Casos Clínicos

011 - ATYPICAL MANIFESTATION OF TUBERCULOSIS: A CASE REPORT

Maria Pontes Ferreira[1], Anita Cunha[1], Catarina Soares[1], Susana Almeida[1], José Tavares-Costa[1]
[1]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal

Introduction: Tuberculosis is a worldwide spread infection caused by Mycobacterium spp, with an estimated incidence of 10.6 million people1. Despite its predilection for lungs and pleura involvement1-2, it can also affect the musculoskeletal system2. We present a case of an atypical manifestation of tuberculosis, posing diagnostic challenges, with which we aim to bring awareness to the importance of considering tuberculosis as a differential diagnosis.
Clinical case: A 59 years-old man with past medical history of latent tuberculosis, treated with 1 year of isoniazid, resected colon adenocarcinoma, atrial fibrillation, hypertension, obesity and history of heavy smoking, was submitted to surgical decompression of the median nerve in 2021 due to paraesthesia of the 1st, 2nd and 3th fingers of the left hand. Afterwards, he noticed pain and swelling of the dorsal surface of the hand and wrist, for which he already was given 80mg/day of prednisolone, with no improvement.

At July 2023 he was referred to our consult, where we observed swelling of soft tissues on the dorsal side of the left hand and forearm, along with arthritis of the wrist, 1st and 4th metacarpophalangeal and 5th proximal interphalangeal joints (picture 1). His blood work showed mild elevation of erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), respectively 58 mm and 1.72 mg/dL, haemoglobin was 12.7 g/dL, rheumatoid factor and anti-citrullinated proteins autoantibodies were negative and there was no reactivity for human immunodeficiency virus, C or B hepatitis virus.

Given the previous lack of response to corticosteroids, chronicity of the clinical manifestations and temporal association with surgery, we performed a synovial biopsy to exclude indolent infection, which showed non necrotizing granulomas with lymphocytic infiltrate; Ziehl-Neelsen stain and Mycobacterium PCR of the synovia were negative. A thoracic computer tomography (CT) was performed, showing centrilobular nodules with ground-glass opacity and a conglomerate of nodules on the upper lobes and mediastinal adenopathies with central calcification.

While waiting for Pneumology consult, he developed constitutional symptoms, such as fever, night sweats, anorexia and weight loss, followed by development of pleural effusion and axillar adenopathy, with ESR and CRP increases (107 mm and 11 mg/dL, respectively). A thoracentesis was performed, showing high levels of adenosine deaminase and lactic dehydrogenase on pleural effusion; axillar adenopathy biopsy showed necrotizing granulomas; and bronchoalveolar lavage was positive for Mycobacterium tuberculosis.

Therefore, the patient begun treatment for active tuberculosis with rifampicin, isoniazid, pyrazinamide and ethambutol, with impressive improvement of the arm and wrist pain and swelling (picture 2) and resolution of pleural effusion and constitutional symptoms.
Discussion: This case unveils the dilemmas brought by unusual presentations of the great mimicker that is tuberculosis and illustrates the importance of the clinical history and comprehensive approach and persistence, even when results are inconclusive.

REFERENCES

1. World Health Organization. (2024). Tuberculosis. Retrieved from https://www.who.int/news-room/fact-sheets/detail/tuberculosis
2. Ngoc CT et al. Clinical characteristics and treatment outcomes of tuberculous tenosynovitis of the hand and wrist. Int J Med Sci. 2023 May 21

026 - WHIPPLE'S DISEASE MIMICKING POLYMYALGIA RHEUMATICA FOLLOWED BY AN IMMUNE RECONSTITUTION INFLAMMATORY SYNDROME

Rodrigo Rei[1], André Florêncio[2], Tiago Marques[3], Catarina Tenazinha[1]
[1]Rheumatology department, Unidade Local de Saúde do Algarve, Faro, Portugal, [2]Internal Medicine department, Hospital de Faro, Unidade Local de Saúde do Algarve, Faro, Portugal, [3]Infectious Diseases department, Unidade Local de Saúde de Santa Maria, Lisboa, Portugal

Introduction: Whipple's disease (WD) is a chronic infectious disease caused by Tropheryma whipplei. It is a rare disease characterized by two clinical stages, starting typically with musculoskeletal symptoms including migratory arthralgia or arthritis. On average, 6 years later, patients develop gastrointestinal (GI) symptoms, such as weight loss, chronic diarrhea and abdominal pain. If the patient receives immunosuppressive therapy, the disease can progress to the latter stage more rapidly.
Clinical report: A 72-year-old man presented to the Rheumatology clinic with a 3-year diagnosis of poly-

myalgia rheumatica (PMR), characterized by bilateral shoulder girdle pain and stiffness persistent throughout the day. He had responded well to therapy with glucocorticoids but the symptoms kept on relapsing for doses of prednisolone under 10mg daily. He had no fever, cranial symptoms or peripheral joint complaints. Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were persistently elevated under prednisolone 10mg daily. Rheumatoid factor was positive in low titer and anti-citrullinated peptide C was negative. The patient had undergone a vascular PET scan in the previous year that described mild uptake in the subclavian and femoral arteries. Methotrexate was started as steroid-sparing agent, up to a maximum dose of 20 mg weekly. Following methotrexate titration, he developed complaints of fatigue, loss of appetite and unintentional weight loss (up to a maximum of 15 kg). There were no other GI symptoms. Blood tests showed a new-onset normocytic anemia and a further increase in the already elevated inflammatory markers. Shoulder US demonstrated bilateral thickening of the subacromial bursa and labral calcifications, with no joint effusion or bicipital tenosynovitis. MRI of the chest and abdomen did not show signs of large vessel inflammation or malignancy. Following the emergence of the constitutional symptoms, he was admitted to inpatient care and underwent digestive endoscopic studies which confirmed the diagnosis of WD after duodenal mucosa biopsy. He started a one-year course of antibiotic-therapy with trimethoprim-sulfamethoxazole with gradual improvement of symptoms and complete resolution of the musculoskeletal complaints. Two months after starting antibiotic therapy, a slow taper in prednisolone was started, followed by an increase in inflammatory markers and resurgence of musculoskeletal complaints with prednisolone doses under 7.5 mg daily. A possible immune reconstitution inflammatory syndrome (IRIS) was assumed and prednisolone maintained at a dose of 10 mg daily. One month later, prednisolone tapering was started without resurgence of complaints and with persistently negative inflammatory markers.

Discussion: Here we present a case of WD with a PMR-like syndrome presentation, with constitutional symptoms triggered by treatment with the immunosuppressant methotrexate. Although migratory arthralgia or arthritis are the typical musculoskeletal complaints, there are a few case reports which describe a former diagnosis of PMR. Hence, we suggest this diagnosis to be considered in refractory cases of otherwise typical or atypical inflammatory joint syndromes.

Furthermore, this patient experienced an IRIS following antibiotic treatment. IRIS associated to WD is poorly described in the literature, but there is emerging evidence regarding its pathophysiology and clinical course, and previous immunosuppressive treatment has been recognized as a risk factor.

027 - EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS PRESENTING WITH PRINZMETAL ANGINA

Rodrigo Rei[1], Joana Massa Pereira[2], Sofia Andraz[2], Dina Bento[2], Catarina Tenazinha[1]
[1]Rheumatology department, Unidade Local de Saúde do Algarve, Faro, Portugal, [2]Cardiology department, Unidade Local de Saúde do Algarve, Faro, Portugal

Introduction: Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare disease with an incidence between 1,3 and 6,8 cases per 1 000 000 patients per year. It is a small and medium vessel vasculitis that also presents with eosinophilia, asthma, nasal polyps and sinusitis and affects multiple organs. Cardiac involvement is the main cause of morbidity and mortality.

Clinical report: We present a 44-year-old woman without known cardiovascular risk factors who was admitted to the cardiology department with anterior chest pain with ST segment elevation on the anterior leads of the electrocardiogram. She underwent invasive coronary angiography that showed a severe left anterior descending artery ostial stenosis that solved with the administration of intracoronary nitrates. Diagnosis of Prinzmetal angina was made. She was discharged from the cardiology department under treatment with triflusal, amlodipine, pitavastatine and transdermal nitroglycerin.

Three days later she presented to the emergency department with a generalized pruritic maculopapular rash involving palms and soles that evolved with tender vesicles. This was admitted as a drug-induced erythema multiforme associated with triflusal. The patient was started on prednisolone 40 mg daily with improvement of symptoms but no complete resolution. Skin biopsy showed an eosinophil-rich infiltrate in the dermis, with no signs of vasculitis.

Three months later, following prednisolone tapering, she was admitted again to the cardiology department with recurrent anterior chest pain with ST segment elevation, which responded to oral nitrates. A recurrent episode of Prinzmetal angina was assumed.

Her past medical history revealed a difficult to treat asthma since childhood and chronic rhinosinusitis with nasal polyps and anosmia. She had previously undergone excision of the polyps, which also showed an eosinophil-rich infiltrate.

At this point, the diagnosis of EGPA was considered. There was leukocytosis with eosinophilia (0,7 x 109/L). and the medical records from the last two decades showed a persistent peripheral eosinophilia,

with maximum values of 2,3 x 109/L. Anti-neutrophil cytoplasmic antibody (ANCA) test was negative

The patient was started on prednisolone 1 mg/Kg/day with complete resolution of the rash, improvement of the nasal symptoms and no new episodes of chest pain or acute asthma exacerbations. Prednisolone was slowly tapered and mycophenolate mofetil was started as a corticoid-sparing agent.

Discussion: Currently, it is recognized that ANCA-negative patients with EGPA are significantly more likely to have cardiac and lung involvement. However, cardiac involvement is the main leading cause of mortality in these patients, with myocarditis being the most common form. A prospective single center study found that 13 of the 52 patients enrolled had cardiac involvement, with 6 being myocarditis and 1 Prinzmetal angina. Prinzmetal angina related to coronary angiitis has been described in other clinical reports in recent years.

Here we report a case of Prinzmetal angina in a patient with a possible diagnosis of EGPA, fulfilling de ACR/EULAR classification criteria5 with 13 points: maximum eosinophil count ≥1×109/L (5), obstructive airway disease (3), nasal polyps (3) and extravascular eosinophilic predominant inflammation (2).

030 - NOTALGIA PARESTHETICA AS A CHALLENGING DIAGNOSIS – CASE REPORT

Inês Genrinho[1], Ana Filipa Gonçalves[2], Miguel Loureiro Guimarães[2], José Luis Carvalho[2]
[1]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [2]Reabilitação Musculo-Esquelética, Centro de Reabilitação do Norte, Vila Nova de Gaia, Portugal

Introduction: Notalgia paresthetica (NP) is a sensory neuropathy characterized by chronic pruritus localized to the upper back, typically in the region of the scapula. Despite being a relatively common condition, NP is often underdiagnosed due to its nonspecific clinical presentation and the overlap with other dermatological and neurological disorders.

Case report: A 81-year-old caucasian woman presented with unilateral chronic pruritus on her left part of the back and pain shoulder. Pruritus started 14 years ago after a fall, being heat the most exacerbating factor. Different therapies with topical and oral medications like opioids, tricyclics, antidepressants and corticosteroids failed to improve this symptom. Itching has been progressively worsening and recently, patient reported shoulder pain and paresthesias with posterior irradiation along the homolateral superior limb.

On admission, physical examination revealed ill-defined hyperpigmented area, with scaly erythematous lesions and slightly lichenification located on the left scapula. Lhermitte's test induced electrical shock-like sensation in trapezius and left shoulder, Hoffman's sign was positive in the left hand, Spurling was negative, motor deficits were absent and deep tendon reflexes were normal. Electromyography showed slight neurogenic atrophy in the left biceps and triceps muscles, suggesting a C6 and/or C7 radicular involvement. Cervical magnetic resonance demonstrated decreased amplitude of the neural foramina due to a disc-osteophyte complex, more prominent on the left, with involvement of the C7 root, and no medular hypersignal.

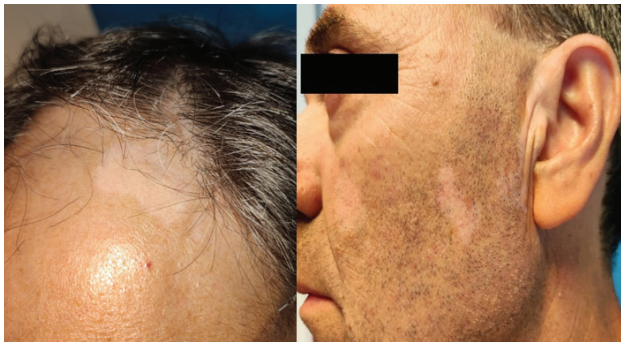
Due to the clinic course, imaging and electrophysiology tests, patient was diagnosed with NP, gabapentin was switched to duloxetine and C7 root ultrasound (US) guided extraforaminal injection with ropivacaine and dexamethasone was performed, with scarce response. We progressed to C7 neuromodulation, with US guided pulsed Radiofrequency plus dexamethasone. It was noted an immediate decrease on the intensity of pain and itching symptoms. The patient was fully satisfied with the treatment, and no adverse events were reported.

Discussion: NP predominantly manifests in middle-aged individuals, with an incidence that is two to three times higher in women. Symptoms usually are unilateral, most commonly on the left side of the body, opposite the dominant hand. The pathophysiology of NP involves compression or irritation of the cutaneous branches of spinal nerves, often associated with degenerative changes in the spine or musculoskeletal compression. Misdiagnosis can lead to prolonged patient discomfort and unnecessary treatments. Accurate diagnosis is critical for effective management, which may include topical and oral therapies, physical therapy and minimally invasive interventions.

