúcia Costa[3], Carlos Vaz[1, 2, 3]
[1]Center for Health Technology and Services Research (CINTESIS), Faculty of Medicine, University of Porto, Porto, Portugal, Porto, Portugal, [2]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal, [3]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [4]Serviço de Nefrologia, Centro Hospitalar Universitário de São João, Porto, Portugal

Introduction: The prolonged use of inhibitors of osteoclast activity, such as bisphosphonates, are associated with atypical femoral fractures (AFF). The pathophysiology of AFF is not completely clear, however it is known that prolonged use of these drugs may led to the development of adynamic fragile bone. Therapeutic options in patients with a previous AFF remains to be clarified. Though, teriparatide is a promising therapy.

Case report: We present a case of a 65-year-old man with a stage 3a chronic kidney disease and osteoporosis with no personal history of fractures. This patient was admitted to the emergency department due to severe left trochanteric region pain following a low impact fall. There were no smoking and alcoholic habits. No current or previous chronic steroid therapy, femoral neck fracture of his parents and prolonged immobility in the last 6 months were reported. He was on alendronate (70 mg weekly) for the past 20 years. Pelvic X-ray revealed a transverse diaphyseal femur fracture with a medial spike. After an osteosynthesis procedure, the patient was referred to our Fracture Liaison Service. On investigation: creatinine is 1.33 mg/dL, with an estimated Glomerular filtration rate (GFR) of 54 mL/min/1.73 m2 (stage 3a). Ionized calcium, inorganic phosphate, 25-OH-vitamin D and parathormone were normal. Regarding bone turnover markers, alkaline phosphatase was 62 U/L (normal range 30-120), osteocalcin was 18 ng/mL (normal <41.3) and beta-crosslaps was 0.17 ng/mL (normal <0.32). Dorsal and lumbar spine radiographs revealed no vertebral fractures. The bone densitometry revealed lumbar spine and femoral neck T-score were -2.9 and -1.9, respectively. For suspicion of atypical femoral fracture, patient performed a tetracycline-labeled bone biopsy. This biopsy showed absent osteoblastic surface, reduced osteoclastic, erosion and osteoid surfaces. Fluorescent microscopy revealed few very weak tetracycline single labels. These findings were compatible with the diagnosis of low turn-

Table I. Histomorphometric parameters of bone biopsy.

Histomorphometric parameters		Reference
BV/TV (%)	12,11	24.60 ± 7.10
OV/BV (%)	0,05	2.30 ± 2.40
OS/BS (%)	0,26	13.30 ± 11.10
ES/BS (%)	0,21	NS
OTh (µm)	4,64	11.2 ± 3.40
TbN (mm)	2,73	NS
Tb.sp (µm)	321,03	NS
ObS/BS (%)	0	1.60 ±3.30
OcS/BS (%)	0,10	0.03±0.09
BFR/BS (µm3/µm2/year)	NS	0.061±0.025
MS/BS (%)	1,14	NS
sL.S/BS	2,28	NS
MLT (days)	NS	17.3±6.5

BV – bone volume; TV – tissue volume; OV – osteoide volume; OS – osteoide surface; BS – bone surface; OTh – osteoide thickness; TbN – trabecular number; Tb.sp – trabecular separation; ObS – osteoblast surface; OcS – osteoclast surface; MS – mineralizing surface; sL.S – single labelled surface; MLT- mineralization lag time. NS – not specified.
Note: BFR/BS and MLT were not calculated due to absence of tetracycline double labels.

over bone disease/ adynamic bone disease. The main histomorphometric parameters are shown in Table 1. The patient received calcium and vitamin D supplementation and started teriparatide (subcutaneous daily administration). After 2 years of teriparatide, patient repeated the bone biopsy showing an improvement of histological findings, namely in bone volume and bone formation parameters. Currently, osteocalcin is 23.4 ng/mL and betacrosslaps 0.26 ng/mL, with improvement of femur neck T-score (-1.7). During this follow-up period no further fractures occurred. Conclusion Treatment of AFF remains to be understood. Our case report suggests that teriparatide could be beneficial with improvement in anabolic bone turnover markers and lumbar spine body mass density. Furthermore, bone histomorphometry showed improvement in bone formation measurements after 2 years of therapy. However, evidence is still scarce about if teriparatide reduce fracture risk over time in these patients and its safety after 2 years of treatment. Hence, prospective studies with large samples are needed to explore this therapeutic option in patients with AFF after a long course of bisphosphonates.

091 - SENSORINEURAL HEARING LOSS AND MONONEURITIS MULTIPLEX IN SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE REPORT

Daniela Santos Oliveira[1, 2, 3], Ana Martins[1, 3], Frederico Rajão Martins[4], Beatriz Samões[5], Diogo Guimarães da Fonseca[5], Carlos Vaz[1, 2, 3], Pedro Madureira[1], Lúcia Costa[1]

[1]Rheumatology Department, Centro Hospitalar Universitário 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]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal, [4]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [5]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Introduction: Peripheral nervous system disease constitutes one of the most important neuropsychiatric systemic lupus erythematosus (SLE) manifestation with a significant negative impact on quality of life. Thus, a case of peripheral nervous system disease with two different findings: sensorineural hearing loss (rarely reported) and mononeuritis multiplex is being reported. **Case report:** We report a case of a 36-year-old female, with SLE with articular involvement and secondary Sjogren for 3 years, medicated with hydroxychloroquine 400 mg/day and prednisolone 5 mg/day. This patient was initially admitted to our hospital for subacute onset of nausea, dizziness and tinnitus and posteriorly left hearing loss. There were no inflammatory arthralgias, systemic complaints, skin rash, Raynaud's Phenomenon, genital or oral ulcers, respiratory, gastrointestinal or genitourinary manifestations. Neurologic examination revealed right rotational nystagmus in all directions of gaze and left head impulse test positive. Otorhinolaryngology examination revealed normal otoscopy, video head impulse test and videonystagmography with left vestibular deficit. On investigation, she had slight elevation of CRP (10 mg/L) and ESR (42 mm/h). Immunological study was positive for antinuclear antibody (>1/1000, homogeneous pattern), anti-dsDNA antibody (737 IU/mL), anti-SSA (strong positive) and anti-RNP (weak positive), with hypocomplementemia (C3 69.6 and C4 9 mg/dL). Antiphospholipid antibodies were negative. Hemogram, platelet, hepatic parameters, renal function and muscle enzymes were normal. Brain magnetic resonance imaging reveled no changes. For suspicion of immunomediated sensorineural hearing loss associated with vestibular symptoms, patient started prednisolone 1 mg/kg/day, though without reversal of hearing loss. After 1 month (M) of this manifestation, patient reported dysesthesia in the right lower limb, with progressively worsening. Neurological examination revealed hypoesthesia of the lateral edge of the right leg and foot, without alteration in muscle strength or proprioception. There was no peripheral arthritis. Motor conduction studies demonstrated low-amplitude potentials in the left deep peroneal nerve and sensory conduction studies low-amplitude potentials in the right superficial peroneal and sural nerves bilaterally. Needle electromyography revealed signs of chronic neurogenic injury in the distal muscles of the lower limbs (sensory and motor involvement). Muscle (right soleus) and nerve (right sural) biopsy showed severe neuropathy with current activity and nonspecific muscle changes, respectively. These findings are consistent with peripheral neuropathy namely mononeuritis multiplex. Hence, patient started pulses of methylprednisolone 1g/day (3 days) and posteriorly prednisolone 1mg/kg/day with partial clinical and analytical improvement. Due to persistent hearing loss and limb sensory deficit, cyclophosphamide was initiated early with a total cumulative dose of 3.95g for 6M (dose reduction required due to leukopenia). After 15M of follow-up, under rituximab (1g, 6/6M), azathioprine (2 mg/kg/day), prednisolone (15 mg/day) and gabapentin (900 mg/day), the patient had stable dysesthesias of lower limbs and left hearing loss. On investigation, she had ESR 27mm/h, CRP 1.5mg/L, anti-dsDNA 425 IU/mL, C3 70.4 and C4 12 mg/dL. **Conclusion:** The involvement of peripheral nervous system in patients with SLE (including rarer forms such as earing and vestibular disorders) must be recognized and treated early in order to prevent disease morbidity.

097 - ANCA-ASSOCIATED VASCULITIS AFTER PFIZER-BIONTECH COVID-19 VACCINATION: TWO CASE REPORTS

Eduardo Dourado[1, 2], Sara Rodrigues Silva[3], Filipe Dias[3], Josiana Duarte[3], António Inácio[4], Catarina Abrantes[4], Liliana Cunha[4], Ana Sofia Costa[5], Cristina Ponte[1, 2], Ana Valido[6]

[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 Medicina Interna, Unidade Local de Saúde do Litoral Alentejano, Santiago do Cacém, Portugal, [4]Serviço de Nefrologia, Centro Hospitalar de Setúbal, Setúbal, Portugal, [5]Serviço de Imagiologia, Hospital Fernando da Fonseca, Amadora, Portugal, [6]Serviço de Reumatologia, Unidade Local de Saúde do Litoral Alentejano, Santiago do Cacém, Portugal

Background: The annual global incidence of anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) has been stable since the early 2000s. However, recent clusters of AAV following COVID-19 vaccination have been reported, most frequently occurring with messenger RNA (mRNA) vaccines such as Pfizer-BioNTech.

Case 1: A 26-year-old woman presented to the emergency department with a one-week history of cough, chest pain, fever and fatigue. The patient was initially diagnosed with community-acquired pneumonia and treated with azithromycin. However, due to worsening of her complaints and onset of anorexia and haemoptysis, the patient was later admitted. She had a background history of sinusitis and active smoking, and two weeks before the onset of symptoms, she received the first dose of the Pfizer-BioNTech COVID-19 vaccine. At admission, the patient had elevated inflammatory markers and acute kidney injury (AKI, Table 1), and chest CT revealed parenchymal consolidations (Figure 1). Despite treatment with IV piperacillin/tazobactam, the patient had increasing fever (up to 38.5ºC), and after a severe episode of haemoptysis, she was admitted to the ICU for transfusional support and ventilation. A new chest CT revealed worsening peripheral consolida-

Table I. Blood workup and urine sample of the two patients at admission

	Case 1	Case 2
Erythrocyte sedimentation rate (mm/h)	102	65
C-reactive protein (mg/dL)	21.0	5.81
White blood count/µL	18.400	11.600
Neutrophils/µL (%)	9.400 (51%)	8.400 (73%)
Eosinophils/µL (%)	7.000 (38%)	0 (0%)
Blood urea nitrogen (mg/dL)	78	169
Serum creatinine (mg/dL)	1.4	8.3
Urine sample	Microscopic haematuria and granular cylinders	Proteinuria (6 g/24h) and haematuria with dysmorphic erythrocytes.
Blood, urine and stool cultures	Negative	Negative
Viral and bacterial serologies, including COVID-19 and IGRA	Negative	Negative

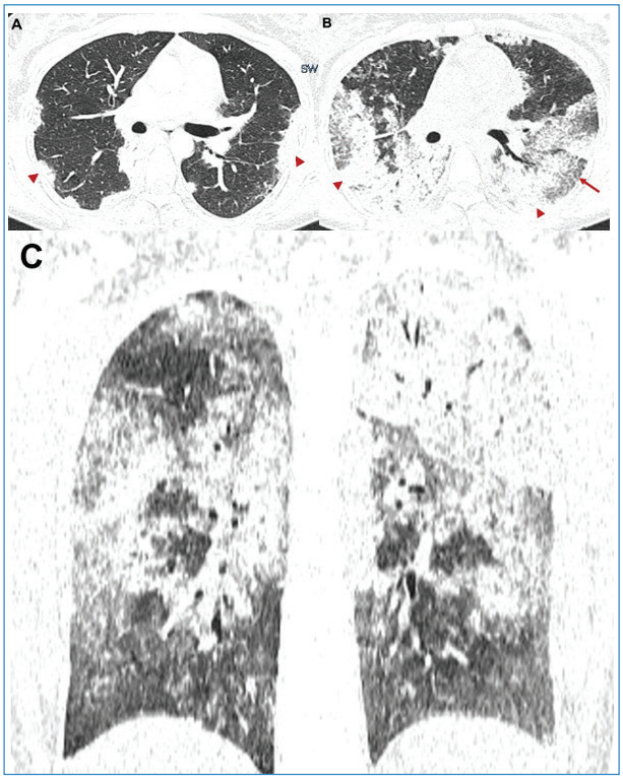


Figure 1. High-resolution thoracic CT scan images. A) Axial image of the first CT scan at the level of the inferior lung lobes demonstrates bilateral and peripheral consolidations. B) Axial image of the second CT scan at the same level shows significant worsening of the parenchymal lung findings. There are more extensive bilateral and diffuse lung consolidations (arrowheads) and new areas of ground-glass opacities with superimposed smooth intralobular and interlobular septal thickening, producing the crazy-paving pattern (arrow). In the clinical scenario, these findings are compatible with acute/subacute diffuse alveolar haemorrhage. C) Coronal image reconstruction shows the extensive craniocaudal involvement of both lungs.

tions involving >50% of the parenchyma. The patient was started on IV methylprednisolone (1000 mg/day for three consecutive days), followed by oral prednisolone 1 mg/Kg/day. Blood workups were positive for anti-proteinase 3 (PR3) ANCA (1610 UI/mL) and negative for anti-myeloperoxidase (MPO) ANCA. Bronchoalveolar lavage was compatible with alveolar haemorrhage. Rituximab was initiated but switched to IV cyclophosphamide due to adverse events. The patient is currently asymptomatic, with normalised kidney function and inflammatory markers, under treatment with methotrexate (15 mg/week) and prednisolone (5 mg/day).

Case 2: A 47-year-old man presented to his general practitioner with increasing fatigue, anorexia and abdominal pain since the administration of the second dose of Pfizer-BioNTech COVID-19 vaccine three months prior. The patient had a background history of active smoking. Laboratory tests revealed AKI with serum creatinine of 4.0 mg/dL and elevated prostate-specific antigen. This was interpreted as prostatitis, and the patient was treated with oral cefuroxime for a month. However, creatinine levels rose to 8.0 mg/dL, and the patient was subsequently admitted for rapidly progressive AKI with elevated inflammatory markers (Table 1). The renal biopsy showed crescentic glomerulonephritis. MPO-ANCA was positive (>134 UI/mL) and PR3-ANCA negative. The patient underwent six plasmapheresis sessions, three administrations of IV cyclophosphamide and methylprednisolone pulse therapy (1000 mg/day for three consecutive days), followed by oral prednisolone 1 mg/Kg/day. There was an initial response to therapy, with improvement of serum creatinine to as low as 5.2 mg/dL, but after three months, the patient became uremic and was started on haemodialysis and rituximab, which was later suspended due to inefficacy. The patient is currently under regular haemodialysis and treated with prednisolone 10 mg/day.

Discussion: mRNA vaccines activate CD8+ and CD4+ T cells and may promote immune-mediated diseases in predisposed individuals. In these two cases, temporal coincidence suggests that AAV was induced by COVID-19 vaccination, although casual association cannot be excluded. Further research into the immune responses following mRNA vaccination may provide better knowledge of the pathophysiology of AAV.

092 - RAPIDLY COGNITIVE IMPAIRMENT: A RARE MANIFESTATION OF SJÖGREN SYNDROME

Rafaela Nicolau[1], Daniela Santos Oliveira[2, 3], Ana Martins[2], Beatriz Samões[4], Frederico Rajão Martins[5], Lúcia Costa[2]

[1]Rheumatology Unit, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [2]Rheumatology Department, Centro Hospitalar 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, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [5]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal

Introduction: Neurological manifestations of Sjögren's Syndrome (SS) are multiple, including peripheral nervous system lesions, in which sensory nerves are the most common affected. On the other hand, central nervous system (CNS) involvement is relatively rare and its clinical presentation varies considerably leading to diagnosis delay.

Case report: We report a case of an 81-year-old man diagnosed with SS 5 years ago, manifested by sensory and motor polyneuropathy at L3 to S1, with gait disturbance and fourth cranial nerve palsy. At this point, patient was medicated with pulses of methylprednisolone and later, azathioprine 75 mg/day, which he discontinued due to gastrointestinal intolerance. The patient denied any symptoms suggestive of infection. Currently, he was medicated with prednisolone 20 mg/day. In June 2021, he was admitted to the Rheumatology department with a 1-month progressive history of impaired gait, loss of strength in the lower limbs and cognitive dysfunction. The patient denied any symptoms suggestive of infection or other focal neurological signs.

On admission general physical examination was unremarkable. He showed time disorientation and impaired attention. Cranial nerve examination was normal. He was unable to walk, muscle strength was globally reduced (grade 4/5) and deep tendon reflexes were week. Laboratory studies revealed mild leukocytosis with neutrophilia, reduced platelet counts, slight reduction of C3 fraction of complement and normal protein eletroctrophoresis. Hepatic, renal and thyroid function, vitamin B12, acid folic, inflammatory makers and urinalyses were normal. The screening for HSV, CMV, HBV, HCV, syphilis, parvovirus and HIV infections was negative. Electromyography of the lower limbs showed a chronic sensory and motor polyneuropathy with a slight aggravation compared to the previous exam and stable chronic neurogenic miopathy. Cranioencephalic CT was performed revealing a clear worsening of the leukoencephalopathy, lacunar lesions in thalamus and corona radiata, and increased dilatation of the supratentorial ventricular system. Magnetic resonance imaging revealed microangiopathic gliotic foci in the bulge and etat cribê pattern in the basal ganglia and countless foci of microhemorrhages (bihemispheric, right cerebellar hemisphere, right thalamus, and left lenticular nucleus). A transthoracic echocar-

diogram, carotid Doppler ultrasonography and holter test were performed, and the cardioembolic cause was excluded. Given the worsening of thrombocytopenia (30,000 /µl), lumbar puncture was delayed and intravenous immunoglobulin therapy (1g/kg/day) was started for 3 days, with recovery of platelet counts. Cerebrospinal fluid cultures were negative and no neoplastic cells were found. Levels of dementia biomarkers (Aβ42 and tau) were normal. Due to those findings, severe CNS involvement was assumed and therapy with Rituximab was initiated. The patient was discharged to a rehabilitation center and after 2-months of follow-up there was a partial improvement of gait and cognitive functions.
Discussion/Conclusion: Neurological system is one of the most common extraglandular involved sites in SS patients, and may precede the sicca symptoms. Neurological manifestations can involve both peripheral and central nervous systems. The authors report a rare case of SS which manifested as rapidly progressive dementia with impaired gait. A careful neurological evaluation is recommended in the global assessment of SS patients and immunosuppression should be attempted before a rapidly progressive cognitive decline.

103 - IMUNOSSUPRESSÃO NA ARTRITE REUMATÓIDE - O OUTRO LADO DA MOEDA

Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Tiago Beirão[1], Joana Aleixo[1]
[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Introdução: O tratamento da Artrite Reumatóide (AR) baseia-se na utilização precoce de fármacos imunossupressores modificadores da atividade da doença. No entanto, estas terapêuticas não são desprovidas de riscos.
Caso clínico: Mulher de 78 anos com antecedentes de AR, artroplastia total do joelho direito, hérnia do hiato esofágico e cataratas. Apresentou ab initio poliartrite crónica e simétrica das pequenas articulações das mãos, atualmente com 30 anos de evolução e sem seguimento há 4 anos, estando em remissão sob deflazacort 30mg/dia. Foi submetida previamente a infiltrações intra-articulares, tratamento com sais de ouro, hidroxicloroquina, cujas dose e duração não sabe precisar, e posteriormente metotrexato 15 mg por via oral. Na primeira observação, além de queixas de dorsalgia mecânica e de poliartralgias de ritmo misto, destacava-se a presença de linfedema exuberante das pernas e pés com hiperqueratose, associado a várias lesões satélite em placa, violáceas, não descamativas, com limites imprecisos, dispersas pelos membros inferiores e a úlcera cutânea da perna direita, a causar marcada limitação na deambulação. Ao exame físico de realçar também acentuação da cifose dorsal e deformidades dos punhos, mãos e pés compatíveis com AR. Foi internada para estadiamento da doença, respetivas sequelas e comorbilidades e para avaliação especializada do linfedema. Revendo o historial clínico, percebeu-se que a doente teria o diagnóstico de Sarcoma de Kaposi há 7 anos, altura em que estaria sob metotrexato e corticóide, e que teria interrompido a radioterapia e o seguimento em consulta de Oncologia por autoiniciativa. Realizou biópsia cutânea que confirmou o diagnóstico de Sarcoma de Kaposi, atribuído à imunossupressão farmacológica. Analiticamente com anemia microcitica e hipocrómica (Hb 10g/dL) ferropénica, PCR e VS normais, hipogamaglobulinemia (IgG 468,0mg/dL e IgM 33,4mg/dL), diminuição de albumina (4.5g/dL) e proteínas totais (5.5g/dL), dislipidemia (colesterol total 240mg/dL, LDL 132mg/dl e triglicerídeos 224mg/dL), défice de vitamina D (17nmol/L) com hiperparatiroidismo secundário (98.2pg/mL), perfil glicémico normal, ausência de proteinúria, serologias de HBV, HAV, CMV, EBV, toxoplasmose, HSV 1/2 e HZV sugestivas de contacto prévio, de HCV, HIV, HTLV I/II, parvovirus e sífilis negativas e Igra, Mantoux e pesquisa de Strongyloides stercoralis negativos. TC do tórax sem doença pulmonar intersticial ou lesões ativas de tuberculose. Estudo radiográfico com múltiplas fraturas vertebrais dorsais e lombares, osteoartrose generalizada, anquilose direita do carpo, diminuição bilateral da interlinha radiocárpica, subluxação das MCF e IFP, polegar em Z e compactação do retropé. Densitometria óssea com valores compatíveis com osteoporose (T-score -2.5 na coluna lombar e T-score -4.4 no colo do fémur). Como a doente se encontrava em remissão clínica foi diminuída progressivamente a dose de deflazacort até 12mg/dia durante o internamento sem flare articular. Foi otimizada a analgesia, ajustados os défices vitamínicos e iniciado tratamento da osteoporose com denosumab.
Conclusão: O Sarcoma de Kaposi é uma doença angioproliferativa decorrente da infeção por vírus Herpes humano 8. Apesar de comumente associado a infeção por HIV, pode ter outras formas, como a associada a imunossupressão, cujo tratamento passa pela redução da mesma. Este caso retrata um exemplo raro dos riscos inerentes à imunossupressão crónica, de como estes podem condicionar o tratamento da AR e as consequências de anos de corticoterapia sem supervisão.

105 - PARANEOPLASTIC SYSTEMIC LUPUS ERYTHEMATOSUS - DESCRIPTION OF A RARE CASE

Ana Raposo[1], Rita Santa Comba[2], João Lagoas Gomes[1], Tiago Meirinhos[1]
[1]Rheumatology Unit, Centro Hospitalar do Tâmega e Sousa, Penafiel, Portugal, [2]Unidade de Saúde da Ilha Terceira, Angra do Heroísmo, Portugal

Background: The expression of neoantigens and the anti-tumor immune response associated with malignancy may elicit an immune response against self-antigens expressed by cancer cells leading to the development of rheumatologic syndromes in predisposed individuals. Despite the description of several types of malignancies presenting with joint, muscle, and soft tissue manifestations, lupus-like paraneoplastic syndromes are a rare entity. It is more frequently described in association with hematologic malignancies, such as hairy cell leukemia, non-Hodgkin's lymphoma, and solid tumors like breast, ovarian and lung cancer.
Clinical Case: A 62-year-old male construction worker, with a history of smoking and peripheral venous insufficiency, was referred to our rheumatology department 2 weeks after transurethral resection of the urinary bladder, performed due to an intra-vesical lesion identified on an ultrasound exam. The patient presented lower urinary tract symptoms for more than 3 months.

One month previous to the tumor identification, during winter months, the patient developed a photosensitive malar rash, papulosquamous skin lesions in UV- exposed areas of the upper extremities (figure 1) and episodes of Raynaud´s phenomenon involving the fingers. No other symptoms were reported. On physical examination, there was no arthritis, cutaneous thickness was normal and there were no digital ulcerations or muscle weakness. Blood analysis identified: positive antinuclear antibodies by indirect immunofluorescence (title of 1:160 with homogenous pattern), leukopenia (total leukocyte count of 3.150 x 10^3/ µL); positive anti-dsDNA antibodies by FEIA (34 UI/mL) and direct Coombs test positivity; antiphospholipid and anti-ENA antibodies were negative. The histological analysis of the tumor confirmed a low-grade papillary urothelial carcinoma. A thoracic, abdominal and pelvic computed tomography scan was performed and did not document distant metastases or the presence of interstitial lung disease. A diagnosis of systemic lupus erythematosus (SLE) was made and the patient was treated with steroids and hydroxychloroquine.
Discussion: Although the association between lupus and cancer is not fully understood, several clinical cases have been previously reported. The association of SLE with bladder cancer was not previously reported. In this case report, due to the temporal relationship between the onset of both disease, it was considered that this may represent a paraneoplastic syndrome.

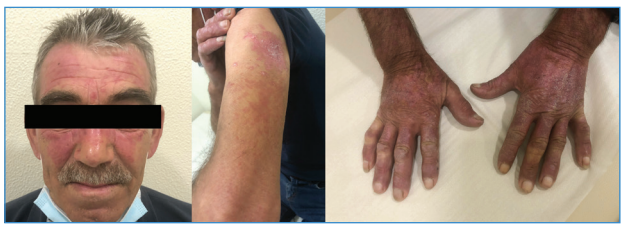


Figure 1.

107 - MACROPHAGE ACTIVATION SYNDROME AS FIRST PRESENTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS

Ana Martins[1, 2], Daniela Santos Oliveira[1, 2, 3], Beatriz Samões[4], Frederico Rajão Martins[5], Diogo Guimarães da Fonseca[4], 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, [3]Faculdade de Medicina da Universidade do Porto, Departamento de Medicina e CINTESIS, Porto, Portugal, [4]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [5]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal

Introduction: Macrophage activation syndrome (MAS) is a potentially life-threatening complication of rheumatic disease characterized by fever, pancytopenia, liver failure, coagulopathy, and neurologic symptoms and is thought to be caused by uncontrolled activation and proliferation of T lymphocytes and macrophages, leading to widespread hemophagocytosis and cytokine overproduction.
Case report: A previously healthy 20-year-old female presented with daily fever in the last 2 weeks, fatigue, anorexia, weight loss, polyarthralgia of the hands and dry mouth. She denied symptoms suggestive of infection, night sweats or swollen lymph nodes. Physical examination revealed normal vital signs except high temperature (38.8ºC), pallor of the mucous membrane and skin, mild tenderness of proximal interphalangeal joints without swelling or deformities. Heart and lung auscultation, abdominal and neurological examination were normal. Small red and painful patches were observed on her fingers. No axillary, cervical and inguinal adenopathy or peripheral edema were noticed. Laboratory workup showed pancytopenia (Hb 8.4 g/dL, WBC 1630/µL, platelet 108000/µL), normal ESR and CRP, normal renal function, mild hypoalbuminemia, high AST and ALT, high ferritin (5075 ng/mL), hypertriglyceridemia (357mg/dL) and hypofibrinogenemia (150mg/dL). High LDH, low haptoglobin, positive direct coombs test, absence of schistocytes on the peripheral blood smear and reticulocytopenia were also observed. Urine analysis was normal. Blood cultures and serological tests for infectious diseases were negative. Autoimmune workup showed positive ANA

(1:1280, homogenous pattern), anti-dsDNA (>800 IU/mL), anti-Sm, anti-SS-A and anti-nucleosome antibodies and low complement levels (C3 54mg/dL, C4 8mg/dL). CT scan of chest, abdomen and pelvis showed cervical, axillary and retroperitoneal lymphadenopathy, hepatosplenomegaly with no focal lesions. No signs of infection or malignancy. Biopsy of axillary node showed non-specific reactive lymphadenitis. Bone marrow aspiration and biopsy were performed but the sample was inappropriate. Soluble CD25 in serum was collected only 10 days after starting therapy. However, the level remained elevated (1187 U/mL, N 158-623).

New-onset of Systemic Lupus Erythematosus associated with MAS was diagnosed. The patient started hydroxychloroquine 400mg daily, intravenous methylprednisolone pulse (1g daily for 3days), followed by 1mg/kg daily of oral prednisolone and cyclosporine 3mg/kg daily. Prophylaxis for Pneumocystis jiroveci, calcium and vitamin D supplementation were also started.

A significant clinical and laboratory improvement was noticed with resolution of fever and constitutional symptoms, improvement of pancytopenia, hyperferritinemia, hypertriglyceridemia and hypofibrinogenemia. The patient was discharged after 3 weeks, with steroid tapering. At one year follow-up, a complete resolution of pancytopenia and other laboratorial parameters was observed. Anti-dsDNA was negative and complement levels were normal. Steroid was stopped and the patient continued taking hydroxychloroquine 400 mg daily and cyclosporine 2.5 mg/kg daily.

Conclusion: MAS is rarely associated with SLE and the incidence is 0.9-4.6%. The mainstay of treatment is steroids. Concomitant use of other agents, such as etoposide, cyclosporine, high-dose IV immunoglobulin and anakinra is useful in patients with severe, corticosteroid-resistant or refractory MAS. Early diagnosis is crucial since mortality rates remain very high in untreated cases.

108 - SÍNDROME DE ERASMUS – A PROPÓSITO DE UM CASO CLÍNICO

Ana Martins[1, 2], 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

Introdução: A silicose resulta da inalação de poeira de sílica e caracteriza-se por fibrose pulmonar irreversível. Esta patologia tem sido associada ao desenvolvimento de outras patologias, tais como a esclerose sistémica (ES). A ES associada à exposição à sílica é indistinguível da ES idiopática, ocorrendo cerca de 5 a 15 anos após a exposição inicial e principalmente em situações de exposição mais intensa.

Caso clínico: Homem de 44 anos, pedreiro desde os 14 anos, fumador, sem antecedentes de relevo, foi referenciado à consulta de reumatologia por fenómeno de Raynaud com vários anos de evolução, dispneia para grandes esforços e anticorpos antinucleares positivos. Negou outra clínica sugestiva de conectivite nomeadamente alopécia, úlceras orais ou genitais, serosite, artralgias de ritmo inflamatório, lesões cutâneas, queixas secas, mialgias, fraqueza muscular, alterações da sensibilidade, disfagia ou úlceras digitais. Referiu pirose e episódios de regurgitação.

Ao exame objetivo, apresentava discreta esclerodactilia, sem puffy hands, pitting scars, úlceras digitais, telangiectasias faciais ou microstomia. A auscultação cardiopulmonar e a palpação abdominal não revelaram alterações. A força muscular estava preservada nos 4 membros. Não se objetivou artrite periférica, edema periférico ou alterações da sensibilidade.

Analiticamente, apresentava hemograma, função renal e hepática sem alterações. Os parâmetros inflamatórios estavam normais. O estudo imunológico revelou anticorpos antinucleares positivos, num título 1/1000 e padrão nucleolar e o painel de esclerose sistémica demonstrou positividade para o antiNOR 90. A videocapilaroscopia do leito ungueal documentou a presença de capilares ectasiados e hemorragia do 4º dedo da mão esquerda, sem megacapilares ou áreas avasculares. A tomografia computorizada de tórax de alta resolução documentou a existência de micronódulos com distribuição aleatória de predomínio nos lobos superiores e segmentos superiores dos lobos inferiores, com densidade elevada e padrão de distribuição característica de silicose. Para além disso, foram identificados múltiplos gânglios hilares e mediastínicos em número superior ao habitual. As provas funcionais respiratórias com capacidade de difusão de monóxido de carbono não revelaram alterações. O ecocardiograma transtorácico também não revelou alterações, nomeadamente a nível da PSAP ou outros sinais indiretos que pudessem sugerir hipertensão pulmonar.

Foi realizado o diagnóstico de ES e silicose. O doente iniciou pentoxifilina 400 mg 2x/dia, nifedipina 30 mg/dia e calcifediol 0.266 mg/mês. Após o início da terapêutica instituída e cessação tabágica, observou-se uma melhoria do fenómeno de Raynaud e da dispneia.

Conclusão: A síndrome de Erasmus, descrita inicialmente em 1957, consiste na associação entre a exposição à sílica (com desenvolvimento ou não de silicose) e o desenvolvimento de ES. Quando comparada com a ES idiopática, a ES associada à exposição à sílica parece surgir mais frequentemente em homens, apresentar maior predisposição para envolvimento pulmo-

nar e positividade para o anticorpo anti-Scl-70 e menor taxa de sobrevida. Assim, consideramos que a exposição à sílica deve ser inquirida a todos os doentes com ES, uma vez que tem implicações no prognóstico. Para além disso, a cessação desta exposição pode conduzir a uma estabilização da progressão da doença ou até, eventualmente em determinados casos, a uma melhoria.

109 - PULMONARY SARCOIDOSIS AND ANTI-SRP IMMUNE-MEDIATED NECROTIZING MYOPATHY: AN UNCOMMON ASSOCIATION

Ana Martins[1, 2], Frederico Rajão Martins[3], Beatriz Samões[4], Rafaela Nicolau[5], Eva Mariz[1, 2], 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, [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]Rheumatology Unit, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Background: Immune-mediated necrotizing myopathy (IMNM) is characterized by acute/subacute and severe proximal muscle weakness, myofiber necrosis with minimal inflammatory cell infiltrate, infrequent extra-muscular involvement and presence of specific antibodies, such as anti-signal recognition particle (anti-SRP) and anti-3-hydroxy-3-methylglutaryl coenzyme A reductase. Sarcoidosis is a multisystemic inflammatory disease of unknown etiology characterised by non-caseating granulomas predominantly in the lungs and intrathoracic lymph nodes. The coexisting IMNM and sarcoidosis has only been reported in 2 previous case reports.
Case report: A 49-year-old woman, previously healthy, non-smoker, presented with severe proximal muscle weakness in the upper and lower limbs in the last 2 months. She also complained about myalgias, Raynaud's phenomenon, anorexia and weight loss. No fever, skin lesions, arthralgia, respiratory, cardiovascular or neurologic complaints. No previous infectious disease, introduction of new drugs or family history of neuromuscular disease.

Laboratory workup showed an elevated CK (15894 U/L), myoglobin (3490 ng/mL), aldolase (145 U/L), aspartate transaminase (AST 513 U/L), and alanine transaminase (ALT 360 U/L). Normal complete blood count, inflammatory markers, renal and thyroid function. Angiotensin-converting enzyme (ECA) level was slightly increased. Results for myositis specific and associated autoantibodies showed positive anti-SRP. Serologic tests for multiple infectious diseases were negative. Electromyography showed abnormal spontaneous muscle activity and a diffuse myopathy, more prominent in the proximal muscles. Magnetic resonance imaging revealed extensive edema of the biceps, triceps, gluteus, quadriceps, hamstrings and paraspinal muscles. Muscle biopsy showed profound myopathic features with numerous necrotic fibers, without significant inflammatory cellular infiltrates and with diffuse overexpression of MHC class I products.

A diagnosis of IMNM was made. Given the association between inflammatory myopathies and malignancy, an investigation was performed. Computed tomography scan of the chest, abdomen, and pelvis was normal except for prominent mediastinal and bilateral hilar lymphadenopathy. Endobronchial ultrasound-transbronchial needle aspiration was performed and biopsy showed non-necrotizing epithelioid cell granulomas with no evidence of malignancy and negative stains and cultures. Hence, a diagnosis of stage I pulmonary sarcoidosis was made.

Patient was started on intravenous methylprednisolone pulse (1g daily, 3 days), followed by 1 mg/kg/day of oral prednisolone and intravenous immunoglobulin (0.4 g/kg/day, 5 days). Prophylaxis for Pneumocystis jiroveci, calcium and vitamin D supplementation were also started. After 25 days, she was discharged on a tapering dose of steroids and exercise-based rehabilitation. Rituximab 500 mg IV on days 1 and 15 was started. At 6-month follow up, patient had a significant improvement in muscle strength and normalization of myoglobin, aldolase, AST and ALT. CK level was almost normal (302 U/L).
Conclusion: Exclusion of other potential causes of granulomatous inflammation is a key before making the diagnosis of sarcoidosis. Muscle biopsy was important in this case, once sarcoidosis can cause myopathy. However, the lack of histological findings suggestive of chronic sarcoid myopathy and the presence of a specific antibody for IMNM makes the diagnosis unlikely. The association between sarcoidosis and other forms of muscle disease warrants further research.