036 - VITILIGO AS A RARE ADVERSE EVENT OF SECUKINUMAB IN PSORIATIC ARTHRITIS: A CASE REPORT

Pedro Miguel Teixeira[1, 2], Carolina Vilafanha[1, 2], Manuela Loureiro[2, 3], Gisela Eugénio[1, 2], Anabela Barcelos[1, 2]
[1]Rheumatology Department, Unidade Local de Saúde da Região de Aveiro, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz Health Alliance, Portugal, Aveiro, Portugal, [3]Dermatology Department, Unidade Local de Saúde da Região de Aveiro, Aveiro, Portugal

Introduction: Secukinumab, an Interleukin (IL)-17A inhibitor, is highly effective in treating both joint and skin manifestations of psoriatic arthritis (PsA). While most adverse events are mild, some paradoxical immune reactions, including cutaneous adverse effects, have been reported.



036 – Figure 1. Vitiligo lesions on scalp and face

Clinical case: We report the case of a 51-year-old man with a history of PsA with peripheral involvement, diagnosed at 33 years of age. Initially treated with methotrexate, he was switched to secukinumab 300mg every 4 weeks due to severe plaque psoriasis, resulting in significant improvement in both skin (PASI 0) and joint (DAPSA 1) symptoms. After five years of therapy, the patient presented to a rheumatology consultation with new hypopigmented, non-scaly patches on his face and scalp, without a prior history of similar lesions. Laboratory results were normal, including negative antinuclear antibodies and normal thyroid function. Initial treatment with oral terbinafine and topical antifungal was ineffective. A skin biopsy revealed a loss of epidermal melanocytes, consistent with vitiligo. The patient was then treated with a topical calcineurin inhibitor, and secukinumab was discontinued, leading to gradual improvement of skin lesions but a worsening of peripheral arthritis. Following a multidisciplinary discussion, it was decided to reinitiate secukinumab, which quickly controlled the articular disease without exacerbating the vitiligo lesions.
Conclusion: Vitiligo associated with IL-17A inhibitors is a rare adverse event, with few cases reported in the literature. It is hypothesized that inhibition of Th17 lymphocytes may enhance a Th1 response mediated by interferon-gamma, resulting in a cytokine dysregulation that favours the Th1 pathway, contributing to the development of vitiligo. Early identification of this adverse effect is crucial to prevent irreversible melanocyte destruction.

038 - SÍNDROME DE CUSHING E OSTEOPOROSE: A PROPÓSITO DE UM CASO CLÍNICO

Rita Rolo de Matos [1], Ana Rita Pereira Fonseca[2]
[1]USF Vale do Vouga, Unidade Local de Saúde de Entre o Douro e Vouga, São João da Madeira, Portugal, [2]Rheumatology Department, Unidade Local de Saúde de Entre o Douro e Vouga, Santa Maria da Feira, Portugal

Enquadramento: A osteoporose é uma doença músculo-esquelética, caracterizada por diminuição da massa óssea e deterioração da microarquitectura do osso, com consequente aumento da fragilidade óssea e risco de fratura. Constitui um dos graves problemas mundiais de saúde pública, pela alta morbi-mortalidade relacionada com as fraturas de fragilidade, afetando mais frequentemente mulheres em idade pós-menopausa e idosos. Até 30% das mulheres pós-menopausa e 50 a 80% dos homens com osteoporose apresentam uma causa secundária, sendo fundamental, nestes casos, tratar a causa subjacente.
Descrição: Mulher de 71 anos, com antecedentes de hipotiroidismo, dislipidemia, insuficiência venosa periférica, depressão, patologia hemorroidária, osteoporose, colecistectomia, histerectomia e anexectomia unilateral aos 36 anos. Refere diagnóstico de osteoporose aos 30 anos de idade, tendo iniciado tratamento com bifosfonato no período pós-menopausa, com pouca adesão à terapêutica. Em 2017, com 64 anos, iniciou denosumab, após fratura de L5, com abandono terapêutico em 2020, por iniciativa própria. Em 2021, por queixas de cervico-dorsalgias, fez radiografia da coluna e posterior ressonância magnética que mostraram fraturas de C6, C7, D1, D2, D5 e D11 e fratura de D9 em TAC de 2022, tendo sido encaminhada a consulta de Reumatologia por osteoporose grave fraturária, em 2023.

Após história clínica mais aprofundada, verificou-se que apresentava aumento ponderal, com predomínio abdominal, equimoses nos membros superiores, edema periférico, plétora facial e diagnóstico recente de Diabetes Mellitus. Foi observada em consulta de Endocrinologia que, após estudo, diagnosticou Síndrome de Cushing. A doente iniciou terapêutica com teriparatida, em março de 2023, que mantém até há data, sem novos episódios de fratura.
Conclusão: Perante uma osteoporose grave fraturária é sempre necessário colocar a hipótese da presença de uma causa secundária para a mesma. Neste caso, a valorização de sintomatologia, inicialmente inespecífica, permitiu o diagnóstico de uma causa rara de osteoporose secundária, sendo que a osteoporose na Síndrome de Cushing se assemelha à osteoporose induzida por glucocorticoides (a causa mais comum de osteoporose secundária).

041 - A20 HAPLOINSUFFICIENCY: UNVEILING THE IMPORTANCE OF FAMILY HISTORY IN ATYPICAL PRESENTATIONS

Margarida Lucas Rocha[1, 2], Roberto Pereira da Costa[2, 3], Patrícia Costa Reis[3, 4], Susana Lopes da Silva[5], Filipa Oliveira Ramos [2, 3, 4]
[1]Rheumatology Department, Unidade Local de Saúde do Algarve, Faro, Portugal, [2]Rheumatology Department,

Unidade Local de Saúde Santa Maria, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, Lisboa, Portugal, [3]Rheumatology Research Unit, Instituto de Medicina Molecular João Lobo Antunes, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [4]Pediatric Rheumatology Unit, Unidade Local de Saúde de Santa Maria, Lisboa, Portugal, [5]Immunoallergology Department, Unidade Local de Saúde de Santa Maria, Lisboa, Portugal

Introduction: A20 Haploinsufficiency (HA20) is a, recently described, early-onset, autoinflammatory disease, with a clinical presentation that mimics Behçet's disease. Frequent features are fever, mucosal ulcers, arthritis and diverse gastrointestinal and skin manifestations. It is an autosomal dominant hereditary disease caused by loss-of-function variants of TNFAIP3, which encodes the protein A20 and causes an activation of NF-KB and NLRP3 pathways. Early diagnosis is crucial for effective management and improving patient outcomes. Here, we describe a patient that highlights the importance of considering genetic causes when there is a family history of immune dysregulation.
Case: We describe a 4-year-old-boy with a previous history of prematurity, global developmental delay, congenital heart septal hypertrophy, first-degree atrioventricular block, an atopic background and IgA deficiency, with frequent upper and lower respiratory infections and gastroenteritis. He was referred to our Pediatric Rheumatology clinic due to polyarthritis affecting the wrists, metacarpophalangeal and proximal and distal interphalangeal joints, knees and ankles, with a significant gait impairment since he was 15 months-old. Additionally, the patient had recurrent abdominal pain and diarrhea and oral aphthosis. The patient had high erythrocyte sedimentation rate (74mm/hr) and C reactive protein (1.66mg/dL) and a mild elevation of fecal calprotectin (148µg/g). Inflammatory bowel disease was excluded and the patient was started on methotrexate.

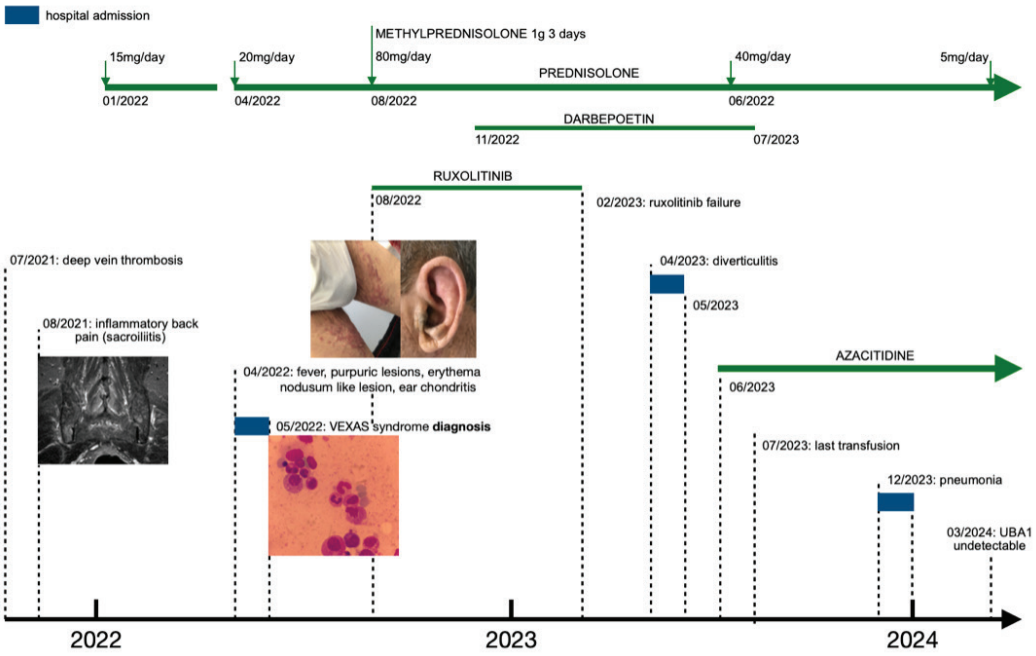
Notably, there was a family history of immune dysregulation spanning three generations. His mother had a diagnosis of common variable immunodeficiency disease with typical recurrent respiratory and gut infections, in association with increased viral susceptibility and predominant immune dysregulation (thyroiditis and type 1 diabetes mellitus). A pathogenic variant in TNFAIP3 (c.811C>T, p.Arg271Ter, non-sense mutation) was detected, and the mother was diagnosed with A20 haploinsufficiency. Interestingly, his grandmother, who had a history of polyarthritis, previously diagnosed as rheumatoid arthritis, was also found to carry the same mutation. Following the diagnosis of the mother and grandmother, genetic testing was conducted on our patient, revealing the same pathogenic variant in TNFAIP3 leading to A20 haploinsufficiency diagnosis.

Despite significant arthritis improvement, methotrexate had to be discontinued due to hepatotoxicity. Afterwards, colchicine and adalimumab were started, but the latter also discontinued due to two episodes of pericarditis, with an assumed probable relation to the drug. The patient was subsequently treated with anakinra followed by tocilizumab, both suspended due to ineffectiveness. For the past 18 months the patient has been treated with colchicine and etanercept. Currently, the patient is in remission, without significant adverse events.
Conclusion: Our patient's detailed anamnesis and family history revealed atypical symptoms and a significant familial background, leading to the diagnosis of A20 haploinsufficiency. This case highlights that the same pathogenic variant in TNFAIP3 can be associated with distinctive phenotypes in members of the same family, which makes HA20 diagnosis even more challenging. It also highlights the importance of expanding differential diagnoses to include genetic causes in atypical disease presentations.

042 - VEXAS SYNDROME: AN ATYPICAL INDOLENT PRESENTATION AS SACROILLIITIS WITH MOLECULAR RESPONSE TO AZACITIDINE

Roberto Pereira da Costa[1, 2], Guilherme Sapinho[2, 3], Matilde Bandeira[1, 2], Joana Infante[2, 3], Tiago Marques[4], Carla Mimoso Santos[4], João Forjaz de Lacerda[2, 3], JE Fonseca[1, 2], José Carlos Romeu[1]
[1]Serviço de Reumatologia, Unidade Local de Saúde Santa Maria (ULSSM), Lisboa, Portugal, [2]Instituto de Medicina Molecular, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Serviço de Hematologia e Transplantação de Medula, Unidade Local de Saúde Santa Maria, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [4]Serviço de Doenças Infeciosas, Unidade Local de Saúde Santa Maria, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introduction: VEXAS syndrome is a recently described autoinflammatory syndrome caused by somatic acquisition of UBA1 mutations in myeloid precursors and is frequently associated with hematologic malignancies, chiefly myelodysplastic syndromes. Disease presentation can mimic several rheumatologic disorders, delaying the diagnosis.
Case: We describe a case of a 74-year-old man with an atypical presentation of VEXAS syndrome resembling late-onset axial spondylarthritis, later progressing to a systemic inflammatory syndrome with fever, chondritis, cutaneous vasculitis and transfusion-dependent



042 – Figure 1. Timeline of symptoms and interventions

anemia, requiring high doses of steroids. Despite being initially diagnosed as relapsing polychondritis, the cause for the disproportionate macrocytic anemia remained unexplained. Therefore, a bone marrow aspiration and biopsy were performed, showing erythroid hypoplasia without significant dysplasia. Upon review of the aspirate, extensive vacuolization of myeloid and erythroid precursors was noted, which raised suspicion of VEXAS syndrome. The UBA1 p.Met41Thr variant was identified through next generation sequencing, with a variant allele fraction of 71.25%.

Ruxolitinib was used as a first steroid-sparing strategy, but, despite titration to the maximum dose, the patient remained dependent on prednisolone≥30mg/day. After six months, a switch to tocilizumab was planned, but it was not carried out as the patient had to be admitted due to a diverticulitis complicated by a local abscess. Therefore, azacitidine was chosen as the second-line steroid sparing agent. This drug allowed for progressive steroid tapering (from 40mg/day to 2.5mg/day), whilst the patient remained in clinical remission, transfusion-free for the past 11 months and without experiencing significant adverse events. Importantly, after nine months of therapy, bone marrow reassessment was consistent with a significant decrease in precursor cell vacuolization with no additional signs of dysplasia and the UBA1 p.Met41Thr VAF decreased from 71.25% to being undetectable, suggesting a deep molecular response.

Discussion: The case presented illustrates the difficulty in diagnosing and managing VEXAS syndrome. Indolent symptom onset with mild nonspecific findings can be misleading. Initial symptoms suggesting an axial spondylarthritis are quite atypical. In the presented case, the development of fever, severe anemia, skin findings, chondritis and a marked inflammatory syndrome widened the differential diagnosis. However, diagnosis was dependent on a second review of the marrow aspirate which identified vacuolization of erythroid and myeloid precursors. This highlights the importance for hematopathologists to be familiar with this entity and the need for physicians across various specialties to communicate and collaborate throughout the diagnostic process.

This case raises the question of whether azacitidine's anti-inflammatory effects are dependent or independent from clonal control. In this case, we demonstrated both success in controlling inflammation and in attaining clonal suppression, as seen by a significant decrease in UBA1 VAF in marrow samples over time, confirming the potential for disease modification with azacitidine in VEXAS syndrome, even in a refractory setting.

Lastly, we emphasize that management of VEXAS syndrome requires a dedicated multidisciplinary team. Most adverse events (including numerous infections) experienced by this patient were attributable to prolonged steroid exposure. Therefore, steroid-sparing strategies to mitigate these risks should be a research priority.

047 - A RARE CASE OF ANCA-ASSOCIATED VASCULITIS AND SCLERODERMA RENAL CRISIS IN MIXED CONNECTIVE TISSUE DISEASE

Mariana Diz-Lopes [1,2], Carlos Marques-Gomes[1,2], Inês Santos[3], Bernardo Fernandes[2,4], Roberto P. Silva[5], Teresa Martins-Rocha[1,2], Pedro Madureira[1], Lúcia Costa[1]
[1]Rheumatology Department, Centro Hospitalar de São João, Porto, Portugal, [2]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal, [3]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [4]Serviço de Nefrologia, Centro Hospitalar de São João, Porto, Portugal, [5]Serviço de Anatomia Patológica, Centro Hospitalar Universitário de São João, Porto, Portugal

Presentation of Case: A 52-year-old woman with Mixed Connective Tissue Disease (MCDT), antinuclear antibodies (ANA) with a 1/1000 titer and strongly positive anti-U1 RNP antibodies, presented in the emergency department with a 1-month evolution of dyspnea, chest pain on minimal exertion and visual disturbances.

On physical examination, she was diaphoretic, had blood pressure (BP) of 186/125mmHg and heart rate of 118bpm. Sclerodactyly and microstomia were evident. Blood tests revealed anemia (hemoglobin 11g/dL), thrombocytopenia (100.000/mm3), elevated serum creatinine (sCr 1.60mg/dL), urea (59mg/dL) and elevated troponin I (maximum 1727ng/mL). Direct fundoscopy confirmed the presence of cotton-wool spots and fresh flame hemorrhages and thus the presence of hypertensive retinopathy. The electrocardiogram had ST segment elevation in V4-V5 and the echocardiogram showed global akinesia and ventricular dysfunction. There was no evidence of coronary disease after percutaneous coronary intervention, compatible with Takotsubo Cardiomyopathy.

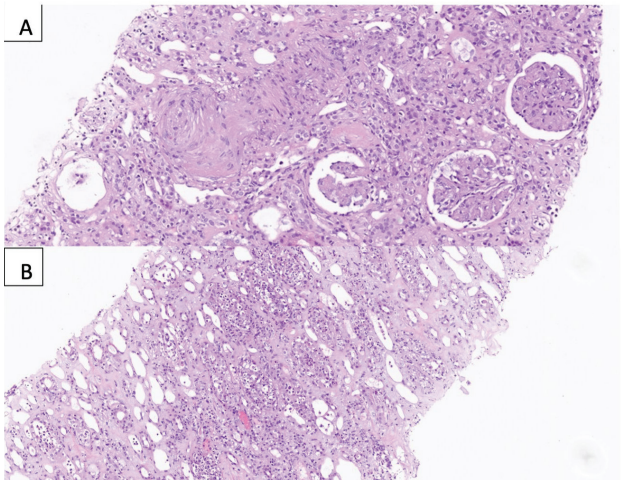
Scleroderma renal crisis (SRC) in the context of MCDT presenting as a hypertensive emergency was the probable diagnosis and captopril was started with progressive titration and adequate BP control. However, despite improvement in hematologic, ophthalmologic and cardiac disturbances, renal function progressively deteriorated (maximum sCr 5.02mg/dL).

Additional testing was pursued: 24-hour urine analysis was remarkable for proteinuria (1.24g/24h); blood tests with anti-double stranded DNA and complement levels were normal; antineutrophil cytoplasmic antibody (ANCA) testing was positive for ANCA-PR3 (>200 U/mL).

A renal biopsy was performed and showed ischemic glomerular changes. Arterioles had intimal proliferation and thickening ("onion-skin" lesion) (Figure 1, A). Inflammatory changes in the medulla with leukocytoclasia of neutrophils and tubular necrosis were present, aspects suggestive of medullar angiitis (Figure 1, B). There was no fibrinoid necrosis and no crescents (Figure 1). Immunofluorescence microscopy was unremarkable except for IgM+ mesangial deposition.

The renal biopsy findings were predominantly explained by SRC. However, the presence of medullar angiitis [1], ANCA-PR3 elevated titers and the progressive renal disfunction with adequate BP control, allowed for the diagnosis of concomitant ANCA-associated vasculitis (AAV).

Treatment was started with 500mg intravenous methylprednisolone for 3 days, followed by prednisolone 50mg/day and rituximab 1g for 2 doses (2 weeks apart). Supporting a glucocorticoids (GC) reduction strategy, she was proposed for Avacopan [2]. Prior to therapeutical approval, she died unexpectedly at home.
Conclusion: SRC is an extremely rare complication of MCTD and is associated with poor outcomes. Progressive renal dysfunction in patients with MCTD requires excluding other aetiologies, including AAV. The concomitant presence of the two entities creates a treatment challenge, particularly with GC, which are essential in AAV treatment but should be avoided in SRC.



047 – Figure 1. Glomeruli with ischemic features and arterioles with intimal proliferation (A - HE 100X). Medulla with lesions characteristic of medullary angiitis (B - HE 100X).

REFERENCES

1. Klein, J., et al., Medullary angiitis and pauci-immune crescentic glomerulonephritis. Proc (Bayl Univ Med Cent), 2017. 30(3): p. 351-352.
2. Jayne, D.R.W., et al., Avacopan for the Treatment of ANCA-Associated Vasculitis. N Engl J Med, 2021. 384(7): p. 599-609.

052 - SILENCING RHEUMATOID NODULES: UPADACITINIB'S QUIET TRIUMPH IN LUNG AND SKIN

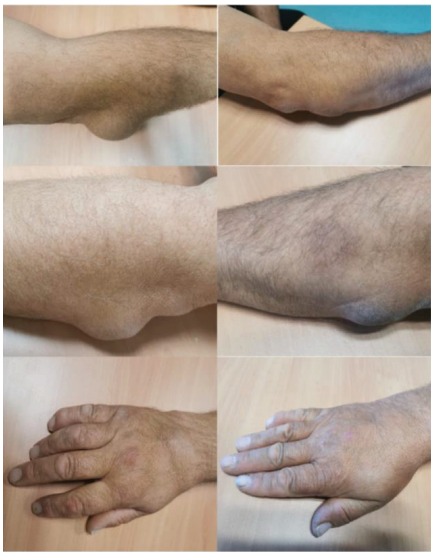
Margarida Lucas Rocha[1], Rodrigo Rei[2], Vítor Teixeira[1]

[1]Rheumatology Department, Unidade Local de Saúde do Algarve, Faro, Portugal, [2]Rheumatology department, Unidade Local de Saúde do Algarve, Faro, Portugal

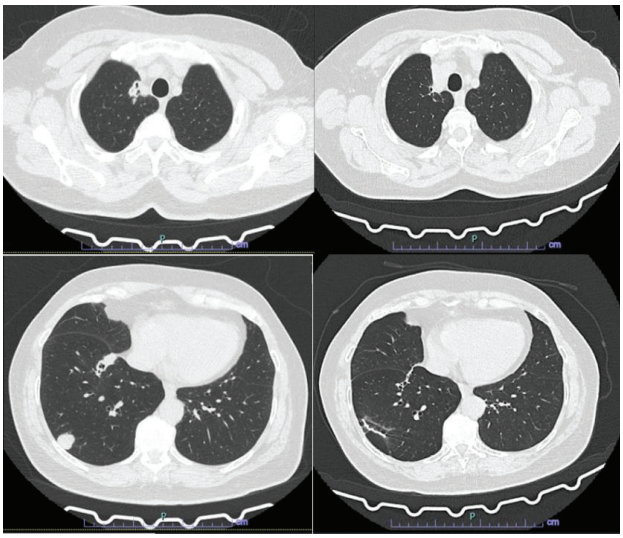
Rheumatoid nodules (RN) are a common extra-articular feature, present in up to 20% of rheumatoid arthritis (RA) patients and are associated with more severe disease and high-titer rheumatoid factor (RF) or anti-citrullinated peptide antibody (ACPA) positivity (Hochberg et al., 2019). RN usually occur over extensor surfaces and joints, in subcutaneous tissue of the fingers and sometimes asymptomatically in other organs. RN can be refractory to rheumatoid arthritis conventional therapy and can paradoxically progress during methotrexate or leflunomide treatment, despite improvements in disease activity. We describe 2 cases of patients with RA whose refractory RN showed significant improvement after treatment with Upadacitinib.

Case 1: 50-year-old male smoker with erosive RA, positive for RF and ACPA and cutaneous RN, with severe functional impairment in daily life. Because of active hepatitis C, sulfasalazine was used as first line treatment for RA, but was unable to control his arthritis. After hepatitis C cure, methotrexate was tried, but RN increased in number and size on the extensor surfaces of his elbows and wrist (the largest on the elbow was 7 cm in diameter without associated bursitis as confirmed by ultrasound). Given the worsening of the nodules under methotrexate therapy, then it was decided to start upadacitinib 15 mg/day. Four months after treatment, there was a significant reduction of RN size by > 50% in both sites and arthritis resolution (image 1).

Case 2: 64-year-old female patient with RA seropositive for RF and ACPA with disease onset 10 years ago. She underwent lung CT scan due to prolonged and recurrent cough with worsening in recent months. Multiple cavitated nodules, subpleural and adjacent to the hilum and bronchi, were detected diffusely and bilaterally. Bilateral subpleural lung lesions had increased nonspecific metabolic activity (SUV 1-3) on PET scan. She underwent a biopsy of the subpleural lesion in the left upper lobe to rule out cancer and tuberculosis. Histology showed necrotizing granulomas with palisading histiocytes, which could be consistent with either tuberculosis infection or rheumatoid nodules. Microbiology exams were negative for tuberculosis. The clinical evolution and the lack of progression of the lesions over one year, as evidenced by serial reassessment CT scans, suggested the diagnosis of lung rheumatoid nodules. Despite intolerance to multiple drugs tried previously (methotrexate, sulfasalazine, hydroxychloroquine) and the ineffectiveness of leflunomide, tocilizumab and tofacitinib, the patient started therapy with upadacitinib and leflunomide with improvement of lung RN



052 – Figure 1. Image sequence showing rheumatoid nodules on the extensor surfaces of both elbows and right wrist at diagnosis (left side) and the successful size reduction of them Just 4 months after starting Upadacitinib and Leflunomlde therapy (right side).



052 – Figure 2. Chest CT scan showing dimensional reduction of subpleural and parenchymal rheumatoid nodules along with bronchiectasis, comparing the initial CT scan (left side) with the latest CT scan 10 months after starting Upadacitinib therapy (right side)

10 months after (image 2) and RA joint remission.

These two cases highlight the challenge in treating RN. Diagnosis can also be challenging, especially when localized to the lungs, as imaging complementary exams are not enough to distinguish between neoplasia or infections like tuberculosis, and neither is their histology. Our findings corroborate recent data on JAK inhibitors efficacy in controlling cutaneous rheumatoid nodules (Rosentein ED, et al, 2024).

073 - EPSTEIN-BARR VIRUS-ASSOCIATED LYMPHOPROLIFERATIVE DISEASE OCCURRING DURING METHOTREXATE THERAPY FOR SJOGREN'S SYNDROME

Laura Gago[1, 2], Mariana Emília Santos[1, 3], Maria Manuela Costa[1], Ana Filipa Mourão[1, 2], Jaime C. Branco[1, 3], MJ Gonçalves [1, 3]
[1]Rheumatology Department, Unidade Local de Saúde de Lisboa Ocidental, Hospital Egas Moniz, Lisboa, Portugal, [2]Comprehensive Health Research Centre (CHRC), NOVA Medical School | Faculdade de Ciências Médicas, NMS|FCM, Universidade Nova de Lisboa, Lisboa, Portugal, [3]Universidade Nova de Lisboa, Nova Medical School, Lisboa, Portugal

Background: Methotrexate (MTX) is an immunosuppressive agent used as a first-line treatment for a variety of rheumatic diseases. Over the years it has been associated with some lymphoproliferative diseases. We describe the case of a 71-year-old woman, followed in our department with primary Sjogren's Syndrome (SS), medicated with MTX, with lymphoproliferative disease associated with reactivation of Epstein Barr virus (EBV).

Clinical case: A 71 years-old-women, followed in our department since 2019 for primary SS with antinuclear antibody 1/320, negative rheumatoid factor, positive anti-SSA antibody and low complement (C4). She was medicated with oral MTX in doses ranging from 7.5-12,5 mg/week and prednisolone 5 mg/week due to persistent arthritis, since 2019.

Due to 1-month history of thoracic opressive pain and dyspnea, the patient underwent a chest CT scan that revealed multiple adenomegalies in the mediastinum and pulmonary hila. The largest had a diameter of 42 mm.

A PET scan was requested, and no malignant lesions were found. The 42 mm mediastinal mass was subsequently biopsied and the pathology report revealed granulomatous lymphadenitis.

Immunophenotyping didn't detect abnormal lymphoid populations and cytology was negative. Lymphadenopathies from the cervical and inguinal regions were also biopsed showing lymphadenitis and EBV was detected in this samples using in situ hybridization (ISH) with EBV-encoded small RNA (EBER).