110 - HERPES ZOSTER COMO POSSÍVEL EFEITO SECUNDÁRIO DO DENOSUMAB

Ana Martins[1, 2], Sofia Pimenta[1, 2], Lúcia Costa[1, 2]

[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

Introdução: O denosumab é um anticorpo IgG2 monoclonal humano que tem como alvo o ligando do recetor ativador do fator nuclear kappa-B (RANKL) ao qual se liga com elevada afinidade e especificidade, prevenindo a ativação do seu recetor, RANK. Este anticorpo ao inibir a interação RANK/RANKL, inibe a formação, função e sobrevivência dos osteoclastos,

reduzindo assim a reabsorção óssea no osso cortical e trabecular. Poucos casos de infeções oportunistas foram reportados previamente, onde se inclui o herpes zoster que apresenta uma incidência aproximada de 1.6 casos/100.000 pessoas por ano.

Caso clínico: Mulher de 90 anos, com antecedentes de hipertensão arterial, fibrilação auricular, insuficiência cardíaca classe I (segundo a escala da New York Heart Association), doença renal crónica estadio 3b e síndrome vertiginoso, seguida em consulta de Reumatologia por polimialgia reumática encontrando-se medicada com prednisolona 7.5 mg/dia, carbonato de cálcio + Colecalciferol 1250 mg +400 UI/dia e calcifediol 0.266 mg/mês. Para avaliação do risco fraturário, realizou densitometria óssea que documentou baixa densidade mineral óssea do colo do fémur e fémur total (0.785 e 0.921, respetivamente), com correspondente T-score de -1.6 e -0.7. Tendo em conta o risco elevado de fratura calculado pelo FRAX e a existência de disfunção renal, optou-se por iniciar denosumab 60 mg subcutâneo de 6 em 6 meses. Dois dias após a primeira toma, a doente desenvolveu múltiplas vesículas dolorosas no hemitórax direito, limitadas a um dermátomo torácico, sugestivas de infeção por herpes zoster.

Foi observada no serviço de urgência e medicada com valaciclovir 1000 mg de 8/8 horas, durante 7 dias e tramadol + paracetamol 37.5 + 325 mg em SOS. Apresentou boa evolução, com resolução das lesões herpéticas e, posteriormente, da dor neuropática.

Face à ausência de sintomatologia sugestiva de processo neoplásico, estudos endoscópicos recentes sem alterações e face à estreita relação temporal com a administração de denosumab, foi assumida infeção por herpes zoster secundária à administração de denosumab. O fármaco foi suspenso e desde então, a doente não teve novos episódios de infeção por herpes zoster.

Conclusão: Ainda que seja um evento raro, a infeção/reativação do herpes zoster pode ocorrer após a administração de denosumab. A coexistência de outros fatores de risco para infeção por herpes zoster, como acontece neste caso clínico, nomeadamente a idade avançada e o uso de corticoides, confere um risco acrescido e merece particular atenção. À semelhança do caso descrito, a maioria das infeções por herpes zoster são auto-limitadas, apresentando resolução espontânea, estando indicado o tratamento antivírico em situações de maior gravidade e para uma resolução mais célere.

122 - CASO CLÍNICO - MELORREOSTOSE

Mariana Rocha Sebastião[1], Luís[1], Carolina Furtado[1], Tomás Fontes[1], David Silva[2], Teresa Novoa[1]

[1]Serviço de Reumatologia, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal, [2]Imagiologia, Hospital Internacional dos Açores, Lagoa, Portugal

Introdução: A Melorreostose é uma doença hiperostótica esclerosante, com uma prevalência de 0.9 casos por milhão de habitantes, cuja etiologia continua por esclarecer. Estudos recentes estabeleceram relação com a mutação somática de ganho-de-função MAP2K1 encontrada nos tecidos afetados. É caracterizada por um crescimento linear anormal do osso cortical que afeta maioritariamente o esqueleto apendicular. A dor, deformidade e diminuição da amplitude articular, alterações da sensibilidade e atrofia cutânea, constituem-se como as manifestações mais frequentes da doença. O diagnóstico resulta da combinação da clínica e imagiologia, sendo patognomônica imagem em "cera de vela derretida" em radiografia convencional.

Caso clínico: Homem de 48 anos, enviado à consulta de Reumatologia por tumefação da face dorsal da mão direita. O quadro clínico inicia-se aos 16 anos de idade, precisamente com aparecimento de tumefação, de consistência pétrea, no dorso do 3º dedo da mão direita, provocando deformação do dedo, na ausência dor, alterações da mobilidade, da sensibilidade ou atrofia cutânea, e sem sintomas referente a outros órgãos e sistemas. Aos 21 anos, ocorre tumefação semelhante no 4º dedo da mesma mão e interrupção do crescimento ungueal do 3º e 4º dedos. Quatro anos após, é observável nova tumefação na face dorsal dos metacarpos. Aos 30 anos com queixas limitação da mobilidade do cotovelo e do punho, com incapacidade de realizar extensão completa do antebraço e do punho. No último ano com surgimento de dor mecânica à flexão do antebraço

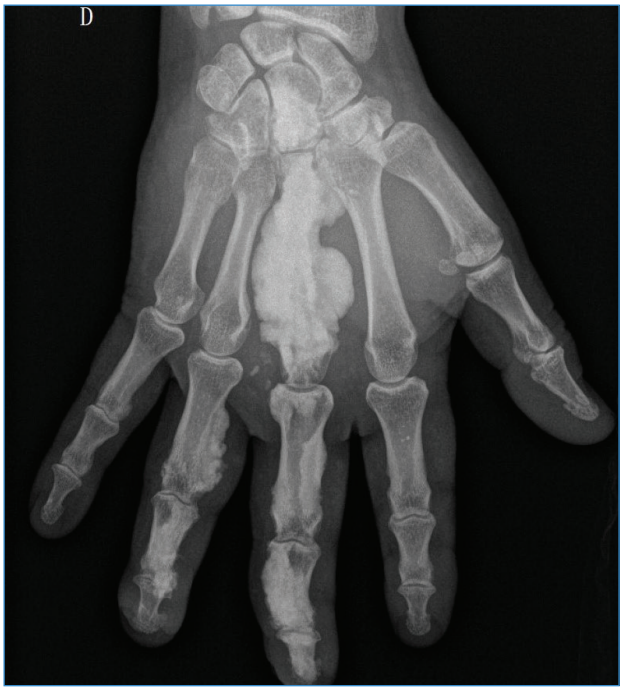


Figura 1. Radiografia da Mão Direita - sinal de "Cera de Vela Derretida"

e flexão dos dedos. Analiticamente todos os marcadores de formação e reabsorção óssea apresentam-se sem alterações, nomeadamente P1NP, CTX, Cálcio e Fósforo séricos, Cálcio na Urina de 24 horas, Razão Cálcio/Creatinina na Urina e Fosfatase Alcalina. As radiografias efetuadas aos segmentos do esqueleto envolvidos mostram espessamento denso, segmentar e ondulante da cortical, organizado e sem irregularidade periosteal, seguindo os esclerótomos, como que "atravessando articulações" sem comprometer o intervalo articular - "sinal do gotejamento da vela" ou de "cera de vela derretida" - com ausência de fraturas destes segmentos.

O doente mantém-se com tratamento analgésico, aguardando estudo genético.

Conclusão: Dada a baixíssima prevalência da doença, a descrição de casos clínicos na literatura é naturalmente reduzida, pelo que assume particular relevância a descrição de novos casos, na confrontação de causas etiológicas, de estudo genético e até do tratamento dirigido que, para já, é centrado no controlo sintomático. Existem, no entanto, casos clínicos publicados que descrevem melhoria sintomática com bifosfonatos.

123 - TACROLIMUS IN REFRACTORY ANTISYNTHETASE SYNDROME – A CASE REPORT

Ana Catarina Duarte[1], Ana Cordeiro[1]
[1]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal

Introduction: Antisynthetase syndrome (ASyS) is defined by the association of inflammatory myopathy, interstitial lung disease (ILD), arthritis, Raynaud's phenomenon (RP), mechanic's hands and the presence of anti-aminoacyl-tRNA-synthetase antibodies. Disease onset can be acute and potentially fatal. There are no specifically approved medications for ASyS, and the drug choice is based on organ involvement and severity of the manifestations. Steroids remain the first-line therapy, but the addition of a steroid-sparing agent is frequently needed. Tacrolimus (tac) has demonstrated to be effective in ASyS, especially in refractory or severe forms of related-ILD. Herein, we report a patient with ASyS, with refractory ILD and myopathy treated with tac in association with rituximab, after incomplete response to cyclophosphamide (CYC).

Case presentation: A 35-year-old previously healthy male presented to Rheumatology clinic with inflammatory arthralgia of both hands, feet and knees, without evidence of arthritis. He had cracked skin on his fingertips, which he associated to his occupation. He had no RP nor muscle weakness. Blood tests had positive antinuclear antibodies (1/640) with positive anti-Jo1 and anti-Ro52 antibodies. Muscle enzymes were normal, as well as muscle magnetic ressonance imaging, chest computed tomography (CT) and echocardiogram.

After 18 months of follow-up he was admitted into hospital with fever and acute, progressively worsening dyspnea, with partial respiratory failure. Physical examination documented proximal muscle weakness. Blood tests had leukocytosis, elevated inflammatory markers and muscle enzymes. Chest CT documented diffuse

Table 1.

	1st appointment	1st flare	After 2 months of CYC	2nd flare	After 3 months of 1st RTX + MTX	3rd flare	Day 0 tac	After 3 months of 2nd RTX	After 10 months of RTX + tac
PDN (mg/dL)	0	0	60	50	40	15	20	12.5	7.5
CK (UI/L)	[80]	4629	368	3195	150	4204	100	181	230
AST (UI/L)	54	323	46	201	15	170	16	19	25
Myoglobin (ng/mL)	<21	4122	310	707.5	88		41.8	64	-
Troponin T (ng/L)	-	-	-	3056	318	478	144	104	121
LDH (UI/L)	-	964	-	785	216	587	189	181	216
ESR (mm/hr)	36	113	91	120	53	98	83	-	32
CRP (mg/dL)	0.42	17.1	1.9	22.3	0.99	7.98	2.85	1.05	0.62
FVC (%)	101.7	-	50.7	-	-	-	-	-	95
DLCO (%)	73.4	-	25	-	-	-	-	-	58
MIP (%)	-	-	25	-	-	-	-	-	73
MEP (%)	-	-	30.5	-	-	-	-	-	32

CYC- cyclophosphamide; RTX – rituximab; MTX – methotrexate; tac – tacrolimus; PDN – prednisolone; CK – creatine kinase; AST – aspartate aminotransferase; LDH - lactate dehydrogenase; ESR - erythrocyte sedimentation rate; CRP – C-reactive protein; FVC – forced vital capacity; DLCO - diffusing capacity for carbon monoxide; MIP - maximal inspiratory pressure; MEP - maximal expiratory pressure

ground glass opacities and consolidation. Pneumonia was assumed and the patient started large-spectrum IV antibiotics. Culture exams were negative. After 4 days of treatment, respiratory failure progressed with need for invasive ventilation. An ASyS flare was assumed and he started IV methylprednisolone (1g/day, 3 days), followed by prednisolone (PDN) 1mg/kg/day, and IV CYC (NIH protocol), with benefit.

Nearly one month after the third CYC administration, under PDN 0.75/mg/kg/day, he developed fever, myalgia, worsening dyspnea and fatigue. Complementary exams revealed new increase in muscle enzymes and normal chest X-ray. At this point refractory myositis was assumed and the patient switched to rituximab (RTX; 2x1g, 2 weeks apart) in association with SC methotrexate 20mg/week. He kept follow-up in outpatient clinic, with PDN tapering and supplementary oxygen therapy suspension.

Twenty-two weeks after RTX administration the patient was readmitted into hospital with fever and acute dyspnea, with severe partial respiratory failure and need for invasive ventilation. Complementary exams showed new increase in muscle enzymes and chest CT documented new bilateral, peripheral, ground glass opacities, with subpleural sparing, consistent with a non-specific interstitial pneumonia pattern. The patient received IV methylPDN (1g/day, 3 days), followed by PDN 1mg/kg/day, and CYC (EuroLES) in association with RTX. There was clinical, radiographic and analytical improvement, and the patient was discharged.

After completing the new induction treatment with CYC, the patient started maintenance therapy with tac (0.075mg/kg/day) in association with RTX. Ten months later he is asymptomatic, working with no restrictions, under PDN 7.5 mg/day and without new flares.

Conclusion: Despite little experience in daily clinical practice, tac seems to be an emerging drug for treating refractory ILD and myositis in ASyS.

127 - OVERLAPPING ANTISYNTHETASE SYNDROME AND RHEUMATOID ARTHRITIS: A RARE ASSOCIATION

Maria Helena Lourenço[1, 2], Ana Bento da Silva[1, 2], Jaime C. Branco[1, 2, 3], Inês Silva[2, 3]

[1]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [2]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [3]NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

Introduction: Antisynthetase (AS) syndrome is a rare condition characterized by myositis, interstitial lung disease, arthritis, Raynaud's phenomenon, fever, mechanic's hands, and a positive AS antibody. Overlapping AS syndrome with other diseases, especially rheumatoid arthritis (RA), is extremely rare, and most of the evidence comes from case reports.1

We present a case of a man with a 7-year-diagnosis of AS syndrome, in clinical remission, who developed an articular flare of the disease and that after a thorough investigation we assumed an overlap syndrome with rheumatoid arthritis.

Clinical Case: 42-year-old man diagnosed with AS syndrome in 2018 (myositis with proximal affection, symmetric and nonerosive polyarthritis, interstitial lung disease with micronodular pattern, Raynaud phenomenon and a positive anti-Jo1 antibody, with both negative anti-citrullinated peptide antibody – anti-CCP - and rheumatoid factor - RF). In remission with prednisolone (PDN) 5mg and azathioprine 200mg/day (hypersensitivity pneumonia with methotrexate) until March 2021. Discontinuation of medication by his own until 2 months ago, when he starts a symmetrical and additive polyarthritis (shoulders, wrists, small joints of the hands, tibiotarsal joints) without others AS syndrome features (Disease Actvity Score - DAS28 – of 5.14 – high disease activity). Blood analysis showed C-reactive protein 1.73mg/dL and erythrocyte sedimentation rate 23 mm/h. X-ray and ultrasound exams did not show joints erosions. PDN was restarted in a 20-30mg/day, along with mycophenolate mofetil (MFM) 2g/day and immunoglobulin (2g/kg/total) to cover joint, muscle and pulmonary issues of AS syndrome activity. MFM was switched to leflunomide 20mg/day because of active polyarthritis. Further investigation detected a new onset elevation of both anti-CCP antibody (306.9 ng/ml) and RF (29.8 UI/mL). He started rituximab in May 2022.

Discussion: Overlapping syndromes are a rare entity in rheumatic diseases and describe conditions where patients meet criteria for the diagnosis of more than one rheumatic diseases. The association between AS syndrome and RA is only described in some case reports, presenting more often as a RA that later develops features of AS.2 AS syndrome flares can start with only articular activity, but in this case, the patient fulfilled the 2010 American College of Rheumatology/European League Against Rheumatism classification criteria for RA. Positive anti-CCP antibodies positive predict a more erosive and refractory arthritis for which rituximab seems to be a good therapeutical approach. This case enhances the importance of not focusing on an only diagnosis, allowing physicians to notice rare conditions and implement a fast and appropriate treatment.

REFERENCES

1. Yasmine M, Saoussen M, Alia F, Mariem S, Kmar O, Souebni L, et al. Antisynthetase Syndrome and Rheumatoid Arthritis: A Rare

Overlapping Disease. Ann Clin Case Rep. 2020; 5: 1860.
2. Meyer A, Lefevre G, Bierry G, et al. In antisynthetase syndrome, ACPA are associated with severe and erosive arthritis: an overlapping rheumatoid arthritis and antisynthetase syndrome. Medicine (Baltimore). 2015;94(20):e523. doi:10.1097/MD.0000000000000523

128 - NECROTIZING MYOCARDITIS AS AN INITIAL PRESENTATION OF AN EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS

Maria Helena Lourenço[1, 2], Rita Amador[3], Ana Bento da Silva[1, 2], Laura Gago[2], Ana Teodósio Chícharo[4], Mariana Santos[2], Inês Silva[2, 5], Jaime C. Branco[1, 2, 5], Maria Manuela Costa[2]

[1]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [2]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [3]Department of Cardiology, Hospital de Santa Cruz, Centro Hospitalar Lisboa Ocidental, Lisboa, Portugal, [4]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal, [5]NOVA Medical School, Faculdade de Ciências Médicas, 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. Rarely, it can affect the cardiovascular system and 15% of patients have eosinophilic myocarditis severe enough to cause cardiomyopathy. 1

Case presentation: A 55-year-old female with a prior history of adult-onset asthma, rhinitis, nasal polyps, and symmetric polyarthritis of the small joints of the hands presented to the emergency room with acute chest pain, fatigue, nausea, and vomiting. Physical examination revealed hypotension, tachycardia, and prolonged capillary refill time. Blood lactate were hight, as so cardiac troponin (2184 ng/L). Electrocardiogram showed a sinus rhythm and a QS pattern from V2 to V4 leads and a transthoracic echocardiogram revealed a left ventricular ejection fraction of 35% with diffuse hypokinesia, no right ventricular dysfunction and a moderate pericardial effusion. A cardiogenic shock was assumed, requiring noradrenaline and dobutamine support. Emergent coronary angiography was performed with no significant epicardial stenoses. The patient was then referred to the Cardiac Intensive Care Unit for further workup. A cardiac magnetic resonance showed suggestive findings of acute myocarditis. Right ventricular endomyocardial biopsy was performed, revealing signs compatible with necrotizing eosinophilic myocarditis; the procedure was complicated with a cardiac tamponade, requiring pericardiocentesis. Moreover, blood analysis revealed peripheral eosinophilia (1900 cells/µL) and elevated erythrocyte sedimentation rate (82 mm/h). Following the development of dry cough, a chest computed tomography presented perilobular inflammation of the inferior pulmonary lobes bilaterally. The case was then discussed with our Rheumatology team and, considering that constellation of features, a clinical diagnosis of EGPA was assumed. Pulses of methylprednisolone 1g/day for 3 days were administered, followed by prednisolone 1mg/kg/day. Cyclophosphamide was also started (CYCLOPS protocol)2. A major clinical and analytical improvement was observed, allowing the weaning of vasopressor and inotropic support and posterior discharge from the Cardiology ward and transference to the Rheumatology department for further therapeutical attitudes. During a 5-month period of follow-up, there was no recurrence of symptoms.

Conclusion: EGPA diagnosis may impose a clinical challenge due to the variety of systemic manifestations. Cardiac involvement in EGPA is rare but one of the most potentially severe and fatal complications. Immunosuppressive drugs are the mainstay of treatment, and a multidisciplinary approach is essential to improve outcomes, as it was seen in this case report, where an aggressive treatment and approach allowed an almost complete recovery.

REFERENCES

1. Firestein, G. S., & Kelley, W. N. (2009). Kelley's textbook of rheumatology. Philadelphia, PA: Saunders/Elsevier.
2. Harper, Lorraine et al. "Pulse versus daily oral cyclophosphamide for induction of remission in ANCA-associated vasculitis: long-term follow-up." Annals of the rheumatic diseases vol. 71,6 (2012): 955-60. doi:10.1136/annrheumdis-2011-200477

135 - TUMOR TENOSSINOVIAL DE CÉLULAS GIGANTES DO PUNHO, A PROPÓSITO DE UM CASO CLÍNICO

Ana Margarida Correia[1], Carla Campinho Ferreira[1], Emanuel Costa[1], Diogo Esperança Almeida[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: Os tumores tenossinoviais de células gigantes são lesões benignas raras que resultam da proliferação da membrana sinovial articular, bursal ou da bainha tendinosa, podendo localizar-se intra ou extra-articularmente. Clinicamente, a sua apresentação é inespecífica, variando quer com a sua localização, quer com a sua progressão, nomeadamente se se trata de um tumor localizado ou difuso. É uma patologia de etiologia não esclarecida, que acomete predominantemente

indivíduos do sexo feminino, apresentando um pico de incidência entre a 3ª e 5ª décadas de vida, sendo mais raramente descrita em crianças. O tratamento atual baseia-se na exérese cirúrgica da lesão tumoral, estando descrita uma elevada taxa de recorrência da lesão.

Caso clínico: Doente do sexo masculino com 17 anos de idade, sem antecedentes pessoais de relevo, referenciado à consulta de Reumatologia por tumefação do punho esquerdo. O doente referia surgimento de tumefação indolor na face dorsal do punho esquerdo há cerca de 4 anos, com crescimento progressivo, sem qualquer outra sintomatologia associada. Ao exame físico apresentava tumefação nodular na face dorsal do carpo esquerdo, com cerca de 3 cm de diâmetro, dolorosa à palpação, sem outros sinais inflamatórios e sem limitação das amplitudes articulares do punho; no restante exame articular não havia evidência de artrite ou limitação da mobilidade articular. Foi realizada ecografia do punho esquerdo que revelou hipertrofia sinovial intercárpica exuberante, com mais de 3 cm de extensão nos planos axial e coronal, com sinal de doppler marcado, sem tenossinovite dos compartimentos extensores e flexores do carpo. Analiticamente, o doente apresentava VS 5 mm/s, PCR < 0,5 mg/L, fator reumatóide e anti-CCP negativos, HLA-B27 negativo, hemograma, função renal e hepática sem alterações e serologias víricas (anti-HCV, anti-HIV e anti-HBV) negativas. O doente realizou também RMN do punho esquerdo, que revelava na face dorsal do punho uma volumosa formação lobulada de contornos irregulares e densidade difusamente heterogénea, desde o plano da face dorsal do hamato até ao plano da articulação do escafoide com o trapézio e o trapezoide, associada a pequenas erosões marginais no escafoide. Estes achados levantavam como principal hipótese diagnóstica um processo proliferativo sinovial de natureza indeterminada. Para melhor caracterização etiológica da lesão descrita, optou-se pela realização de biópsia sinovial da tumefação do carpo esquerdo. Os exames microbiológico, micobacteriológico e micológico da membrana sinovial foram negativos. Em termos histológicos, a lesão era constituída por células mononucleadas com fenótipo histiocitário, sem atipia acentuada, macrófagos xantelasmizados, por vezes com pigmento hemossidérico e células gigantes multinucleadas de tipo osteoclasto, sendo os aspetos morfológicos compatíveis com tumor tenossinovial de células gigantes. O doente foi orientado para consulta de Ortopedia, tendo sido inscrito para cirurgia de exérese tumoral.

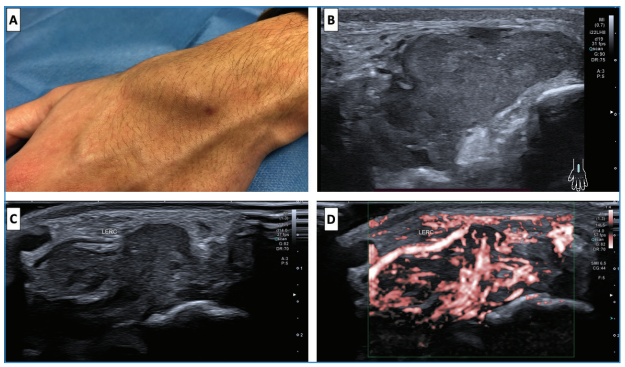


Figura 1. A – Tumefação da face dorsal do punho esquerdo; B a D – Ecografia do punho com hiperrofia sinovial intercárpica e sinal de doppler marcados

Conclusão: Apesar de raro, o tumor tenossinovial de células gigantes deve ser considerado no diagnóstico diferencial de uma tumefação articular ou peri-articular, particularmente na ausência de patologia reumática inflamatória.

137 - EFEITOS ADVERSOS RAROS DOS AINES EM DOENTES COM PATOLOGIA REUMÁTICA INFLAMATÓRIA CRÓNICA SOB AGENTES BIOLÓGICOS

Bruna Silva[1], Bruno Miguel Fernandes[1], Miguel Bernardes[1, 2], Hugo Fernandes Diniz[3], José Silvano[3], Pedro Rodrigues Pereira[4], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar de São João, Porto, Portugal, [2]Faculdade de Medicina da Universidade do Porto, Porto, Portugal, [3]Serviço de Nefrologia, Centro Hospitalar de São João, Porto, Portugal, [4]Serviço de Anatomia Patológica, Centro Hospitalar de São João, Porto, Portugal

Introdução: A retenção de sódio é o efeito adverso renal mais frequentemente associado à toma de anti-inflamatórios não esteróides (AINEs). No entanto, outros efeitos adversos mais incomuns poderão ocorrer.

Caso clínico 1: Doente do sexo masculino, 45 anos, com antecedentes de espondilartrite axial radiográfica com sete anos de evolução, medicado com golimumab mas com história prévia de toma de AINEs durante 4 anos (etoricoxib 90mg id durante 2 anos e celecoxib 200mg id durante 4 anos); e cirrose biliar primária (CBP) diagnosticada aos nove meses de tratamento com golimumab e sob ácido ursodesoxicólico. Aos 2 meses de retoma de tratamento com golimumab e após interregno terapêutico de 12 meses, por apresentar astenia marcada com um mês de evolução, agravamento da função renal (creatinina sérica de 3.06 mg/dL; basal de 1.39 mg/dL) e proteinúria (720mg/24h), assim como IgM aumentada (3510 mg/dL), foi enviado ao serviço de urgência de Nefrologia, tendo-se optado por internamento para estudo. O estudo complementar não mostrou evidência de patologia hematológica e a biópsia renal mostrou achados compatíveis com nefrite tubulo-intersticial, que se considerou em provável associação à toma crónica de AINEs. Contudo, já os tinha descontinuado há cerca de 15 meses por elevação da creatinina sérica, não se podendo excluir nefrite intersticial associada à própria CBP ou nefrite intersticial não-granulomatosa associada ao golimumab, dada a estreita relação temporal com o seu

reinício e à semelhança de outros casos raros mas já descritos com adalimumab, etanercept e infliximab. Foi medicado com prednisolona oral 1mg/kg/dia em esquema de desmame durante dois meses e com melhoria marcada da azotemia (creatinina 1.99 mg/dL) e da proteinúria. Atualmente mantem seguimento em Nefrologia com doença renal crónica estadio 3, estável, e sem proteinúria; não retomou o golimumab e mantem o ácido ursodesoxicólico.

Caso clínico 2: Doente do sexo feminino, de 67 anos, com antecedentes de artrite reumatóide estabelecida, medicada com rituximab, naproxeno 500mg/dia e prednisolona 5mg/dia e hipertensão arterial controlada. Por apresentar hematoproteinúria de novo em sumária de urina e proteinúria de 2g em urina de 24h, foi pedida observação por Nefrologia. O AINE foi suspenso e o estudo complementar não mostrou outras alterações de relevo (nomeadamente pesquisa de substância amilóide na gordura abdominal negativa, imunologia negativa, eletroforese de proteínas séricas e imunoglobulinas normais). Iniciou lisinopril 5mg/dia e foi submetida a biópsia renal que demonstrou achados compatíveis com glomerulonefrite membranosa PLA2R (phospholipase A2 receptor) negativa (marcador relacionado com glomerulonefrite membranosa idiopática). A proteinúria reduziu após a suspensão do naproxeno (<500mg/24h), pelo que foi assumido um provável quadro de glomerulonefrite membranosa relacionada com AINE, mantendo-se com função renal normal e sob vigilância.

Conclusão: A nefrite tubulo-interesticial e a glomerulonefrite membranosa são efeitos adversos raros relacionados com a toma de AINEs. As alterações do sedimento urinário (proteinúria e hematúria) são as manifestações mais frequentes, sendo possível haver alterações na função renal. Como é possível verificar nestes dois casos, é importante que o Reumatologista esteja sensibilizado para a importância da monitorização da função renal e do sedimento urinário nos doentes sob AINEs, visto que estes efeitos adversos, ainda que raros, poderão beneficiar de atitudes terapêuticas atempadas com o intuito de prevenir o dano renal irreversível.

144 - ARTRITE E GOTA – UM CASO DE SOBREPOSIÇÃO DAS DUAS PATOLOGIAS

Laura Gago[1, 2], Margarida Lucas Rocha[3], Maria Helena Lourenço[1, 2], Alexandre Sepriano[1], Maria Manuela Costa[1], Jaime C. Branco[1, 2], Ana Filipa Mourão[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]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [4]Comprehensive Health Research Center (CHRC), Universidade NOVA de Lisboa, Portugal, Lisboa, Portugal

A Gota e a Artrite Reumatoide (AR) são artropatias inflamatórias com prevalência semelhante no nosso país, sendo que cada vez mais são descritos casos de doentes com AR que posteriormente desenvolvem gota.

Descrevemos um caso de um doente do sexo masculino, com diagnóstico prévio de AR seropositiva e erosiva, com posterior desenvolvimento de gota.

Homem, 82 anos, com antecedentes de insuficiência cardíaca com fração de ejeção reduzida, hipertensão, doença pulmonar obstrutiva crónica com necessidade de oxigenoterapia no domicílio, excesso de peso e AR do idoso com fator reumatoide (283 mg/dL) e anticorpo anti peptídeo citrulinado cíclico (127.9) positivos, medicado com metotrexato (MTX) oral 15 mg/semana e leflunomida (LFN) 20 mg/dia, furosemida 20 mg e ivabradina 7,5 mg, que cumpria com irregularidade. Apresentava quadro com 4 meses de evolução de dor, edema e rubor do pé direito associado a úlcera do dorso do pé sem febre associada. Recorreu ao serviço de urgência (SU), sendo medicado com ciprofloxacina, sem resolução do edema e da dor e com aparecimento de nova úlcera no dorso do pé direito, retomando ao SU um mês depois. Radiologicamente existia nova erosão sobre o osso navicular direito. Nas análises apresentava anemia normocrómica normocítica (NN), proteína C Reativa (PCR) de 3.91 mg/dL, sem outras alterações. Foi realizado novo ciclo de antibioterapia com amoxicilina+ácido clavulânico e claritromicina com resolução das úlceras, porém, manteve edema e rubor do pé direito, motivo pelo qual foi internado no serviço de Reumatologia. Mantinha anemina NN, PCR 0.93 mg/dL, ácido úrico de 4.7 mg/dL e hemoculturas negativas. A ecografia articular do pé direito mostrava sinovite da segunda e terceira metatarsofalângica, hipertrofia da membrana sinovial da tibio-társica direita, sinal de duplo contorno e a presença de tofos gotosos (Figura 1). Foi também realizada RMN do pé direito com osteomielite dos ossos do tarso e artrite séptica talo-navicular. Procedeu-se a artrocentese diagnóstica, observando-se ao microscópio óptico de luz polarizada cristais de monourato de sódio. No exame cultural foi isolado Staphylococcus aureus oxacilina sensível. Tendo em conta os resultados descritos, assumiu-se como hipóteses diagnósticas artrite gotosa, osteomielite dos ossos do tarso e artrite séptica talo-navicular. Foi iniciada terapêutica com colchicina 1 mg/dia e antibioterapia com flucloxacilina 2 g. endovenosa, mantendo terapêutica com MTX e LFN. Verificou-se melhoria do edema e dos sinais inflamatórios associados, todavia, houve franco agravamento da função renal com desenvolvimento de síndrome cardio-renal, anasarca e agravamento da insuficiência respiratória com necessidade de diálise e ventilação não invasiva. Contudo, o doente não respondeu à terapêutica e viria a falecer.

Alguns autores defendem que a gota e a AR são mutuamente exclusivas, porém, já foram descritos vários casos de doentes com ambas as patologias.

Num estudo desenvolvido por Merdler-Rabinowicz et al. os principais fatores de risco identificados para desenvolvimento de gota em doentes com AR foram o sexo masculino, idade avançada, hipertensão arterial e excesso de peso. Neste estudo, os doentes apresentavam níveis de ácido úrico dentro dos parâmetros de normalidade, o que levou à formulação da hipótese que em doentes com AR o limiar de ácido úrico para desenvolvimento de uma crise gotosa é mais baixo.

Este caso alerta-nos para a importância de um diagnóstico precoce de gota, para instituição correta de terapêutica e assim evitar destruição articular e outras complicações.

147 - ARTRITE PSORIÁTICA E SARCOIDOSE: UM DESAFIO DIAGNÓSTICO

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

Introdução: A sarcoidose é uma doença inflamatória multissistémica, com apresentação clínica heterogénea, caracterizada pela formação de granulomas não caseosos. Afeta mais frequentemente o pulmão mas qualquer órgão pode estar envolvido. O envolvimento articular pela sarcoidose num doente com psoríase pode levar erradamente ao diagnóstico de artrite psoriática.
Caso clínico: Doente de 32 anos, sexo feminino, natural da Alemanha, seguida na consulta de Reumatologia por artrite psoriática, refere astenia, febre de predomínio vespertino, hipersudorese noturna e tosse seca com 2 meses de evolução e início após um mês da suspensão de etanercept, por pretender engravidar. Como antecedentes pessoais destaca-se psoríase do couro cabeludo, diagnosticada aos 14 anos de idade. Aos 18 anos, refere quadro de artralgias dos punhos, joelhos e tornozelos, lombalgia de ritmo inflamatório, astenia e febre. O fator reumatóide era negativo, as radiografias osteoarticulares não apresentavam alterações de relevo e foi descartada etiologia infeciosa. Estabelecido o diagnóstico de artrite psoriática no seu país de origem, a doente foi medicada com etanercept com melhoria clínica. Na investigação do quadro atual, apresentava anemia (Hb 10,1g/dL), linfopenia (900/mm), elevação de parâmetros inflamatórios, das enzimas de colestase (GGT 482U/L, FA 228U/L) e da enzima conversora da angiotensina (261U/L). Realizou TC do tórax que evidenciou múltiplos micronódulos com distribuição peri-bronco-vascular e formações ganglionares mediastínicas milimétricas. O estudo do lavado broncoalveolar revelou uma razão linfócitos CD4+/CD8+ elevada e exame cultural negativo. Foi realizada biópsia transbrônquica que evidenciou granulomas epitelioides. A ressonância magnética abdominal e a biópsia hepática foram sugestivas de sarcoidose hepática. Estabeleceu-se o diagnóstico de sarcoidose multissistémica e iniciou-se prednisolona 1mg/kg/dia e posteriormente etanercept com melhoria clínica franca.
Conclusão: Embora o acometimento articular na sarcoidose seja raro, é essencial um elevado nível de suspeição para esta entidade na presença de manifestações sistémicas. Tratando-se a sarcoidose de uma grande mimetizadora, o diagnóstico inicial deve ser oportunamente revisto.