The EBV viral load was measured and found to be positive (19550 IU/mL). Thus, the most likely diagnostic hypothesis was lymphoproliferative disease due to EBV reactivation in the context of MTX therapy, so we decided to discontinue this therapy.

Two months after the withdrawal of MTX, a chest CT scan was repeated, showing a marked reduction in the size of the mediastinal adenopathies, with improvement in chest pain and dyspnea. Also, the EBV viral load is decreasing (4385 IU/mL).

Conclusion: This case shows that MTX therapy can play a role in the development of EBV-associated lymphoproliferative disease in patients with rheumatic diseases. More studies and case reports are needed to confirm this causal link.

074 - NOT YOUR USUAL SUSPECTS: BEHIND PULMONARY NODULES IN RHEUMATOID ARTHRITIS

Duarte Augusto[1], Sara Paiva Dinis [1], Filipe Cunha Santos[1], Nathalie Madeira[1], Joana Fonseca Ferreira[1, 2], Catarina Quinaz[3], Cláudia Vaz[1, 2]
[1]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, [2]Faculdade de Ciências da Saúde, Universidade da Beira Interior, Covilhã, Portugal, [3]Serviço de Doenças Infeciosas, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal

Introduction: Rheumatoid arthritis (RA) is a chronic inflammatory disorder that primarily affects the joints but can also lead to extra-articular manifestations such as rheumatoid nodules. Although they can present elsewhere, these typically occur in areas of mechanical stress.

Mycobacterium avium complex encompasses multiple nontuberculosus mycobacterial species, which often present with tuberculosis-like symptoms but can also manifest as solitary pulmonary nodules resembling lung cancer.

In this case report, we describe a patient with RA who developed pulmonary nodules during routine follow-up. Initially suspected to be rheumatoid nodules, further investigation revealed these nodules to be caused by another entity other than RA.

Clinical report: A 55-year-old woman with seropositive RA diagnosed at age 34, treated with 10mg of methotrexate and 20 mg of leflunomide, presented with a 3 mm pulmonary nodule, annually monitored through imaging. The patient was referred to an oncologic pulmonology consultation due to the suspicion of primary lung neoplasia when the most recent chest CT scan revealed a suspicious 7 mm nodule in the internal segment of the right middle lobe and micronodular subpleural nodules in the left lung.

Since 2021, the nodule had been under surveillance and showed an increase in size. A PET scan with FGD-F18 revealed a 15mm nodule in the right lower lobe, with central cavitation and irregular contours, showing mild 18FDG contrast uptake, initially presumed to be a necrobiotic nodule associated with RA. Bilateral pulmonary nodules were also noted, with the

largest being 6.5 mm on the right and 6 mm on the left. These findings initially suggested rheumatoid pulmonary nodules, and escalation to JAK inhibitor therapy was proposed.

During this period, the patient was clinically stable and without respiratory symptoms. In preparation for the introduction of immunosuppression, Interferon Gama Release Assay and Mantoux tests were performed, both returning negative results. Due to the cavitated nodule and the proposed therapy, a mycobacteriological and bacteriological sputum examination were performed, revealing positive results for Mycobacterium intracellulare in two samples. Analysis at the Instituto Nacional de Saúde Doutor Ricardo Jorge confirmed sensitivity to clarithromycin. Previously thought to be rheumatoid nodules, they were thus attributed to Mycobacterium intracellulare infection. Treatment with azithromycin, weight-adjusted ethambutol, and rifampicin was initiated. The patient remains asymptomatic and is expected to recover fully while continuing RA treatment.

Conclusion: This case report highlights not only the importance of a thorough investigation and the complexity of diagnosing rheumatoid nodules in patients with RA. While rheumatoid nodules are a well-recognized manifestation of RA, it is crucial to consider alternative diagnoses, particularly infectious or cancerous causes. The identification of Mycobacterium intracellulare as the probable causative agent in this patient underscores the importance of thorough microbiological evaluation in cases of pulmonary nodules, especially prior to the initiation of immunosuppressive therapy. We believe that the patient will improve, as the root cause is now being treated. In the end, it serves as a reminder of the diverse etiologies of rheumatoid nodules and the importance of a comprehensive and multidisciplinary approach in our patients with systemic diseases.

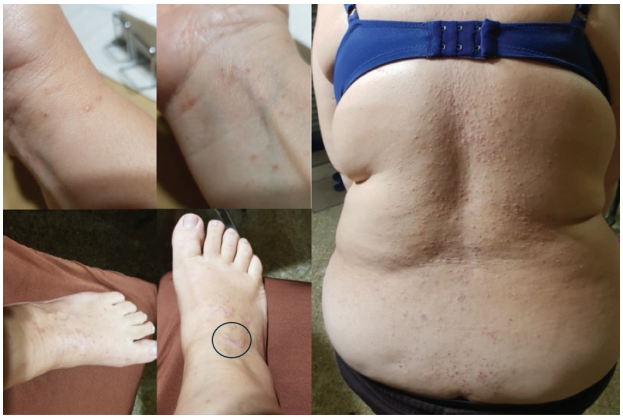
076 - A RARE CASE OF LICHENOID DRUG ERUPTION IN A PATIENT WITH SPONDYLARTHRITIS RECEIVING SECUKINUMAB

Carlos Marques-Gomes[1, 2], Bárbara Granja[2, 3], Mariana Diz-Lopes [1, 2], Miguel Bernardes[1, 2], Lúcia Costa[1]
[1]Rheumatology Department, Centro Hospitalar de São João, Porto, Portugal, [2]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal, [3]Dermatology Department, Centro Hospitalar de São João, Porto, Porto, Portugal

Introduction: Lichen planus (LP) is an idiopathic disease whose pathogenesis is poorly understood, but it appears to represent a T-cell-mediated autoimmune disease. LP is characterized by pruritic, violaceous papules and plaques, commonly found on the wrists, lower back, and ankles. Several drugs have been associated with skin LP (drug-induced lichen planus, DI-LP). Cutaneous lichenoid reaction is rarely reported as an adverse effect of interleukin-17A inhibitors.

Presentation: A 51-years-old woman was diagnosed with spondylarthritis, axial and peripheral involvement [inflammatory low back pain with good response to non-steroidal anti-inflammatory drugs, oligoarthritis, sacroiliitis on MRI, elevated C-reactive protein (CRP) and HLA-B27 positivity], at the age of 35, in 2008. She was treated with naproxen, prednisolone, sulfasalazine, adalimumab (40 mg weekly, from 2017 to 2019) and certolizumab (CTZ, 200 mg every other week, from 2019 to 2022). In December 2022, due to secondary failure, the patient switched from CTZ to secukinumab (SEKU), 300 mg every 28 days. In November 2023, she presented to the emergency department with a 5-months history of a pruritic rash on her back, which later spread to her ankles, wrists, suprapubic region and lower abdominal region. She had no fever and there were no respiratory, genitourinary or gastrointestinal symptoms. There was no other relevant past medical history other than obesity. She was also taking acemetacin (60 mg id), duloxetine (30 mg id), rabeprazole (20 mg, id) and samaglutide (0.5 mg/0.37 mL, weekly). On physical examination, there were pruritic, violaceous papules and plaques (clusters of shiny, raised, purple-red blotches) scattered over the lumbar region, ankles, feet and wrists, without transudate or signs of bacterial infection (Fig. 1). She also had lacy-like lesions on oral mucosa and tongue. Laboratory results showed elevated CRP (11.1 mg/L) and ESR (65 mm/1st hour). The patient was observed by Dermatology department, and the diagnosis of lichen planus was made. She was discharged and received topical betamethasone dipropionate and tacrolimus. SEKU was stopped and the patient started on treatment with etanercept (50 mg, weekly), on January 2024, after skin lesions resolution. At six months of treatment with tumor-necrosis-alpha (TNF) inhibitor, no new lesions had appeared, and the patient had no articular symptoms.

Discussion: With the increasing use of biological disease modifying anti-rheumatic drugs (bDMARDs), there have been reports of lichenoid drug eruptions after initiation of biologic agents. In a systematic review, TNF-inhibitors were the most implicated bDMARDs and only 3 (of 67) cases of DI-LP were associated with SEKU. In our review, we found 1 additional case report of LP secondary to SEKU. The period between biologic drug administration and lichenoid eruption can range from 3 to 8 months. This woman developed LP after



076 – Figure 1. Itchy lichenoid lesions (clusters of shiny, raised, purple-red blotches) scattered over the lumbarregion, ankles, feet and wrists. Note a violet plaque on the right ankle (circle).

6 months of treatment with SKU. DI-LP can usually be managed by discontinuing the offending agent and, sometimes, with corticosteroids or other immunosuppressors (such as tacrolimus). Although it can be challenging in rheumatic patients, as it may contribute to delaying disease remission, switching biologic class does not appear to lead to resurgence of LP, as is evident in our patient. This case highlights the importance of recognizing skin conditions potentially associated with bDMARDs. SEKU is usually used in the rheumatologist daily clinical practice. We report a rare cutaneous adverse effect associated with this biological agent.

077 - DEFORMIDADE ARTICULAR E OLHO "EM CABEÇA DE MEDUSA" - UM CASO DE EHLERS-DANLOS

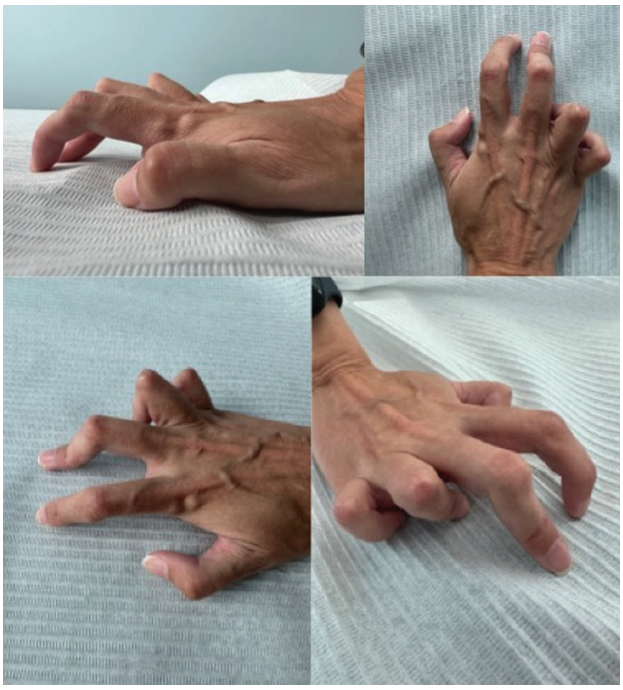
Carlos Marques-Gomes[1, 2], Mariana Diz-Lopes [1, 2], Paula Freitas[2, 3], João Oliveira[4], Ana Aires Silva[5], Miguel Bernardes[1, 2], Lúcia Costa[1]

[1]Rheumatology Department, Centro Hospitalar de São João, Porto, Portugal, [2]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal, [3]Endocrinologia, Centro Hospitalar e Universitário de São João, Porto, Portugal, [4]Centro Hospitalar Universitário de São João, Porto, Portugal, [5]Serviço de Neurologia, Centro Hospitalar Universitário de São João, Porto, Portugal

Caso clínico: Sexo feminino, 33 anos, observada na consulta de Reumatologia (março de 2018) por limitação na extensão ativa dos dedos da mão direta com 18 meses de evolução. Negava poliartralgias inflamatórias e alterações cutâneas e referia história de equimoses espontâneas ou resultantes de traumatismos menores. Ao exame objetivo, apresentava baixa estatura (1,52 metros), pavilhões auriculares com hélice fina, mal-oclusão dentária (de tipo mordida aberta), acantose nigrans das axilas, mãos pequenas, com dedos finos e adelgaçados, veias superficiais das pernas visíveis na região pretibial, pés pequenos com procidência posterior dos calcâneos, deformidade em flexão de todos os dedos da mão direita (Fig. 1), com limitação franca da sua extensão (contraturas tendinosas em flexão em maior grau nos 4º e 5º dedos), discreto halo esclerótico azulado, elasticidade cutânea aumentada e lipodistrofia. Dos antecedentes pessoais, destacava-se correção cirúrgica de pé boto à direita (4 meses de idade) e seguimento em consulta de endocrinologia por suspeita de lipodistrofia familiar parcial (porém, com estudo genético molecular normal). O estudo analítico não apresentou alterações. Realizou ressonância magnética da mão direita que revelou luxação do tendão do extensor cubital do carpo, sem outras alterações. Em dezembro de 2019, na sequência de cefaleia fronto-parietal direita foi internada no serviço de Neurologia tendo sido diagnosticada fístula carótido-cavernosa direita com refluxo venoso cortical e marcada drenagem venosa anterior para as veias oftálmicas, que condicionava arterialização dos vasos sanguíneos conjuntivais e episclerais num aspeto em "cabeça de medusa". Foi solicitada consulta de genética com estudo molecular e o diagnóstico de Síndrome de Ehlers-Danlos do tipo vascular (EDV) foi estabelecido (variante C.1149+3A>T do gene COL3A1 em heterozigotia). A patogenicidade desta variante foi confirmada em estudo funcional em cultura de fibroblastos obtidos em biopsia de pele. Foram excluídas outras alterações vasculares com o angio-TC toraco-andomino-pélvico. A doente continua em vigilância sem outras complicações até ao momento.

Discussão: A Síndrome de Ehlers Danlos corresponde a um grupo de doenças hereditárias do tecido conjuntivo cuja apresentação clássica engloba hiperelasticidade cutânea, hipermobilidade articular, cicatrizes atróficas e fragilidade vascular, podendo a baixa estatura ou contraturas tendinosas dos dedos estar presentes. O diagnóstico genético é necessário; a identificação do tipo (clássica, hipermobilidade, vascular, cifoescoliose, artrocalasia, dermatosparaxis) torna-se relevante para a orientação do doente. As complicações são variáveis: difícil cicatrização cutânea, perfuração do globo ocular, (sub)luxação articular, cifoescoliose, pé equinovaro, pé plano, displasia congénita da anca, hérnias gastrointestinais, osteoartrose precoce (pelas deformidades articulares persistentes). Na EDV (mutações nos genes COL3A1 e COL1A1), o risco de complicações vasculares (rotura arterial ou visceral) é elevado e tem impacto na mortalidade.

Apresentamos uma doente com suspeita de lipodis-



077 – Figure 1. Deformidades dos dedos da mão direita correspondentes a contraturas tendinosas em flexão.

trofia desde os 28 anos de idade em que o surgimento de deformidades articulares e as alterações vasculares (incluindo, o surgimento da fístula carótido-cavernosa) levaram à reconsideração do diagnóstico e à repetição do estudo genético que confirmou a existência de EDV. Este caso, realça, assim, a importância da revisitação diagnóstica e traduz, mais uma vez, a influência do fator "tempo" no estabelecimento do diagnóstico reumático definitivo.

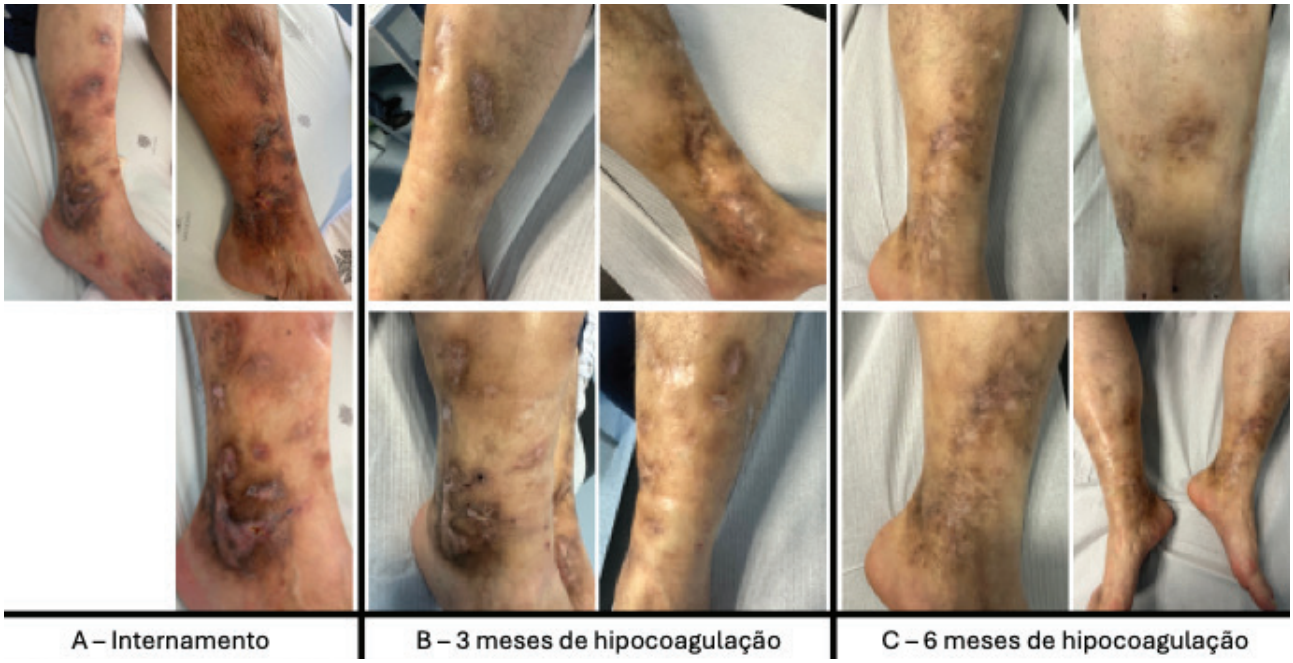
078 - LESÕES VASCULÍTICAS RECORRENTES NOS MEMBROS INFERIORES COM 3 DÉCADAS DE EVOLUÇÃO - UM CASO DE VASCULITE LIVEDÓIDE

Carlos Marques-Gomes[1, 2], Mariana Diz-Lopes [1, 2], Miguel Bernardes[1, 2], Lúcia Costa[1]
[1]Rheumatology Department, Centro Hospitalar de São João, Porto, Portugal, [2]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal

Caso clínico: Doente do sexo feminino, 50 anos de idade, admitida a internamento de Reumatologia para estudo de lesões purpúricas e ulceradas nos membros inferiores (MI), associadas a parestesias, com 2 meses de evolução. A doente era portadora do diagnóstico de panarterite nodosa desde os 18 anos de idade, após realização de 5 biopsias cutâneas e avaliação por Dermatologia. Aos 25 anos, por recorrência de lesões ulceradas no terço distal dos MI, em consulta de Reumatologia privada, iniciou tratamento com azatioprina (50 mg/dia) e deflazacort (6 mg/dia), que auto-suspendeu aos 45 anos. Na admissão, negou poliartralgias, úlceras orais, rash malar de novo, fotossensibilidade, história de trombose venosa e/ou arterial e de abortamentos espontâneos. Apresentava múltiplas lesões ulceradas no terço distal de ambas as pernas associadas a zonas de hiperpgimentação com região central hipopgimentada/cicatricial, semelhantes a lesões vasculíticas (Fig. 1); não era evidente artrite periférica. Do estudo inicial, destacavam-se os aumentos da velocidade de sedimentação (51 mm/1ªh) e da proteína C reativa (13.7 mg/L). O estudo imunológico (incluindo os fatores do complemento e os anticorpos anti-nucleares, anti-dupla cadeia do DNA, anti-fosfolipídicos, anti-citoplasma de neutrófilo) foi negativo. A eletromiografia foi compatível com polineuropatia axonal sensitiva, mas com franca limitação técnica. A biópsia da pele revelou a presença de um infiltrado inflamatório linfocítico num vaso da derme, no entanto, sem depósitos imunes, nem necrose fibrinoide, compatível com vasculopatia do tipo obliterativo. O estudo protrombótico não revelou alterações relevantes e a pesquisa de mutações no gene CECR1, codificante da adenosina deaminase 2 foi negativo. Dada a ausência de achados sugestivos de vasculite imunomediada, o diagnóstico de vasculopatia livedóide foi estabelecido e a doente iniciou riavaroxabano na dose de 10 mg/dia. Após 1 ano de tratamento, houve resposta quase completa, com diminuição franca das lesões cutâneas (Fig. 1).

Discussão: A vasculite livedóide é uma doença crónica caracterizada pela ulceração recorrente da pele (após a formação de lesões purpúricas), sobretudo, nos MI, tendo um impacto negativo significativo na qualidade de vida dos doentes afetados. Trata-se de uma patologia rara (incidência de 1 para 100 mil indivíduos), mais frequente no sexo feminino. Pode surgir isoladamente ou associada a conectivite (nomeadamente, como manifestação da síndrome antifosfolipídica ou do lúpus eritematoso sistémico).

Até ao momento, não existe um tratamento estabelecido, no entanto, a utilização de novos anticoagulantes orais (NOACs) tem sido frequente. É também descrito o uso de heparina de baixo peso molecular, varfarina, corticopterapia, imunoglobulinas endovenosas, antiagregantes plaquetares, radiação ultravioleta e trombolíticos. Sendo uma doença rara, não existem ensaios clínicos e estudos observacionais robustos, pelo que a descrição de séries e de casos clínicos na literatura são a principal fonte de evidência no que respeita à melhor definição do tratamento. Neste caso clínico fica patente a eficácia dos NOACs e, em particular, do rivaroxabano, no tratamento da vasculite livedóide.



078 – Figura 1. Evolução das lesões ulceradas de vasculite lievedóide. A - Lesões ativas em internamento. B - Resolução parcial das lesões após 3 meses de tratamento com rivaroxabano. e - Resolução quase completa das lesões 12 meses após início do tratamento com rivaroxabano.

Podendo as lesões cutâneas características da vasculite livedóide ser potencialmente confundidoras quanto ao diagnóstico diferencial, ou até mesmo serem manifestação, de várias doenças reumáticas, este caso realça a importância da revisitação diagnóstica, destacando o papel do estudo histológico e imunológico na (exclusão) caracterização das doenças inflamatórias.

090 - LEPROSY: A FORGOTTEN MIMIC

Daniel Melim[1, 2], Ana Catarina Moniz[2], Mariana Emília Santos[2, 3], Laura Gago[2, 3], João Borralho[4], Cristina Gonçalves Castro[5], Isabel Viana[5], Maria João Gonçalves[2, 3], Manuela Costa[2], Jaime C. Branco[2, 3]

[1]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal, [2]Rheumatology Department, Unidade Local de Saúde de Lisboa Ocidental, Hospital Egas Moniz, Lisboa, Portugal, [3]Universidade Nova de Lisboa, Nova Medical School, Lisboa, Portugal, [4]Department of Infectious Disease, Unidade Local de Saúde de Lisboa Ocidental, Hospital Egas Moniz, Lisboa, Portugal, [5]Department of Dermatology and Venereology, Unidade Local de Saúde de Lisboa Ocidental, Hospital Egas Moniz, Lisboa, Portugal

Introduction: Leprosy is a chronic infectious disease caused by Mycobacterium leprae, primarily affecting the skin and peripheral nerves. Rheumatological symptoms are common, and Leprosy can mimic various diseases such as Relapsing Polychondritis (RP), due to findings of symmetric chronic polyarthritis, perichondritis and autoantibodies.

Clinical Case: We report the case of a previously healthy 26-year-old Brazilian man living in Portugal for 1 year. Over the past 6 months, he developed progressive deforming facial edema and erythema, as well as pain and swelling in the small joints of his hands. The physical exam revealed arthritis in the metacarpophalangeal and proximal interphalangeal joints of both hands. His ears showed nodular infiltrates and erythema thus RP was considered as a diagnosis. Diffuse infiltration and enlargement of his face, particularly nose and lips, as well as rarefaction of eyebrows and eyelashes were also noted. He had nodular lesions on his legs, some of them ulcerated, and non-pitting edema and petechiae on his feet. Due to involvement of both pinna and lobules of the ears the hypothesis of RP was refuted. Laboratory studies showed an elevated ESR, a positive rheumatoid factor and triple positive antiphospholipid autoantibodies. An electromyogram showed axonal sensorimotor polyneuropathy and a skin biopsy confirmed the presence of numerous acid-fast bacilli with positive Fite and Ziehl-Neelsen staining, establishing the diagnosis of Leprosy. He was started on corticosteroids and a multidrug antibiotic regimen.

Conclusion: Leprosy can mimic rheumatologic diseases and, although it has been mostly eliminated in Portugal, the increase in migration from countries where it is still endemic, such as Brazil, means that this potential diagnosis should be considered. Therefore, a high

level of suspicion and thorough physical examination are necessary for an accurate diagnosis and to avoid unnecessary immunosuppressive therapy and promptly initiate antimycobacterial therapy.

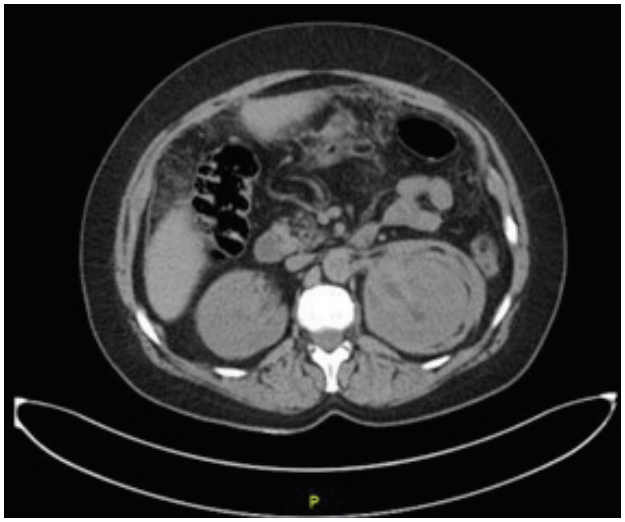
093 - ERDHEIM-CHESTER DISEASE: AN ELUSIVE DIAGNOSIS

Daniel Melim[1, 2], Ana Catarina Moniz[2], Sara Dias Rodrigues[2], Mariana Emília Santos[2, 3], Laura Gago[2, 3], Maria João Gonçalves[2, 3], Manuela Costa[2], Jaime C. Branco[2, 3], Carina Lopes[2, 3]

[1]Rheumatology Department, Centro Hospitalar do Funchal, SESARAM, Funchal, Portugal, [2]Rheumatology Department, Unidade Local de Saúde de Lisboa Ocidental, Hospital Egas Moniz, Lisboa, Portugal, [3]Universidade Nova de Lisboa, Nova Medical School, Lisboa, Portugal

Introduction: Erdheim-Chester disease (ECD) is a rare non-Langerhans cell histiocytosis characterized by the clonal expansion of foamy histiocytes in various organs and tissues. The diagnosis is often delayed due to its diverse clinical presentation, although it typically affects the bones, retroperitoneum, heart, vessels, central nervous system, and lungs. This multisystemic involvement can lead to confusion and misdiagnosis of other rheumatic diseases.

Clinical Case Report: We report a case of a 45-year-old woman who presented with additive and symmetric polyarthritis in 2011. She was diagnosed with seronegative Rheumatoid Arthritis and started on Methotrexate with subsequent addition of Sulfasalazine and Infliximab due to refractory polyarthritis. In 2018, she experienced abdominal pain partially relieved by analgesics and an abdominal and pelvic CT scan disclosed a thickened greater omentum suggesting peritoneal carcinomatosis. All immunosuppressive treatment was discontinued, and she underwent a diagnostic laparoscopy, ruling out malignancy. The greater omentum biopsy revealed sclerosing peritonitis with xanthogranulomatous infiltration. In 2021, a follow-up abdominal and pelvic MRI showed thickening of the pararenal fascia in the left kidney consistent with "hairy kidney" (Figure 1). This was associated with heterogeneous peritoneal thickening, displaying an omental cake morphology previously observed in the CT scans and peri-aortic segmental thickening, para-renal, and proximal ureteral wall thickening were also reported. A PET scan revealed inflammatory activity in multiple locations including the aorta and adjacent tissue, kidneys, adrenals, and lymph nodes. After case review and excluding other causes of aortitis, "seronegative" IgG4-related disease was considered the most likely diagnostic hypothesis and she was started on Prednisolone 1mg/kg/day and Rituximab. The patient maintained elevated inflammatory parameters and a worsening inflammatory involvement in the aortic region was evident on the follow-up PET scan. The lack of response to this usually efficacious treatment led to consideration of other non-rheumatic infiltrative diseases, such as ECD. Biopsy revision confirmed xanthomatous histiocytes (CD68+, CD1a-) and a whole-body bone scan showed uptake of Tc-99m in the distal femurs and proximal tibias, consistent with ECD. The patient was referred to the Hematology department and started on interferon alpha therapy.