148 - ARTRITE PSORIÁTICA E SÍNDROME DE ALPORT: UMA ASSOCIAÇÃO INESPERADA

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

Introdução: As alterações do sedimento urinário em doentes com poliartrite podem ser um verdadeiro desafio clínico. Podem traduzir uma manifestação da doença reumática subjacente, um efeito adverso das terapêuticas utilizadas ou uma doença renal primária. O síndrome de Alport é uma doença hereditária da membrana basal glomerular, da cóclea e do olho, resultante de mutações no gene do colagénio do tipo IV.
Caso clínico: Doente do sexo masculino, de 34 anos, recorreu à consulta de Reumatologia por poliartralgias das pequenas articulações das mãos e pés com um ano de evolução. Ao exame objetivo apresentava artrite das

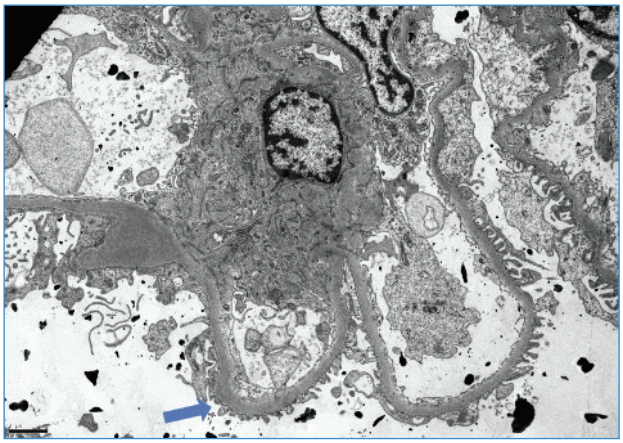


Figura 1. Microscopia eletrónica de biópsia renal a evidenciar irregularidade da espessura da membrana basal com áreas de membrana fina e áreas espessadas com esboço de lamelação (seta azul)

articulações interfalângicas distais, dactilite do 3º dedo do pé direito e lesões de psoríase em placas, com atingimento do couro cabeludo e unhas. Dos exames complementares de investigação diagnóstica apresentava elevação da proteína C reativa. A tipagem HLA-B27, o anticorpo antipeptídeo citrulinado e o fator reumatóide eram negativos. Foi estabelecido o diagnóstico de artrite psoriática, segundo os critérios de classificação CASPAR, e o doente foi medicado com sulfassalazina 2g/dia, com franca melhoria clínica. Durante o primeiro ano de seguimento apresentava persistentemente hematúria microscópica e proteinúria subnefrótica (1,4g/24h). Os restantes dados laboratoriais revelaram creatinina sérica, complemento e imunoglobulinas dentro dos parâmetros da normalidade. Os anticorpos antinucleares e anti-citoplasma de neutrófilos foram negativos. Foi realizada biópsia renal cuja microscopia eletrónica mostrou achados compatíveis com síndrome de Alport (Figura 1). O teste genético revelou uma variante molecular no gene COL4A5. Foi referenciado a consulta de Otorrinolaringologia tendo sido realizado o diagnóstico de hipoacusia neurossensorial moderada bilateral. O doente foi medicado com losartran, sem agravamento da proteinúria e mantendo função renal preservada em 3 anos de seguimento.

Conclusão: A associação da artrite psoriática e do síndrome de Alport não está previamente descrita. Permanece por esclarecer se existe um mecanismo fisiopatológico comum entre ambas ou se apenas se trata de um epifenómeno.

149 - GRAVIDEZ, INIBIDOR XA E PARTO – SIMBIOSE FRACTURÁRIA.

Daniel Carvalho[1], Lídia Teixeira[1], Tiago Sardinha[2], Daniano Caires[3], Pedro Martins[3], Gabriela Fernandes[3], Margarida Faria[1], Agna Neto[1], Ricardo Figueira[1], Alberto Quintal[1], Mário Rodrigues[1]

[1]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [2]Centro de Saúde do Caniço, Funchal, Portugal, [3]Hospital Central do Funchal (Serviço de Medicina Física e De Reabilitação), Funchal, Portugal

A osteoporose é um dos efeitos adversos graves associados à utilização prolongada de fármacos anticoagulantes como a heparina não fracionada (HNF) e as heparinas de baixo peso molecular (HBPM). Em mulheres com o diagnóstico conhecido de trombofilia, as HBPM são utilizadas no tratamento e profilaxia de eventos trombóticos durante a gravidez. A farmacocinética e longa semivida das HBPM possibilitam uma menor frequência de administrações comparativamente à HNF, o que torna a sua utilização mais prática durante a gestação. A gravidez e amamentação são também períodos particularmente sensíveis à perda de DMO.

Mulher de 26 anos, caucasiana, sem hábitos toxifílicos ou restrições alimentares conhecidas, apresenta história de trombose venosa da veia ilíaca comum às 25 semanas de gestação, secundária a trombofilia hereditária por mutação do fator V de Leiden em homozigotia, pela qual realizou anticoagulação terapêutica com Enoxaparina s.c. 60mg q12h durante 15 dias e posteriormente, com Tinzaparina sódica 10,000 s.c. U.I./dia. Às 39 semanas de gestação, foi submetida a cesariana eletiva, tendo sido necessária a execução da manobra de Kristeller. No pós-parto imediato, referiu dor intensa dorso-lombar com pouca resposta à analgesia. Três dias após a alta hospitalar, recorreu ao serviço de urgência por manutenção do quadro álgico e marcada incapacidade funcional, tendo realizado ressonância magnética (RM) da coluna dorso-lombar, que detetou fracturas recentes dos corpos vertebrais de D8, D9 e D10, tendo ficado internada ao cuidado da Neurocirurgia para controlo álgico. Dois meses mais tarde, regressou ao serviço de urgência por agravamento da dorso-lombalgia sem trauma associado, tendo realizado nova RM que revelou fracturas vertebrais compressivas recentes de D12 a L5, bem como a presença de microfracturas trabeculares sagradas bilaterais de predomínio direito. Na avaliação da densitometria óssea apresentava um Z-score do colo femoral e coluna lombar de -1.5 e -3.3, respetivamente. Tendo sido nesta altura referenciada à consulta de Reumatologia. Objetivamente, apresentava agravamento da cifose dorsal. Após exclusão de causas como o hipertiroidismo, hiperparatiroidismo, hipercortisolismo, doença celíaca, doença renal ou hematológica, bem como utilização de outros fármacos, assumiu-se osteoporose fracturária secundária à HBPM e gravidez, tendo sido iniciada terapêutica osteoformadora com Teriparatida s.c. 20µg/dia e proposta a utilização de ortótese dorso-lombar (Spinomed®). A doente iniciou ainda seguimento em consulta de Medicina Física e de Reabilitação e Neurocirugia. Atualmente, após 6 meses de Teriparatida e 3 meses de fisioterapia, a doente apresenta melhoria significativa do controlo álgico, sem ocorrência de novas fracturas.

Na maioria dos casos não é possível estabelecer uma relação causa-efeito linear no diagnóstico da osteoporose secundária, tendo em conta o seu caráter multifatorial. Apesar da raridade da osteoporose fracturária em mulheres jovens, os riscos de perda de massa óssea na gravidez e amamentação são conhecidos, e em casos como este, são agravados pela utilização de anticoagulantes por longos períodos. Ao apresentar este caso clínico pretendemos alertar para o risco particular das HBPM, nomeadamente a Tinzaparina, que embora mais recente do que a Enoxaparina, não é isenta de efeitos adversos, requerendo a mesma atenção para a sua utilização no período gestacional e puerperal.

152 - ESOPHAGEAL INVOLVEMENT IN BEHÇET DISEASE: A CASE REPORT

Maria Pontes Ferreira[1], Francisca Guimarães[1], Hugo Parente[1], Emanuel Costa[2], Catarina Soares[1], Soraia Azevedo[1], Daniela Peixoto[1], Filipa Teixeira[1], Daniela Santos-Faria[1], Carmo Afonso[1], José Tavares-Costa[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: Behçet's disease (BD) is a rare multisystemic disorder characterized with mucosae ulcers, and eye and skin lesions1. Gastrointestinal (GI) involvement, although variable, has been reported in 1-60% of patients, however esophageal involvement is thought to be rarer2. We present a case report of a patient with BD with serious esophageal involvement.
Clinical Case: A 53 years old women followed in our department since 2013 with the diagnosis of BD, with mucocutaneous involvement with oral, genital and cutaneous, folliculitis and recurrent thrombophlebitis, that had an episode of hematemesis and rectal bleeding. The colonoscopy did not find any changes in colon mucosae, but the esophagogastroduodenoscopy (EGD) showed esophageal ulcer (histology with inflammatory polymorphic infiltrates and some eosinophiles, and no vasculitis lesions). She begun treatment with 125mg/day of azathioprine, 12mg/day of deflazacort and 1mg/day of colchicine. She was referred to gastroenterology consult, and was treated with sucralfate and proton pump inhibitor (PPI). Since she had improved of the mucocutaneous and gastroesophageal symptoms, glucocorticoids were progressively reduced and azathioprine increased to 150mg/day.

However, in 2020 she manifested heartburn and pyrosis. Esophageal ulceration and esophagitis were found on EGD, biopsies showed esophageal mucosae with lymphocyte exocytosis and polymorphonuclear cells, with microabcess formation and negative PAS stain; therefore, chronic esophagitis with signs of activity, in the clinical context of Behçet's disease.

In November 2021, the dysphagia worsens and she is unable to eat solid food, therefore she was submitted to new EGD, which was unable to be completed due to progressive luminal narrowing and mucasae friability. Barium esophageal transit study revealed important stenosis of proximal esophagus. For this reason, she is scheduled for esophageal dilation, in the perspective of improving her quality of life.
Discussion: Behçet's disease is a multisystemic vasculitis, characterized by recurrent mucosal ulceration, ocular lesions like uveitis, gastrointestinal tract ulceration, skin lesions, pulmonary, cardiovascular, neurologic and also musculoskeletal involvement1. Low intestinal involvement is rather frequent (up to 60%), but not esophageal involvement; the latter can lead to severe complications such as stenosis, perforation and fistula formation2,3. The treatment of esophageal BD remains uncertain but it is usually responsive to PPIs, mesalazine, and/or colchicine2. In this clinical case, the patient had a history of chronic esophagitis and ulceration, despite medical treatment, that eventually led to clinically significant proximal esophageal stenosis.

REFERENCES

1. Zou J, Shen Y, Ji DN, Zheng SB, Guan JL. Endoscopic findings of gastrointestinal involvement in Chinese patients with Behcet's disease. World J Gastroenterol. 2014 Dec 7;20(45):17171-8. doi: 10.3748/wjg.v20.i45.17171.
2. Yi SW, Cheon JH, Kim JH, Lee SK, Kim TI, Lee YC, Kim WH. The prevalence and clinical characteristics of esophageal involvement in patients with Behçet's disease: a single center experience in Korea. J Korean Med Sci. 2009 Feb;24(1):52-6. doi: 10.3346/jkms.2009.24.1.52. Epub 2009 Feb 28.
3. Ideguchi, Haruko; Suda, Akiko; Takeno, Mitsuhiro; Miyagi, Rumiko; Ueda, Atsuhisa; Ohno, Shigeru; Ishigatsubo, Yoshiaki (2014). Gastrointestinal manifestations of Behçet's disease in Japan: a study of 43 patients. Rheumatology International, 34(6), 851–856. doi:10.1007/s00296-013-2838-5

154 - DISSEMINATED GONOCOCCAL INFECTION VS REACTIVE ARTHRITIS

Daniel Carvalho[1], Lídia Teixeira[1], Nancy Faria[2], Francisco Barreto[3], brandon allan[4], Margarida Faria[1], Agna Neto[1], Daniano Caires[5], Ricardo Figueira[1], Alberto Quintal[1], Mário Rodrigues[1]
[1]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [2]Hospital Central do Funchal (Serviço de Infecciologia), Funchal, Portugal, [3]Serviço de Medicina Interna, Hospital Central do Funchal, Funchal, Portugal, [4]Serviço de Medicina Física e De Reabilitação, Hospital de Cascais, Dr. José de Almeida, Cascais, Portugal, [5]Hospital Central do Funchal (Serviço de Medicina Física e De Reabilitação), Funchal, Portugal

Disseminated gonococcal infection (DGI) results from the bacteremic spread of Neisseria gonorrhoeae. DGI develops in up to 3% of patients infected with Neisseria gonorrhoeae, and comprises two major clinical syndromes: localized septic arthritis and arthritis-dermatitis syndrome, the latter characterized by the triad of polyarthralgia, tenosynovitis and skin lesions. Reactive arthritis (ReA) is an inflammatory syndrome that can result from gastrointestinal and genitourinary infections, and includes an acute inflammatory arthritis, conjunctivitis, and dysuria. Nevertheless, the majority of patients don't present with this classic triad. Gonococcal infection is seldom associated with ReA.

A 45-year-old man, with a 12 years personal history of syphilis, presented to the emergency department

reporting a 2-week history of dysuria and urethral purulent discharge, followed by the development, after 1 week, of bilateral knee, right ankle and mid-foot pain. At this time, the patient was discharged with oral cefuroxime and azithromycin, and reassessed 5 days later showing no signs of improvement, with new symptoms of malaise and diaphoresis, resulting in admission to the Infectious diseases ward. Lab results revealed leukocytosis (12.3x103/µL), elevated ESR 63mm and protein C-reactive 48.53mg/L. He started on i.v. ceftriaxone after blood and urine sampling, as well as urethral and rectal swabs for culture and nucleic acid amplification testing (NAAT). Due to persistent arthralgia after 3 days of antibiotic, Rheumatology department was contacted. Physical examination showed swollen and painful joints involving both knees, right tibiotarsal and metatarsophalangeal, as well as tenosynovitis of 4th and 5th flexors tendons of the right hand. Closer examination didn't reveal any visible skin lesions. Ultrasound showed synovitis of both knees, right ankle and right metatarsophalangeals, as well as tenosynovitis of the 4th and 5th flexor tendons, both right peroneal tendons and posterior tibialis. Arthrocentesis of the right knee revealed translucent and low viscosity synovial fluid with a cell count of 10545 WBC/mm3, predominantly mononuclear type. Neisseria gonorrhoeae DNA was detected in urethral and rectal samples by NAAT, also VDRL was reactive. Chlamydia trachomatis co-infection was excluded. Blood, urine and synovial cultures and NAAT were all negative. DGI was assumed, he completed a 10-day course of i.v. ceftriaxone and oral NSAIDs, with significant clinical and analytical improvement.

On the follow-up visit, 1 month later, the patient presented with arthralgia exacerbation. Objectively, he had several swollen joints including left knee, right tibiotarsal and bilateral metatarsophalangeals, as well as right arm flexors tenosynovitis. He denied any dysuria or urethral discharge. By suspicion of ReA, he started on oral prednisolone for 3 weeks, showing complete clinical remission.

This case raises awareness for sexual transmitted infections, an increasingly public health problem. It´s also relevant because of the diagnostic ambiguity between DGI and ReA, arisen by the return of arthritis after antibiotic completion and later clinical and analytical resolution with prednisolone. Follow-up is important to further clarify the definitive diagnosis.

158 - DACTILITE E INFEÇÃO POR FUSARIUM SPP

Bruna Silva[1], Daniela Santos Oliveira[1], Frederico Rajão Martins[2], Rafaela Nicolau[3], Daniela Cerqueira[4], Miguel Bernardes[1, 5], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar de São João, Porto, Portugal, [2]Rheumatology Department, Hospital de Faro, CHA, Faro, Portugal, [3]Rheumatology Unit, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [4]USF Esposende Norte, Esposende, Portugal, [5]Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Introdução: A dactilite e a distrofia ungueal são manifestações muito frequentes de Artrite Psoríática (Apso). Sabe-se que há um risco aumentado de infeções em doentes com Apso, nomeadamente Osteomielite ou Onicomicose.
Caso clínico: Apresentamos o caso clínico de uma doente do sexo feminino, caucasiana, de 27 anos, com antecedentes pessoais de Doença de Crohn, diagnosticado em 2019 e atualmente sob Ustecinumab 90mg subcutâneo, de 8 em 8 semanas. Previamente à terapêutica atual, realizou Azatioprina, que suspendeu por toxicodermia, e Infliximab, suspenso por suspeita de Lupus Eritematoso Sistémico Induzido (poliartralgias, ANA 1/1000 e Anticorpos Anti-Histonas persistentemente positivos). Recorre à nossa consulta por dor, calor, rubor e edema súbitos do hálux direito com cerca de 2 meses de evolução. Apresentava ainda alterações ungueais em ambos os háluxes, desde há 2 anos. Ao exame objetivo, apresentava sinais inflamatórios exuberantes no hálux direito associado a alterações distróficas da unha, sugestivas de dactilite. Foi realizado estudo analítico que evidenciou: Hemograma e Velocidadade de Sedimentação (VS) sem alterações, Proteína C Reativa (PCR) cerca de 10 mg/L. Após 1 mês, estes achados analíticos mantiveram-se semelhantes. Realizou Ressonância Magnética do pé direito (Anexo 1) com identificação de edema medular ósseo extenso na falange distal, associado a inflamação com origem provável na matriz ungueal, que se apresentava espessa e com inflamação sobreposta. Assim, foram colocadas as hipóteses de Psoríase Ungueal ou eventual etiologia infecciosa (Osteomielite). Por este motivo, foi realizada biópsia óssea, que evidenciou amostra insuficiente para diagnóstico. Foi também realizado exame cultural da amostra colhida na biópsia, o qual não obteve isolamento de microorganismos até à data. Na análise cultural da unha, foi identificado Fusarium spp. Dado estes achados, optou-se por manter Ustecinumab e iniciar Prednisolona 10 mg/dia e tratamento anti-fungico tópico, com melhoria dos sinais inflamatórios do dedo e das alterações ungueais.
Discussão: Este caso clínico remete para uma doente com LES induzido, com dactilite e alterações distróficas ungueais, manifestações muito sugestivas de APso. Contudo, em casos com apresentação exuberante, é necessário excluir osteomielite, pelo que a RMN é fundamental. Caso esta não seja conclusiva, a realização de biópsia óssea e exame cultural devem ser consideradas. Neste caso, a biópsia óssea foi insuficiente para diagnóstico e o exame cultural da biópsia encontra-se neg-

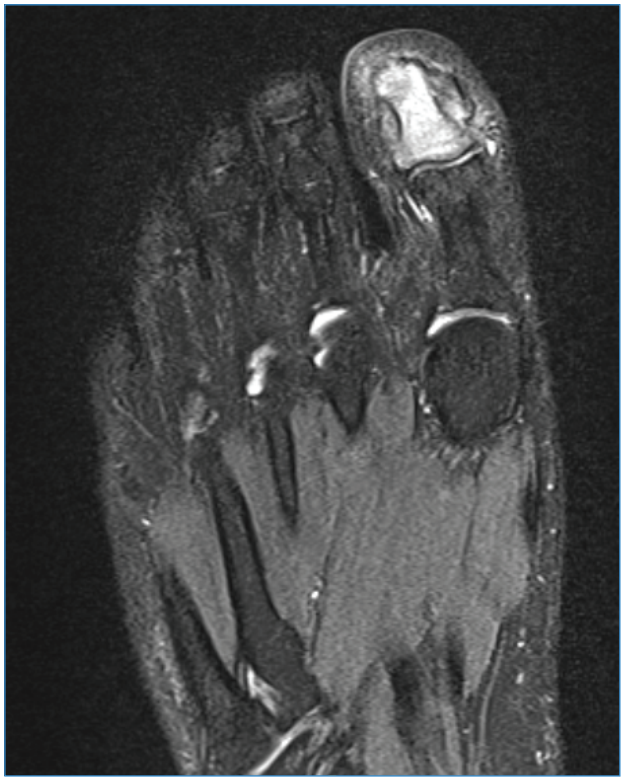


Figura 1. RMN Sequência T2 de pé direito

ativo até à data. Na amostra cultural da unha, isolou-se *Fusarium spp*, frequente em doentes com psoríase[1] e atualmente considerado uma infeção oportunista emergente, que pode causar desde infeção superficial a infeções mais graves e disseminadas em doentes imunodeprimidos[2]. Deste modo, a sua identificação e tratamento precoces tornam-se imprescindíveis. Dado o isolamento do microorganismo apenas na unha, não nos permite inferir que este esteja implicado no quadro clínico, pelo que se optou por retomar o biológico e iniciar Prednisolona associado a anti-fungico tópico.

Conclusões: Em casos selecionados de dactilite é necessário a exclusão de osteomielite. A identificação de *Fusarium* pode ser a causa da distrofia ungueal, ou ser fator confundidor no diagnóstico de psoríase ungueal, sendo que a sua identificação e tratamento precoces evita a progressão para doença mais invasiva.

164 - CARDIOPATIA VENTRICULAR DIREITA INDUZIDA PELA HIDROXICLOROQUINA EM DOENTE COM LÚPUS ERITEMATOSO SISTÉMICO

Inês Santos[1], Vanda Neto[2], Nádia Martins[1], Paulo Monteiro[1], Costa Cabral[2], Lina Carvalho[3]
[1]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [2]Serviço de Cardiologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [3]Serviço de Anatomia Patológica, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

Introdução: A hidroxicloroquina (HCQ) é usada frequentemente no tratamento de várias doenças reumáticas inflamatórias, como é o caso do Lúpus Eritematoso Sistémico (LES). Além das propriedades imunomoduladoras, estão também descritos efeitos antitrombóticos, estando a sua suspensão associada a maior probabilidade de exacerbação da doença. A toxicidade retiniana é a complicação mais comum, com uma prevalência de cerca de 7,5%, ocorrendo geralmente após 5 anos de tratamento. A neuropatia (0,3%), a miopatia (0,3%) e a toxicidade cardíaca estão igualmente associadas ao efeito cumulativo da dose mas estão descritas como ocorrências muito raras. Esta última, pode no entanto aumentar de forma significativa a morbimortalidade nos doentes afetados.

Caso clínico: Doente de 71 anos com antecedentes de LES inativo sob HCQ na dose de 400 mg id desde há 10 anos (dose cumulativa de 1460 g e 6,3 mg/Kg), fibrilhação auricular paroxística, bronquiectasias difusas, hipotiroidismo e osteoporose, recorre ao Serviço de Urgência por episódio de pré-síncope de difícil caracterização. A doente negou dor torácica, palpitações, dispneia, fadiga e edemas periféricos. A tomografia computadorizada crânio-encefálica não apresentou alterações de relevo. Os D-dímeros foram negativos mas apresentou elevação da troponina I de alta sensibilidade (572 ng/L) associada a alterações eletrocardiográficas de bloqueio completo de ramo direito com supradesnivelamento do segmento ST anterior, bem como elevação do NTproBNP (3164 pg/mL). Foram realizados ecocardiograma transtorácico e coronariografia, que não revelaram alterações. Realizou estudo complementar com ressonância magnética cardíaca que objetivou um importante espessamento da parede livre do ventrículo direito com sinais de inflamação difusa local, bem como realce difuso do tipo não isquémico com hipocontratilidade global, a condicionar depressão ligeira a moderada da função sistólica. Para melhor caracterização, foi submetida a biópsia endomiocárdica que concluiu cardiotoxicidade associada à HCQ, que foi entretanto suspensa. Por síncopes de repetição nos 3 meses subsequentes, houve necessidade de colocação de um pacemaker provisório devido à presença de um bloqueio auriculoventricular completo, seguido de um cardiodesfibrilhador implantável (CDI). No mês seguinte, a doente apresentou um novo episódio de síncope no contexto de fibrilhação ventricular, revertido com sucesso pelo seu dispositivo. Iniciou terapêutica com 200 mg de amiodarona id, não se verificando até à data (4 meses após a síncope pós implantação de CDI) o registo de novos eventos.

Discussão: Embora raro, a HCQ pode associar-se a

cardiotoxicidade de forma direta. Apesar de mal compreendida, pensa-se que a sua patofisiologia envolve a disfunção lisossomal através da inibição das enzimas lisossómicas. Esta alteração leva à acumulação cardíaca de produtos metabólicos tóxicos, associando-se a várias manifestações: cardiomiopatia com hipertrofia concêntrica e distúrbios da condução, culminando na insuficiência cardíaca. A dose cumulativa e a duração da terapêutica descritas na literatura são bastante díspares, com respetivos valores medianos reportados de 1235 g e 8 anos, semelhantes ao caso exposto.

Dependendo da gravidade, as manifestações cardíacas podem ser parcial ou completamente reversíveis com a suspensão do antimalárico ou, pelo contrário, irreversíveis e até com desfecho fatal.

Assim, tendo em conta a potencial morbimortalidade associada e a falta de terapêuticas específicas eficazes, o reconhecimento da cardiotoxicidade induzida pela HCQ é fundamental.

165 - DOENÇA RELACIONADA COM IGG4 – UM DESAFIO DIAGNÓSTICO

Inês Santos[1], Nádia Martins[1], Paulo Monteiro[1]
[1]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Introdução: A doença relacionada com IgG4 (DRIgG4) é uma síndrome fibroinflamatória imunomediada. Caracteriza-se pelo aparecimento de massas pseudotumorais e/ou aumento indolor de vários órgãos. A presença de um infiltrado linfoplasmocítico rico em plasmócitos IgG4+ acompanhada por fibrose e flebite obliterativa é altamente sugestiva. Os níveis séricos de IgG4 estão aumentados em 60-70%. A terapêutica com corticoides é eficaz na maioria dos casos.

Caso clínico: Doente do sexo feminino de 73 anos, com antecedentes de hipertensão arterial e dislipidemia, avaliada em consulta de diversas especialidades por quadro de hipogeusia e xerostomia associado a astenia e perda ponderal de 5 Kg em 3 meses, sem outras queixas. Analiticamente, sem alterações: hemograma normal, VS 21 mm/h (<30), glicemia em jejum 86 mg/dL, serologias virais VHC, VHB e HIV não reativas; ECA normal, ANA negativos, dsDNA negativo, C3 e C4 normais, ENA screen negativo (SSA, SSB, entre outros), FR e aCCP negativos, proteinograma eletroforético, B2 microglobulina e LDH normais; IGRA negativo. Realizou ainda estudo complementar com AFP, CEA, CA 19.9 e CA 125 que se revelou negativo, endoscopia digestiva alta que demonstrou pangastrite crónica com predomínio de plasmócitos (não tendo sido efetuado estudo imunohistoquímico) e tomografia computadorizada (TC) toraco-abdomino-pélvica (TAP) que revelou apenas um nódulo pulmonar calcificado. Cerca de 4 meses depois, surgiram múltiplas adenopatias cervicais palpáveis com suspeita de tumefação da glândula submandibular direita. Repetiu TC torácica onde foram identificados dois nódulos pulmonares de novo e realizou TC cervical que confirmou a presença de múltiplas adenopatias com distribuição bilateral e extensa, a destacar a mais volumosa (17 mm) na região submandibular direita, bem como uma imagem nodular expansiva (19 mm) localizada na amígdala direita. A doente veio a ser submetida a amigdalectomia bilateral e linfadenectomia cervical esquerda, cujo estudo anatomopatológico permitiu excluir neoplasia linfoide. O estudo imunohistoquímico confirmou o diagnóstico de linfadenite por IgG4. O doseamento sérico de IgG4 não foi feito numa fase inicial mas foi normal após a corticoterapia. A doente foi encaminhada à consulta de Reumatologia e iniciou terapêutica com prednisolona (PDN) na dose de 20 mg id. Por agravamento clínico na tentativa de redução da dose, houve necessidade de novo aumento temporário, estando atualmente assintomática e a recuperar peso (do total perdido de 15 Kg em 2 anos) sob PDN 7,5 mg id. Fez TC-TAP que confirmou a resolução das linfadenopatias e dos nódulos pulmonares.

Discussão: A DRIgG4 pode constituir um desafio diagnóstico visto mimetizar várias patologias neoplásicas, inflamatórias e infeciosas. No caso exposto, podemos verificar envolvimento ganglionar e provável amigdalino, gástrico e pulmonar. A linfadenopatia é das manifestações mais frequentes, assim como o envolvimento das glândulas salivares. Neste caso, na ausência de confirmação histológica, não é possível excluir este último, o que justificaria as queixas de xerostomia e hipogeusia. Os envolvimentos gástrico e pulmonar também estão reportados com frequência. Pelo contrário, o envolvimento amigdalino é raro.

Em linha com a literatura, verificou-se boa resposta inicial à corticoterapia, estando recidivas descritas na tentativa da sua redução, tal como no caso descrito.

Não tratada, esta patologia pode levar à fibrose e disfunção de órgão, pelo que o seu reconhecimento precoce é essencial.

167 - LOW BACK INFLAMMATORYPAIN IN A PATIENT WITH PERIPHERAL SPONDYLARTHRITIS - NOT ALWAYS SACROILIITIS!

Joao Paulo Vilas-Boas[1, 2], Gisela Eugénio[1, 2], Anabela Barcelos[1, 2]
[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz Health Alliance, Portugal, Aveiro, Portugal

Introduction: Spondylarthritis (SpA) is a group of inflammatory diseases that may have axial or peripheral involvement. Inflammatory back pain (IBP) is the key

clinical symptom of axial SpA. A wide variety of conditions can present with this symptom, and should therefore be considered in the differential diagnosis.

Clinical Case: A 35 years old female followed regularly in a rheumatology outpatient center due to a spondylarthritis HLA-B27 positive with peripheral, enthesopatic and ocular involvement complained about a chronic inflammatory back pain (IBP) with insidious onset that worsened with prolonged rest. She also mentioned a burning sensation in the lumbosacral area that radiated to both knees. Sacroiliac joint provocative tests were negative and no other relevant signs were found on physical examination. The pain was resistant to NSAIDs. Pelvic radiograph was normal. Pelvic MRI showed no signs of sacroiliitis; however, it showed a large intra-osseous sacral lesion with well-defined limits and heterogeneous signal intensity on T2-weighted image, compromising the left S2 root. Biopsy was performed and histologic analysis demonstrated a schwannoma. Later the patient was submitted to surgery to remove the neoplastic lesion and she is recovering at the moment.

Discussion: The differential diagnosis of IBP is extensive and it is crucial to rule out pain sources such as vertebral body compression fractures or bone tumours. Intraosseous schwannomas are rare neoplastic disorders, compromising less than 0,2% of the primary bone tumours, being the most common locations the mandibula and the sacrum. Schwannomas are mostly benign and less than 1% become malignant. Studies show a female dominancy, most often between the 2nd and 5th decades of life. Sacral schwannomas are usually found incidentally or when patients present with pain and neurological symptoms such as lower back pain, numbness or paresthesia. Its early diagnosis is difficult to make because of patient´s' vague aches and minimal symptoms. CT and MRI are the most useful imaging modalities for preoperative diagnosis of this tumour. Local recurrence and transformation to malignancy are very rare. For this reason, the frequently preferred treatment is subtotal removal of the mass.

Conclusion: This case shows that immediate diagnostic assumptions of sacroiliitis shouldn´t be made and an MRI study is important to confirm or exclude the presence of sacroiliitis when no radiographic signs are present. Neoplastic lesions should be considered in the differential diagnosis of IBP, especially when neurologic symptoms are present. Early recognition allows for earlier interventions and thus a better prognosis.

169 - SÍNDROME DE PSEUDO FOSTER-KENNEDY – APRESENTAÇÃO RARA DE ARTERITE DE CÉLULAS GIGANTES

Ana Teodósio Chícharo[1], Mariana Santos[2], Margarida Lucas Rocha[1], Laura Gago[2], Maria Helena Lourenço[2], Ana Bento Silva[2], Carina Lopes[2, 3], Miguel Cordeiro[4], Alexandre Sepriano[2, 3], Jaime C. Branco[2, 3], Manuela Costa[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, [3]CEDOC, NOVA Medical School. Faculdade de Ciências Médicas da Universidade NOVA de Lisboa., Lisboa, Portugal, [4]Oftalmologia, Centro Hospitalar de Lisboa Ocidental - Hospital Egas Moniz, Lisboa, Portugal

Introdução: A Arterite de Células Gigantes (ACG) é uma vasculite caracterizada pela presença frequente de sintomas visuais (20-50%). Não obstante, o atingimento binocular é menos frequente, estando presente em 9% dos doentes na apresentação e em mais 9% após sintomas iniciais monoculares.

A síndrome de Foster Kennedy (SFK) traduz-se na presença de edema do disco óptico com atrofia do nervo óptico contralateral, devido a uma massa intracraniana a nível frontal. A compressão axonal provocada pela lesão ocupando espaço (LOE) leva a atrofia do nervo óptico ipsilateral e o aumento da pressão intracraniana leva ao edema do disco óptico contralateral. A presença destes achados fundoscópicos na ausência de LOE em exames de neuroimagem, define a síndrome de Pseudo Foster Kennedy (SPFK).

Caso clínico: Descrevemos um caso de um homem de 77 anos, com antecedentes de hiperplasia benigna da próstata, que referia quadro com 3 semanas de evolução caracterizado por perda de acuidade visual no olho esquerdo (OE), seguido, uma semana depois, de diminuição da acuidade visual no olho direito (OD), com agravamento progressivo. Simultaneamente, relata cefaleia temporal bilateral, astenia e perda de peso não quantificada. Negava febre, claudicação da mandíbula ou sintomas compatíveis com polimialgia reumática.

Recorre à urgência de Oftalmologia, onde se objetivou acuidade visual de 1/10 no OD e apenas sensível a movimentos da mão no OE. A fundoscopia revelou edema da papila pálido chalky-white no OD e atrofia óptica total pós-NOIA (neuropatia óptica isquémica anterior) no OE. O doente apresentava ainda hipersensibilidade do couro cabeludo na região temporal esquerda e pulsos temporais não palpáveis bilateralmente. Os restantes pulsos eram palpáveis e simétricos e não se auscultavam sopros cardíacos ou vasculares.

Laboratorialmente, a destacar uma anemia normocítica normocrómica (Hb 10.5 mg/dL), leucocitose ligeira (Leuc 11.1x109/L) e elevação dos parâmetros inflamatórios (Vs 62 mm/h, PCR 1.72 mg/dL).

Tendo em conta a clínica sugestiva, foi assumido o diagnóstico de ACG e iniciada terapêutica com pulsos

de metilprednisolona de 1g durante 3 dias consecutivos. Ao 4º dia, iniciou prednisolona 1mg/kg/dia per os (70mg/dia), cálcio, vitamina D, alendronato e cotrimoxazol. O doente melhorou clínica e analiticamente, com resolução da cefaleia. O eco-doppler das artérias temporais foi realizado ao sétimo dia de corticoterapia, não tendo sido identificado o sinal do halo.

Perante os achados na fundoscopia sugestivos do SFK, o doente realizou TC-CE, que não revelou massas intracranianas. Assim, foi feito o diagnóstico de SPFK de causa arterítica, no contexto de ACG, com NOIA arterítica sequencial do OD após o OE.

Teve alta após 9 dias de internamento com acuidade visual de 2/10 no OD e com contagem de dedos a 1 metro no OE. Os parâmetros inflamatórios normalizaram (Vs 16 mm/h, PCR <0.1 mg/dL).

Conclusão: Este caso reforça a multidisciplinaridade inerente ao doente reumático, sendo a colaboração com a Oftalmologia indispensável para a formulação de todas as hipóteses diagnósticas num doente com perda da acuidade visual.

Apesar de o envolvimento ocular ser comum na ACG, os achados fundoscópicos apresentados são muito pouco frequentes, tendo os autores identificado na literatura raros casos semelhantes, sem estimativas de prevalência. Este caso ganha ainda especial relevância pelo diagnóstico diferencial com neoplasias intracranianas num doente com clínica de ACG.

171 - THE EVOLUTION AND IMPACT OF PYCNODYSOSTOSIS ON AN ADULT FEMALE: A 40-YEAR ANALYSIS

Brandon Allan[1], Daniel Carvalho[2], Agna Neto[2], Lídia Teixeira[2], Ricardo Figueira[2], Alberto Quintal[2], Carolina Falcão[1], Mónica Bettencourt[1], Mário Rodrigues[2]

[1]Serviço de Medicina Física e De Reabilitação, Hospital de Cascais, Dr. José de Almeida, Cascais, Portugal, [2]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal

Introduction: Pycnodysostosis is a rare autosomal recessive disorder, with estimated incidence of 1-1.7 per million births, caused by the mutation in the cathepsin K gene. Deficiency of this enzyme causes poor resorption of bone matrix and results in diffuse osteosclerosis. The disease is characterized by short stature, increased bone fragility, acro-osteolysis of terminal phalanges, clavicular dysplasia, delayed closure of the cranial sutures and typical craniofacial dysmorphism. Typical features become more prominent with age and complications may appear over the years.

Clinical Case: We present the case of a 40-year-old Portuguese female, independent in activities of daily living, with previous history of hypothyroidism, hirsutism, migraines, and retinitis pigmentosa. During her late childhood, she was diagnosed with pycnodysostosis through clinico-radiographic features.

Our first contact with the patient was in a rheumatology consultation at the age of 40, due to complaints of bilateral shoulder pain from rotator cuff tendinopathy. A closer look at the history revealed she has had to date 24 non-traumatic fractures since the age of 1. She complained of generalized chronic pain and rated her overall health to be weak.