090 – Figure 1. Left kidney with a "Hairy kidney" appearance

Conclusion: Herein, we present a case of ECD that presented with skeletal, retroperitoneal, and vessel involvement and, to our knowledge, arthritis is scarcely reported in the literature. The diagnosis was possible after a careful review of the clinical presentation, imaging, and tissue biopsy with concordant immunohistochemistry.

The authors wish to raise awareness among fellow clinicians about this rare disease as it can mimic other rheumatic conditions such as IgG4-related disease, which may lead to unnecessary and potentially harmful treatments.

095 - EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS TREATED WITH MEPOLIZUMAB AND RITUXIMAB COMBINATION THERAPY – A CASE REPORT

Ana Catarina Moniz[1, 2], Laura Gago[1, 2], Mariana Emília Santos[1, 2], Daniel Melim[2, 3], Sara Dias Rodrigues[1, 2], Paula Araújo[1, 2], Jaime C. Branco[1, 2], MJ Gonçalves [1, 2]

[1]Rheumatology Department, Unidade Local de Saúde de Lisboa Ocidental, Hospital Egas Moniz, Lisboa, Portugal, [2]Universidade Nova de Lisboa, Nova Medical

School, Lisboa, Portugal, [3]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal

Eosinophilic granulomatosis with polyangiitis (EGPA) is a multisystemic disease characterized by eosinophilic necrotizing vasculitis of small and medium sized blood vessels, mainly affecting the lungs, skin and peripheral nervous system (PNS). Both rituximab and mepolizumab are recommended in the treatment of EGPA2. However, the use of both drugs in combination is not usually performed. To date, there are only six reported cases of both mepolizumab and rituximab in combination and two cases of sequential therapy.

We report a case of a 46-year-old man with a past medical history of mild asthma and chronic rhinosinusitis. The patient presented in the emergency department with worsening dyspnoea, numbness of the feet and purpuric lesions of the lower limbs, causing severe functional impairment and complete inability to walk. Laboratory studies showed eosinophilia of 16%, ESR of 58 mm/h and negative ANCA. The chest CT showed ground glass areas in both lungs and the head CT showed inflammatory mucosal thickening of all perinasal sinus. Cutaneous punch biopsy of the skin lesions revealed eosinophilic infiltrate with granulomas. Electromyography revealed severe axonal polyneuropathy in upper and lower limbs. The diagnosis of ANCA-negative EGPA was established. Patient received treatment with methylprednisolone pulse therapy 1 g/day for five days and cyclophosphamide 750mg/m2 monthly for six months. Maintenance therapy included prednisolone 1 mg/kg/day in a tapering scheme and azathioprine 150 mg/day. After two years of treatment, despite significant clinical (mainly neurological and cutaneous) and analytical improvement, it was not possible to taper prednisolone below 10mg/day because of respiratory symptoms recurrence. He developed osteoporosis with multiple vertebral fractures as corticoid complication. When mepolizumab was approved for EGPA , about seven years after the diagnosis, the patient started this treatment, allowing to decrease prednisolone to 5 mg/day with no respiratory exacerbations. However, he began experiencing polyarthritis, which only partially responded to optimising subcutaneous methotrexate and joint synoviorthesis. In a multidisciplinary meeting, it was considered to start Rituximab for both articular and pulmonary involvements, but pneumonology specialists preferred mepolizumab due to its positive effects on lung manifestations. Hence, he started a combination of mepolizumab (300mg every four weeks) and rituximab (2 infusions of 1000 mg each, 15 days apart, every 6 months). After 6 months of this therapy, there was a significant improvement in joint symptoms, and no other disease exacerbations were reported, even with a gradual reduction of glucocorticoid dosage to 5 mg/day, with no apparent adverse effects to report.

Herein, we report a case of a patient with ANCA-negative EGPA experiencing severe respiratory and PNS symptoms that required multiple treatment adjustments. As described, only mepolizumab was effective in controlling respiratory symptoms but had minimal effect in articular disease, justifying the use of rituximab for refractory arthritis in the setting of low disease activity, resulting in sustained remission and improved quality of life. This case highlights the complexity of this multiorgan disease, with multiple flares and corticoid dependency, which needed treatment associations with different mechanisms of action to control all disease involvements. Despite the scarce cases reported, the association between rituximab and mepolizumab seemed safe and effective.

097 - COULD CGRP MABS FOR MIGRAINE TRIGGER RHEUMATOID ARTHRITIS? INSIGHTS FROM A CASE REPORT

Catarina Rua[1], Tiago Beirão[1], Catarina Silva[1], Tiago Meirinhos[1], Patrícia Pinto[1], Romana Vieira[1], Joana Abelha-Aleixo[1], Flávio Campos Costa[1], Diogo Fonseca[1], Ana Sofia Pinto[1], Beatriz Samões[1], Taciana Videira[1]
[1]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Background: Calcitonin gene-related peptide (CGRP) is involved in pain pathways and is primarily released from sensory nerves and the central nervous system. Monoclonal antibodies (mAbs) targeting CGRP have shown promising outcomes for migraine prevention. However, reports suggest that CGRP mAbs may trigger inflammatory flares in patients with pre-existing autoimmune diseases. We present a case of new-onset rheumatoid arthritis (RA) diagnosed three months after initiating fremanezumab therapy, highlighting RA as a potential adverse event of CGRP mAb treatment.
Case Description: A 56-year-old woman with a history of severe migraines, hypertension, and depression experienced significant improvement in migraine frequency and intensity after starting fremanezumab 225 mg monthly. However, three months into the treatment, she developed symmetric inflammatory polyarthritis. Physical examination confirmed polyarthritis, and laboratory tests showed elevated inflammatory markers and positive anti-cyclic citrullinated peptide antibodies. Initial X-rays of hands, feet, and knees were normal. Diagnosed with RA, she was treated with prednisolone, methotrexate, folic acid, calcium carbonate, and cholecalciferol, resulting in clinical improvement. Despite recommendations, she continued fremanezumab

due to its efficacy in reducing migraine episodes. Her articular symptoms persisted after discontinuation of fremanezumab. She temporarily ceased methotrexate due to a bacterial infection, resulting in a flare-up of RA characterized by polyarthritis and elevated inflammation markers. Later, changed to leflunomide 20 mg due to gastrointestinal intolerance and is currently in remission from her RA symptoms.

Discussion/Conclusions: CGRP mAbs are effective for migraine prevention, but there are documented cases of immune-mediated disease flares, such as in psoriatic arthritis, psoriasis, and granulomatosis with polyangiitis, triggered by them. This case is unique as it suggests a potential association between CGRP mAbs and RA onset, underscoring the importance of ongoing monitoring and reporting of adverse events. Further research is essential to fully comprehend the impact of CGRP modulation in rheumatic/autoimmune diseases.

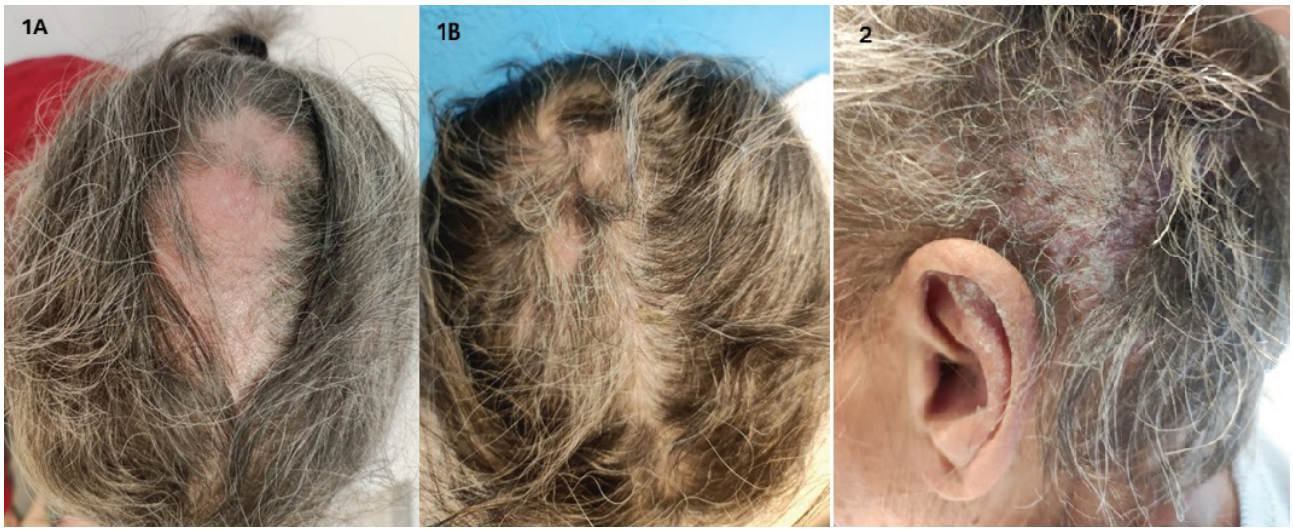
100 - SCALP INVOLVEMENT IN DERMATOMYOSITIS: REPORT OF TWO CLINICAL CASES

Pedro Miguel Teixeira[1, 2], C. Pinto Oliveira[1, 2], Carolina Vilafanha[1, 2], Eduardo Dourado[1, 2], Ana Rita Prata[1, 2], Gisela Eugénio[1, 2], Manuela Loureiro[2, 3], Anabela Barcelos[1, 2]
[1]Rheumatology Department, Unidade Local de Saúde da Região de Aveiro, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz Health Alliance, Portugal, Aveiro, Portugal, [3]Dermatology Department, Unidade Local de Saúde da Região de Aveiro, Aveiro, Portugal

Introduction: Dermatomyositis is an idiopathic inflammatory myopathy known for its distinctive cutaneous manifestations, often accompanied by multisystemic manifestations. While classic skin findings such as Gottron's papules and the heliotrope rash are well recognized, scalp involvement is less frequently reported and can be easily overlooked.

Case 1: A 60-year-old woman was referred to Rheumatology with a 6-month history of inflammatory arthralgia of the wrists, hands, and knees. She exhibited a photosensitive erythematous rash in the palpebral, infraorbital, and anterior cervical regions, along with erythematous lesions on the dorsal and palmar aspects of the proximal and distal interphalangeal joints. Additionally, a large nonscarring alopecia plaque with surrounding scaling was evident on the scalp (Figure 1A). She had 5 tender/swollen joint counts out of 28 on physical examination, confirmed by ultrasound, and an MMT8 score of 146/150. There was no reported history of muscle weakness, dysphagia, or dyspnoea. Laboratory workup showed mildly elevated acute phase reactants, normal muscle enzymes, and a positive anti-MDA5 antibody. A nonspecific interstitial pneumonia (NSIP) pattern was found on chest computed tomography (CT). A scalp biopsy revealed interface dermatitis, leading to a diagnosis of pauci-myopathic anti-MDA5 dermatomyositis. Initial treatment included topical and oral glucocorticoids and hydroxychloroquine, followed by rituximab due to refractory joint and lung involvement. Four months post-rituximab infusion, notable improvements included hair regrowth, reduction of facial lesions, and stability of systemic involvement (Figure 1B.)

Case 2: An 82-year-old woman diagnosed with clinically amyopathic dermatomyositis 17 years before, sought a Rheumatology consultation due to Dermatology loss of follow-up. The patient presented with new hypopigmented patches on the scalp, hyperkeratotic



100 – Figure 1. Non-scarring alopecia, before and after 4 months of treatment. Figure 2. Psoriasiform changes in the scalp

plaques, hair thinning, and significant scaling (Figure 2.). A scalp biopsy revealed paucicellular interface dermatitis with dermal fibrosis and focal mucin deposition, indicative of active dermatomyositis. Treatment with topical steroids and azathioprine markedly improved the cutaneous lesions.

Conclusion: Scalp involvement is an important yet often neglected manifestation in patients with dermatomyositis. It may be present at the time of diagnosis or develop years after the initial symptoms, which creates additional challenges in diagnosis and therapeutic management.

105 - REFRACTORY RS3PE IN A PATIENT WITH INTESTINAL LEISHMANIASIS

Vanessa Fraga[1], Catarina Abreu[1], Susana Matias[1], Sandra Sousa[1], Maria José Santos[1, 2]

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

Introduction: RS3PE stands for Remitting Seronegative Symmetric Synovitis with Pitting Edema, which is a rheumatic syndrome that usually responds well to low-to-intermediate doses of corticosteroids. Leishmaniasis is an infectious zoonotic disease caused by a protozoan, typically presenting with fever, hepatosplenomegaly, and pancytopenia. Atypical disease manifestations include intestinal and articular involvement1.