Physical examination showed several common features of the disease: short stature (140cm; 50kg), brachydactyly (figure 1A), micrognathia, obtuse mandibular angle, prominent nose with convex nasal ridge, and frontal and parieto-occipital bossing (figure 1B). Previous imagiological studies demonstrated acro-osteolysis of the distal phalanges, several sclerotic bone lesions, open fontanelles, thoracolumbar scoliosis, hypo-pneumatized mastoids, and dental anomalies. Bone densitometry was normal.

Due to the rarity of pycnodysostosis and to confirm the diagnosis, we requested a genetic test that detected a pathogenic variant of CTSK: c.436G>C p. (Gly146Arg) in probable homozygosity. Genealogical data was then collected. The patient is an only child born to non-consanguineous parents and is the only known family member with the disease. She is single and has no intention of having offspring.

Figure 1. Typical clinical characteristics of pycnodysostosis.

There are no published treatment or surveillance guidelines, therefore management emphasized multidisciplinary care. Over the past years, she was treated with many craniofacial and orthopedic surgeries, including intramedullary nailing for femoral shaft fractures and pseudoarthrosis repair after metatarsophalangeal fractures. Severe teeth abnormalities have led to multiple dental procedures, one of which complicated with osteonecrosis of the jaw, requiring mandibular reconstruction with a fibular graft. When general anesthesia was applied, intubation was difficult. It is noteworthy that the patient enrolled in numerous rehabilitation programs to regain physical function and had environmental and occupational modifications to increase independence and promote emotional and social well-being. Growth hormone therapy was not instituted during childhood. Bisphosphonate therapy was incorrectly used for a short period of time. Genetic counselling was proposed as well as polysomnography to screen for obstructive sleep apnea.

Conclusion: Our case draws attention to the recognition of this entity and to appropriate follow-up in order to facilitate the prevention and treatment of common complications. This study aims to highlight the phenotypic abnormalities, the radiological signs, the genetic characterization, the therapeutic, the evolutionary features of the disease and its impact on quality of life.

175 - DOENÇA INTERSTICIAL PULMONAR COMO POSSÍVEL FACTOR DE RISCO PARA NEOPLASIA DO PULMÃO EM DOENTES COM ESCLEROSE SISTÉMICA – A PROPÓSITO DE 3 CASOS CLÍNICOS

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

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

Vários trabalhos têm demonstrado um aumento do risco de vários tipos de neoplasia na esclerose sistémica (ES), nomeadamente para a neoplasia do pulmão, documentado de forma consistente em vários estudos, em particular em doentes com doença pulmonar intersticial (DPI).

Os autores apresentam três casos de ES com diagnóstico de neoplasia do pulmão durante o curso da doença. As três doentes eram do sexo feminino, com idades compreendidas entre os 66 e os 76 anos à data do diagnóstico da neoplasia. Todas apresentavam ES com envolvimento cutâneo limitado, envolvimento pulmonar e anticorpos anti-topoisomerase I positivos. O intervalo de tempo entre o diagnóstico de ES e o de neoplasia variou entre 7 meses e 11 anos. Nenhuma das doentes tinha história de tabagismo ou outros factores de exposição.

Doente 1: Diagnóstico de ES aos 66 anos na sequência da investigação de alterações do interstício pulmonar, condicionando insuficiência respiratória (IR) parcial. Em tomografia computorizada (TC) torácica apresentava alterações sugestivas de pneumonia organizativa bilateral, com áreas em favo de mel. Foi inicialmente medicada com glucocorticóides (GC) sistémicos e micofenolato de mofetil (MMF), com agravamento clínico e progressão radiográfica ao fim de 3 meses, pelo que foi alterada terapêutica para ciclofosfamida (CF) endovenosa (EV) mensal. Após 3 ciclos, objectivou-se agravamento da IR, com presença em TC de extensas consolidações bilaterais e inúmeras opacidades nodulares em vidro despolido, algumas cavitadas. Foi submetida a biópsia aspirativa transtorácica (BATT), que revelou adenocarcinoma mucinoso do pulmão. Atendendo ao performance status não foi candidata a quimioterapia (QT), acabando por falecer.

Doente 2: Queixas de fenómeno de Raynaud (FR) desde os 54 anos, e diagnóstico de ES aos 60 anos com envolvimento pulmonar (padrão pneumonia intersticial não específica [NSIP], fibrótica) desde então, condicionando IR parcial crónica. Medicada inicialmente com GC sistémicos e indução com CF EV, seguida de manutenção com MMF e posterior associação de nintendanib. Após 11 anos de doença foi documentado em TC aumento dimensional de nódulo pulmonar do lobo superior esquerdo, pelo que foi submetida a BATT, com histologia compatível com adenocarcinoma do pulmão. A doente foi recusada para cirurgia e radioterapia estereotáxica, tendo iniciado QT. Contudo, verificou-se progressão da neoplasia, tendo a doente falecido cerca de 2 anos e meio após o diagnóstico.

Doente 3: Doente com queixas de FR desde os 25 anos, com diagnóstico de ES aos 69 anos. Envolvimento pulmonar desde o diagnóstico com padrão inicial de NSIP fibrótica e posterior progressão para pneumonia intesrsticial usual, com IR parcial, estando medicada com ácido micofenólico. Sete anos após o diagnóstico inicia quadro de perda ponderal e anemia de agravamento progressivo, sendo posteriormente internada por agravamento agudo da IR. Realizou TC que documentou lesão nodular no lobo superior esquerdo com invasão da parede costal e do mediastino, e nódulo subpleural direito compatível com metástase. Foi submetida a BATT, que revelou carcinoma pavimento-celular do pulmão. Durante o internamento observou-se agravamento da IR, condicionando o óbito.

Com este trabalho pretendemos alertar para o risco de neoplasia do pulmão nos doentes com ES, em particular na presença de DPI. É fundamental que os clínicos se mantenham atentos a sinais e sintomas de novo, podendo ser pertinente nestes doentes a reavaliação com TC tórax, independentemente do agravamento clínico ou funcional respiratório.

176 - SARCOIDOSE NUM DOENTE COM LÚPUS EM REMISSÃO

António Barreto Violante[1], Ana Filipa Rocha Águeda[1], Miguel Guerra[1], José Marona[1], Joana Ramos Rodrigues[1], Margarida Oliveira[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário Cova da Beira, Covilhã, Portugal

Introdução: A sarcoidose é uma doença inflamatória de causa desconhecida caracterizada pela formação de granulomas em vários órgãos e tecidos. Apresenta enorme variedade de manifestações clínicas, muitas delas de instalação insidiosa e baixa especificidade, tendo por isso um diagnóstico diferencial desafiante. As manifestações pulmonares são as mais frequentes, podendo incluir infiltrados e adenopatias hilares. Dentro das manifestações extratorácicas, destacam-se as cutâneas em até um terço dos doentes, nomeadamente eritema nodoso e lesões papulares.

Caso clínico: Apresenta-se o caso de uma doente do sexo feminino, de 54 anos, com antecedentes pessoais de depressão, tremor essencial e diagnóstico de lúpus eritematoso sistémico (LES) desde 1995, na sequência de um internamento por glomerulonefrite classe V com podocitopatia lúpica e desde então seguida em Nefrologia. Atualmente medicada com hidroxicloroquina 200 mg id e ácido acetilsalicílico 100mg id. Sem seguimento prévio em Reumatologia, foi referenciada por quadro de astenia de longa data e, desde há 6 meses, queixas de dispneia para esforços moderados e artralgias das pequenas articulações das mãos e metatarsofalângicas direitas, de ritmo misto. Sem outras queixas na revisão por órgãos e sistemas. À observação inicial apresentava lesões papulares de pequenas dimensões em fundo eritematoso e com descamação ligeira, localizadas aos antebraços, região dorsal alta e orla de implantação capilar. Ao exame reumatológico com dor à palpação da 4ª articulação metatarsofalângica direita. Auscultação cardiopulmonar com discretas crepitações na base esquerda. Sem outras alterações.

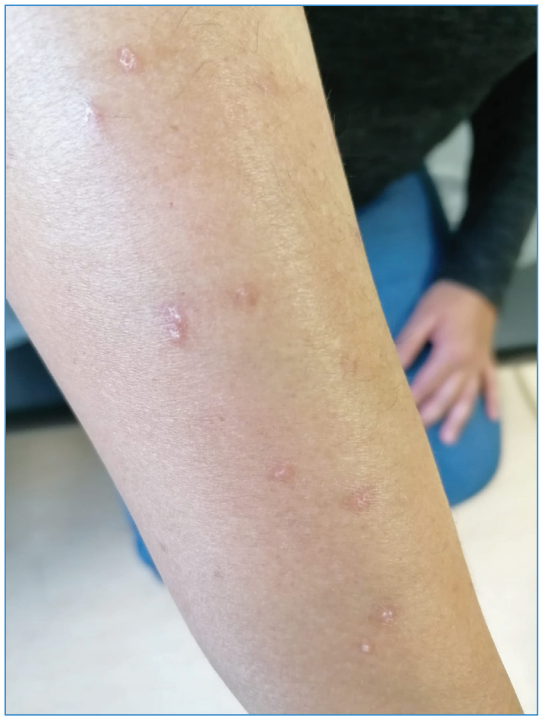


Figura 1. Lesões papulares no antebraço de doente com sarcoidose

Analiticamente com ANAs, anti-dsDNA e ENA's negativos, anticoagulante lúpico positivo em duas medições e anticorpo antiB2 glicoproteína IgG borderline, sem consumo de complemento, função renal preservada, sem proteinúria, e hipercalcemia (9.8 mg/dL). A radiografia do tórax demonstrava um reforço hilar direito, tendo sido solicitada tomografia computorizada do tórax que evidenciou múltiplas adenopatias mediastínicas e subcarinais. Ecocardiograma e provas de função respiratória sem alterações de relevo.

Apesar dos antecedentes de LES, os achados descritos permitiam colocar o diagnóstico diferencial de sarcoidose. Efetivamente, objetivou-se elevação da enzima conversora da angiotensina (134.8 UI/L) e da relação CD4/CD8 no lavado broncoalveolar (10.62) Procedeu-se a broncoscopia e mediastinoscopia, mas as mesmas foram inconclusivas por colheita insuficiente. A tomografia por emissão de positrões revelou atingimento ganglionar múltiplo. Realizou posteriormente biópsia cutânea das lesões papulares que evidenciou granulomas epitelióides compatíveis com o diagnóstico de Sarcoidose.

A doente veio depois a apresentar progressiva melhoria sintomática após o início de corticoterapia sistémica.

Discussão: No caso em discussão a doente apresentava já um diagnóstico prévio de LES, cuja presença levou à não valorização dos sinais e sintomas apresentados de novo, os quais não eram totalmente concordantes com o diagnóstico de base. O caso descrito salienta a importância da observação crítica dos doentes mesmo quando existe um diagnóstico sistémico prévio.

177 - HIPEREOSINOFILIA - PARA ALÉM DAS VASCULITES

Ana Teodósio Chícharo[1], Mariana Santos[2], Margarida Lucas Rocha[1], Laura Gago[2], Maria Helena Lourenço[2], Ana Bento da Silva[2], MJ Gonçalves [2], Manuel Neves[3], Sara Casanova[4], Isabel Antunes[4], Kamal Mansinho[4], Jaime C. Branco[2, 5], Manuela Costa[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, [3]Champalimaud Research, Champalimaud Centre for the Unknown, Lisbon, Portugal, Lisboa, Portugal, [4]Serviço de Doenças Infecciosas - Hospital Egas Moniz, Centro Hospitalar Lisboa Ocidental, Lisboa, Portugal, [5]CEDOC, NOVA Medical School. Faculdade de Ciências Médicas da Universidade NOVA de Lisboa., Lisboa, Portugal

O síndrome hipereosinofílico (SHE) é caracterizado por eosinofilia periférica (>1.500/uL) sustentada por mais de 6 meses, associada a lesão de órgão. É uma doença rara e com uma apresentação heterogénea que pode ter envolvimento multissistémico. Consiste num diagnóstico de exclusão, após o estudo de causas secundárias ou clonais de eosinofilia.

Apresentamos o caso de uma mulher melanodérmica, sem antecedentes pessoais ou familiares relevantes, que aos 42 anos inicia quadro de hemiparesia direita e disartria. Objetiva-se AVC isquémico a nível dos gânglios da base e cortico-subcortical esquerdo e uma infeção por HIV-1 previamente desconhecida. É medicada com ácido acetilsalicílico, estatina, co-trimoxazol e terapêutica antiretroviral.

Ao longo do seguimento, é objetivada eosinofilia em perfil ascendente (máximo 12.620 cel/uL, 76.5%). O hemograma apresenta ainda uma anemia normocítica (Hb 10.7 g/dL), sem alterações no valor absoluto dos leucócitos ou plaquetas. O esfregaço de sangue periférico revela formas maduras de eosinófilos, sem blastos. Clinicamente, a doente refere astenia e apresenta lesões cutâneas maculopapulares pruriginosas nos membros superiores, tendo-se objetivado ainda um episódio de angioedema da face. Nega outras alterações da pele, sintomas do trato respiratório superior e inferior, anorexia, perda de peso, artralgia e outras queixas.

É excluída causa farmacológica da eosinofilia, após suspensão temporária de todos os fármacos. É também excluída causa infecciosa, através de exame bacteriológico e parasitológico das fezes, cultura de fezes frescas para larvas, serologias para parasitas e IgE específica para Aspergillus spp. É prescrita terapêutica empírica de eventual parasitose com albendazol, sem impacto na eosinofilia.

No seguimento do estudo de outras causas de eosinofilia destaca-se hipergamaglobulinémia (IgG 2410 mg/dL), aumento da IgE (812 UI/mL) e da vitamina B12 (842 mg/dL), triptase normal (9.4 mg/dL). Estudo imunológico negativo, nomeadamente ANCAs, ANA, anti-dsDNA, ENA e AAF, sem consumo de complemento e crioglobulinas <1%. Sem alterações da função renal ou do sedimento urinário, sem elevação dos parâmetros inflamatórios (Vs 1mm/h e PCR 0.18mg/dL). A TC-TAP revela pequenos nódulos pulmonares e infiltrado em vidro despolido. As PFR são inconclusivas e o ecocardiograma não revela alterações. Sem evidência de quistos hepáticos ou lesões ocupantes de espaço sugestivas de causa neoplásica. Realiza ainda biópsia medular com "hiperplasia eosinofílica acentuada".

Com a colaboração da Reumatologia, Infecciologia e Hematologia, a doente é diagnosticada com SEH idiopático. O estudo das mutações PDGFR (Platelet-derived growth factor receptors) foi negativo.

Em termos de terapêutica, inicia prednisolona 60mg/dia durante uma semana, com posterior redução gradual e suspensão. Observa-se marcada resposta da eosinofilia (5.000 cel/uL) após 7 dias e normalização do leucograma após 14 dias.

Este caso retrata uma doente com diagnóstico diferencial extenso, na exclusão de causas secundárias de eosinofilia, das quais do foro reumatológico se destaca a granulomatose eosinofílica com poliangeíte.

A apresentação inicial com acidente vascular cerebral, eosinofilia exuberante com histologia medular com acentuada hiperplasia eosinofílica, presença de angioedema e alterações pulmonares favorecem o diagnóstico de SEH, bem como a ausência de asma ou rinite, parâmetros inflamatórios normais e ANCAS negativos. O tratamento inicial do SEH passa pela corticoterapia nos doentes sem a mutação PDGFR, e pelo Imatinib, nos portadores da mutação.

180 - KIKUCHI-FUJIMOTO'S DISEASE AND ADULT ONSET STILL DISEASE – A DIFFERENTIAL DIAGNOSIS OR COEXISTING DISEASES?

Margarida Faria[1], Lídia Teixeira[1], Daniel Carvalho[1], Bela Machado[2], Mário Rodrigues[1]

[1]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [2]Serviço de Medicina Interna, Hospital Central do Funchal, Funchal, Portugal

Kikuchi-Fujimoto's disease (KFD) and Adult Onset Still disease (AOSD) are both rare inflammatory diseases that share typical clinical and laboratorial features. KFD is characterized by cervical lymphadenopathy, fever and often arthritis and cutaneous rash. Laboratorial tests usually reveal anaemia, erythrocyte sedimentation rate elevation (ESR) and leukopenia. The diagnosis is based on lymph node biopsy. AOSD typically presents with fever accompanied with evanescent salmon coloured rash, arthritis, lymphadenopathy, hepatosplenomegaly, ESR elevation and leucocytosis.

We present a 20-year-old man with a history of recurrent episodes of fever accompanied with evanescent exanthem, sore throat, night sweats and inflammatory arthralgias evolving medium size joints.

The first episode was in march 2018, in which the patient was medicated with oral prednisolone with

complete remission after one moth. Similar episodes occurred with a free of disease interval of one month. In august 2018 the patient was bed admitted in Internal Medicine for investigation of fever of unknow origin and an enlarged cervical lymph node was identified and biopsied. In the meantime, due to the suspicion of hepatosplenic t-cell lymphoma, the patient was transferred to the Haematology service and started chemotherapy. However, the biopsy revealed necrotizing lymphadenitis suggestive of KFD. Therefore, the diagnosis of KFD was assumed and the patient had hospital discharge. After 2 years of prednisolone therapy with disease recurrence in every corticoid suspension, the patient was bed admitted in the Internal Medicine because of a new episode with increased resistance to corticosteroids. In face of persistent fever and increased acute phase reactants after broad-spectrum antibiotics, the Rheumatology service was contacted. At the physical examination the patient had evanescent salmon-coloured rash, arthritis in his left wrist and ankle and in both knees. Blood analysis revealed leucocytosis, neutrophilia, ferritin 21834 ng/mL, ESR 44 mm and protein C-reactive 141 mg/L. After the exclusion of infectious, lymphoproliferative and autoimmune diseases, the diagnosis of AOSD with a polycyclic pattern was assumed. In the following analysis there was a new onset of anaemia, leukopenia, hyperfibrinogenaemia, hypertriglyceridemia, elevated lactate dehydrogenase and a ferritin rise to 52451 ng/mL. In this scenario, it was decided to start 1000mg/day of methylprednisolone followed by 1mg/kg/day of prednisolone and anakinra 100mg/day with an important clinical and analytical improvement.

After 1 month of anakinra, the patient presented thrombocytopenia that was resolved with the suspension of the drug 1 day in every 2 days. Nowadays, the patient is medicated with anakinra 100mg/day, methotrexate 15mg/day, folic acid 10mg/day and remains asymptomatic with normal blood cell counts and normal acute phase reactants.

A diagnosis of AOSD was made in a patient that fulfils the 1992 Yamaguchi criteria and that had a previous diagnosis of KFD based upon a histopathological finding of necrotizing lymphadenitis. KFD and AOSD are both inflammatory conditions with overlapping clinical that can coexist in the same patient. Therefore, a histopathological diagnosis of KFD does not exclude AOSD. The onset of thrombocytopenia after 1 month of anakinra led to the suspicion of a drug side effect and the increase of the administration interval ran to a normalization of the platelets count.

182 - ACUTE SARCOIDOSIS: A CASE OF LOFGREN'S SYNDROME

Sara Paiva Dinis [1], Ana Sofia Pinto[1], Filipe Cunha Santos[1], Nathalie Madeira[1], Cláudia Vaz[1, 2], 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

Sarcoidosis is a systemic inflammatory disorder that can potentially affect any organ system, notable for great variability in clinical presentation and clinical course. Lofgren syndrome is a clinically distinct phenotype of sarcoidosis specifically characterized by a self-limiting disease course.

A 45-year-old male patient with no previous relevant clinical history presented to our Rheumatology outpatient clinic complaining of symmetrical and inflammatory joint pain of ankles and skin rash of the anterior surface of both legs for the last month. He mentioned a progressive evolution, initially with pain and swelling of both ankles, which improved with physical therapy and non-steroidal anti-inflammatory drugs (NSAIDs) for 14 days. Nevertheless, symptoms got worse in the following week, and a skin rash involving the anterior surface of both legs appeared. He had experienced a fever (38º C) lasting one day.

On physical examination, he had erythematous tender nodules on the anterior side of both legs, suggestive of erythema nodosum (EN), and arthritis in both ankles, in addition to painful palpation and movement of the tibialis and peroneus tendons. The remaining physical exam was unremarkable.

Blood work showed elevated erythrocyte sedimentation rate (ESR, 82 mm/hr) and C-reactive protein level (CRP, 2.58 mg/dL). His renal and liver function tests, TASO, uric acid, calcium and ACE levels were within

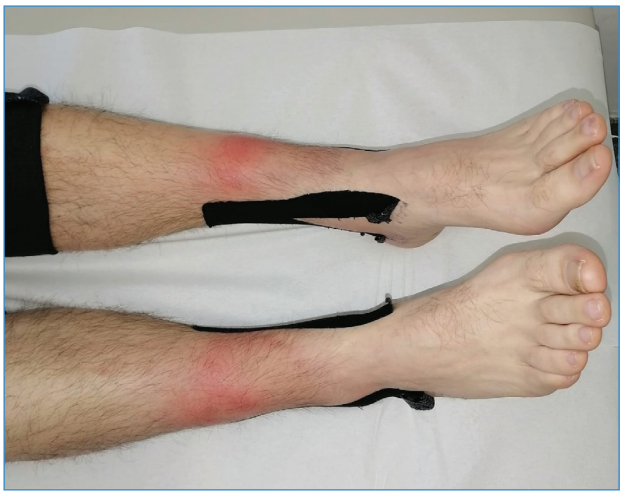


Figure 1. Clinical photograph shows erythematous, tender nodules on the anterior side of both legs (erythema nodosum)

normal range. Rheumatoid factor, anti-cyclic citrullinated peptide (anti-CCP), antinuclear, and other specific antibodies were not detected. Chest X-ray revealed bilateral hilar lymphadenopathy and a chest computed tomography scan demonstrated multiple enlarged lymph nodes of the mediastinum and both hila with numerous, predominantly peripheral subpleural and pericissural nodules and scattered ground-glass areas, mainly in the lower zones. Ankle ultrasonography findings suggested bilateral tenosynovitis of the tibialis posterior and peroneus brevis.

Based on the triad of acute periarthritis and ankle arthritis, bilateral hilar adenopathy, and erythema nodosum, the patient was diagnosed with Lofgren's syndrome. He was prescribed 90 milligrams of etoricoxib once a day for twenty days. After one week, skin lesions had partially subsided, joint pain improved and levels of ESR and CRP decreased (29 mm/hr, 0.28 mg/dL). In the second week, both pain and rash disappeared.

Lofgren syndrome typically does not require a histologic diagnosis, yet the case was presented to multidisciplinary discussion with the Pulmonology team, which decided against mediastinoscopic lymph node biopsy.

Treatment of Lofgren syndrome is largely supportive with spontaneous resolution occurring over 1 to 2 years. Constitutional symptoms and arthralgias are treatable with non-steroidal anti-inflammatory drugs or colchicine.

Arthralgias and EN are often observed in various connective tissue diseases, including rheumatoid arthritis. Thus, Lofgren's syndrome, an acute form of sarcoidosis, should also be taken in consideration in differencial, since a strong clinical suspicion and prompt identification of classic clinical features can lead to an early diagnosis and treatment. A chest radiograph to confirm hilar adenopathy is mandatory.

187 - DERMATOMIOSITE INDUZIDA PELAS ESTATINAS

Bruna Silva[1], Miguel Bernardes[1, 2], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar de São João, Porto, Portugal, [2]Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Introdução: As estatinas são dos fármacos mais utilizados na prática clínica e apresentam um bom perfil de segurança. O efeito adverso mais comum é a miotoxicidade. Contudo, estudos recentes têm demonstrado que estes fármacos podem associar-se também a disturbios de auto-imunidade, tais como Dermatomiosite.

Caso clínico: Apresentamos o caso de um doente do sexo masculino, caucasiano, de 52 anos, com antecedentes pessoais de: doença coronária assintomática (rastreada pela existência de história familiar em idade jovem), dislipidemia, tabagismo passado (8 UMA), trombocitopenia minor, esteatose hepática e hemangiomas hepáticos. Em maio de 2020, alterou a sinvastatina 20mg id, iniciada em outubro de 2019, para rosuvastatina 20mg id e ezetimiba 10mg id. Em fevereiro de 2021, iniciou quadro de astenia e mialgias proximais dos membros inferiores incapacitantes, associado a lesões cutâneas nas mãos, pálpebras e hélices de novo. Foi medicado com betametasona intramuscular, com melhoria clínica. Em agosto de 2021, mudou para atorvastatina 20mg id e ezetimiba 10mg id, passando a apresentar poliartralgias simétricas de ritmo inflamatório, envolvendo as articulações metacarpofalângicas (MCFs) e interfalângicas proximais (IFPs) das mãos, e acrocianose das mãos. Em fevereiro de 2022, foi medicado com deflazacorte 3mg bid, naproxeno 500mg bid e pentoxifilina 400mg bid, com alguma melhoria clínica; sendo encaminhado para a Reumatologia para ulterior investigação. Ao exame objetivo apresentava sinovite de várias IFPs (sobretudo 2ª e 3ª), pápulas de Gottron no dorso das MCFs e IFPs das mãos (Figura 1), rash heliotrópico nas pálpebras e hélices, esclerodactilia nas falanges intermédias e perioníquia em todos os dedos das mãos, com força muscular conservada. Analíticamente apresentava VSG e PCR normais, linfopenia, trombocitopenia, função renal normal, discreta citólise hepática e marcadores de lesão muscular aumentados. Do estudo imunológico destacavam-se as positividades para os ANAs (1/100, padrão mosqueado) e os anticorpos Anti-Scl70 e Anti-MDA5. Ainda em março de 2022, foi realizada ressonância magnética das coxas, que foi compatível com miosite/miopatia, e videocapilaroscopia do leito ungueal, que revelou a presença de megacapilares e hemorragias em todos os dedos, compatíveis com o diagnóstico clínico de dermatomiosite. Em maio de 2022, foi realizada biópsia muscular do reto femoral e iniciada terapêutica com metrotexato (12,5mg, per os, semanais), mantendo seguimento na consulta.

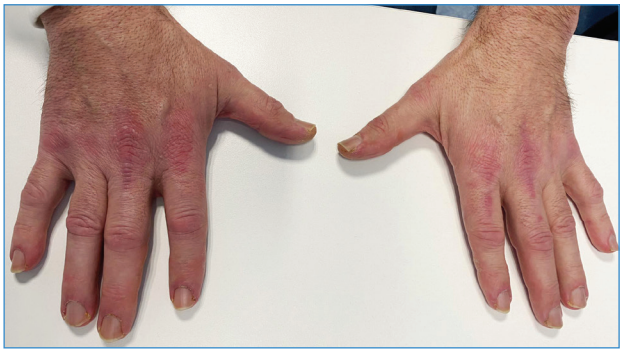


Figura 1. Pápulas de Gottron no dorso das articulações metacarpofalângicas e interfalângicas proximais das mãos.

Discussão: Este caso clínico remete para a diagnóstico de dermatomiosite induzida pelas estatinas, uma enti-

dade rara e de fisiopatologia desconhecida. Até 2020, encontravam-se descritos cerca de 25 casos clínicos em literatura médica. Este diagnóstico passa pela identificação dos sinais e sintomas típicos de dermatomiosite, aliada à presença de elevação sérica das enzimas musculares, alterações eletromiográficas sugestivas e/ou histologia muscular compatível. A identificação de anticorpos especificos para a dermatomiosite suporta o diagnóstico. Acresce ainda a necessidade de se verificar uma relação temporal entre a introdução da estatina e o ínicio da clínica, após exclusão de outras etiologias.
Conclusões: Este caso realça o facto de que, apesar de raro, a dermatomiosite poder ser secundária a fármacos de uso frequente na prática clínica, pelo que devemos estar atentos já que o diagnóstico e tratamento precoces parecem melhorar significativamente o prognóstico destes doentes.

191 - TWO CASE REPORTS OF HEART CONDUCTION SYSTEM DEFECTS IN SYSTEMIC SCLEROSIS

Catarina Abreu[1], Vanessa Fraga[1], Ana Cordeiro[1], Alice Castro[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

Cardiac manifestations in systemic sclerosis (SSc) can be primary due to involvement of the myocardium, pericardium, valves and conduction system; or secondary to pulmonary arterial hypertension (PAH), interstitial lung disease (ILD) and renal disease. The prevalence of primary cardiac involvement is difficult to determine since it is probably underestimated due to subclinical disease. Cardiac manifestations are more frequent in patients with anti-topoisomerase I antibodies (Scl70) positivity and together with pulmonary manifestations are the main cause of death in patients with SSc.
We aim to report 2 clinical cases of primary cardiac involvement in SSc patients.

The first case reports to a 63-year-old Caucasian female with the diagnosis of SSc since 2014 with anti-Ro60 positivity. Clinically she presented with cutaneous (sclerodactily); vascular (Raynaud phenomenon and digital ulcers); pulmonary (usual interstitial pneumonia); upper and lower gastrointestinal (gastroesophageal reflux (GER) and fecal incontinence) and musculoskeletal (arthritis and myositis) involvement. On diagnosis she performed a 24-hour Holter monitoring (24hHm) that documented bigeminy. In 2016 she presented with progressive exertional fatigue and increased N-terminal prohormone of brain natriuretic peptide (NT-proBNP) on work-up. The echocardiogram had a non-previously documented reduced ejection fraction due to hypokinesis of the apex, posterior and inferior walls and septum. She was referred to a cardiology appointment and placed drug eluting stents due to sub occlusive lesions of 3 coronary arteries, with clinical improvement. In 2017, although asymptomatic, complete right bundle branch block and frequent ventricular extrasystole with bigeminy and trigeminy (less than 10.000/24 hours) were present on 24hHm. In 2021 she implanted a definitive pacemaker due to a new onset second degree atrioventricular block on a routine 24hHm.

The second case reports to a 20-year-old Caucasian male. In 2020 he developed distal skin thickness and Raynaud phenomenon with digital pitting and ulcers six months later. He also had significant weight loss and GER complaints. In 2021 he was diagnosed with limited cutaneous SSc with Scl70 positivity. On diagnosis he was asymptomatic with a normal echocardiogram but with a raised NT-proBNP. Six months after diagnosis, he was admitted to another hospital complaining of dyspnea on exertion and thoracic pain. He performed a cardiac magnetic ressonance (CMR) that described extensive biventricular transmural late gadolinium enhancement, suggestive of myocardial fibrosis confirmed by myocardial biopsy, without inflammation. He did a 24hHm that demonstrated right bundle branch block with bigeminy and a 7-day register monitoring that could not correlate symptoms with the register findings. Due to maintenance of symptoms with elevated troponin T (TnT) a loop recorder was implanted to monitor events.

These cases highlight the occurrence of less common cardiac manifestations early in the course of SSc. A careful clinical evaluation may not be enough to identify myocardial involvement. It is important to regularly perform appropriate screening exams, including soluble biomarkers (TnT, NT-proBNP), echocardiogram and electrocardiogram. If alterations are documented, a 24hHm, exercise testing or CMR may be needed.

193 - ATYPICAL PRESENTATION OF CALCIUM PYROPHOSPHATE CRYSTAL DEPOSITION DISEASE AFTER COVID-19 INFECTION

Maria Pontes Ferreira[1], Francisca Guimarães[1], Hugo Parente[1], Catarina Soares[1], Emanuel Costa[2], Soraia Azevedo[1], Daniela Peixoto[1], Filipa Teixeira[1], Daniela Santos-Faria[1], Carmo Afonso[1], José Tavares-Costa[1]
[1]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [2]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

Background: Calcium pyrophosphate crystal deposi-

tion disease (CPPD) is a microcrystalline arthropathy, caused by deposition of calcium pyrophosphate crystals (CPP) in the articular space and soft tissues, with consequent joint damage1. The disease mainly presents after the age of 60, and can have several presentations, including asymptomatic phenotype, CPPD with osteoarthritis, acute arthritis and chronic inflammatory arthritis 1-3. We present a clinical case of CPPD after COVID-19 infection, with atypical features.

Case: A 68 years old woman developed a mildly symptomatic COVID-19 infection in 26th January 2022, with non-productive cough, rhinorrhoea, myalgias, low fever and dyspnoea for moderate efforts. At the end of February, she noticed a painless axillar tumefaction, which was progressively increasing in size and unspecific dorsal pain. At physical examination she had inspiratory crackles on left hemithorax, and right axillar tumefaction of 4 cm in diameter. She had no abnormalities in blood work, besides a negligible C-Reactive Protein (CRP) of 0.70 mg/dL and Erythrocyte Sedimentation Rate (ESR) of 26 mm, and bilateral infiltrates in thoracic radiographies. A thoracic computerized tomography (CT) scan revealed ground glass opacities in left hemithorax, covering less than 50% of lung, with no pleural effusion. Therefore, she begun antibiotherapy with amoxicillin and clavulanic acid, with improvement of respiratory symptoms. Axillar ultrasound (US) scan was performed, showing a liquid collection of 41x15x39mm, which extended laterally to the glenohumeral joint. In April 2022 the patient complained of significant inflammatory shoulder and cervical pain, with limitation in range of motion of both shoulders and the cervical spine. In blood work, there was CRP of 1.44 mg/dL and ESR of 44 mm, shoulder US showed signs of bilateral glenohumeral effusion and radiographies exhibited calcification of the glenohumeral labrum bilaterally and calcification of the intervertebral discs. US-guided arthrocentesis of glenohumeral articulation was performed, with aspiration of inflammatory synovial fluid, with presence of CPP crystals on optic microscopy. When reviewing the clinical process, it was possible to ascertain that, although previously asymptomatic, the patient already had calcification of the transverse ligament of atlas on a cranioencephalic CT scan performed in another context. The diagnosis of CPPD with axial involvement was then confirmed, and the patient begun treatment with NSAIDs and colchicine 1mg/day, with symptomatic and ESR/CRP important improvements.

Discussion: CPPD is an inflammatory crystal-induced arthropathy that can have several presentations, ranging from asymptomatic to chronic osteoarthritis2. In this clinical case we present an atypical presentation of CPPD, since the first manifestation of the disease was in post-COVID-19 infection context and was initially a painless glenohumeral effusion, developing afterwards a flare of significant inflammatory shoulder and cervical pain.

REFERENCES

1. Abhishek A, Doherty M. Update on calcium pyrophosphate deposition. Clin Exp Rheumatol. 2016 Jul-Aug;34(4 Suppl 98):32-8. Epub 2016 Jul 22. PMID: 27586801.
2. Sidari A, Hill E. Diagnosis and Treatment of Gout and Pseudogout for Everyday Practice. Prim Care. 2018 Jun;45(2):213-236. doi: 10.1016/j.pop.2018.02.004. PMID: 29759121.
3. Zamora EA, Naik R. Calcium Pyrophosphate Deposition Disease. 2022 May 4. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan–. PMID: 30986010.

199 - BILATERAL ISCHIUM CONDROCALCINOSIS ON PATIENT WITH WALDENSTROM'S MACROGLOBULINEMIA WALDENSTROM: A CASE REPORT

Tiago Beirão[1], Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Catarina Rua[1], Flávio Campos Costa[1], Romana Vieira[1], Joana Abelha-Aleixo[1], Taciana Videira[1], P Pinto[1]

[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Objective: Waldenstrom's macroglobulinemia is a rare B-cell lymphoproliferative neoplasm, in which lymphoplasmacytic cells infiltrate the Bone Marrow, along with an IgM monoclonal gammopathy in the serum. 4% of patients have been reported for having hypercalcemia by either binding of calcium to IgM leading to pseudohypercalcemia, or induced by PTHrP. Due to this, it can lead to higher local levels of calcium, which may facilitate calcium pyrophosphate dihydrate crystal deposition disease.

Methods: Based on clinical observation of 81 years old male with acute polyarthritis and previous diagnosis of

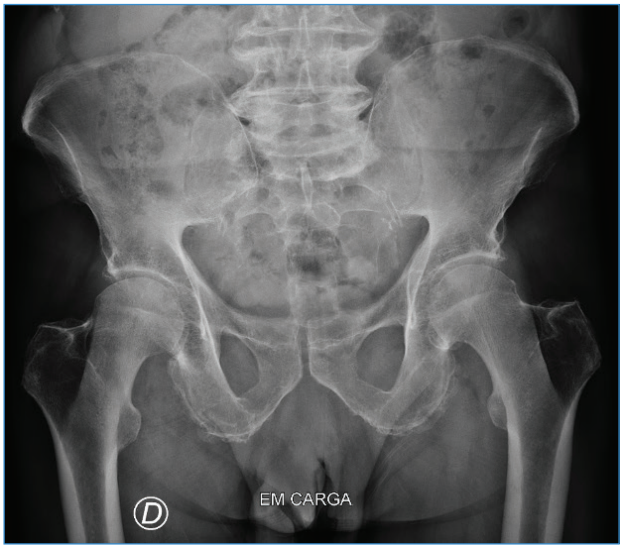


Figure 1. Bilateral Ischium Chondrocalcinosis

Waldenstrom's macroglobulinemia
Results: A 81-year-old male, with previous diagnosis of Waldestrom's macroglobulinemia, is referenced to the rheumatology department due to polyarthritis. Patient describes inflammatory arthralgias of wrist, knees, elbows, and metacarpophalangeal and proximal interphalangeal joints. On physical exam, arthritis was found on these joints, with no erythema. Previous medical records showed an increased calcium level (10.8 mg/dL, with normal albumin) and normal phosphates levels. Clinical suspicion of calcium pyrophosphate dihydrate crystal deposition disease was raised and X rays were taken, which confirmed presence of chondrocalcinosis of right knee and bilateral ischium (figure 1). Colchicine was started with excellent clinical result, resolving patient's complaints.
Conclusion: Rheumatologists should be aware that lymphoproliferative diseases, such as Waldenstrom's macroglobulinemia, may lead to hypercalcemia. In patients with acute polyarthritis, calcium pyrophosphate dihydrate crystal deposition disease should be excluded.

200 - ALÉM DE UM ANTICORPO POSITIVO

Tiago Beirão[1], André Araújo[2], Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Catarina Rua[1], Flávio Campos Costa[1], Romana Vieira[1], Joana Abelha-Aleixo[1], Taciana Videira[1], P Pinto[1]
[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [2]Neuroradiology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Introdução: A trombose venosa cerebral é uma doença cerebrovascular provocada por oclusão dos seios venosos e/ou veias cerebrais por trombos, associada a múltiplos fatores de risco, tais como contracetivos orais ou a gravidez. Esta manifestação pode estar associada a doenças reumáticas complexas, sendo importante a avaliação clínica e analítica destes doentes.
Breve descrição: Doente do sexo feminino, de 20 anos, sem antecedentes médico-cirúrgicos de relevo, recorre ao serviço de urgência por episódios de dor e edema do membro inferior direito, tendo realizado eco-doppler que confirmou o diagnóstico de trombose venosa profunda tratada com heparina durante 6 meses. 2 anos depois, inicia quadro de cefaleia aguda, tendo sido efetuado o diagnóstico de trombose venosa cerebral (figura 1) com inicio de hipocoagulação. Da avaliação realizada, destaca-se duas medições de anti-Beta2Glicoproteina I positivas IgM + IgG - (valor mais alto: 15,0 U/mL). Doente teve alta com diagnóstico de Síndrome Antifosfolipidico. Recorrencia de Trombose Venosa Cerebral após 3 anos do primeiro evento, apesar de manter terapêutica com varfarina com INR de 2-3. Após o segundo episódio, doente é referenciada para a consulta de reumatologia para estudo etiológico. Da anamnese realizada, a doente refere episódios de aftose oral de repetição e história familiar de primeiro grau de Doença de Behçet. Apresentava HLAB51 positivo. Foi efetuado o diagnóstico de Doença de Behçet e iniciou azatioprina. A doente não teve novos episódios de trombose.

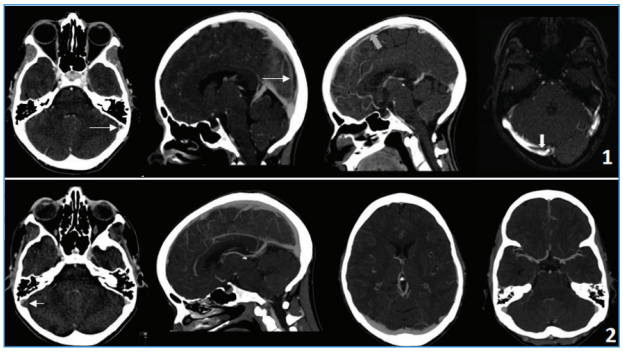


Figura 1.

Legenda da figura: Figura 1. Após administração de contraste iodado, o estudo de venograma nos planos axial (a) e sagital (b) demonstra imagens de substração no seio sigmóide esquerdo e no terço posterior do seio longitudinal superior (setas), sugestivas de trombose de seios venosos cerebrais; (c) 1 ano depois, verificam-se sinais de trombose "de novo" no terço médio do seio longitudinal superior (seta espessa cinzenta), previamente patente; (d) o estudo por RM (2D-TOF) revelou ainda sinais de trombose da vertente proximal do seio transverso direito (seta espessa branca); Figura 2. (a) No venograma 3 anos após o primeiro evento, observa-se preenchimento subtotal do seio sigmóide direito, "de novo", sugerindo trombose em nova localização (seta); (b-d) o venograma 5 anos após o primeiro evento demonstra repermeabilização completa do seio longitudinal superior (a), da vertente proximal do seio transverso direito (b) e dos seios sigmóides (d))
Conclusão: A doença de Behçet é uma doença inflamatória trombofilica, sendo a trombose venosa a principal complicação vascular. Este caso realça a importância da avaliação clínica, para estabelecer o diagnóstico correto, apesar da presença de um anticorpo positivo.

206 - OSTEOMALÁCIA SECUNDÁRIA A HIPOFOSFATÉMIA – PAPEL DO FGF 23: RELATO DE DOIS CASOS CLÍNICOS

Emanuel Costa[1], Carla Campinho Ferreira[1], Ana Margarida Correia[1], Diogo Esperança Almeida[1], Joana Leite Silva[1], Marcos Cerqueira[1], Joana Sousa-Neves[1], Ana Ribeiro[1]
[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

Introdução: A osteomalácia é um distúrbio ósseo, caracterizado pela diminuição da mineralização do osso recém-formado. Várias etiologias distintas podem causar osteomalácia sendo as mais frequentes o défice de vitamina D e a hipofosfatémia.

O fator de crescimento de fibroblastos 23 (FGF23) é uma hormona secretada pelos osteócitos em resposta ao aumento do calcitriol e fosfato sérico. A principal função do FGF23 é a regulação da fosfatémia atuando no túbulo contornado proximal onde diminui a expressão do cotransportador sódio-fosfato. Assim, diminui a reabsorção de cálcio e aumenta a excreção de fosfato na urina. O FGF23 também pode suprimir a 1-alfa-hidroxilase, reduzindo a sua capacidade de ativar a vitamina D e, consequentemente, diminuindo a absorção de cálcio.

Caso 1: Osteomalacia oncogénica: Foi pedida a avaliação por Reumatologia de um homem de 63 anos internado no serviço de Ortopedia para cirurgia eletiva de artroplastia total da anca direita por fratura do colo bilateral não traumática.

Segundo o doente, o quadro clínico ter-se-á iniciado 6 anos antes, após paraparésia progressiva dos membros inferiores. Na sequência da investigação etiológica, identificado tumor mesenquimatoso do corpo de D9 com efeito expansivo e compressão medular. Nesse contexto, o doente terá sido avaliado por equipa multidisciplinar que considerou a lesão irressecável mas, tendo em conta o compromisso neurológico, foi submetido a cirurgia descompressiva. Foi ainda proposta radioterapia local que o próprio doente recusou.

À data de observação, apurada historia de múltiplas fraturas não traumáticas nos últimos 5 anos (arcos costais, tornozelo, bacia, colo do fémur e omoplatas), sem queixas de dor óssea ou osteoarticular.

Analiticamente apresentava fosforo sérico indoseável, défice de vitamina D, hipocalcemia, com hormona paratiroideia normal, aumento da fosfatase alcalina sérica sem fosfaturia. O doseamento posterior de FGF23 revelou um valor 6x acima do limite superior do normal (LSN).

Assim, concluiu-se tratar de osteomalácia oncogénica por tumor mesenquimatoso produtor de FGF23. O doente iniciou reposição de fosfato endovenoso em unidade de cuidados intermédios durante o internamento e foi orientado para ambulatório com suplementação oral de fosforo, cálcio e vitamina D. Foi orientado para consulta de endocrinologia e oncologia e está proposto para inicio de burosumab (anticorpo dirigido a FGF23).

Caso 2: Osteomalácia secundária a administração de carboximaltose férrica: Mulher de 57 anos, com antecedentes de sindrome de Rendu-Osler-Weber com anemia ferripriva sob suplementação crónica de ferro endovenoso (carboximaltose férrica). Referenciada por fratura do 2º metatarso à direita e alterações do metabolismo fosfocálcico com hipofosfatémia, défice de vitamina D, elevação de fosfatase alcalina sérica 3xLSN e PTH 2xLSN com valores normais de calcémia. O estudo subsequente confirmou a suspeita de hipofosfatémia mediada por FGF23 (elevação de 3xLSN).

A doente iniciou suplementação de fosforo oral, vitamina D e foi alterada a formulação deferro endovenoso. Seis meses após a avaliação, os resultados analiticos revelam normalização dos valores prévios.

Pensa-se que o mecanismo chave responsável por este efeitoresulte da inibição desproporcional da degradação de FGF23 pelos hidratos de carbono presentes nas formulações de ferro endovenoso com consequente aumento da sua concentração e a atividade. O risco de hipofosfatemia e osteomalácia parece ser maior com carboximaltose férrica do que com outras formulações intravenosas de ferro.

REFERÊNCIAS

1. Schaefer, B., Tobiasch, M., Wagner, S., Glodny, B., Tilg, H., Wolf, M., & Zoller, H. (2022). Hypophosphatemia after intravenous iron therapy: Comprehensive review of clinical findings and recommendations for management. Bone, 154, 116202. doi: 10.1016/j.bone.2021.116202
2. Klein, K., Asaad, S., Econs, M., & Rubin, J. (2018). Severe FGF23-based hypophosphataemic osteomalacia due to ferric carboxymaltose administration. BMJ Case Reports, bcr-2017-222851. doi: 10.1136/bcr-2017-222851

208 - GOTA TOFÁCEA – ENVOLVIMENTO DA COLUNA DORSAL

Carla Campinho Ferreira[1], Ana Margarida Correia[1], Emanuel Costa[1], Diogo Esperança Almeida[1], Hugo Pereira[2], Joana Sousa-Neves[1], Marcos Cerqueira[1], José Redondo[1], Ana Ribeiro[1], Joana Leite Silva[1]

[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal, [2]Serviço de Neurocirurgia, Hospital de Braga, Braga, Portugal

Introdução: A artropatia gotosa é uma doença inflamatória causada pela deposição de cristais de monourato de sódio (MUS) nos tecidos. A hiperuricemia é um fator preponderante para o desenvolvimento de gota, sendo que quando se ultrapassa a capacidade de dissolução do ácido úrico (AU) no sangue, formam-se cristais de MUS que se podem depositar nas articulações, cartilagens, tendões e tecidos moles. Normalmente estão envolvidas as articulações periféricas, no entanto, o atingimento da coluna vertebral, embora raro, pode ocorrer. Uma revisão de 131 casos de envolvimento axial por gota, revelou que o local mais comum é a coluna lombar, seguido da coluna cervical e, menos frequente, da coluna dorsal.

Caso clínico: Doente do sexo feminino, 77 anos de

idade, previamente autónoma, com antecedentes pessoais de acidente vascular cerebral isquémico, hipertensão arterial e gastrite autoimune. Recorreu ao serviço de urgência por quadro insidioso e progressivo de dificuldade na marcha por défice de força e alteração da sensibilidade dos membros inferiores (MIs), com 3 meses de evolução. Referia ainda perda ponderal e diminuição da massa muscular dos MIs. Negava alterações nos membros superiores (MSs), esfincterianas, dorsalgia ou lombalgia. Ao exame objetivo destacava-se paraparésia espástica, com força muscular (FM) grau 3 à direita e grau 4 à esquerda na flexão da coxa, FM grau 3 à direita e grau 5 à esquerda na extensão/flexão do joelho e FM grau 4 à direita e grau 5 à esquerda dorsiflexão/flexão plantar; atrofia muscular de predomínio proximal nos MIs; reflexos osteotendinosos hipercinéticos e com área alargada nos MIs; reflexo cutâneo-plantar indiferente bilateralmente; hipostesia à picada com nível por D4; e alterações da propriocepção. Realizou ressonância magnética da coluna dorsal que revelou lesão ocupante de espaço intracanalar epidural direita, no plano de D2-D3 na dependência da articulação posterior, com 1,2 cm por 0,74cm de diâmetro, marcadamente hipointensa em T2, a condicionar desvio da haste medular e compressão, com hipersinal tradutor de mielopatia (fig. 1a). Colocadas as hipóteses de meningioma ou quisto sinovial calcificado, foi decidido remover cirurgicamente a lesão. O resultado da avaliação anatomopatológica revelou material amorfo compatível com cristais de AU. A doente foi posteriormente enviada à consulta de Reumatologia onde referia quadro compatível com crises de artrite envolvendo essencialmente o cotovelo e punho direitos e história prévia de podagra, com vários anos de evolução, alimentação rica em purinas no passado e consumo de 1 copo de vinho a cada refeição. Sem insuficiência renal conhecida ou fármacos causadores de hiperuricemia. Ao exame objetivo apresentava tofos gotosos no cotovelo direito (fig. 1d), pavilhão auricular esquerdo (fig. 1e) e em várias articulações metacarpofalângicas (fig. 1f); sem artrite objetivável. Apresentava uricemia de 11,1 mg/dl, nunca tendo realizado terapêutica hipouricemiante previamente.

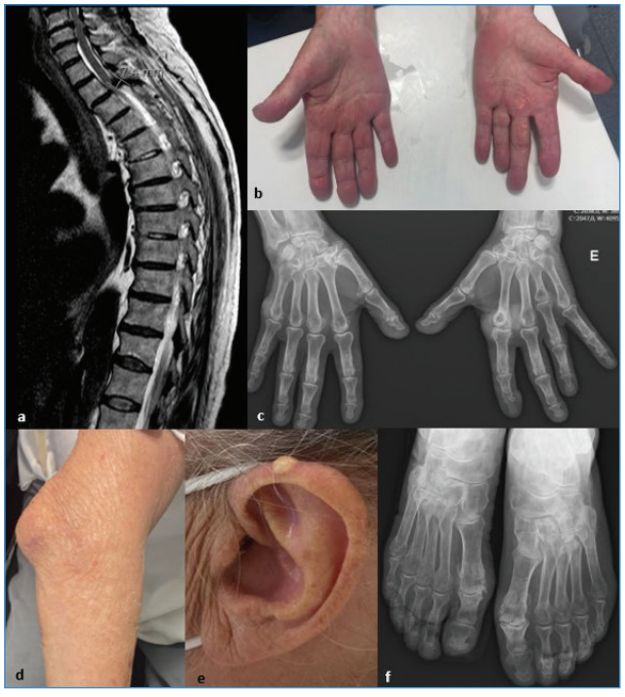


Figura 1. a) Imagem de RM dorsal ponderada em T2 com lesão ocupante de espaço intracanalar, epidural, hipointensa; b) tofos gotosos na face palmar de ambas as mãos; c) radiografia das mãos com alargamento de tecidos moles ao redor da articulação metacarpofalângica (MCF) 5 à direita e MCF 1 à esquerda; d) tofo gotoso no cotovelo direito; e) tofo gotoso no pavilhão auricular esquerdo; f) radiografia dos pés com provável erosão na falange proximal do primeiro dedo à direita.

Discussão: O envolvimento axial por depósitos de MUS é raro e pode manifestar-se de múltiplas formas, desde dor não controlada a défices neurológicos, com tempo de evolução variável. O diagnóstico é difícil, porque além de raro, as manifestações clínicas e imagiológicas podem mimetizar múltiplos outros diagnósticos como tumores, abcessos ou lesões císticas. Este diagnóstico deve ser equacionado nos diagnósticos diferenciais de lesões vertebrais epidurais, nomeadamente em doentes com diagnóstico prévio de gota, sendo necessária a cirurgia para diagnóstico definitivo.

209 - SARCOIDOSE, A GRANDE MIMETIZADORA

Maria João Cadório[1], João Alexandre Oliveira[1], Liliana Saraiva[1], Mariana Luis[1, 2], Cátia Duarte[1]
[1]Serviço de Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [2]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal

Introdução: A Sarcoidose é uma doença multissistémica caracterizada pela presença de granulomas não caseosos. Trata-se de uma entidade rara de etiologia desconhecida, cuja apresentação clínica pode mimetizar outras doenças, como Linfomas ou Tuberculose Pulmonar.

Caso clínico: Homem caucasiano, 48 anos, sem antecedentes ou contexto epidemiológico relevantes, iniciou quadro de febre, sudorese profusa, astenia e tosse não produtiva com meses de evolução. Analiticamente: anemia normocítica normocrómica, sem outras alterações do hemograma, ECA 73 U/L (N<52 U/L), VS e PCR, LDH e imunoeletroforese normais. Ecoendoscopia brônquica inconclusiva, medulograma e biópsia óssea normais. Realizou TC toraco-abdomino-pélvica,

que revelou a presença de conglomerados adenopáticos mediastínicos, cuja biópsia identificou granulomas epitelioides com células gigantes multinucleadas, não caseosos, compatíveis com o diagnóstico de Sarcoidose, sem achados de linfoma ou outra neoplasia. Iniciou tratamento com prednisolona (PDN) na dose máxima de 40 mg/dia, com necessidade de terapêutica adjuvante com metotrexato (MTX) 15 mg/semana e Ácido Fólico 5 mg/semana. Por ter apresentado boa resposta ao tratamento instituído, cumpriu esquema de desmame de corticoterapia até suspensão total, que decorreu sem intercorrências, mantendo terapêutica de manutenção com MTX e Ácido Fólico. Seis meses depois, iniciou quadro de fadiga, febre de predomínio vespertino e sudorese noturna. Apresentou episódios recorrentes de olho vermelho com diminuição aguda da acuidade visual, tendo sido feito o diagnóstico de uveíte posterior. Analiticamente, apresentava anemia normocítica normocrómica, sem outras alterações (incluindo PCR, VS, CK, ECA, calcémia e calciúria). Apesar dos antecedentes de Sarcoidose, foi feito estudo para exclusão de neoplasia e tuberculose: IGRA negativo; PET-FDG a evidenciar envolvimento ganglionar extenso por Sarcoidose com alto grau de atividade, esplenomegalia, rins e musculatura glútea com captação aumentada de FDG-F18. Realizou ainda TC Pulmonar de Alta Resolução que evidenciou alterações sugestivas de Sarcoidose pulmonar estádio II, com Provas de Função Respiratória normais. Foi admitido o quadro de recidiva de Sarcoidose com envolvimento ganglionar, pulmonar, ocular e sistémico, tendo sido reintroduzido PDN na dose máxima de 60 mg/dia (em adição à terapêutica de manutenção). Atualmente, encontra-se em esquema de desmame de PDN, sem novas intercorrências clínicas e com normalização dos parâmetros analíticos.

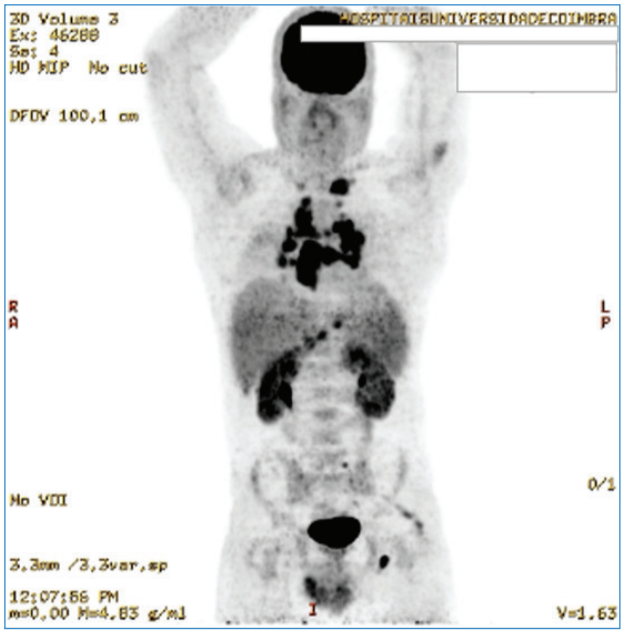


Figura 1. PET-FDG: adenopatias hipermetabólicas; baço, rins, musculatura glútea e deltóide esquerdos com captação difusamente aumentada

Conclusão: Com este relato de caso, os autores pretendem salientar a importância de manter elevada suspeita diagnóstica para outras doenças potencialmente graves, apesar do diagnóstico já estabelecido de Sarcoidose. Além da possibilidade de coexistência de 2 entidades distintas, a terapêutica imunossupressora pode conferir risco aumentado de infeções oportunistas e neoplasias hematológicas, pelo que estas devem ser sempre excluídas.

211 - ENVOLVIMENTO CERVICAL NA DOENÇA POR DEPOSIÇÃO DE CRISTAIS DE PIROFOSFATO DE CÁLCIO

Emanuel Costa[1], Carla Campinho Ferreira[1], Maria Pontes Ferreira[2], Ana Margarida Correia[1], Catarina Soares[2], Hugo Parente[2], Filipe Cunha Santos[3], Diogo Esperança Almeida[1], Francisca Guimarães[2], Joana Leite Silva[1], Joana Sousa-Neves[1], Marcos Cerqueira[1], Carmo Afonso[2], Ana Ribeiro[1], Soraia Azevedo[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: A doença por deposição de pirofosfato de cálcio (DPPC) é uma artropatia microcristalina que afeta o espaço articular, fibrocartilagem e tecidos periarticulares com consequente inflamação e dano estrutural. Afeta preponderantemente o esqueleto apendicular tem diferentes fenótipos que lhe valem os pseudónimos de pseudogota, pseudo artrite reumatoide e pseudo osteoartrose. O envolvimento do esqueleto axial é menos frequente e pouco reconhecido, com um amplo espectro de apresentações clínicas.

Caso 1: Deposição assintomática de PPC no ligamento transverso do atlas: Mulher de 68 anos, com diagnóstico de DPPC com apresentação semelhante a pseudogota. Tinha previamente sido realizada artrocentese de um joelho aquando de episódio de monoartrite e, na análise de líquido sinovial ao microscópio ótico de luz polarizada, foram identificados cristais rombos, com fraca birrefringência e elongação positiva, compatíveis com cristais de PPC. Na sequência de um episódio de urgência por traumatismo craniano, realizou Tomografia Computorizada (TC) cerebral onde foi possível observar a presença de calcificação do ligamento transverso do atlas – Fig 1 A., sem sintomatologia atual ou pregressa associada.

Caso 2: Síndrome do dente coroado: Uma mulher de

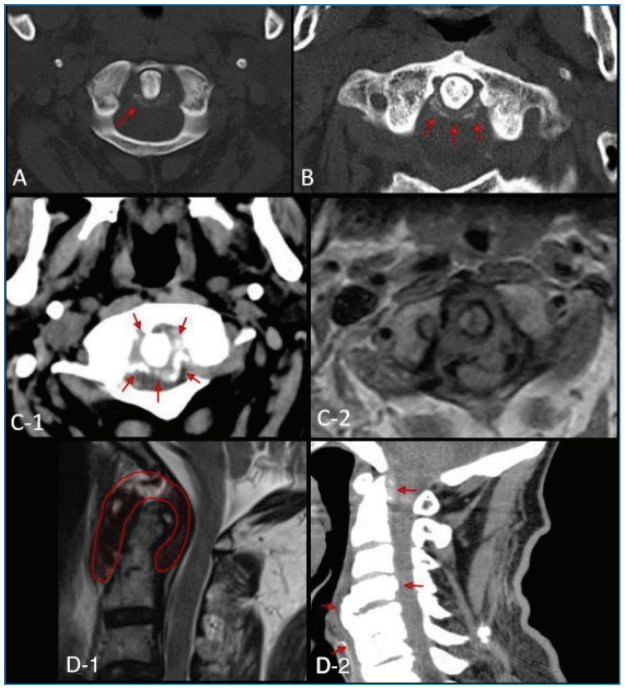


Figura 1.

59 anos, com o diagnóstico definitivo de DPPC secundária a hipomagnesémia iatrogénica por inibidor da bomba de protões. Solicitou avaliação urgente por cervicalgia de ritmo inflamatório com 3 dias de evolução. Referia início súbito durante a noite e limitação franca das amplitudes de movimento do segmento cervical. Associadamente, descrevia sensação febril com Tax 38.1ºC. Referia suspensão autónoma da medicação em curso 1 mês antes do início das queixas (colquicina e magnésio oral). Analiticamente, apresentava elevação dos parâmetros de fase aguda. O TC da coluna cervical demonstrava mineralização dos ligamentos transverso do atlas e alar. A doente cumpriu esquema de corticoterapia em baixa dose e retomou a medicação habitual, com rápida e completa recuperação– Fig 1B.

Caso 3: Pseudotumor retro-odontoideu: Mulher de 85 anos, referenciada por neurocirurgia onde mantinha seguimento por clínica sugestiva de mielopatia cervical. Do estudo imagiológico realizado, dispunha de TC e RM da coluna cervical e de crânio onde foi identificada calcificação e espessamento marcado do complexo ligamentar atlantoaxial e tecidos moles periodontoideus, com luxação anterior da odontoide e estenose do canal vertebral na transição bulbo medular. Clinicamente, referia episódios de longa data, caracterizados por cervicalgia de ritmo misto, mas sem história sugestiva de artrite periférica. Evidência radiográfica e ecográfica de condrocalcinose da fibrocartilagem triangular do carpo, meniscos e imagens hiperecoicas intra cartilagíneas em múltiplas localizações sugestivas de deposição de PPC. A doente iniciou terapêutica com colquicina 1mg/dia e uso de ortotese cervical para controlo sintomático – Figuras 1C-1 e 2.

Caso 4: Discite inflamatória: Homem de 78 anos internado por cervicalgia de ritmo inflamatório, artrite de punhos e MCF, disfagia, dispneia, incontinência urinária e febre. Apresentava elevação marcada de parâmetros de fase aguda sem leucocitose. Após investigação exaustiva de etiologia infeciosa e refractariedade a múltiplos antibióticos, iniciou corticoterapia em dose alta com rápida resolução das queixas. A imagem de TC demonstrava achados sugestivos de discite C4-C5 e C5-C6 com exuberante osteofitose condicionando distorção anatómica faríngea. Associadamente com calcificação periodontoideia e ligamento longitudinal posterior – Figuras 1D-1 e 2.

REFERÊNCIAS

1. Moshrif, A., Laredo, J., Bassiouni, H., Abdelkareem, M., Richette, P., Rigon, M., & Bardin, T. (2019). Spinal involvement with calcium pyrophosphate deposition disease in an academic rheumatology center: A series of 37 patients. Seminars In Arthritis And Rheumatism, 48(6), 1113-1126. doi: 10.1016/j.semarthrit.2018.10.009
2. Rosenthal, A., & Ryan, L. (2016). Calcium Pyrophosphate Deposition Disease. New England Journal Of Medicine, 374(26), 2575-2584. doi: 10.1056/nejmra1511117

223 - LATE ONSET PERIPHERAL SPONDYLARTHROPATHY (LOPS) – A RARE AND CHALLENGING DIAGNOSIS

Vanessa Fraga[1], Catarina Abreu[1], Sandra Sousa[1]
[1]Reumatology Department, Hospital Garcia de Orta, EPE, Almada, Portugal

Introduction: Spondylarthropathies are generally observed in young male patients, but can also occur in older patients. Late-onset peripheral spondylarthropathy (LOPS) was first described in 1989 by Dubost and Sauvezie1. In their series, the patients were all male, HLA-B27 positive and the disease was characterized by oligoarthritis, moderate involvement of the axial skeleton, pitting oedema of the lower limbs, poor response to NSAIDs and elevated inflammatory markers. There are no specific radiological abnormalities.2

Case report: A 75-year-old men presented with a one month history of cervical spine inflammatory pain and prolonged morning stiffness with partial response to NSAIDs. The patient denied fever, anorexia, weight loss, cutaneous lesions, arthralgia, inflammatory back or hip pain, headache, visual disturbances, jaw claudication or other organ-specific symptoms.

The physical exam showed limited cervical spine movements, but otherwise was unremarkable. Laboratory results revealed markedly elevated inflammatory markers (ESR 120mm/h and CRP 7.30 mg/dl), negative auto-immunity tests and HLA-B27 positivity. Cervical, dorsal and lumbar spine x-ray revealed multiple de-

generative alterations and hip x-ray bilateral narrowing across the sacroiliac joints. CT scan showed "Continuous dense lamina along the anterior aspect of the dorsal vertebrae, which may correspond to extensive degenerative or syndesmophytic changes"

Assuming the probable diagnosis of polymyalgia rheumatica, the patient started prednisolone 15mg/day, but had limited response. Thoraco-abdomin-pelvic CT scan, echocardiography and PET-scan were performed and excluded malignancy, infection and vasculitis.

In an eight-month period, the patient developed asymmetrical oligoarthritis of the ankle, wrist and two metacarpophalangeal joints as well soft tissue swelling with pitting oedema of the left ankle and dactylitis of the 2nd finger of the left hand. There was no uveitis, enthesitis, intestinal bowel disease, psoriasis or family history of psoriasis. The diagnosis of LOPS was made and patient started a cDMARD (methotrexate).

Conclusion: LOPS is characterized by late onset of oligoarthritis, pitting oedema of the lower limbs, moderate involvement of the axial skeleton and markedly elevated inflammatory markers. A clinical presentation such as LOPS is a challenging diagnosis, as the clinician may contemplate other diagnosis such as polymyalgia rheumatica, RS3PE and even rheumatoid arthritis.

REFERENCES:

1. Dubost JJ, Sauvezie B. Late onset peripheral spondyloarthropathy. J Rheumatol. 1989 Sep;16(9):1214-7. PMID: 2810278.
2. Toussirot E. Late-onset ankylosing spondylitis and spondylarthritis: an update on clinical manifestations, differential diagnosis and pharmacological therapies. Drugs Aging. 2010 Jul 1;27(7):523-31. doi: 10.2165/11315970-000000000-00000. PMID: 20583847.

224 - DOENÇA DE DERCUM COMO MIMETIZADORA DE FIBROMIALGIA

Maria Seabra Rato[1], Alexandra Bernardo[1], Filipe Oliveira Pinheiro[1], Lúcia Costa[1]

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

Introdução: A doença de Dercum (DD) é uma doença rara, de etiologia desconhecida, caracterizada por lipomas subcutâneos dolorosos. Está associada ao excesso de peso/obesidade e a vários distúrbios psiquiátricos, tais como ansiedade, depressão e alterações do sono. O seu diagnóstico é baseado na apresentação clínica e na exclusão de outros distúrbios associados a lipomas. O diagnóstico diferencial entre fibromialgia (FM) e DD pode ser difícil de estabelecer, uma vez que ambas se caracterizam por dor crónica, principalmente no doente obeso.

Caso clínico: Doente do sexo feminino, de 58 anos, foi enviada à consulta de Reumatologia por dor músculo-esquelética generalizada incapacitante, fadiga e parestesias dos membros com cerca de 6 anos de evolução. Apresentava como antecedentes pessoais de relevo diabetes mellitus tipo 2, dislipidemia e perturbação de ansiedade. Relativamente aos antecedentes cirúrgicos destacava-se história de exérese de lipoma intramuscular da coxa direita, com 6 cm de maior eixo, que recidivou 2 anos após a excisão. A doente havia sido observada, 3 anos antes, em consulta de Neurologia, onde foi diagnosticada FM e medicada com pregabalina e tramadol. Por ausência de melhoria clínica suspendeu estes fármacos. Ao exame objetivo, os sinais vitais eram normais, o peso de 80kg e o índice de massa corporal de 30.9, correspondendo a obesidade. Apresentava hiperalgesia à palpação das massas musculares e eram palpáveis 4 pequenas formações nodulares dolorosas, de limites bem definidos e consistência mole, na face medial do braço e antebraço esquerdo, face ântero-lateral da coxa esquerda e região occipital. Não eram visíveis lesões cutâneas e o exame neurológico não apresentava alterações. Analiticamente, o hemograma, os reagentes de fase aguda, as enzimas musculares, o cálcio sérico e a função tiroideia não evidenciavam alterações. A eletromiografia dos membros superiores e inferiores era normal. Realizou ecografia da coxa esquerda que revelou múltiplas formações nodulares de aparência lipomatosa no tecido celular subcutâneo e ecografia mamária (por mastalgia bilateral) que também mostrou várias formações nodulares sólidas e de aparência lipomatosa. A apresentação clínica, juntamente com os achados dos exames complementares foram compatíveis com DD. A doente foi medicada com duloxetina e paracetamol e incentivada para a prática regular de exercício físico, com discreto alívio das queixas, mantendo seguimento em consulta.

Conclusão: A DD é uma doença enigmática, com vários aspetos da sua etiopatogenia, apresentação clínica e terapêutica ainda não totalmente esclarecidos. Devido à sobreposição de sintomas é muitas vezes confundida com FM. Embora as diferenças entre estas duas entidades possam ser muito subtis, deve suspeitar-se de DD em doentes com nódulos dolorosos no tecido subcutâneo. Atualmente o tratamento tem como principais objetivos o alívio da dor e a recuperação de uma aparência normal (com procedimentos cirúrgicos e/ou de lipoaspiração). Este caso alerta para a importância do reconhecimento desta entidade nosológica, assim como para necessidade de realização de estudos que levem à sua melhor compreensão e abordagem.

227 - THE WINDOW OF OPPORTUNITY IN RHEUMATIC DISEASES: MORE THAN A THEORETICAL CONCEPT

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

[1]Rheumatology Department, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal

Background: The therapeutic "window of opportunity" is a critical concept in rheumatoid arthritis (RA) management. It states that early treatment can change the outcome of the disease. Some studies suggest that the concept of a "window of opportunity" is relevant not only to RA but also to spondyloarthritis. We describe 3 case reports in which this window of opportunity may have been missed.
Clinical cases: The first description relates to a 74-year-old male, diagnosed with ankylosing spondylitis. He presented to our department, at the age of 70, with a history of inflammatory back pain since the age of 20. He also had symptoms suggestive of Achilles tendon enthesopathy, and longstanding inflammatory knee and hip pain. He was never previously assessed by any doctor for this reason and had never taken nonsteroidal anti-inflammatory in full dose.

On the physical examination, the patient presented loss of lumbar lordosis, thoracic hyperkyphosis and anterior projection of the neck with markedly decreased spine range of motion. He also had limited range of movement on hips and crepitus in the knees. The results of his analyses revealed slight increase in erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) and positive HLAB27. Plain radiographs showed "bamboo spine" appearance, bilateral sacroiliitis grade IV, ankylosis of the coxofemoral joints and uniform joint space narrowing with bony proliferation in the knees. Some of these radiographic features are illustrated on figure 1 A-C.

The second case report refers to a 62-year-old woman diagnosed with psoriatic arthritis at the age of 32 with axial and peripheral joint involvement.

Since diagnosis, her behavior was marked by non-adherence to medication, both regarding the onset of new drugs and also with lack of treatment persistence.

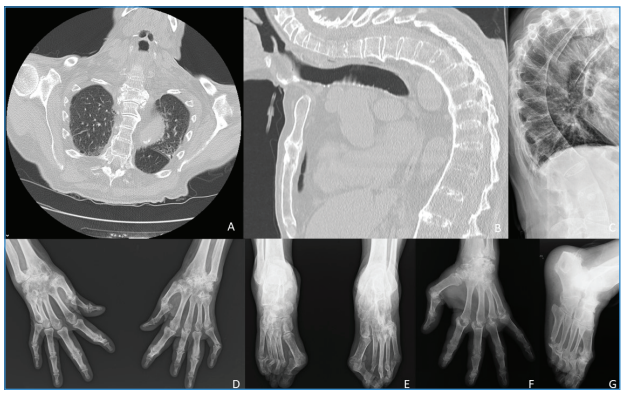


Figure. Axial and sagittal CT scans (A-B) and X-rays (C-G) of patients in which the window of opportunity may have been missed.