Case report: A 66-year-old male smoker, with a previous history of peptic ulcer and hypertension presented to the emergency department with sudden onset of severe inflammatory arthralgia involving small and medium joints with impact on his daily activities. The patient denied having fever or other associated symptoms. There were no previous episodes of arthritis or recent infections. Physical examination revealed polyarthritis of the small joints of the hands, wrists, ankles and exuberant pitting edema of hands and feet. Remaining examination was unremarkable. The laboratory workup showed high inflammatory markers (ESR 70mm/h and CRP 3.9mg/dL). Hemogram, renal and hepatic function were normal. Viral serologies for EBV, HIV 1/2, HCV, HBV and CMV were negative. Autoimmunity including ANA, ENA, rheumatoid factor and ACPA were negative. The diagnosis of RS3PE syndrome was assumed and treatment with prednisolone (PDN) 20mg/d was started with benefit. During corticosteroids tapering, the patient experienced a worsening of symptoms with persistent polyarthritis, requiring a higher dose of prednisolone (max 60mg/d) and hydroxychloroquine (400mg/d) for symptom control. Additionally, there was new-onset of anorexia and weight loss.

Complementary study to exclude neoplasia was performed. PET-scan showed bilateral mediastinal-hilar and bilateral axillary lymph nodes, with probable inflammatory etiology. Thoraco-abdominal-pelvic-CT revealed no additional findings. A mediastinal lymph node biopsy guided by Endobronchial Ultrasound (EBUS) was negative for malignancy and granulomas. Two months later, while on 20mg/d of prednisolone and with no evidence of arthritis, the patient developed daily fever and hematoquezias and was admitted to the infectious diseases ward. A colonoscopy revealed colitis with multiple erosions and ulcerations. Biopsies of the intestinal lesions revealed the presence of intrahistiocytic microorganisms with morphological characteristics compatible with Leishmania amastigotes. Other infections, including tuberculosis, were extensively excluded. The patient was treated with Amphotericin B (60mg/Kg) and corticosteroids were tapered until suspension. The resolution of colitis was confirmed by colonoscopy and there was no recurrence of arthritis. However, during hospitalization, the patient developed multiple nosocomial infections leading to respiratory failure and death.

Conclusion: Although rare, Leishmaniasis has been associated with arthritis2,3. In our case, the articular symptoms can be interpreted as the first manifestation of Leishmaniasis. A synovial biopsy could have been helpful to confirm the diagnosis histologically. This case highlights the importance of the differential diagnosis of a seronegative polyarthritis, particularly when it is refractory to standard treatment. Moreover, it emphasizes that immunosuppression is a risk factor for potential life-threatening infections, particularly in hospitalized patients.

REFERENCES

1. Marinoni B. et al, Endoscopy, 2020
2. Guidelli, G.M. et al, Synovial Leishmaniasis, Arthritis & Rheumatology, 2016
3. Perelló-Roso A. et al, Clin Infect Dis., 1996

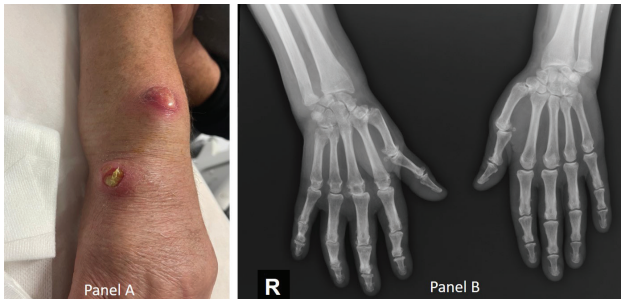
106 - POLYARTHRITIS MIMICS - LESSONS FROM A CASE REPORT

Catarina Rua[1], Tiago Beirão[1], Catarina Silva[1], Tiago Meirinhos[1], Patrícia Pinto[1], Taciana Videira[1], Romana Vieira[1], Joana Abelha-Aleixo[1], Flávio Campos Costa[1], Ana Sofia Pinto[1], Beatriz Samões[1], Diogo Guimarães da Fonseca[1]

[1]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Background: Polyarthritis in the elderly includes a wide range of differential diagnoses, including rheu-

matic diseases, infections, and malignancy.
Case description: A 72-year-old woman with a 2-year history of inflammatory persistent asymmetrical additive hand polyarthralgias with good clinical response following a brief 5mg corticosteroid trial. She denied inflammatory back pain, skin lesions, and a family history of psoriasis. There was no history of uveitis or gastrointestinal symptoms. She reported occasional xerophthalmia and xerostomia but denied constitutional symptoms. Her past medical history included hypothyroidism, dyslipidemia, and left deep venous thrombosis. On physical examination, she exhibited polyarthritis of hand and wrist joints sparing the distal interphalangeal. Blood tests revealed iron-deficiency anemia, reactive C protein of 0.55 mg/dL and erythrocyte sedimentation rate of 60 mm/Hr, negative rheumatoid and anti-citrullinated antibodies, antinuclear antibodies titer of 1/320, with a positive anti-SSA antibody. Serology for infectious diseases, including screening for latent tuberculosis were negative. X-rays of hands showed bilaterally reduced radiocarpal joint space. Salivary gland biopsy and Schirmer test were normal. The right hand X-ray showed marked joint space narrowing, especially with ankylosis of the 2nd and 3rd MCP joints (see figure 1 - panel B). Musculoskeletal ultrasound (MUS) revealed marked tenosynovitis of the 5th and 6th extensor compartments on the right hand, with microcalcifications, and marked tenosynovitis of the flexors of the 4th, 5th, and 1st fingers. Right hand MRI revealed extensive inflammation in the wrist and carpal joints, with significant bone edema involving the distal epiphysis of the radius, ulna, all midcarpal bones, and the bases of the 2nd to 4th metacarpals and confirmed tenosynovitis findings previously found on MUS. Seven months after the initial appointment, she developed two painful cutaneous lesions on the right forearm (figure 1 - see panel A). She started prednisolone 20 mg/daily with a slight improvement in inflammatory markers. The patient was placed on hydroxychloroquine 400 mg daily and the prednisolone dosage was reduced to 10 mg daily. A cervico-thoracic abdomino-pelvic CT scan was performed to exclude malignancy, which identified several enlarged lymph nodes in the right axilla, the largest measuring approximately 15 mm in short axis. A biopsy of the lymph node revealed a positive culture for Mycobacterium tuberculosis. Consequently, anti-tubercular therapy resulted in clinical improvement, normalization of inflammatory markers, and healing of the cutaneous lesions.
Discussion/Conclusions: This case illustrates the importance of considering infectious etiologies, including tuberculosis, in elderly patients presenting with polyarthritis, particularly in the absence of classical autoimmune/rheumatic disease markers. Timely diagnosis and appropriate management of tuberculosis can prevent complications and improve patient outcomes in such cases.
Conclusion. Eye involvement in rheumatic diseases is common and may have various presentations. PUK is an urgent situation and a quick referral for corneal consultation is necessary. Having excluded other etiologies and after double laboratory confirmation of positivity for anti-centromere antibodies, this was the only analytical and immunological change present in this case. Despite not having signs of systemic rheumatic disease, systemic immunosuppressants are sometimes necessary, like in this case. The patient will maintain follow-up and surveillance in a Rheumatology appointment. This case is especially interesting as it highlights the rare association between PUK and anti-centromere antibodies positivity. In the future, the patient's temporal and clinical evolution may help us draw new conclusions.



106 – Figure 1. Right forearm and wrist Musculoskeletal tuberculosis (Panel A - Skin lesions / Panel B - Bilateral Hand X-ray)

107 - "ORANGE PEEL APPEARANCE" AND "GROOVE SIGN": CLUES IN A CASE OF EOSINOPHILIC FASCIITIS

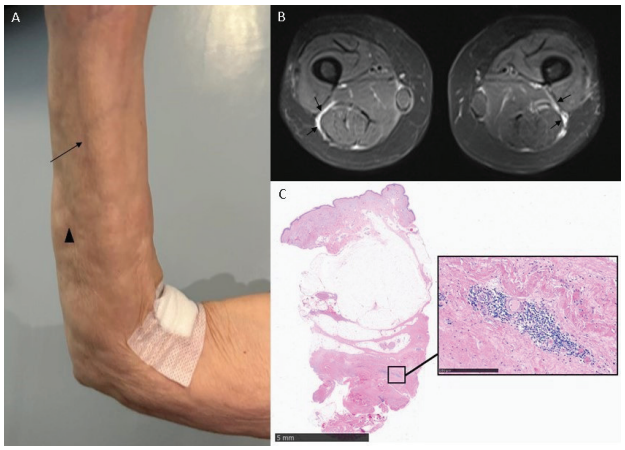
Inês Almeida[1, 2], Mariana Rocha Sebastião[2, 3], Bárbara Fernandes Esteves[2], Carlos Marques-Gomes[2, 4], Mariana Diz-Lopes [2, 4], Miguel Correia Natal[2], Sofia Pedrosa[5], Sofia Pimenta[2, 4], Eva Mariz[2, 4], Lúcia Costa[2]
[1]Rheumatology Department, Unidade Local de Saúde de Viseu Dão-Lafões, Viseu, Portugal, [2]Rheumatology Department, Unidade Local de Saúde de São João, Porto, Portugal, [3]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal, [4]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal, [5]Pathology Department, Unidade Local de Saúde de São João, Porto, Portugal

Introduction: Eosinophilic fasciitis (EF) is a rare connective tissue disease characterized by full-circumference swelling, erythema and progressive thickening of the skin, mainly affecting the distal limbs. A puckering

of the skin (resembling "orange peel" texture) and a linear depression along the course of veins (known as the "groove sign") may occur. The diagnosis relies on clinical findings, peripheral blood eosinophilia and a full-thickness biopsy showing inflammatory cell infiltration, particularly in the fascia.

Case report: A 72-year-old woman was admitted to the rheumatology department due to erythematous skin induration on all 4 extremities with 22months of evolution. She complained of a feeling of tension in her elbows, wrists, fingers and ankles, along with limited mobility and an unintended loss of 7kg over a year. She denied other systemic complaints, Raynaud's Phenomenon, oral or genital ulcers, respiratory, gastrointestinal or genitourinary manifestations. No trauma, increased physical activity, previous infection or exposure to rural environments were reported. Her medical history was positive for dyslipidaemia and arterial hypertension, for which she was on antihypertensive and lipid-lowering medications. Physical examination revealed symmetrical thickened skin on the forearms, dorsum of the hands, lower legs and dorsum of the feet, sparing the fingers, face and trunk. Joint contractures were noted in the elbows and proximal interphalangeal joints, with limited joint range in the wrists and ankles. The characteristic "orange peel" appearance and the "groove sign" were present on the forearms (figure 1A). There were no other mucocutaneous changes or peripheral arthritis. Laboratory investigations at symptom onset showed peripheral eosinophilia (840 cells/µL), elevated C-reactive protein (CRP) 8.6 mg/L and erythrocyte sedimentation rate (ESR) 49 mm/h. Subsequent tests during hospitalization revealed polyclonal hypergammaglobulinemia (20.6%, N 10.5-19.5). Hepatic parameters, renal function and muscle enzymes were within normal ranges. Antinuclear antibodies, antineutrophil cytoplasmic antibodies and serological tests for infectious agents were negative. The magnetic resonance imaging (MRI) documented thickening and increased uptake of the contrast agent in the intermuscular fascial planes involving the posterior compartments of the thigh, all compartments of the legs and arms, with no muscle involvement (figure 1B). Extensive workup for occult malignancies, including CT scans of the thorax, abdomen and pelvis, gastrointestinal endoscopies, mammography and breast ultrasound were unrevealing. A full-thickness biopsy showed epidermal atrophy, thickening of the superficial fascia and perivascular lymphoplasmacytic infiltrate, compatible with EF (figure 1C). Prednisolone 0.75mg/kg/day was started, associated with calcium carbonate, calcifediol and oral bisphosphonate for osteoporosis prevention. She was referred for nutritional counselling and physiotherapy.

Conclusions: EF is a rare and poorly understood disease, requiring a high index of clinical suspicion to establish the diagnosis. Ideally, glucocorticoids and physiotherapy should be initiated early to prevent skin thickening and preserve joint mobility. In this case, despite the long disease duration, there are signs of active inflammation in the complementary diagnostic tests performed, so immunosuppressive therapy may still provide clinical benefit.



107 – Figure 1. Findings on physical examination (A), axial STIR MRI sequences of both thigh (B) and histopathological examination (C).

110 - UMA TEMPESTADE LÚPICA

Carlos Marques-Gomes[1, 2], Mariana Diz-Lopes [1, 2], Inês Santos[3], Tiago Beirão[4], Miguel Bernardes[1, 2], Lúcia Costa[1]
[1]Rheumatology Department, Centro Hospitalar de São João, Porto, Portugal, [2]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal, [3]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [4]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Introdução: Embora o atingimento cutâneo e articular sejam o hallmark do lúpus eritematoso sistémico (LES), os envolvimentos renal e, nomeadamente, neuropsiquiátrico afetam, sobremaneira, o prognóstico e a qualidade de vida dos doentes, particularmente, quando surgem de forma quase concomitante.