Thus, several treatments, including methotrexate (MTX), leflunomide, sulfasalazine and ciclosporin were ineffective or not tolerated. Many years after being proposed, she accepted adalimumab treatment, however, paradoxical worsening of cutaneous psoriasis was noted. Thereafter, biological treatment was changed to secukinumab, yet again discontinued a few months later by her own initiative. Joint damage, such as multiple erosions, "pencil-in-cup" changes and joint subluxations, are shown in figure 1 D-E.

The last report refers to a 55-year-old man with a diagnosis of RA. He described pain and swelling of small and medium joints by the age of 35. The first evaluation in our department was only at 44 years of age, with a clinical presentation of polyarthritis, and already with established joint deformities. Laboratory results showed increased ESR and CRP and positive rheumatoid factor (710 UI/mL) and anti-citrullinated protein antibodies (1432 UI/mL). Some of his plain radiographies are shown in figure 1 F-G.
Discussion: The treatment of rheumatic inflammatory diseases has made significant progresses in the last decades. Markers of poor prognosis and consequent destructive course are being increasingly identified. Early identification of these patients and timely treatment start can mitigate joint damage. Beyond that, treatment adherence in these diseases remains an ongoing challenge for successful outcome.

229 - ANCA-ASSOCIATED VASCULITIS: REPORT OF A CASE WITH INAUGURAL PULMONARY HAEMORRAGE AND LITERATURE REVIEW

Beatriz Mendes[1], Adriana Carones[1], João Rovisco[1, 2], Margarida Coutinho[1, 2], Maria João Salvador[1, 2], Tânia Santiago[1, 2]

[1]Serviço Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [2]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal

Introduction: Antineutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis (AAV) frequently involve the lung. However, diffuse alveolar hemorrhage (DAH) is a rare manifestation and is exceptionally the first manifestation of the disease. Our case report and literature review provide insight into the epidemiology and management of this severe condition.
Case report: A 52-year-old previously healthy female was referred to the emergency department due to de novo microcytic hypochromic anemia, with hemoglobin of 7.0 g/dL. She reported dry cough, bloody nose discharge and nasal obstruction for 3 months. She denied dyspnea, hemoptysis, or chest pain. She also referred diffuse pain in her limbs, worse in the evening,

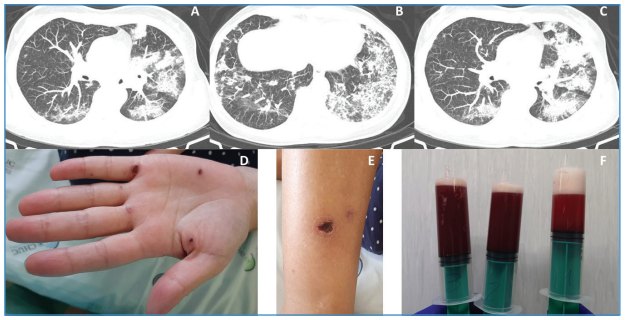


Figure 1. A, B and C: Chest CT scans showing ground glass opacities and heterogeneous parenchymal consolidations; D and E: Purpuric and ulcerated skin lesions in the left hand and in the right leg. F: Bronchoalveolarlavage revealing a progressively (right to left) hemorrhagic fluid.

associated with distal paresthesia. Moreover, she described weight loss of 7%, loss of appetite and fatigue, with no fever or night sweats. On physical examination she was breathing normally on room air and had normal vital signs. Purpuric and ulcerated skin lesions in her hands and right leg were noted. Lung auscultation revealed fine crackles. She had distal grade 4+ muscle weakness in both her upper and lower right limbs and a decreased pain sensitivity in the lateral side of her right leg. Laboratory tests revealed a leucocyte count of 13100 x106/L, with 3970 x106/L eosinophils, serum creatinine 1.22 mg/dL, erythrocyte sedimentation rate 99 mm/h and C-reactive protein 11.66 mg/dL. Chest radiography showed diffuse alveolar opacities. She was admitted to the rheumatology department due to the hypothesis of AAV associated DAH. Chest computerized tomography (CT) showed ground glass opacities and heterogeneous parenchymal consolidations, suggesting DAH. Bronchoscopy with bronchoalveolar lavage (BAL) revealed a progressively hemorrhagic fluid, with 40% hemosiderin-laden macrophages, confirming the diagnosis of DAH. The diagnostic workup revealed positivity for perinuclear ANCA and anti-proteinase 3 antibodies (96 IU/mL). Pulmonary function tests were normal and the diffusing capacity for carbon monoxide was moderately reduced (57% of predicted). The urinalysis revealed hematuria with proteinuria (urinary protein/creatinine ratio of 2618 mg/g). Renal biopsy demonstrated crescentic glomerulonephritis with mild interstitial fibrosis; immunofluorescence was inconclusive due to the low number of glomeruli in the biopsy specimen. Electromyography showed a sensorimotor polyneuropathy. CT scan of the paranasal sinuses revealed inflammatory filling of the right frontal sinus. Electrocardiography and transthoracic echocardiography were normal.

A diagnosis of granulomatosis with polyangiitis (GPA) was established. The patient received pulse intravenous methylprednisolone at a dose of 1 g/day for three days, followed by oral prednisolone 1 mg/kg/day. Remission induction was started with pulsed intravenous cyclophosphamide (CYCLOPS regimen), with gradual improvement of symptoms, laboratory, and radiologic parameters. After 6 months of follow-up, the patient remains asymptomatic and mycophenolate mofetil was started.

Conclusion: DAH is a prominent and life-threatening pulmonary manifestation of AAV. From a diagnostic reasoning point of view, this case exemplifies the importance of pursuing a unifying diagnosis in a patient who has developed symptoms in multiple organ systems over a short period of time. An active diagnostic workup, intensive observation, and aggressive immunosuppressive treatment are cornerstones of the management of this condition.

232 - O PAPEL DAS IMUNOGLOBULINAS INTRAVENOSAS NO SÍNDROME ANTI-SINTETASE

Hugo Parente[1], Catarina Soares[1], Maria Pontes Ferreira[1], Emanuel Costa[2], Francisca Guimarães[1], Soraia Azevedo[1], Daniela Santos-Faria[1], Daniela Peixoto[1], José Tavares-Costa[1], Carmo Afonso[1], Filipa Teixeira[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: O síndrome anti-sintetase (SAS) pertence às miopatias inflamatórias, podendo apresentar-se com doença pulmonar intersticial (DPI), artrite, fenómeno de Raynaud, febre e mãos de mecânico. É uma entidade rara e pouco estudada, cujas opções terapêuticas se extrapolam de relatos de casos, estudos pequenos e de outras miopatias inflamatórias. Para a miosite, conhecem-se dados1 que apontam um benefício do uso de imunoglobulinas intravenosas (IgIV). Os anticorpos anti-PL7 e PL122 têm maior associação a DPI e pior prognóstico.

Objetivo: Apresentação de um caso de SAS grave, refratário ao tratamento com vários imunomoduladores, e com resposta positiva às IgIV.

Homem de 45 anos, enviado a consulta de Reumatologia, após internamento em Medicina Interna, por quadro de astenia, febre, perda ponderal, fraqueza muscular, dispneia, tosse, e artralgias periféricas e assimétricas, de ritmo misto. Após exclusão primordial de COVID-19 e tuberculose, foi interpretado como de etiologia infeciosa indeterminada, mas dada a refratariedade a vários antibióticos, foram investigadas outras causas. Nessa circunstância, iniciou corticoterapia (CCT - 1mg/kg/dia), com melhoria acentuada. Os exames efetuados permitiram um diagnóstico de SAS – VS 65mmHg, PCR 13mg/dL, aumento de enzimas musculares (creatinaci-

nase – CK - e mioglobina na ordem dos 2000U/L e aldolase 15 UI/L), alterações inflamatórias residuais (pelos CCT já iniciados) em ressonância magnética (adutores e retos femorais), eletromiografia e biópsia musculares compatíveis, tomografia torácica e biópsia pulmonar com pneumonia organizativa, alterações hipoxémicas gasimétricas e funcionais respiratórias (síndrome restritivo e DLCO 37%), imunologia com ANA positivos (título 1/320), anti-Ro52 e anti-PL7 positivos. Iniciou, pelo envolvimento pulmonar, micofenolato de mofetil, mas com o desmame de CCT, reagravou o seu estado clínico, tendo tido indicação para pulsos de CCT e ciclofosfamida. A resposta muscular foi satisfatória, contudo, do ponto de vista respiratório, a melhoria foi parca. Do seguimento em consulta de Pneumologia, foi otimizada a oxigenoterapia (O2) de deambulação e CPAP noturno. Nesta conjuntura, iniciou tratamento com rituximab (RTX). Apesar de sintomaticamente estável, piorou a DLCO para 23%. Ao fim de 4 meses do primeiro ciclo, voltou a haver recidiva da componente muscular (CK 1009). Por essa razão, e aguardando o 2º ciclo de RTX, realizou tratamento com imunoglobulinas intravenosas (IgIV - 400mg/kg/dia durante 5 dias), por 2 períodos intervalados de 15 dias, com benefício parcial (CK 454). Passados 2 meses, ocorreu nova recidiva (CK 1493), e nova intervenção com IgIV, com normalização das enzimas musculares, associado a melhoria respiratória sintomática e possibilidade de redução do débito de O2 suplementar, pelo que tem mantido regime mensal de IgIV.

Conclusão: Este caso evidencia o potencial terapêutico e poupador de CCT das IgIV, no componente inflamatório muscular e pulmonar do SAS, nomeadamente após falência de vários imunomoduladores.

REFERÊNCIAS

1. Farini A, Villa C, Tripodi L, Legato M, Torrente Y. Role of Immunoglobulins in Muscular Dystrophies and Inflammatory Myopathies. Front Immunol. 2021 Jul 14;12:666879. doi: 10.3389/fimmu.2021.666879. PMID: 34335568; PMCID: PMC8316973.
2. Jiang M, Dong X, Zheng Y. Clinical characteristics of interstitial lung diseases positive to different anti-synthetase antibodies. Medicine (Baltimore). 2021;100(19):e25816. doi:10.1097/MD.0000000000025816.

248 - SHRINKING LUNG SYNDROME IN SYSTEMIC LUPUS ERYTHEMATOSUS

Adriana Carones[1], Beatriz Mendes[1], João Rovisco[1], Ines L[1]

[1]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

Introduction: Shrinking Lung Syndrome (SLS) is a rare complication of systemic autoimmune diseases, most commonly Systemic Lupus Erythematosus (SLE). Its pathophysiology is largely unknown but may involve respiratory muscle weakness, phrenic nerve dysfunction and diaphragmatic myopathy. Pulmonary function tests (PFT), chest x-ray and computed tomography (CT) can be helpful to identify SLS.

The authors report a case of a patient with SLE who developed SLS.

Clinical Case: A 66 year-old woman with SLE for 24 years, with previous hematologic, musculoskeletal and mucocutaneous involvement, treated with prednisolone 5mg/day, methotrexate and belimumab, presented with progressive shortness of breath, orthopnea, paroxysmal nocturnal dyspnea and pleuritic chest pain. Prominent findings on physical examination included polypnea, decreased breath sounds and tele-inspiratory crackles on both lung bases.

Chest radiography and CT revealed bilateral decreased lung size, elevated right hemidiaphragm and minimal pleural effusion at the right costophrenic recess. PFT results were compatible with a restrictive pattern. These findings led to the diagnosis of SLS. Treatment was started with iv pulses of 500mg of methylprednisolone for 3 days, followed by oral prednisolone 0.5mg/kg/day, and a cycle of rituximab (2 doses of 1g with an interval of 15 days). Belimumab was discontinued and reintroduced 4 weeks following rituximab cycle. There was a progressive and sustained clinical improvement, with reduction of SLEDAS from 19.32 to 2.08 at 6 months of follow-up.

Conclusion: It has recently been suggested that the combined use of the two drugs targeting B cells (rituximab as induction) and belimumab (as maintenance therapy) may be more effective than using the drugs individually. The rationale for using two consecutive therapies targeting B cells lies in the observation that serum levels of lymphocyte stimulator are significantly higher and promote disease flares during B cell repopulation after rituximab therapy.

The BEAT-Lupus study, a double-blind, randomized, placebo-controlled, phase II clinical trial aimed to evaluate the safety and efficacy of belimumab after rituximab. The results suggested that belimumab after rituximab is a safe and effective treatment for patients with SLE and further supports the use of this combination as a novel therapeutic strategy.

In our patient we observed clinical remission at 6 months of follow-up after combined therapy with rituximab and belimumab.

249 - ANCA ASSOCIATED VASCULITIS: AN UNUSUAL PRESENTATION

Catarina Abreu[1], Vanessa Fraga[1], Ana Catarina Duarte[1], Alice Castro[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

Vasculitis is a diverse group of disorders where inflammation of the blood vessel walls leads to organ damage. Due to the wide distribution of blood vessels, it can affect any organ with multiple different clinical presentations, which makes its diagnosis a huge challenge.

We present a case of a 70-year-old Caucasian female with previous history of hypertension, dyslipidaemia, gastroesophageal reflux disease and hypothyroidism. Since 2021 she presented with recurrent syncope without prodromes, associated with peripheral vertiginous syndrome, for which she had an extensive etiologic study performed with no significant findings.

In august 2021 she refers hearing loss on the left side and one month later progressive deafness on the right side. One week later she complained of nausea, vertigo and tinnitus, with documented sensorineural hearing loss with peripheral vertigo in an urgent otolaryngology appointment. During observation she had a syncope with sudden visual loss of the left eye. On ophthalmoscopy observation diffuse vascular retinal sclerosis with retinal pallor and cherry red spots were identified, suggestive of retinal artery occlusion. Physical examination was otherwise unremarkable. On work-up she had normocytic/normochromic anemia (haemoglobin 10,1 g/dL), thrombocytosis (604 000/uL), leucocytosis (14200x109); elevated erythrocyte sedimentation rate (ESR) (111 mm 1ºH) and C-reactive protein (CRP) (18,79 mg/dL), creatinine 2,1 mg/dL, and urinalysis with erythrocyturia, leukocyturia and proteinuria. She was admitted for further diagnostic evaluation. Electrocardiogram, transthoracic echocardiogram, chest X-ray, cranioencephalic computed tomography, and lumbar puncture were normal. A brain magnetic resonance was also performed, which showed anterior flattening of the optic papilla with hypersignal in T2/FLAIR. The clinical presentation with new onset elevated creatinine and active urinary sediment was suggestive of rapidly progressive glomerulonephritis. On extensive blood workup she was positive for anti-proteinase 3 anti-neutrophil cytoplasmic antibody with a cytoplasmatic pattern on immunofluorescence. The diagnosis of granulomatosis with polyangiitis (GPA) with vestibulocochlear, retinal and kidney involvement was made. The patient started treatment with intravenous (IV) methylprednisolone 1g/day for 3 days followed by oral prednisolone (PDN) 1mg/kg/day, in association with IV cyclophosphamide. After 5 days her hearing capacity, creatinine (1,6mg/dL) and acute phase reactants (ESR 45 mm 1ªH; CRP 1.19 mg/dL) had substantially improved. Oral PDN was progressively tapered and after 8 months of follow-up she maintains diminished visual acuity on the left side, but kidney function and hearing capacity improved significantly, and no new episodes of vertigo or syncope occurred.

The involvement of the central nervous system with cranial neuropathies is rare in GPA, with sensorineural hearing loss being the most frequent. The presentation with syncope and peripheral vertigo has been reported in few patients, but its physiopathology is still not understood. It is important to distinguish ischemic optic neuropathy from central retinal artery occlusion for the differential diagnosis of the causes of amaurosis fugax, such as large vessel vasculitis.

We want to highlight the importance of recognizing a less common presentation of vasculitis in order to establish an early diagnosis and start prompt treatment.

ARP Rheumatology 2022;3:183-200 (SUP)

Imagens em Reumatologia

001 - SUDDENLY I AM OLD

Ana Margarida Duarte-Monteiro[1, 2], Rita Pimenta[3], Carla Macieira[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, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Dermatologia, Hospital Garcia de Orta E.P.E, Almada, Portugal

Introduction: Hydroxychloroquine is an antimalarial drug that is commonly used in the treatment of Lupus Erythematosus. It has a favorable safety profile and can be used in pregnancy and lactation. A common side effect is hyperpigmentation of skin, sometimes nails and mucosa. It can also affect hair follicles, with hair loss being most frequently reported. Although it is rarely described in the literature, hair bleaching is a possible side effect of both chloroquine and hydroxychloroquine and it is usually reversible.

Clinical vignette: A 57-year-old Caucasian male, previously healthy, presented to the dermatology clinic after developing an erythematous rash affecting his face, torso, and arms. Symptom onset had happened a month earlier and was progressively worsening. He denied any other complaints or having started any new medications before symptom onset. There was no history of fever, night sweats, weight loss or anorexia, as well as no other mucocutaneous manifestations, such as oral ulcers. He denied relevant sicca symptoms, eye complaints or Raynaud phenomena. Erythematous papules and plaques were evident in sun-exposed areas in the face, neckline, hands, and forearms, and physical exam was otherwise unremarkable. The patient was initially treated with oral prednisone 20 mg/day for two weeks, and topical betamethasone 1mg/g once daily, with partial skin improvement. A skin biopsy was performed prior to treatment and was inconclusive. Laboratory evaluation revealed positive ANA with a title of 1/320 and a nuclear speckled pattern, a positive anti-SSA Ro52 with a title of 27348.1 UQ, anti-SSA Ro60 (6115.2 UQ), and anti-SSB La (736.9 UQ). Complement was low (C3c 70 mg/dL; C4 4mg/dl) and anti-dsDNA was negative.

A diagnosis of probable subacute cutaneous lupus was made. Treatment with hydroxychloroquine 400mg/day was started, with significant improvement of skin lesions. Two months after beginning treatment, the patient developed discoloration of hair and all body hair, including lashes and eyebrows, which became white

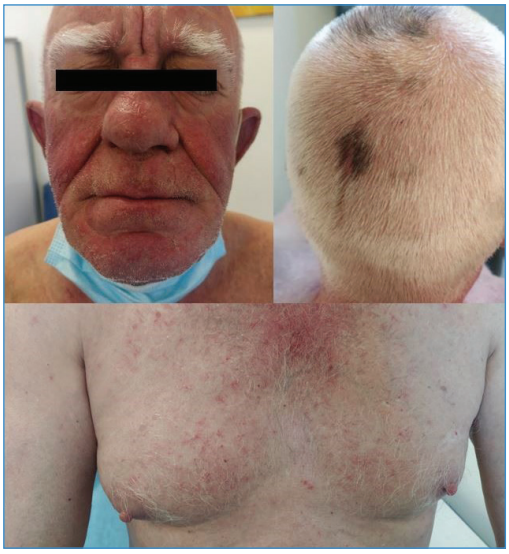


Figure 1. Generalized poliosis induced by hydroxycloroquine

in color. Hydroxychloroquine was discontinued after six months. After 1 year the patient was evaluated by Dermatology and Rheumatology in a multidisciplinary outpatient appointment; even after stopping hydroxychloroquine, the discoloration had not completely regressed, and generalized poliosis was still evident, except for small circular areas in the back of the patient's head that had regained its original brown color. Considering maintenance of bleached hair despite treatment being discontinued for 12 months and worsening of skin lesions, the multidisciplinary decision was to resume hydroxychloroquine.

Conclusions: We present a rare side effect of commonly used hydroxychloroquine. Hair bleaching by hydroxychloroquine was first described in 1948, around the same time antimalarials started being used for the treatment of rheumatic diseases. Despite its rareness, it is a side effect to be considered, especially in young patients, taking into account its aesthetic implications and possibility of irreversibility.

048 - CALCINOSIS IN SYSTEMIC SCLEROSIS – AN EXTREME PORTRAYAL OF A CHARACTERISTIC FINDING

Matilde Bandeira[1, 2], Eduardo Dourado[1, 2]

[1]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

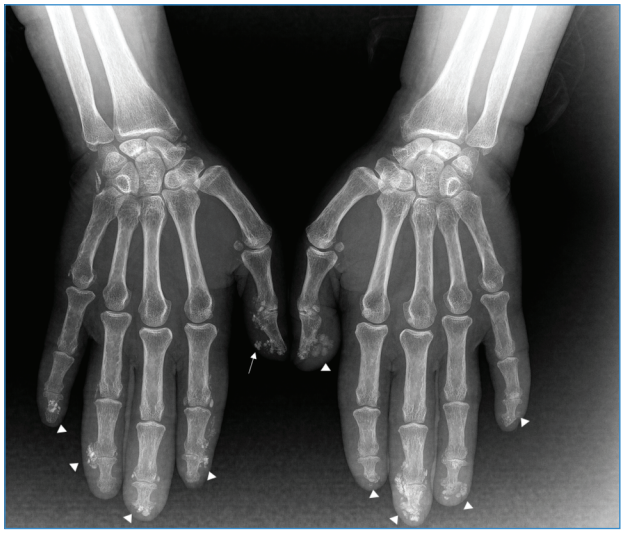


Figure 1. Hand radiographs showing calcinosis in all fingers

Hand radiographs showing calcinosis in all fingers. The patient suffers from anticentromere-positive limited cutaneous systemic sclerosis with severe gastrointestinal involvement, including a complete absence of oesophageal motility on manometry. Calcinosis was palpable on the first right finger, where it seems more superficial on the radiograph (arrow). However, calcinosis was not evident on any other finger by physical examination, but is evident in plain hand radiographs (arrowheads).

056 - LEUKOCYTOCLASTIC SMALL VESSEL VASCULITIS POS-COVID-19 INFECTION

Sofia Ferreira Azevedo[1, 2], Carolina Mazeda[1, 2], Susana P. Silva[1, 2], C. Pinto Oliveira[1, 2], Inês Cunha[1, 2], Manuela Loureiro[2, 3], Anabela Barcelos[1, 2]

[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz Health Alliance, Portugal, Aveiro, Portugal, [3]Dermatologia, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal

Background: Since the beginning of the covid-19 pandemic, multiple dermatological manifestations have been reported, such as urticaria, livedo reticularis, chilblain lesions, and various exanthems. Covid-19 induced cutaneous small vessels vasculitis has also been described in adults and children with recent/ current covid-19 infection. In children or young adults, this condition typically occurs in the presence of mild lung disease.

We present a case of a 17-year-old boy with leukocytoclastic, small vessels vasculitis post-covid-19 infection.

Case description: A 17-year-old otherwise healthy boy presented to the emergency department of our center with a 6-day history of petechial and purpuric skin lesions in the lower limbs, with progressive worsening over the last days (Figure 1). He also had myalgias and fatigue that began 2-weeks before the appearance of the lesions, and swollen and painful right wrist and tibiotarsal joints. He denied weight loss, fever, nocturnal sudoresis, headaches, and abdominal or urinary complaints. He had mild covid-19 infection 2-weeks before the begging of the symptoms. The patient also denied taking any drug or having a history of sick cohabitants.

On physical examination, the patient was hemodynamically stable and apyretic, had a slight bilateral conjunctival hyperemia, lower limbs purpuric lesions, and swollen tibiotarsal and right wrist joints. Laboratory data showed high leucocyte count with monocytosis, high levels of inflammatory markers (erythrocyte sedimentation rate (ESR) 37mm and C-reactive protein (CRP) 2.43 mg/dl), and altered coagulation times (a prothrombin time of 13.70 seconds, an activated partial thromboplastin time of 41.70 seconds, and an international normalized ratio of 1.20). The patient was admitted in our hospital for further study and vigilance.

A complete laboratory study for vasculitis causes (with antinuclear antibodies, antineutrophil cytoplasmatic antibodies, atypical antineutrophil cytoplasmatic antibodies, anti-double-stranded DNA antibodies, rheumatoid factor, immunoglobulins, anti-cardiolipin antibodies, and beta-2-microglobulin, complement C3 and C4, and serologies for cytomegalovirus, Epstein-Barr, human immunodeficiency virus, hepatitis A, B and C virus, and VDRL test) was negative. Urinary sediment was negative for proteins and showed the presence of rare erythrocytes. The total protein/

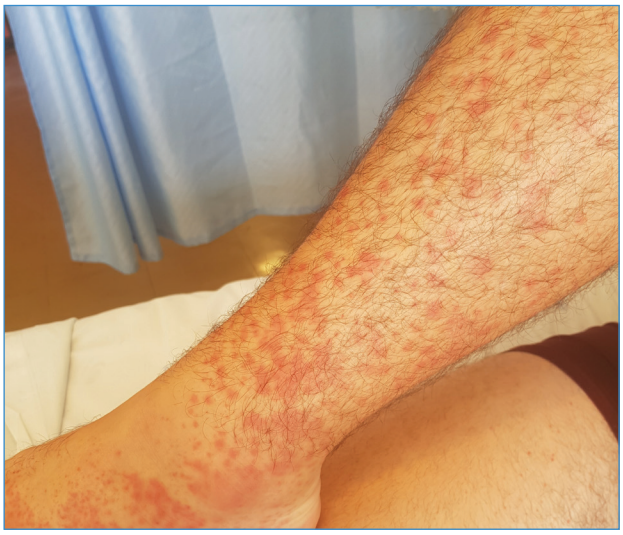


Figure 1. Image depicting palpable purpuric violaceous lesions in the lower limb of a 17-year-old boy after covid-19 infection

creatinine urinary ratio was normal. An abdominal and kidney ultrasound was also performed, with no abnormal aspects found. In collaboration with Dermatology, a biopsy of the skin lesions was also performed.

The patient was discharged after 3-days, with marked improvement of the symptoms and skin lesions.

Histology results of the skin biopsy later revealed the presence of a leukocytoclastic small vessels vasculitis, with negative immunofluorescence for IgA deposits.

Currently, the patient maintains follow-up in our Rheumatology Department and shows complete resolution of the complaints.

Conclusion/discussion: Covid-19 infection, nowadays a frequent disease, may be a cause of leukocytoclastic small vessel vasculitis. Despite that, even with a history of recent covid-19 infection, other causes for this dermatological manifestation must be ruled out before a diagnosis is established.

058 - PROGRESSIVE ACRO-OSTEOLYSIS IN SYSTEMIC SCLEROSIS

Eduardo Dourado[1, 2], Ana Valido[3]

[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 do Litoral Alentejano, Santiago do Cacém, Portugal

A 66-years-old woman presented to the Rheumatology clinic complaining of metacarpophalangeal and proximal and distal interphalangeal arthralgia, Raynaud's phenomenon, pelvic dysfunction, and asthenia. She had no sicca symptoms. On physical examination, the patient had acral telangiectasias, onycholysis, and distal phalange shortening but no swollen joints, skin thickening, calcinosis or digital ulcers. Hand radiographs revealed significant acro-osteolysis (Figure 1 panel A – arrows). HEp-2 immunofluorescence assay was positive for antinuclear antibodies, showing a fine granular pattern. Extractable nuclear antigen antibodies panel was positive for anti-Ro52 antibodies.

Systemic sclerosis sine scleroderma was diagnosed, and the patient was treated with nifedipine. Although Raynaud's phenomenon frequency and severity decreased, a follow-up hand radiograph two years after presentation showed progression of the acro-osteolysis (Figure 1 panel B, arrowheads).

Acro-osteolysis is usually associated with acral ischaemia or severe calcinosis in systemic sclerosis patients. In this case, significant progression occurred, despite no calcinosis and only mild Raynaud's phenomenon.

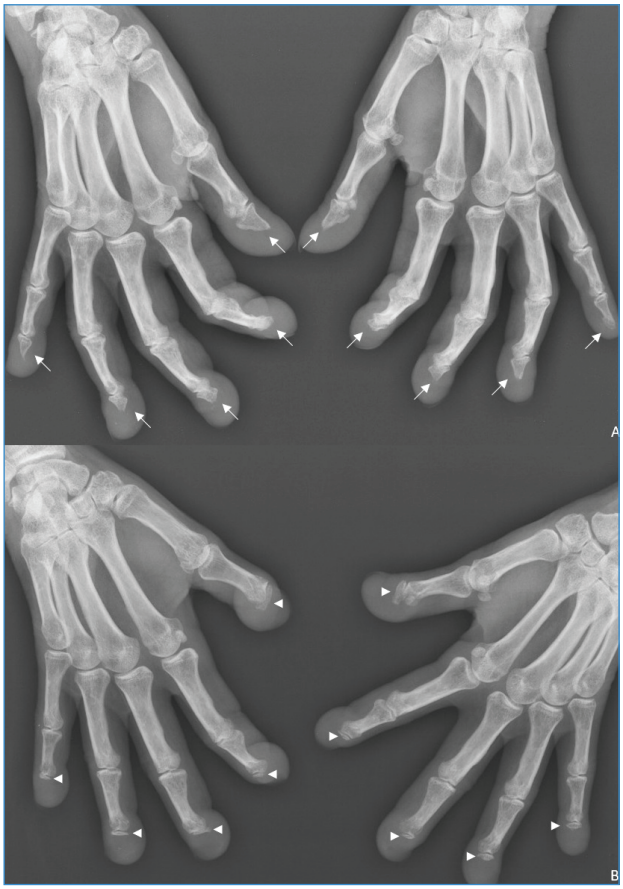


Figure 1. Hand radiographs revealing significant acro-osteolysis

059 - 30 YEARS OF UNATTENDED LOW BACK PAIN: AN IMAGE WE SHOULD NOT SEE IN THE 21ST CENTURY

Eduardo Dourado[1, 2], Ana Valido[3]

[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 do Litoral Alentejano, Santiago do Cacém, Portugal

A 74-year-old man presented to the Rheumatology clinic with a 30-year history of inflammatory back pain that improved with non-steroid anti-inflammatory drugs (NSAIDs). On physical examination, the patient had severely impaired cervical and lumbar rotation, flexion, and extension, as well as cervical hyperkyphosis and lumbar lordosis rectification. Radiographic findings included vertebral fusion by syndesmophytes (bamboo spine, Figure 1 panel A – arrows), ossification of the supraspinous and interspinous ligaments (dagger sign, Figure 1 panel B – arrowheads), and complete

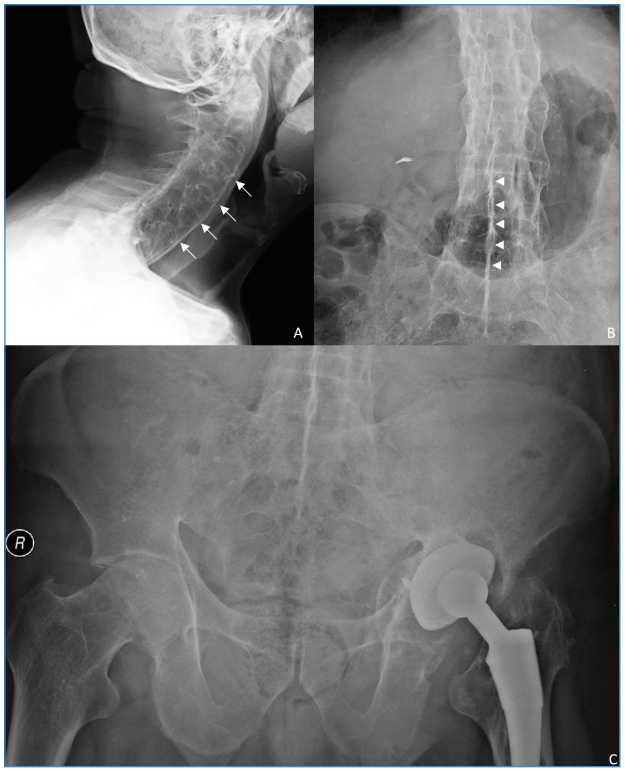


Figure 1. Classical spondyloarthritis radiographical findings.

ankylosis of the sacroiliac joints (ghost joints, Figure 1 panel C). A diagnosis of radiographic axial spondyloarthritis (axSpA) was made.

Patients with untreated axSpA may develop structural changes that manifest as typical radiographical signs. These changes are irreversible but can be prevented by timely treatment of the disease. Therefore, early diagnosis is critical for preventing these highly disabling lesions.

088 - FREIBERG DISEASE: A RARE CAUSE OF CHRONIC FOOT PAIN

Daniela Santos Oliveira[1, 2, 3], Lúcia Costa[1], Carlos Vaz[1,2, 3]
[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]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal

Non-weightbearing (Figure 1A) and weightbearing (Figure 1B) anteroposterior radiographs demonstrated a Freiberg Disease in a 78-year-old female patient with forefoot chronic pain, swelling and restricted motion of the third metatarsophalangeal joint. Laboratory results revealed normal inflammatory parameters. Radiographs images showed avascular necrosis of right third metatarsal head. The patient was improved with daily

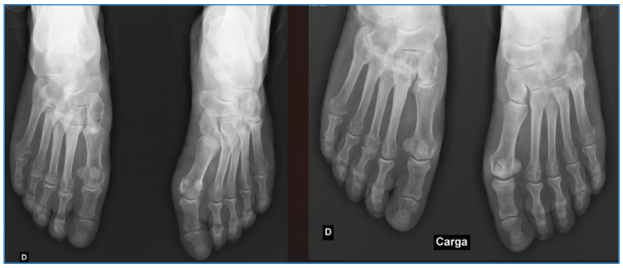


Figure 1. Non-weightbearing (Figure 1A) and weightbearing (Figure 1B)anteroposterior radiographs demonstrated a Freiberg Disease

activity and shoe wear modifications combined with an oral nonsteroidal anti-inflammatory drug. Freiberg disease is a rare clinical condition chatacterized by avascular necrosis of metatarsal head, most commonly the second metatarsal. The etiology of this condition is multifactorial, involving traumatic causes, vascular compromise and systemic disorders such as systemic lupus erythematosus. The differential diagnostic based on clinical presentation and radiograph findings is crucial and include stress fracture, neuroma, rheumatoid arthritis and gout. Conservative management, namely rest, activity and/or shoes modifications and analgesia aims to control pain and prevent progression. However, when these interventions are ineffective, surgical treatment may be indicated.

102 - OSTEOPOIQUILOSE: ACHADO INCIDENTAL

António Barreto Violante[1], Ana Filipa Rocha Águeda[1], Miguel Guerra[1], José Marona[1], Joana Ramos Rodrigues[1], Margarida Oliveira[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário Cova da Beira, Covilhã, Portugal

Mulher de 26 anos, inicialmente observada em Medicina Interna por noção da própria de aumento ganglionar ao nível das cadeias inguinais e cervicais com cerca de 1 mês de evolução e lombalgia mecânica ocasional. Sem outra sintomatologia acompanhante, nomeadamente constitucional, que pudesse sugerir causa sistémica, neoplásica ou infeciosa. Apesar das queixas veiculadas, ecograficamente sem critério de adenopatias. Foi referenciada à consulta de Reumatologia após realização de uma tomografia computorizada (TC) axial com evidência de múltiplas lesões osteoescleróticas ovaladas na bacia e fémur (figura 1), sugestivas de osteopoiquilose, uma displasia óssea rara, hereditária, benigna e de diagnóstico frequentemente incidental em exames de imagem.

Na observação em consulta de Reumatologia, sem alterações ao exame objetivo ou no estudo analítico.

O caso em apreciação demonstra a importância de estar alerta para a existência desta doença na análise de lesões

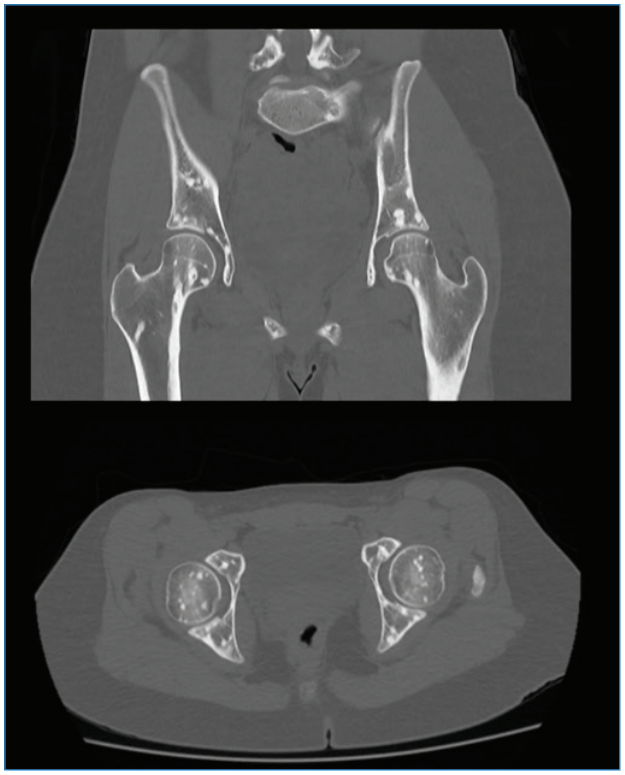


Figure 1. Osteopoiquilose: achado incidental

ósseas incidentais, recorrendo à conjugação dos achados clínicos e imagiológicos no sentido de evitar procedimentos diagnósticos e/ou terapêuticos excessivos.

104 - HIPERPIGMENTAÇÃO GENGIVAL – EVENTO INVULGAR MAS CONSTRANGEDOR

Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Tiago Beirão[1], Joana Aleixo[1]

[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Mulher de 76 anos, seguida em consulta de Reumatologia por Artrite Reumatóide com 3 anos de evolução, reporta o aparecimento de hiperpigmentação da mucosa oral. As lesões, inicialmente punctiformes, passaram despercebidas durante a avaliação médica pelo uso da máscara e pelo facto de a doente não ter feito referência às mesmas. No espaço de 6 meses as lesões evoluíram com atingimento de praticamente toda a região gengival superior anterior. Aquando da consulta subsequente, a doente foi referenciada com urgência a Estomatologia onde foi submetida a biópsia da mucosa cujo resultado histológico revelou nevo melanocítico juncional por provável toxicidade medicamentosa, secundária a metotrexato e/ou a hidroxicloroquina, iniciados há 2 e 3 anos, respetivamente. Os dois fármacos foram suspensos e substituídos por leflunomida 15 mg id. A doente

Figura 1. Hiperpigmentação gengival em doente com Artrite Reumatóide

apresentou melhoria progressiva da hiperpigmentação, atualmente com lesões apenas na periferia dos dentes incisivos superiores (figura).

O aparecimento de nevos melanocíticos na mucosa oral é pouco frequente [1]. A hiperpigmentação da mucosa oral pode ser induzida por vários tipos de fármacos, estando descritos casos associados a hidroxicloroquina e a metotrexato[2]. A sua patogénese não está bem definida e o diagnóstico é complexo, sendo importante excluir outros diagnósticos diferenciais como malignidade[2]. A melhoria do quadro após suspensão dos fármacos suporta a etiologia iatrogénica.

REFERÊNCIAS

1. M Lambertini, A Patrizi, PA Fanti, B Melotti, U Caliceti, C Magnoni, C Misciali, C Baraldi, GM Ravaioli, E Dika. J Eur Acad Dermatol Venereol 2018 Feb;32(2):209-214
2. García RMG, Molina SG. Drug-Induced Hyperpigmentation: Review and Case Series. The Journal of the American Board of Family Medicine Jul 2019, 32 (4) 628-638

115 - SKIN THICKENING – JUST SCLERODERMA OR ANOTHER DISEASE?

Maria Helena Lourenço[1, 2], Ana Bento da Silva[1, 2], Laura Gago[2], Maria João Gonçalves[2], Jaime C. Branco[1, 2, 3], Inês Silva[2, 3]

[1]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [2]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [3]NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

A thirty-year-old male, evacuated from Guinea Bissau, was admitted to the Rheumatology inward due to mechanic polyarthralgias and skin thickening (hands, elbows, knees – figures 1-8). The patient also reported muscle weakness since childhood and skin depigmentation over the metacarphalangeal and interphalangeal joints and elbows. Examination revealed skin thickening, sclerodactyly, lesions similar to Gottron papules and generalized muscle atrophy with normal muscle strength. Inflammatory markers were negative and there was a positive anti-NXP2 antibody. Juvenile dermatomyositis was assumed. Methotrexate 15mg/week

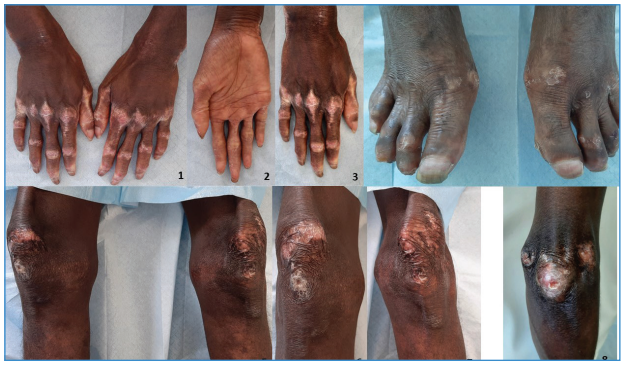


Figura. 1-3 - Dorsal and palmar views of hands; 4 - Dorsal anterior feet; 5-7 - Knees; 8- Elbow

and prednisolone 5mg/day were started, with no improvement of the lesions or pain, so their suspension was decided. The patient started physical therapy. This case reinforces the need of an early diagnosis as the delay may convey irreversible outcomes.

116 - DESTRUCTIVE RHEUMATOID ARTHRITIS: AN INITIAL PRESENTATION OF A YOUNG PATIENT

Maria Helena Lourenço[1, 2], Ana Bento da Silva[1, 2], Laura Gago[2], Jaime C. Branco[1, 2, 3], Inês Silva[2, 3]

[1]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [2]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [3]NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

A previously healthy thirty-one-year-old female presented to the Rheumatology outpatient clinic with inflammatory polyarthralgias of the small joints of the hands, wrists and feet since she was 18 years-old, accompanied by morning stiffness lasting more than one hour. She had been previously treated with non-steroidal anti-inflammatory drugs. Physical exam revealed polyarthritis of the small joints of the hands (metacarpophalangeal and interphalangeal joints), wrists, knees and tibiotarsal joints. There was a clear limitation of the flexion of the wrists (about 20º). Blood analysis showed negative inflammatory markers and a both positive rheumatoid factor and anti-cyclic citrullinated peptides antibody. The wrists x-ray showed major erosion with loss of the ulnar styloid process, narrowing of the joint space and erosions of the carpal bones (figures 1,2). A diagnosis of erosive and seropositive rheumatoid arthritis was made. Subcutaneous methotrexate 15mg per week and prednisolone 15mg per day in a weaning scheme were started, achieving low activity disease. The patient was referred to the Orthopaedics outpatient

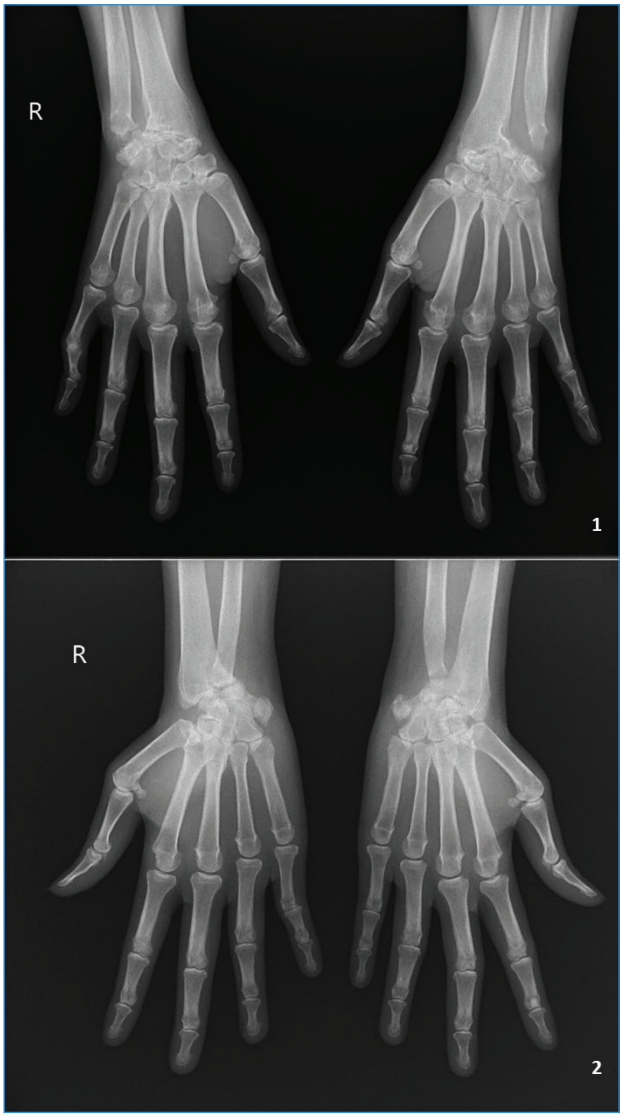


Figure. Radiograph of both hands (2 incidences)

department to evaluate the need of arthrodesis of the wrists. This case reports an advanced and destructive stage of rheumatoid arthritis, which is nowadays rare, and which presented as an initial evaluation of a young female, enhancing the need of being aware of the symptoms and the importance of early referral and treatment.

117 - SKIN RHEUMATOID NODULES: A COMPLICATION OF THE DISEASE OR THE THERAPY?

Maria Helena Lourenço[1, 2], Ana Palmeiro[3], Alexandre Catarino[3], Isabel Viana[3], Jaime C. Branco[1, 2, 4], Inês Silva[2, 4]

[1]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal, [2]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz

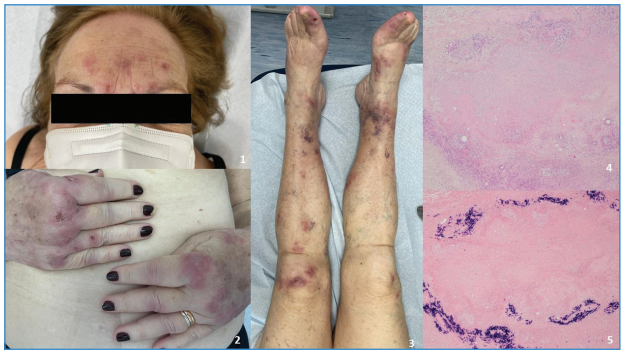


Figure. 1-3 – Erythematous nodules on the patients' forehead, hands and legs; 4 – Histopathological features of punch biopsies

EPE, Lisboa, Portugal, [3]Dermatology, Centro Hospitalar de Lisboa Ocidental EPE, Hospital de Egas Moniz, Lisboa, Portugal, [4]NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

A 79-year-old woman with a 26-year history of rheumatoid arthritis (positive for both rheumatoid factor and anti-cyclic citrullinated peptides antibody) and under methotrexate (MTX) 15mg/week, leflunomide 10mg/day and prednisolone (PDN) 2.5mg/day, with low disease activity, presented to the outpatient clinic with a 2-month history of a progressive disseminated pruritic dermatosis. Physical exam showed a disseminated bilateral and symmetrical dermatosis, with numerous erythematous papules, plaques, infiltrated nodules, and traumatic excoriations, mainly affecting the face and extensor surface of the limbs (elbows, forearms, legs, and hands) (figures 1-3). A high erythrocyte sedimentation rate was seen, with no others relevant changes. Histopathology revealed a very extensive confluent area of necrobiosis associated with palisading of histiocytes, giant multinucleated cells, lymphoplasmacytic infiltrate and an increased number of neutrophils with nuclear dust (figures 4-5). Other aetiologies were excluded, as perforating annular granuloma and erythema elevatum diutinum. Furthermore, chromogenic in situ hybridization showed kappa chains restriction, that, together with an increased kappa chain IgG in immunofixation, led to an investigation to exclude monoclonal gammopathy. Other aetiologies, such as vasculitis, infection or paraneoplasic, were also excluded. A diagnosis of MTX-induced accelerated nodulosis was assumed. Methotrexate was suspended, and prednisolone was increased to 20mg/day, with a great response after four months allowing tapering of prednisolone to 10mg/day. Accelerated nodulosis is a recognized complication of some immunomodulatory drugs. Further investigation is needed to better understand the physiopathology and enable the creation of universal guidelines.

121 - MIOSITE OSSIFICANTE TRAUMÁTICA

Laura Gago[1, 2], Maria Manuela Costa[1], Jaime C. Branco[1, 2], Ana Filipa Mourão[1, 2, 3]

[1]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [2]CEDOC, NOVA Medical School. Faculdade de Ciências Médicas da Universidade NOVA de Lisboa., Lisboa, Portugal, [3]Comprehensive Health Research Center (CHRC), Universidade NOVA de Lisboa, Portugal, Lisboa, Portugal

Doente, 70 anos, com história de acidente de viação do qual resultou fratura da tíbia direita em 2015, enviada à consulta de reumatologia por dor na face anterior da perna direita com irradiação à tibio-társica.

À observação a doente apresentava apenas dor à palpação da face anterior da perna direita, sem deformações ou massas associadas. Não apresentava atrofias musculares, ou défices de força ou da sensibilidade, nem articulações tumefactas ou dolorosas.

Foi solicitado estudo radiográfico da perna direita onde era evidente uma imagem hiperdensa nos tecidos moles entre a tíbia e o perónio (Figura 1). Foi solicitada TC da perna que mostrou extensa calcificação que envolvia o corpo muscular do tibial anterior e o longo extensor dos dedos com atrofia muscular associada. Estas alterações eram compatíveis com miosite ossificante, consequência do traumatismo prévio. Analiticamente não apresentava alterações e não apresentava presença de anticorpos anti-miosites negativos. Foi medicada com naproxeno 500 mg e iniciou fisioterapia dirigida com melhoria da sintomatologia.

A miosite ossificante ou ossificação heterotópica é uma ossificação do músculo que se desenvolve raramente após traumatismo com contusão ou estiramento das fibras musculares estando o seu desenvolvimento relacionado com a gravidade do trauma. Ocorre mais frequente no quadricípite e bicípite. É importante fazer o diagnóstico diferencial com osteossarcoma. Recomenda-se a realização de fisioterapia e o uso de anti-inflamatórios não esteróides. Se a lesão for muito extensa e causar dor incapacitante está recomendada a sua excisão cirúrgica.

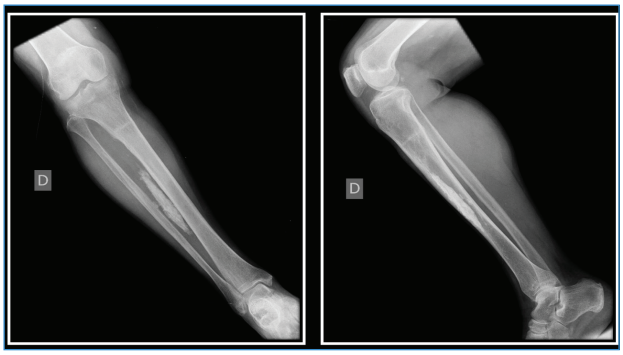


Figura 1. Radiografia perna direita - miosite ossificante

129 - TEMPORAL ARTERY ENLARGEMENT - BEFORE AND AFTER

Vanessa Fraga[1], Catarina Abreu[1], Ana Catarina Duarte[1], Sandra Sousa[1]
[1]Reumatology Department, Hospital Garcia de Orta, EPE, Almada, Portugal

A 74-year-old man presented to the emergency department with sudden vision loss of the right eye and a 2-month history of new-onset bilateral temporal headache, self-limited episodes of vision loss, jaw claudication and muscle pain/stiffness in the shoulders and hips. Physical exam showed enlarged temporal arteries and ophthalmic examination showed pallor right optic disc.

Laboratory studies showed normocytic/normochromic anemia and elevated inflammatory markers. The diagnosis of Giant cell arteritis was made and the patient started methylprednisolone pulses (1g/day, 3 days) followed by prednisolone 1mg/Kg/day. Temporal artery ultra-sound showed typical non-compressible "halo sign". Here we present the temporal artery enlargement at hospital admission (left) and after 1-month of treatment (right).

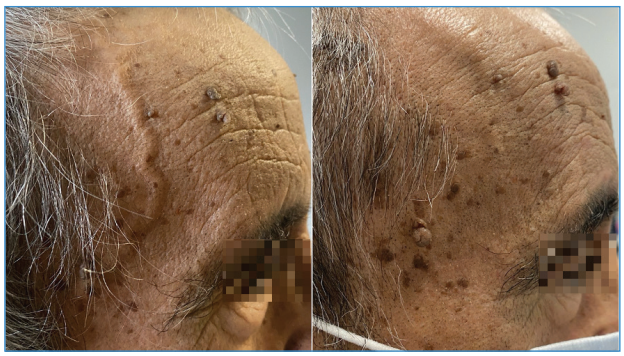


Figure 1. Temporal artery enlargement in a 74-year-old patient with Giant cell arteritis – before (left) and after treatment (right)

132 - OSTEOFITOSE CERVICAL COMO CAUSA DE DISFAGIA E ROUQUIDÃO

Ana Margarida Correia[1], Carla Campinho Ferreira[1], Emanuel Costa[1], Diogo Esperança Almeida[1], Joana Leite Silva[1], Joana Sousa-Neves[1], Marcos Cerqueira[1], Ana Ribeiro[1]
[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

Apresenta-se o caso de um doente do sexo masculino com 69 anos de idade, seguido em Consulta de Reumatologia por Espondilite Anquilosante, em remissão clínica sob terapêutica com anti-inflamatório não-esteróide on demand e Hiperostose Esquelética Idiopática Difusa (DISH). No final de 2018, iniciou queixas de disfagia para sólidos e líquidos e rouquidão, após realização de endoscopia digestiva alta (EDA), sem

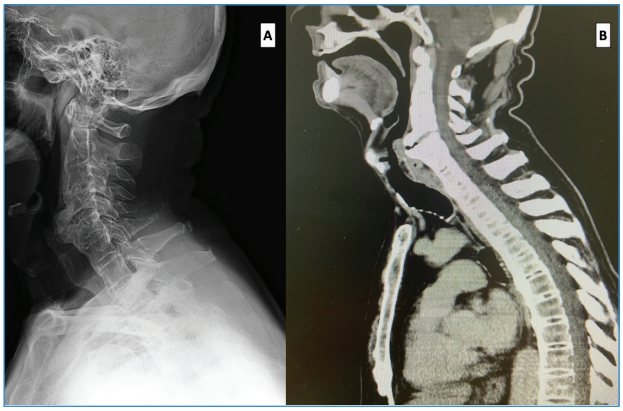


Figura 1. A e B - Radiografia da coluna cervical e TC pescoço, respetivamente, a revelar volumoso osteófito cervical C4-C5.

qualquer outra sintomatologia associada. O estudo endoscópico não apresentava qualquer alteração que justificasse a clínica do doente, tendo sido posteriormente avaliado por Otorrinolaringologia. A nasofaringolaringoscopia revelou edema da região retro-cricóideia, com abaulamento da respetiva parede posterior da faringe envolvente. Atendendo a estes achados e para melhor caracterização, realizou Tomografia Computorizada (TC) do pescoço que demonstrou a existência de volumoso osteófito anterior na coluna cervical ao nível de C4-C5, também evidente em radiografia da coluna cervical, que provocava o desvio anterior das estruturas laríngeas no plano da cartilagem cricoide (Imagem 1). Inicialmente foram propostas medidas de tratamento conservador, nomeadamente alterações dietéticas, mas pela persistência dos sintomas foi orientado para Ortopedia para ponderação de intervenção cirúrgica.

A osteofitose cervical anterior é um achado radiográfico comum na população idosa, devido a processos de espondilartrose, trauma, cirurgia prévia ou no contexto de DISH. Contudo, raramente se associa a sintomas, podendo estes surgir quando os osteófitos são volumosos, resultando em disfagia, disfonia, dispneia ou apneia obstrutiva do sono. Estudos prévios revelam que cerca de 1% dos osteófitos cervicais conduzem a disfagia e que em apenas 1,7% dos casos de disfagia esta se pode dever à presença de osteofitose. O tratamento conservador é na maioria dos casos a opção terapêutica de eleição correspondendo a adaptações dietéticas, miorrelaxantes, anti-inflamatórios ou alterações posturais durante a alimentação. Nos casos de refratariedade ao tratamento conservador ou de sintomatologia significativa e incapacitante, a opção cirúrgica deve ser considerada.

138 - A GLIMPSE INTO THE PAST: A CASE OF DESTRUCTIVE RHEUMATOID ARTHRITIS

Mariana Santos[1], Ana Teodósio Chícharo[2], Maria João Gonçalves[1], Maria Manuela Costa[1], Jaime C. Branco[1, 3],

Alexandre Sepriano[1, 3]
[1]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [2]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, Faro, Portugal, [3]CEDOC, NOVA Medical School, Lisboa, Portugal

Introduction: Patients with rheumatoid arthritis (RA) can gave severe disease which, if left untreated, leads to joint destruction. Disease-modifying anti-rheumatic drugs (DMARDs) are efficacious in preventing joint damage in RA and their widespread use has rendered 'destructive RA' uncommon nowadays.
Cases report: We report a case of a 52-year-old woman with the diagnosis of RA for over 15 years and with poor adherence to treatment. The patient was a smoker (7 pack per year) and seropositive for both rheumatoid factor and anti-cyclic citrullinated peptide with high titles (145,4 UI/mL and > 340 UA/mL, respectively). During the entire disease duration, the patient has often missed her appointments with her treating rheumatologist with no more than 5 visits within 15 years. The distance between the patient home and the hospital has made treatment and monitoring difficult. Despite several attempts for starting conventional synthetic DMARDs (methotrexate and sulfasalazine), the patient had always ended up withholding treatment, keeping only oral glucocorticoids which she tailored according to the severity of the complaints. Several years after her last visit, she was again referred to our department in April 2022 by the general practitioner because of severe pain, disability and severe limitation in the activities of daily living.

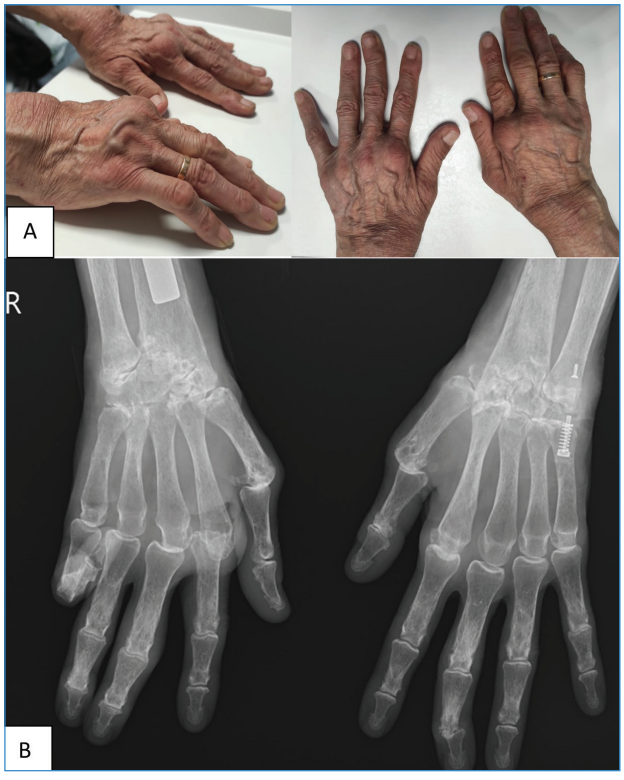


Figure 1. Findings on (A) physical examination and on (B) hand radiographs

At physical examination, the patient had arthritis of 5 joints (both wrists, 2nd and 3rd right interphalangeal joints and right knee) and elevated erythrocyte sedimentation rate (37 mm/1h) and C-Protein Reactive (1.25 mg/dL) (DAS28-VS: 3.2). In addition, she also had severe hand deformities typical of longstanding and untreated RA, namely atrophy of the interosseous muscles, subluxation of the radiocarpal and of various metacarpophalangeal joints (Figure 1A). Mobility was also impaired with complete loss of the flexion and extension of the wrist, and decreased amplitude of flexion of the metacarpophalangeal joints. Hands radiographs revealed severe and erosive RA, with damage involving carpal bones bilaterally, as well as various metacarpophalangeal and interphalangeal joints (Figure 1B). ankylosis of some metacarpophalangeal and articulations.

The patient also had osteoporosis and cataracts due to chronic use of glucocorticoids.
Discussion: This case is a sharp (and unfortunate) remainder of the management of RA in the pre-DMARD era. Even though glucocorticoids are often considered DMARDs, these drugs should serve as a bridging therapy to safer and more effective drugs. Strategies for improving adherence to therapy, including education and social support (when appropriate) are key components of the management of RA.

143 - KNEE PAIN IN HIV PATIENT – RED FLAGS

Inês Genrinho[1, 2, 3], Anabela Barcelos[2, 3]
[1]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [2]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [3]Centro Académico Clínico Egas Moniz, Aveiro, Portugal

Osteonecrosis (ON), also termed ischemic or avascular necrosis, can result from multiple secondary aetiologies. HIV infection increase 100-fold ON risk and the precise mechanism remains unclear, since HIV, antiretroviral therapy and others risk factors are commonly present in HIV patients. The most frequent locations are hip and knee followed by shoulder joint, affecting multiple sites in over two-thirds of cases.

A 31 year-old caucasian woman, presented 8 years after HIV type 1 infection diagnosis, with bilateral knee pain during last 6 months, with no trauma history.

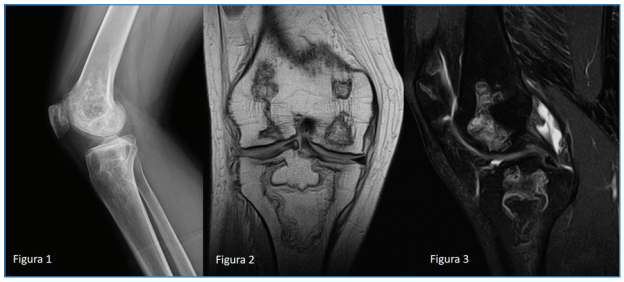


Figure 1-3. Knee ON

The pain has a mechanical character, worsening with weightbearing and knee flexion movements. No joint swelling or evidence of synovitis was seen on musculoskeletal examination and there was no other evidence of connective tissue disease. At the time of presentation, blood tests demonstrated a normal CD4 count and a viral load < 20. Routine blood tests were normal, including serum calcium, phosphate, and cholesterol. Knee radiographs demonstrated patchy sclerosis and lucencies in both femoral condyles, patellae and proximal tibia (Fig. 1). Magnetic resonance imaging revealed irregular, serpiginous subchondral low signal intensity in all pulse sequence and intense surrounding bone marrow edema pattern with multiple areas of bone infarction in the distal femora, proximal tibia and the upper patellae pole (Figs. 2–3).

Additional risk factors for secondary ON identified from the patient's history were chronic treatment with protease inhibitors, hypertriglyceridaemia, tobacco abuse and previous ON in the distal tibia.

This case highlights a significant musculoskeletal comorbidity linked to HIV infection that is not regularly described and remains poorly understood. Patient risk factors management and early diagnosis can have a positive impact on patient outcomes.

156 - SARCOIDOSE INAUGURAL EM ENXERTO CUTÂNEO

António Barreto Violante[1], Ana Filipa Rocha Águeda[1], Miguel Guerra[1], José Marona[1], Joana Ramos Rodrigues[1], Neide Pereira[1], Margarida Oliveira[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário Cova da Beira, Covilhã, Portugal

Homem de 65 anos com antecedentes de queimaduras corporais de 2º e 3º grau, atingindo 66% da superfície corporal e ditando necessidade de enxertos cutâneos autólogos, há cerca 4 anos. Encaminhado da consulta de Dermatologia por apresentar lesões infiltrativas eritematosas, de bordos elevados, na zona de transição entre o tegumento íntegro e o tegumento cicatrizado de ambos os antebraços, mãos e membros inferiores, com

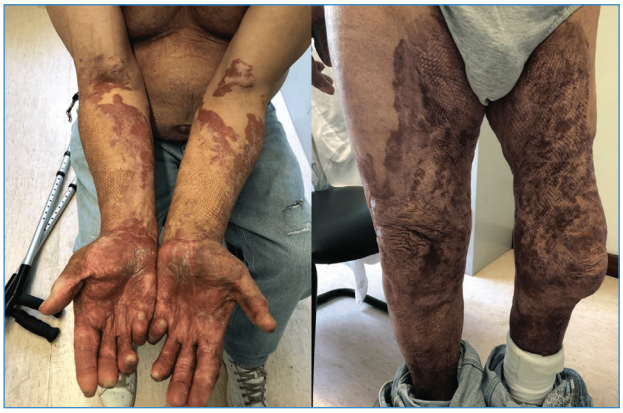


Figura 1. Sarcoidose inaugural em enxerto cutâneo

cerca de 7 meses de evolução.

Procedeu-se a biópsia das lesões, a qual revelou um infiltrado granulomatoso na derme reticular, compatível com o diagnóstico de sarcoidose.

Para além do envolvimento cutâneo, o doente apresentava também atingimento ocular sob a forma de uveíte anterior do olho direito, já com sinequia. Sem outras manifestações de órgão-alvo, nomeadamente pulmonar.

A imagem aqui exibida exemplifica uma sarcoidose com manifestação inaugural cutânea de características singulares, sob a forma de um infiltrado heterogéneo que segue a transição entre pele cicatrizada e íntegra.

173 - VÉRTEBRAS EM SANDWICH E "BONE WITHIN BONE" – SINAIS IMAGIOLÓGICOS DE OSTEOPETROSE

Diogo Esperança Almeida[1], Torcato Meira[2], Carla Campinho Ferreira[1], Ana Margarida Correia[1], Emanuel Costa[1], Ana Ribeiro[1], João Fernandes[2], Marcos Cerqueira[1]
[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal, [2]Serviço de Neurorradiologia, Hospital de Braga, Braga, Portugal

Uma mulher de 38 anos de idade apresentou-se em consulta de Reumatologia por queixas de lombalgia progressiva com cerca de 3 meses de evolução. Não tinha antecedentes médicos relevantes e apresentava história familiar de Osteopetrose (pai e tias paternas). Ao exame físico, não eram evidentes quaisquer alterações. A realização de tomografia computorizada da coluna dorsolombar mostrou a presença de múltiplas "vértebras em sandwich", com bandas escleróticas nas plataformas vertebrais, e também aparência de "bone within a bone". Não foram detectadas fracturas ou neuropatias compressivas. O estudo genético subsequente confirmou o diangóstico de Osteopetrose autossómica dominante, também conhecida como doença de Albers-Schönberg.

174 - ÚLCERAS CUTÂNEAS NA ARTITE REUMATÓIDE – O QUE NÃO ESQUECER

Ana Teodósio Chícharo[1], Mariana Santos[2], Margarida Lucas Rocha[1], Laura Gago[2], Maria Helena Lourenço[2], Ana Bento da Silva[2], Alexandre Sepriano[2, 3], Paula Araújo[2], Jaime C. Branco[2, 4], Manuela Costa[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, [3]CEDOC, NOVA Medical School. Faculdade de Ciências Médicas da Universidade NOVA de Lisboa., Lisboa, Portugal, [4]Centro de Estudos de Doenças Crónicas (CEDOC), NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

Introdução: A artrite reumatóide (AR) cursa frequentemente com manifestações extra-articulares. O compromisso cutâneo ocorre em 27.5% dos doentes, sendo muito diversificado. Dentro deste, as dermatoses neutrofílicas são raras, encontrando-se em 0.9-1.8% dos doentes.

Caso clínico: os autores apresentam uma mulher de 60 anos com AR seropositiva e erosiva com 25 anos de evolução. Durante o curso da sua doença efetuou vários DMARDs biológicos e convencionais. De momento está medicada com metotrexato 20mg/sem oral, leflunomida 20mg/dia e deflazacorte 6mg bid, com baixa atividade da doença (DAS28 Vs 3.17).

Nos antecedentes pessoais, a destacar insuficiência venosa periférica crónica, episódios prévios de erisipela, hipertensão arterial, diabetes tipo 2, dislipidemia e osteoporose não fraturária.

No final de 2021, e após um período de imobilização por lombociatalgia, a doente refere agravamento do edema dos membros inferiores e o aparecimento de pápulas eritematosas em ambas as pernas, que ulceraram e aumentaram de dimensões em poucas semanas. Refere dor intensa associada, mas nega prurido, febre, sintomas constitucionais, traumatismo ou picada de inseto prévios.

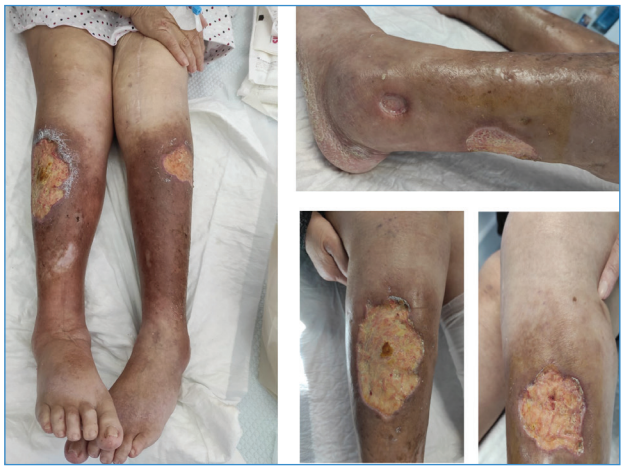


Figura 1. Aspeto macroscópico das lesões ulceradas à admissão da doente.

A doente fez antibioterapia empírica com doxiciclina durante 14 dias e cuidados de enfermagem no centro de saúde, sem melhoria. Neste contexto, é internada para esclarecimento diagnóstico e terapêutica.

Ao exame objetivo, apresenta uma úlcera na face antero-lateral da perna direita (10x6cm), e três úlceras na perna esquerda, uma na face externa (6x5 cm), uma no maléolo externo (2x3 cm) e uma na face posterior (7x7cm). As lesões têm base necrótica e bordos irregulares e violáceos (Figura 1). Dos exames complementares efetuados, salienta-se o aumento dos parâmetros inflamatórios (Vs 93 mm/h, PCR 5.4 g/mL) e o isolamento de Staphylococcus aureus sensível à oxacilina no exsudado das úlceras. A doente realizou eco-doppler arterial e venoso dos membros inferiores, sem alterações de relevo, excluindo possível contributo de estase venosa ou doença arterial periférica. Neste contexto, foi inicialmente assumida sobreinfeção bacteriana e prescrita flucloxacilina endovenosa de acordo com o teste de sensibilidade aos antibióticos, mas sem melhoria. Tendo em conta as características macroscópicas das úlceras, foi colocada a hipótese de Pioderma Gangrenoso (PG), que foi confirmado pela Dermatologia, sendo os isolamentos microbiológicos considerados apenas como colonização das lesões.

Tendo em conta o diagnóstico, a doente iniciou tratamento sistémico com prednisolona 0.75mg/kg/dia e tratamento local com betametasona e ácido fusídico, para além da optimização da analgesia. Objetivou-se melhoria clínica e analítica ao fim do primeiro mês, com diminuição do rubor peri-lesão, ligeira redução das dimensões das úlceras e parâmetros inflamatórios em perfil descendente. A doente foi posteriormente proposta para iniciar infliximab como poupador de corticoide, em regime de hospital de dia.

Conclusão: Consideramos que este caso demonstra um desafio diagnóstico pela presença de diferentes fatores de risco para úlceras nos membros inferiores. O PG é uma manifestação extra-articular rara da AR mas que deve ser considerada perante úlceras cutâneas com as características apresentadas, para um diagnóstico atempado e tratamento adequado.

186 - SPINE FRACTURE IN ANKYLOSING SPONDYLITIS

Sara Paiva Dinis [1], Ana Sofia Pinto[1], Filipe Cunha Santos[1], Joana Fonseca Ferreira[1, 2], Cláudia Vaz[1, 2], Nathalie Madeira[1]
[1]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda,

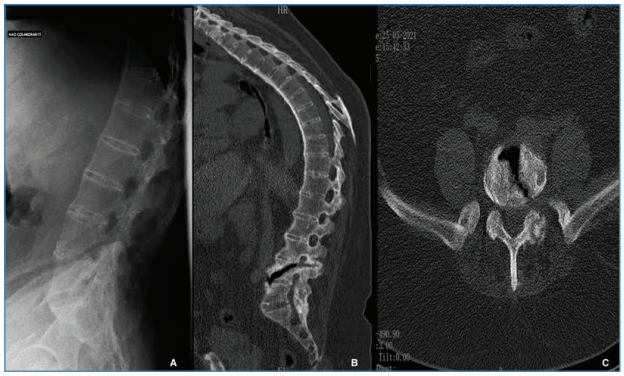


Figure 1. X-ray and computed tomography of the lumbar spine

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

A 66-year-old man was referred to our outpatient clinic with a long-standing history of anquilosing spondylitis (AS), who presented with worsening low back pain after a low impact fall 12 months ago. After the injury, he was admitted to the emergency room complaining of severe low back pain with no neurological signs. The lumbar spine X-ray (Figure A) performed on admission revealed a "bamboo spine", without evidence of fracture. The patient was discharged and treated conservatively with nonsteroidal anti-inflammatory drugs with partial pain relief. Nevertheless, for persisting pain further investigation was requested and computed tomography (CT) of the lumbar spine (Figures B,C) confirmed mass syndesmophyte formation and articular ossification and revealed a transvertebral extensive oblique fracture line crossing L4 and L5 vertebral bodies and extending through the posterior vertebral elements of L3 and L4.

All trauma patients with an extensive axial ankylosis must be carefully evaluated, since nondisplaced vertebral fractures can be easily missed on conventional X-rays. Thus, CT scans are recommended for fracture screening in any case of relevant injury to patients with an ankylosed spine.

188 - UMA FOTOGRAFIA DO PASSADO AINDA PRESENTE

Tiago Beirão[1], Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Catarina Rua[1], Flávio Campos Costa[1], Romana Vieira[1], Joana Abelha-Aleixo[1], Taciana Videira[1], P Pinto[1]

[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Breve Descrição: Doente do sexo feminino de 78 anos, parcialmente autónoma e cognitivamente integra, recorre à consulta de reumatologia por queixas de artralgias de longa duração, mas com agravamento recente. Tem como antecedentes dislipidemia, insuficiência cardíaca e diabetes mellitus tipo 2. Em 1984, foi observada na consulta de reumatologia, tendo sido efetuado o diagnóstico de artrite reumatoide e medicado com corticoide. Posteriormente, perdeu seguimento, mantendo a medicação inicialmente prescrita durante 36 anos – 8mg de metilprednisolona diários. Ao exame físico, apresentava desvio cubital dos dedos, subluxação das MCFs e flexo irreversível. O estudo imunológico e apresentava fator reumatoide e anticorpos anti-citrulinados positivos. Apresentava estudo radiográfico das mãos e dos pés.

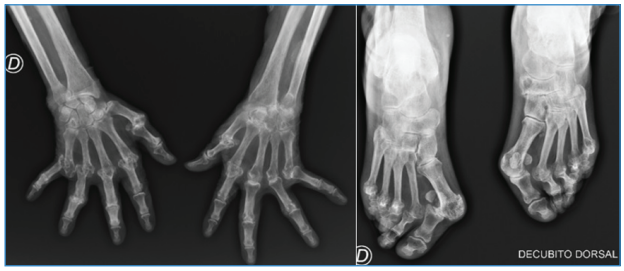


Figura 1. Radiografia convencional das mãos e punhos e dos pés

Radiografia convencional das mãos e punhos: Subluxação de todas as articulações Metacarpo falângicas com desvio ulnar mais evidente nos 4º e 5º raios bilateralmente. Erosões marginais múltiplas de todas as metacarpofalângicas e das IFP's do 4º raio bilateralmente. Sinais de dissociação escafo-semilunar bilateralmente. Várias erosões dispersas pelos ossos do carpo e das articulações radiocárpica e carpo-metacárpicas, predominantemente à esquerda.

Radiografia convencional dos pés: Dismorfia da arquitetura do pé bilateralmente, com antepé triangular. Subluxação com desvio lateral das articulações MTF.

Conclusão: Este caso representa uma realidade ainda presente nas consultas de reumatologia. Embora a maior sensibilização por parte da comunidade médica e um maior arsenal terapêutico, ainda se observam doente com deformidades irreversíveis, causadores de incapacidade. Deste modo, reforça-se a importância do diagnóstico e tratamento precoce, com um seguimento regular.

192 - DEFORMIDADES ARTICULARES NO ATRASO DE TRATAMENTO DE ARTRITE IDIOPÁTICA JUVENIL

Tiago Beirão[1], Beatriz Samões[1], Diogo Guimarães da Fonseca[1], Catarina Rua[1], Flávio Campos Costa[1], Romana Vieira[1], Joana Abelha-Aleixo[1], Taciana Videira[1], P Pinto[1]

[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

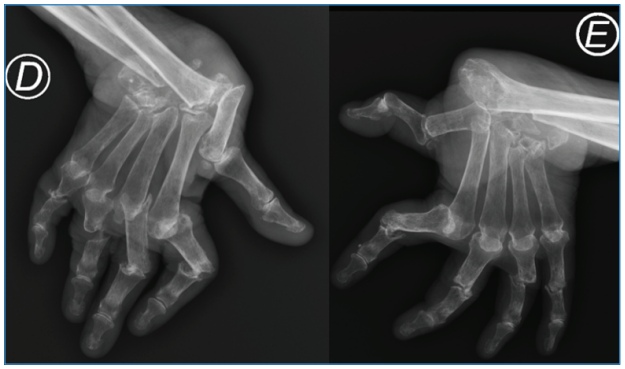


Figura 1.Radiografia convencional das mãos e punhos

Breve Descrição: Doente do sexo feminino de 53 anos, autónoma e cognitivamente integra, recorre à consulta de reumatologia para seguimento de doença reumática crónica. Aos 16 anos, a doente apresentou-se com um quadro de poliartrite, compatível com artrite idiopática juvenil poliarticular com fator reumatoide positivo, tendo sido iniciado sais de ouro. Aos 25 anos, foi diagnosticada neoplasia mamária, com suspensão da terapêutica imunossupressora, mantendo apenas tratamento sintomático. Em consequência da interrupção, houve evolução para poliartrite erosiva e deformante. Aos 42 anos, a doente é referenciada a nossa consulta de reumatologia, tendo iniciado por doença ainda ativa, metotrexato, com posterior switch para leflunomida. Foi pedido controlo radiográfico das mãos.

Radiografia Convencional das mãos e punhos: objetiva-se poliartropatia com componente mutilante, apresentando subluxação das articulações MCFs em direção palmar, bem como da articulação IFPs do 4º dedo mão esquerda em direção radial. Alterações degenerativas dos ossos do carpo com desvio ulnar das articulações CMC de todos os dedos. Luxação das articulações radiocárpica e ulnocárpica. Dissociação da articulação distal radioulnar com impactamento do rádio nos ossos carpais.

Conclusão: A remissão da artrite idiopática juvenil ocorre em 50 a 70% dos casos. Contudo, os doentes com artrite idiopática juvenil poliarticular fator reumatoide positivo tem prognóstico menos favorável. Este caso clínico demonstra a importância da manutenção de tratamento imunossupressor nesta patologia, já que a sua ausência dita, a longo prazo, deformidades irreversíveis causadoras de incapacidade, tal como evidenciado neste caso.

201 - DISH – A CAUSE OF POLYARTICULAR PAIN

Ana Margarida Duarte-Monteiro[1, 2], Filipe Ramos[3], Ana Rita Cruz-Machado[1, 2], Joaquim do Brito [3], José Padin[3], Rodrigo Goes[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]Serviço de Ortopedia, Centro Hospitalar e Universitário de Lisboa Norte, Lisboa, Portugal

Introduction: Diffuse Idiopathic Skeletal Hyperostosis (DISH) is a non-inflammatory musculoskeletal disorder that causes calcification mainly of spinal ligaments (more commonly the anterior longitudinal ligament). The most common sites of involvement in the spine are the thoracic and lumbar regions. Peripheral ossification at the entheses may also be present, more commonly in the elbows, wrists and pelvis.

It is more common in males and its prevalence is higher after the age of 60. Evidence has shown that DISH is associated with metabolic disorders such as obesity, hypertension, glucose intolerance or Diabetes, hyperuricemia and dyslipidemia, and these patients also have a significantly higher likelihood of being affected by Metabolic Syndrome. The pathogenesis of the disease is not fully understood, and while there is no effective treatment, conservative measures are encouraged, such as physiotherapy programs and symptomatic medication with pain killers, in order to improve quality of life. Conventional treatments for comorbidities are also a mainstay. Surgery may be warranted for certain complications such as severe symptomatic cer-

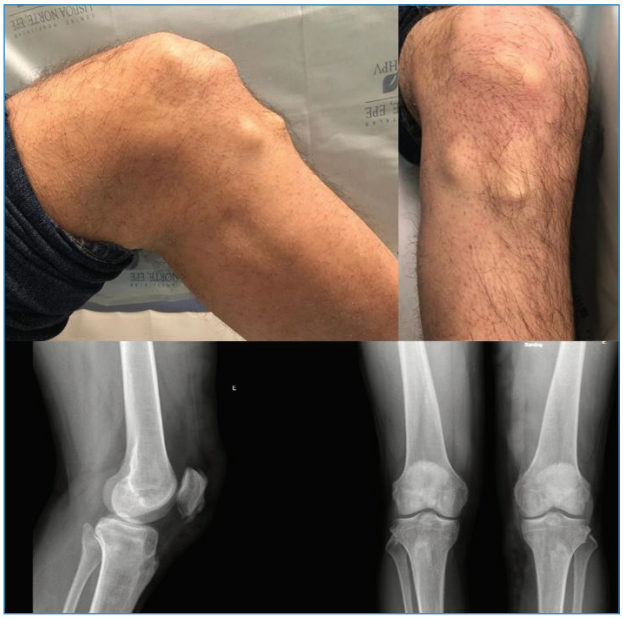


Figure 1. Bony prominences were evident at the knees, with corresponding radiographic calcifications

vical DISH and for unstable spinal fractures.

Clinical Vignette: A 53-year-old man presented in the orthopedics department complaining of mechanical pain referred to his elbows, knees, ankles and dorsal and lumbar spine. These symptoms started 5 years ago and have worsened progressively. He reported a Visual Analog Scale (VAS) for pain of 8/10 in the previous week and was using pain killers daily. HAQ-DI was also performed and a score of 1.13 was obtained. Physical examination revealed bilateral symmetrical bony prominences over both olecranons and Gerdy tubercles. There was no joint swelling or tenderness. Range of motion was mildly limited in the spine. The patient was overweight (IMC=28.5 kg/m2) and had an abdominal perimeter of 113cm. There was a personal history of diabetes, hypertension and dyslipidemia, and he fulfilled the diagnosis criteria for Metabolic Syndrome.

Radiographic evaluation of the spine showed bridging osteophytes in all segments. Also, exuberant calcifications were observed at the quadricipital, proximal and distal patellar entheses bilaterally, as well at the tricipital and Aquilian entheses. Laboratory evaluation revealed elevated total cholesterol. Parathormone, calcium, phosphate and uric acid levels were all normal.

Ankylosing spondylitis was considered as a differential diagnosis, but the patient reported no inflammatory back pain or prolonged morning stiffness. He had no history of gastrointestinal or genitourinary infection prior to symptom onset and there was no history of inflammatory bowel disease, psoriasis or eye inflammation. Radiographs of Sacroiliac joints showed no erosions, sclerosis or ankylosis. There was no elevation of inflammatory markers and HLA-B27 was negative. This way, a diagnosis of DISH was assumed.

Conclusions: We present a case of a patient with polyarticular pain that was diagnosed with DISH with both axial and peripheral involvement. Besides affecting the spine, DISH frequently involves the appendicular skeleton, mainly at bony eminences such as the Gerdy tubercles and the olecranon. Thus, the disease can present as generalized joint pain and should be considered in the differential diagnosis of patients with this presentation.

203 - UM ACHADO ECOGRÁFICO CURIOSO E RARO

Susana P. Silva[1, 2], Carolina Mazeda[1, 2], Sara Ribeiro Silva[3], Rita Cunha[1, 2], Gisela Eugénio[1, 2], Anabela Barcelos[1, 2]

[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz Health Alliance, Portugal, Aveiro, Portugal, [3]Physical Medicine and Rehabilitation, Centro Hospitalar Entre Douro e Vouga, Santa Maria da Feira, Portugal

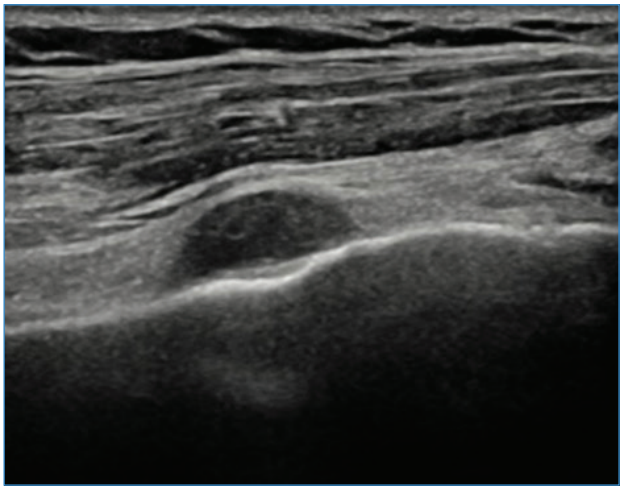


Figura 1. Imagem ecográfica de lesão anecogénica no interior do tendão da longa porção do bicípite correspondente a quisto gangliónico

Introdução: Os quistos gangliónicos são lesões pseudo-tumorais, de tecido mole, que surgem a partir da membrana sinovial de articulações ou bainhas tendinosas, sendo mais frequentemente encontrados nas mãos, punhos e pés. Raramente, estes quistos podem surgir dentro do tendão. A Ressonância Magnética é o método de diagnóstico mais utilizado para a visualização e diagnóstico destas lesões, contudo a ecografia também permite diferenciar estas lesões e apresenta-se como um método adjuvante ao seu tratamento.

Caso Clínico: Mulher de 78 anos, sem antecedentes pessoais de relevo, com quadro clínico de omalgia direita, de ritmo misto, com 2 meses de evolução, não responsiva a tratamento com anti-inflamatório não esteróide. Negava história de trauma ou cirurgias ao ombro. Ao exame objetivo, apresentava dor à mobilização apenas ativa do ombro direito, na flexão e abdução, sem limitação na amplitude dos movimentos. Foi realizada ecografia ao ombro, que revelou uma lesão anecogénica no interior do tendão da longa porção do bicípite, com 8.5x5.6x4.8mm, com alguns focos hiperecogénicos no seu interior, de contornos bem definidos (figura 1). Após discussão com a doente, optou-se por realização de fisioterapia, com melhoria sintomática.

217 - THE "CHARLOT SIGN/MOUSTACHE SIGN": A NEW SIGN OF PAGET'S DISEASE OF BONE

Carolina Ochôa Matos[1], José Carlos Romeu[1]

[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

Paget's disease of bone (PDB) is a chronic disease char-

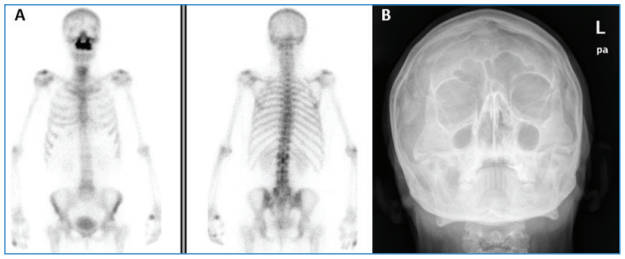


Figure 1. The "Charlot Sign/Moustache Sign"

acterized by focal areas of increased and disorganized bone remodeling. It can be monostotic or, more commonly, polyostotic. All bones of the skull can be affected (42% of cases of PDB), with a predilection for the bones of the calvarium and the base of the cranium. The diagnosis is mostly based on radiological examination and on biochemical markers of bone turnover, such as elevated serum alkaline phosphatase (SAP). PDB progresses through phases: first, lytic activity predominates, causing focal osteolytic lesions. Subsequently, areas of sclerosis develop, leading to the characteristic appearances of mixed lytic/sclerotic phase. Later, the sclerotic phase predominates, often associated with deformation and increased volume of the affected bone. There are several classical radiographic presentations associated with the different phases of Paget's bone disease, such as the osteoporosis circumscripta of the skull, the V-shaped resorption front of long bone, the cotton wool appearance of the skull, the Tam o'Shanter sign (skull), the picture frame vertebral body, and the pelvic brim sign (iliopubic/ilioischial lines).

The bone scintigraphy with 99mTc-MDP, with an intense and homogeneous uptake of the marker by the involved bones, allowing the identification of the affected bones and the extent of the disease, as well as bone deformity and expansion, is an effective method in the evaluation of the patient with or suspected of having Paget's bone disease. There are some signs associated with Paget's disease on bone scintigraphy, such as Abe Lincoln sign (also known as black beard sign, when the disease involves the mandible), Mickey Mouse sign (spinous process and pedicles), and Yarmulke sign (inhomogeneous in the skull).

We present the case of a 61-year-old female patient with history of ulcerative colitis, initially suspected of Paget's disease of bone due to an elevation of SAP (236 U/L) and high bone mineral density in L1 in bone densitometry. The bone scintigraphy with 99mTc-MDP and with single-photon emission computed tomography/computed tomography (SPECT/CT) revealed high-intensity diffuse uptake involving the upper jaw bilaterally and the spinous apophysis and right lateral aspect of L1 vertebral body, according to the diagnosis of PDB. These findings were corroborated with the radiography which showed maxilla's sclerosis. She was treated with a single administration of zoledronate (5 mg) with normalization of SAP, remaining asymptomatic.

With this case, we want to highlight the imaging features of the rare involvement of maxilla in PDB and suggest a new interesting finding: the "Charlot/Moustache sign", due to the uptake pattern in scintigraphy resembling a moustache like the one used by Charlot, the famous character played by Charlie Chaplin.
Figure: 99mTc-MDP bone scintigraphy image (A) with diffuse high intensity tracer uptake involving the upper jaw bilaterally, and radiograph image of the face (B) with sclerosis of the maxilla due to PDB.

222 - SCLERODERMA-LIKE NAIL FOLD CAPILLAROSCOPY PATTERN IN A DERMATOMYOSITIS PATIENT

Margarida Lucas Rocha[1], Ana Teodósio Chícharo[1], Frederico Rajão Martins[1], Ana Lúcia Fernandes[1], Célia Ribeiro[1], Vítor Teixeira[1], Graça Sequeira[1]
[1]Serviço de Reumatologia, Centro Hospitalar e Universitário do Algarve, Faro, Portugal

Nailfoldcapilaroscopy (NFC) of a 40 years-old patient with dermatomyositis (DM) associated with positive anti-Mi2 antibodies. The exam revealed a scleroderma-like pattern: capillary loss including avascular areas and tortuous capillaries and capillary ectasia (*) and several morphologic alterations of the capillaries including crossed, ramified and bush shaped capillaries compatible with neoangiogenesis phenomena (**) (Figure 1).

Scleroderma-like pattern could be present in up to 60-87.1% of DM patients. It could be also present in undifferentiated connective tissue disease and it does not have an obligatory association with a scleroderma overlap syndrome.

According to some authors, NFC alterations reflect disease activity in DM, especially muscle disease activity. O'Callaghan et al. found a relation between interstitial pulmonary disease and high capillary scores.

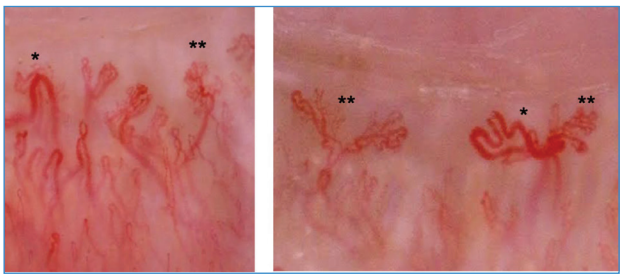


Figure 1. Nailfoldcapilaroscopy

228 - UMA LOCALIZAÇÃO MENOS COMUM DE DOENÇA ÓSSEA DE PAGET

Maria Seabra Rato[1], Alexandra Bernardo[1], Filipe Oliveira Pinheiro[1], Lúcia Costa[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário de São João, Porto, Portugal

A doença óssea de Paget (DOP) é um distúrbio benigno do osso, no qual existe um aumento da reabsorção óssea seguido do aumento da formação de osso, de forma desorganizada. É a segunda doença óssea metabólica mais comum, afetando 1-2% dos adultos acima dos 55 anos. Esta entidade é frequentemente assintomática, mas pode associar-se a deformidade e dor, aumento do risco de fratura e a complicações tais como surdez e osteossarcoma. Embora qualquer osso possa ser afetado, as localizações mais comuns são a bacia (58-80%), a coluna (40%), o fémur (32%) e a tíbia (16-20%), sendo a clavícula um local menos comum. Descreve-se o caso de um doente do sexo masculino, de 71 anos, com deformidade indolor da clavícula direita com 6 anos de evolução (Figura 1A e 1B). Não tinha história prévia de trauma nem de neoplasia. Analiticamente, apresentava elevação da fosfatase alcalina (220 U/L, valor normal entre 30-120 U/L) com restante estudo analítico incluindo hemograma, reagentes de fase aguda, creatinina, cálcio e fósforo sem alterações. A radiografia revelou um aumento difuso do tamanho da clavícula e alteração da estrutura trabecular óssea, com alternância entre áreas osteoscleróticas e osteopénicas (Figura 1C) e a tomografia computorizada heterogeneidade da trabeculação da clavícula, com expansão óssea e espessamento cortical. A cintigrafia óssea mostrou hipercaptação do radiofármaco na projeção de toda a clavícula direita e também da 1ª vértebra dorsal (Figura 1D). Estes achados foram compatíveis com DOP poliostótica. O doente foi medicado com ácido zoledrónico (5mg, endovenoso) apresentado normalização dos níveis da fosfatase alcalina um ano após o tratamento.

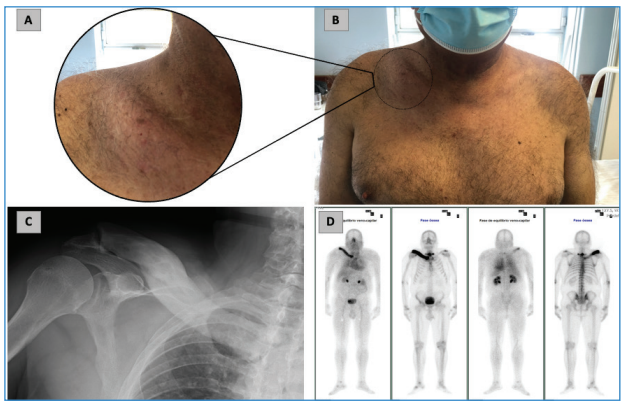


Figura 1. Doença óssea de Paget

231 - HIPEROSTOSE ESQUELÉTICA IDIOPÁTICA DIFUSA – UMA CAUSA DE DISFAGIA

Inês Santos[1], Nádia Martins[1], Paulo Monteiro[1], Ângela Domingues[2], Ricardo Araújo[2], Filipe Nunes Rodrigues[3]
[1]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [2]Serviço de Gastrenterologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [3]Serviço de Otorrinolaringologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

A hiperostose esquelética idiopática difusa (HEID), também conhecida por doença de Forestier, é uma doença caracterizada pela ossificação de ligamentos, tendões e enteses. Quando atinge a coluna cervical, pode causar disfagia por compressão extrínseca da faringe e do esófago.

Apresentamos o caso de um doente do sexo masculino, de 82 anos, com antecedentes pessoais de pancreatite aguda litiásica, hipertrofia benigna da próstata e hidrocelo direito, encaminhado para a consulta de Otorrinolaringologia (ORL) por disfagia para sólidos persistente, não progressiva, com 3 anos de evolução. Negou odinofagia, disfonia e dispneia, assim como sintomas constitucionais e outras queixas de órgãos ou sistemas. A palpação cervical foi negativa para massas ou adenopatias. A laringoscopia indireta revelou um abaulamento significativo da parede posterior da hipofaringe, de contorno regular e arredondado, sem alterações mucosas associadas. Tais achados foram confirmados por nasolaringofibroscopia. Analiticamente, sem alterações de relevo. Pelo mesmo motivo, foi também solici-

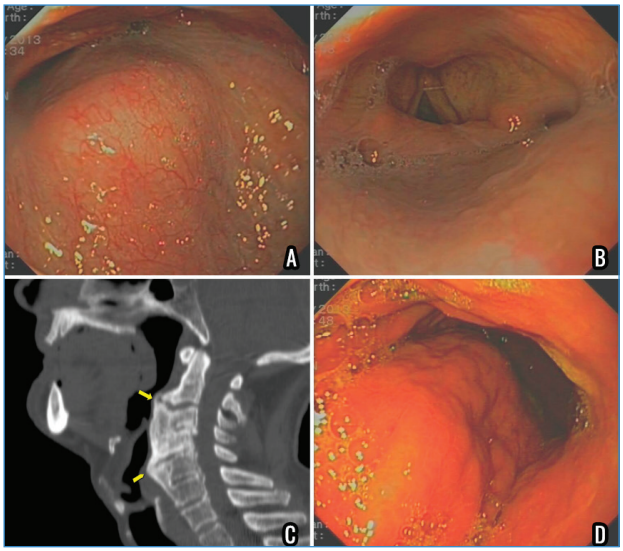


Figura 1. Compressão osteofitária na região supraglótica (A-C) e na grande curvatura gástrica (D)

tada consulta de Gastrenterologia, tendo sido realizada endoscopia digestiva alta que objetivou o abaulamento da face posterior da faringe, com revestimento mucoso de aspeto normal, por prováveis compressões extrínsecas a condicionar distorção da anatomia local até ao nível cricofaríngeo e abaulamento da grande curvatura do corpo gástrico. A tomografia computadorizada (TC) cervical revelou compressão extrínseca na região supraglótica, secundária a proliferação osteofitária anterior dos corpos vertebrais da coluna cervical, particularmente nos níveis de C2 a C6, associada a calcificação do ligamento longitudinal anterior (Fig. 1). A TC toraco-abdomino-pélvica não revelou alterações de relevo. A eco-endoscopia não demonstrou alterações gástricas. Concluiu-se o diagnóstico de HEID. O doente recusou terapêutica cirúrgica e mantém vigilância clínica regular com ORL, encontrando-se estável desde há 9 anos.

A HEID é uma patologia degenerativa, mais comum em homens, com um pico de incidência entre os 60-70 anos. Afeta de forma mais frequente a coluna torácica, sendo o achado típico a presença de calcificação linear e ossificação ao longo da porção antero-lateral dos corpos vertebrais. Quando se verifica envolvimento da coluna cervical, este ocorre predominantemente no seu nível inferior (abaixo de C4), sendo a disfagia o sintoma mais frequente, ocorrendo em 17-28% dos doentes. Além da disfagia, o envolvimento cervical pode associar-se a outros sintomas mais graves como: estridor, pneumonia de aspiração, apneia do sono e inclusivé subluxação atlanto-axoideia. Nos casos graves e progressivos, a ressecção cirúrgica dos osteófitos e calcificações pode ser um tratamento eficaz.

Apresentamos assim um caso que achamos relevante pelo envolvimento cervical superior que tem sido reportado na literatura como pouco comum.

233 - RECURRENT MECHANICAL LOW BACK PAIN CAN BE MORE THAN COMMON BACK PAIN

Augusto Silva[1], Maria João Saavedra[2], JE Fonseca[2]

[1]Rheumatology and Bone Metabolic Diseases Department, Centro Hospitalar Lisboa Norte, EPE - Hospital de Santa Maria, Lisbon Academic Medical Centre, 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

Images of two men with various visits to the emergency department due to recurrent mechanical rhythm low back pain and always medicated with analgesics, without further evaluation (Figure).

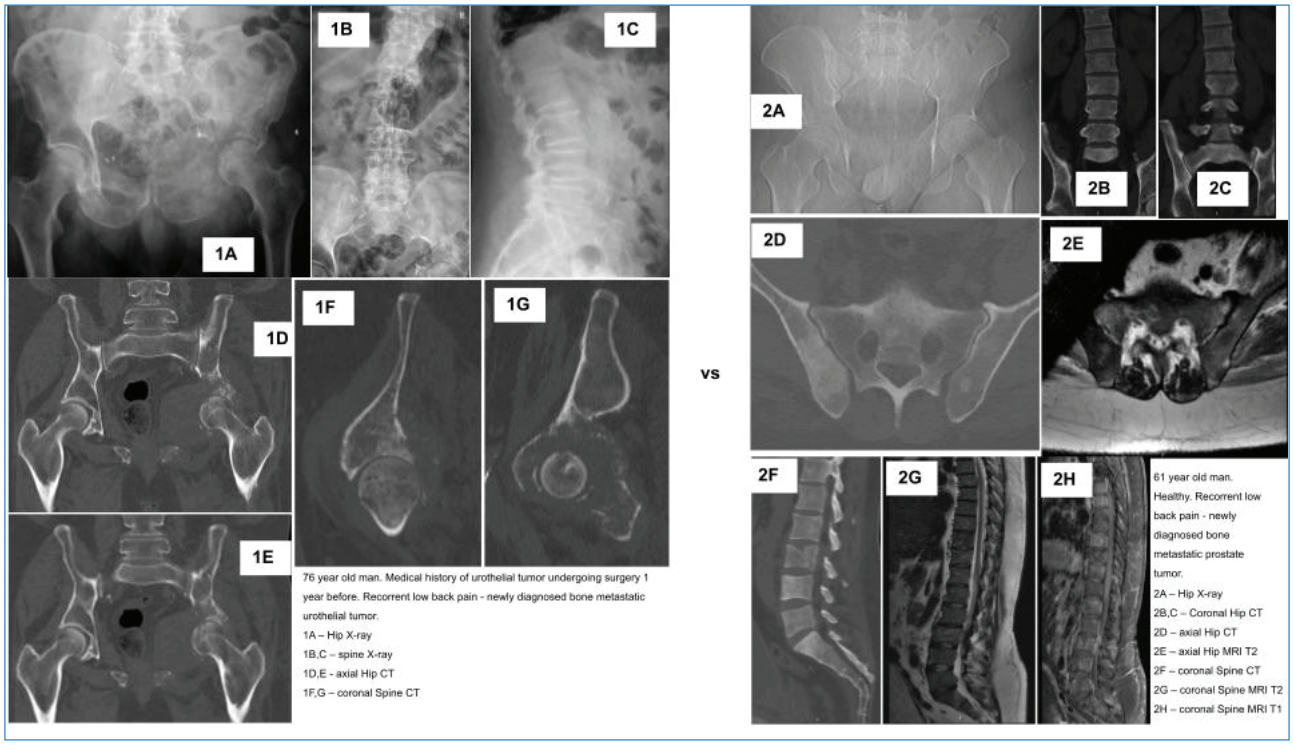


Figure 1. Two men with recurrent low back pain observed in urgency department with a diagnosis of metastatic prostate and urothelium tumors

250 - CUTANEOUS REACTION DUE TO COVID-19 VACCINATION

Carolina Mazeda[1, 2], Sofia Ferreira Azevedo[1, 2], C. Pinto Oliveira[1, 2], Manuela Loureiro[2, 3], Barcelos A.[1, 2]

[1]Serviço de Reumatologia, Centro Hospitalar do Baixo Vouga, E.P.E., Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz, Aveiro, Portugal, [3]Dermatologia, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal

Introduction: Since the outbreak of coronavirus disease 2019 (COVID-19), caused by severe acute respiratory syndrome coronavirus 2 (SARSCoV-2) in 2019, different strategies have been developed to control it. Vaccination is currently the most powerful medical approach against the pandemic. Just as COVID-19 has been associated with a wide variety of cutaneous manifestations, these post-vaccination symptoms have also been reported. Most of these cutaneous side effects are mild and transient manifestations of uncertain clinical significance.

Clinical case: A 68-year-old female patient, retired, autonomous, with a personal history of invasive ductal carcinoma of the right breast (T2N1M0) fifteen years ago, arterial hypertension, hypothyroidism and dyslipidemia, being treated with perindopril 5 mg, ezetimibe 10 mg and levothyroxine 0.088 µg daily. Admitted to the emergency department for erythematous, non-pruritic rash on both legs with petechiae up to the root of the lower limb with about five days of evolution. She also referred to knee and tibiotarsal arthralgias. Patient denied fever, nausea, abdominal pain, recent travel, introduction of new drugs or similar episodes in the past. She had been vaccinated with the 2nd dose of the ChaAdOx1 nCoV-19 vaccine five days before the onset of symptoms. Three days earlier she had been evaluated by her physician who prescribed topical corticosteroids and oral antihistamines that were ineffective. Physical examination revealed bilateral and symmetrical dermatosis affecting the lower limbs with the presence of violet papules that converge in plaques and do not blanch on diascopy (Figure 1). Initial investigations revealed elevation in acute phase reactants (VS 30 mm in the 1st hour, normal < 20) and mild thrombocytopenia (121x109/L, normal 150-500x109/L). Immunologic studies showed a negative cryoglobulins, antinuclear antibodies and anti-neutrophil cytoplasmic antibodies, as well as immunoglobulin assays. High-resolution chest computed tomography scan, mammography and breast ultrasound without changes. A skin biopsy was performed and revealed signs of acute vascular damage, with a peripheral lymphomononuclear infiltrate with small lymphocytes and eosinophils and blood extravasation compatible with the pathological diagnosis of small-sized vessel vasculitis. Direct immunofluorescence test was negative. The lesions resolved with rest without the need other treatment and without recurrence.

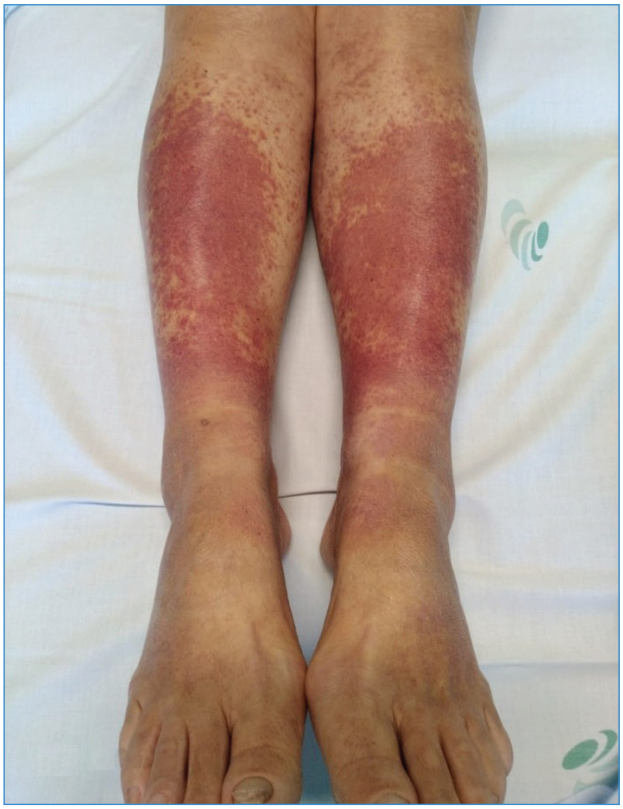


Figure 1. Cutaneous lesions present on lower extremity.

Conclusion: The temporal relationship of COVID-19 vaccine induction with dermatosis onset in the absence of other triggers renders the vaccine most likely responsible for our patient's presentation. Most post-vaccination reactions are mild and limited to the site of inoculation but cases of vasculitis have been reported. The most common subtypes of vasculitis reported are leukocytoclastic vasculitis and IgA and are generally transient, benign, self-limited, and usually not a contraindication to further doses of the vaccine. Clinicians should be aware the possibility of cutaneous reactions that may develop after the COVID-19 vaccination.

SPR

SOCIEDADE
PORTUGUESA DE
REUMATOLOGIA