Caso Clínico: Os autores apresentam uma doente de 42 anos de idade, com o diagnóstico de LES [anticorpos anti-nucleares (ANA), anti-cadeia dupla do DNA (anti-dsDNA) e anti-cardiolipina IgG positivos] com envolvimento cutâneo (rash malar; lúpus cutâneo subagudo; fotossensibilidade), articular (poliartrite) e hematológico (linfopenia e neutropenia), aos 30 anos (2011). Cumpriu tratamento com hidroxicloroquina (HCQ 400mg/dia) até 2019 (suspensa por toxicidade oftalmológica) e prednisolona (PDN 10mg/dia em es-

quema de desmame), encontrando-se sob ácido acetilsalicílico (100mg/dia) e metotrexato (20mg/semana). Dos restantes antecedentes pessoais, incluía-se perturbação depressiva. A doente manteve-se estável até Fevereiro/2023, altura em que apresentou aumento da velocidade de sedimentação (VS, 89 mm/1ªh), proteína C reativa (PCR, 91 mg/L), anti-dsDNA (258,1 UI/mL), leucoeritrocitúria e proteinúria (0,29g/24h) de novo. Realizou biópsia renal que foi compatível com nefrite lúpica classe III, tendo aumentado a PDN para 0,5 mg/kg/dia e iniciado micofonelato de mofetil (até 2g/dia), em Julho de 2023. Na sequência de intercorrências infeciosas e má compliance terapêutica (também motivada por intolerância gastrointestinal), a doente foi readmitida 3 vezes a internamento. No último (Dezembro/2023), e com 1 mês de evolução, apresentava-se com delírio persecutório, humor profundamente deprimido e progressivamente menos colaborante, com recusa de cuidados médicos. Mantinha proteinúria (1,095g/24h), leucoeritrocituria, elevação de VS (80 mm/1ªh), PCR (39 mg/L) e anti-dsDNA (205.7 UI) e consumo de C3 (45 UI). Não apresentava alterações iónicas nem da função renal e o estudo microbiológico do sangue e urina foi negativo. Após avaliação por Psiquiatria e Neurologia, foi considerado tratar-se de episódio psicótico no contexto de doença inflamatória. O estudo do líquor revelou proteinorráquia (0.8 g/L), índice IgG normal e glicose 42 mg/dL. A ressonância magnética cerebral não demonstrou alterações significativas. Assim, foi admitido o diagnóstico de psicose lúpica e iniciado tratamento com metilprednisolona (500 mg/dia durante 3 dias). Dois dias após a doente já se encontrava consciente, orientada e totalmente colaborante, iniciando o protocolo de ciclofosfamida segundo o esquema NIH, pela nefrite lúpica. Após 6 ciclos, manteve-se com discurso adequado e coerente, sem proteinuria/leucoeritrocituria e sem outros sinais clínicos de atividade de doença.

Discussão: Após anos de estabilidade clínica, esta doente apresentou um agravamento rápido e progressivo da doença, que se traduziu no diagnóstico de nefrite lúpica. A não adesão ao tratamento aliada à persistência de sinais imunológicos de atividade culminaram no desenvolvimento de um quadro de neurolupus. Embora a análise do líquor possa ser normal no contexto de psicose lúpica, a presença de proteinorráquia e a exclusão de outras causas ajudaram a estabelecer o diagnóstico que foi corroborado pela melhoria clínica franca com a corticoterapia em alta dose. Neste caso clínico, é descrita uma manifestação neuropsiquiátrica, cada vez menos reportada (prevalência 1,5%), do LES, sublinhando-se a sua heterogeneidade clínica, o seu potencial de severidade e a importância da adesão ao tratamento instituído.

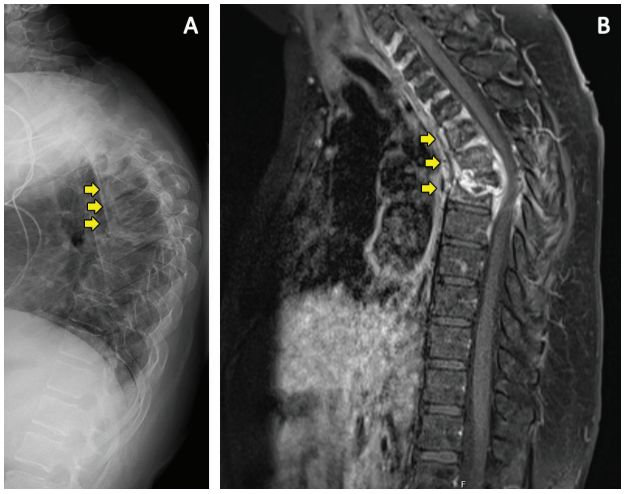
111 - FROM FLAMES TO INFECTION: A CASE OF POST-BURN INFECTIOUS SPONDYLODISCITIS

Inês Santos[1], Tiago Beirão[2], Carlos Marques-Gomes[3, 4], Mariana Diz-Lopes [3, 4], Eva Mariz[3, 4], Ana Martins[3, 4], Lúcia Costa[3]

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

Introduction: Spondylodiscitis is a rare but severe infectious disease that affects the intervertebral disc and adjacent vertebrae, which may also involve peri-vertebral structures and lead to extensive destruction. Its nonspecific nature of symptoms is associated with a frequent delay in diagnosis. The mainstay of treatment is prolonged antibiotic therapy, but surgical intervention may be needed to eliminate the infection focus and restore spinal stability.

Case report: We report the case of a 49-year-old female patient who presented to the emergency department due to aggravation of upper back pain with several days' duration. The pain was constant and radiated to the anterior thoracic and epigastric regions with pleuritic characteristics. It had begun 7 months prior, about 1 month after being discharged from a prolonged hospitalization in a Burn Unit. In the meantime, she visited several emergency departments due to frequent pain exacerbations, despite being on tapentadol 150 mg and acemetacin 90 mg. Her previous complementary study in primary care revealed mildly elevated inflammatory markers and positive HLA-B27, which together with history of niece and nephew with axial spondylarthritis motivated referral to Rheumatology consultation. Her past medical history also included hysterectomy with adnexectomy at the age of 12 due to ovarian dysgerminoma, severe aortic stenosis, complete atrioventricular block requiring a pacemaker, diabetes mellitus, dyslipidemia and overweight. Physical examination revealed short stature, marked adipose deposition on the back with dorsal hyperkyphosis, and an extensive burn scar in the left dorsal region without inflammatory signs. Dorsal mobility was severely limited due to intense pain. Laboratory studies revealed negative troponins and D-dimers, an erythrocyte sedimentation rate of 96 mm/h, C-reactive protein of 166.6 mg/L and procalcitonin of 0.13 ng/mL. Chest X-ray did not present any pleuroparenchymal pulmonary changes. Dorsal spine X-ray showed fracture with anterior wedging of D5 to D7 vertebral bodies (Fig. 1A), later characterized by contrasted MRI that was suggestive of



111 – Figure 1. Sagittal dorsal spine X-ray (A) showing fracture with anterior wedging of D5 to D7 vertebral bodies. Sagittal T1 (...)

spondylodiscitis with extensive bone destruction and associated epidural empyema (Fig. 1B). The patient underwent transpedicular posterior fixation D3-D4-D5 and D7-D8-D9, and material was collected for microbiological study. She completed 11 days of vancomycin and cefepime, later adjusted to 24 days of ceftazidime after isolation of Pseudomonas aeruginosa in the material collected intraoperatively. Blood cultures were negative. After clinical and analytical improvement, the patient was discharged with optimized analgesia and oral levofloxacin 750 mg daily which she took for 83 days, totalizing over 16 weeks of antibiotic therapy. From the outpatient study, bone densitometry revealed a lumbar spine T-score of -3.1 and subcutaneous denosumab 60 mg every six months along with calcium carbonate and vitamin D supplementation were initiated. Sacroiliac joint X-ray was normal. Due to Turner-like traits, a karyotype was performed with normal result and exome sequencing is in progress.

Discussion: The association of chronic back pain with elevated inflammatory markers, HLA-B27 positivity and family history of axial spondylarthritis was suggestive of the latter diagnosis. However, the presence of spine fractures prompted further investigation, culminating in the diagnosis of spondylodiscitis.

Since back pain is a very common symptom, a high degree of clinical suspicion for spondylodiscitis is required due to its rarity and severity.

117 - THERAPEUTIC COMPLEXITY AND EVOLUTION IN AUTOIMMUNE DISEASES: A CASE STUDY OF ANKYLOSING SPONDYLITIS, MULTIPLE SCLEROSIS, AND CROHN'S DISEASE

Miguel Correia Natal[1], Bárbara Fernandes Esteves[1], Lúcia Costa[1], Georgina Terroso[1]
[1]Serviço de Reumatologia, Unidade Local de Saúde de São João, Porto, Portugal

Introduction: The treatment of inflammatory rheumatic diseases has significantly evolved in recent years. Today, numerous therapeutic options are available, particularly since the advent of biologic agents and, more recently, Janus kinase inhibitors. However, these treatments carry risks, and their selection must consider not only their efficacy in managing the rheumatic condition but also the patient's comorbidities.

Case presentation: A 38-year-old male patient was diagnosed with ankylosing spondylitis (AS) at the age of 18 and has been under regular rheumatology follow-up since. He commenced adalimumab (40 mg biweekly) in 2015 for axial disease control and recurrent anterior uveitis.

In October 2018, he presented with right-sided retro-orbital pain and ipsilateral reduced vision. The neurological examination revealed a right eye relative afferent pupillary defect and severe visual loss. Initial laboratory investigations, including comprehensive blood tests, immune studies and viral serologies, were unremarkable. Magnetic resonance imaging (MRI) showed findings consistent with optic neuritis and demyelinating changes ("Dawson's fingers"). Optic neuritis secondary to anti-TNF therapy was assumed, leading to the discontinuation of adalimumab and treatment with intravenous methylprednisolone pulses, resulting in symptom resolution. A 2019 follow-up MRI revealed a new demyelinating lesion, fulfilling criteria for relapsing-remitting multiple sclerosis. The patient was started on dimethyl fumarate (240 mg/day), which was later switched to teriflunomide, with disease stabilization.

Despite ongoing treatment, the patient experienced persistent inflammatory low back pain and recurrent uveitis. Secukinumab (150 mg monthly, later increased to 300 mg) was initiated in August 2019, but did not yield significant improvement. Consequently, a switch to ixekizumab (80 mg monthly) was made in May 2021, which effectively controlled joint symptoms.

However, by late 2021, the patient developed new-onset abdominal pain, diarrhea, and 30% weight loss. Hospitalization for etiological work-up in Gastroenterology led to a diagnosis of penetrating ileal Crohn's disease (CD). Ustekinumab (90 mg every 8 weeks) was commenced, which resolved gastrointestinal complaints but led to a flare in AS. Treatment was then switched to guselkumab (100 mg every 8 weeks, later adjusted to every 4 weeks) in October 2022.

In August 2023, once again the patient experienced diarrhea and weight loss. In collaboration with Gas-

troenterology, upadacitinib (15 mg daily) was initiated, achieving notable improvement in joint symptoms and normalization of weight. Despite this, the patient continued to have 2-3 liquid bowel movements per day, and follow-up colonoscopy in March 2023 indicated active CD. Therefore, the dose was escalated to 45 mg daily (induction dose for CD) in June 2024, which he maintains. Currently, the patient is stable from both rheumatologic and gastroenterologic perspectives.

Discussion: The patient's treatment journey highlights the complexity of managing multiple autoimmune conditions with overlapping and potentially conflicting therapies. The therapeutic approach required careful balancing to control the disease activity of AS, anti-TNF-induced multiple sclerosis (a rare, but previously described entity), and CD. Hence the importance of tailored therapeutic strategies and the integration of care across specialties to achieve optimal outcomes in complex clinical scenarios, with close monitoring for potential treatment-related complications.

121 - IMMUNITY'S PRICE: UNCOVERING PEMBROLIZUMAB-INDUCED PSORIATIC ARTHRITIS

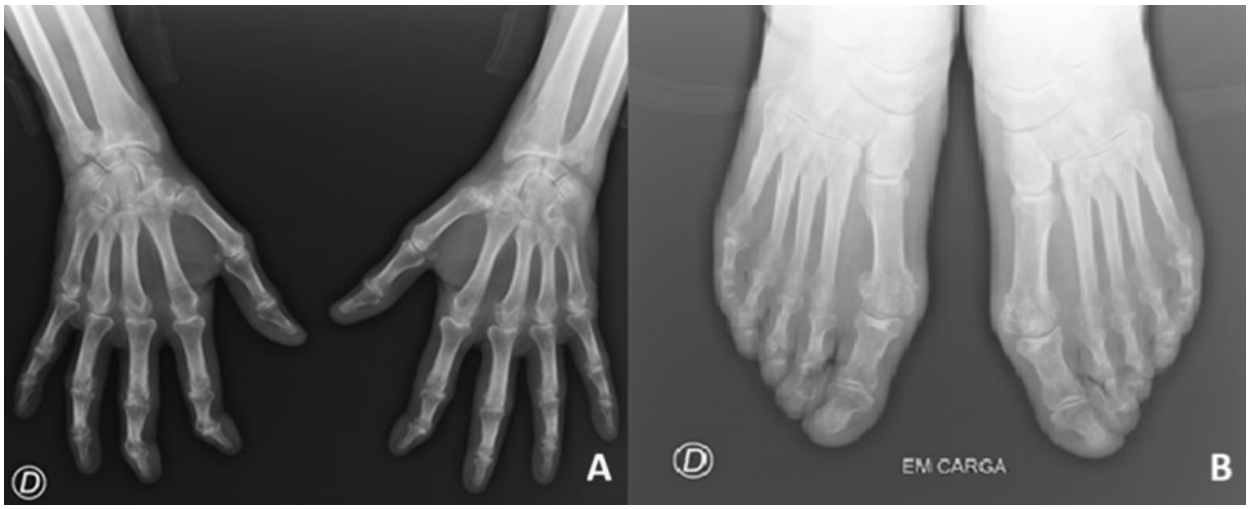
Tiago Beirão[1], Catarina Rua[1], Catarina Silva[1], Tiago Meirinhos[1], Taciana Videira[1], Diogo Guimarães da Fonseca[1]
[1]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Introduction: Pembrolizumab, a programmed death receptor-1 (PD-1) inhibitor, has revolutionized the treatment of various malignancies, including non-small cell lung cancer (NSCLC). Its mechanism of action enhances the body's immune response against tumor cells, but can also lead to immune-related adverse events (irAEs). Among these, articular manifestations are common, but pembrolizumab-induced psoriatic arthritis (PsA) is a rare condition, particularly in older patients. This case study documents the occurrence of PsA in an 87-year-old patient undergoing pembrolizumab therapy for NSCLC.

Clinical Case: An 87-year-old male with a history of NSCLC presented with new-onset joint pain and skin lesions after receiving three cycles of pembrolizumab 6/6weeks. The patient reported symmetrical inflammatory polyarthralgia involving the hands, knees, and ankles. Physical examination confirmed the presence of onycholysis and subungeal hyperkeratosis, with symmetric polyarthritis of the elbows, wrists, metacarpal joints, proximal interphalangeal joints, knees and ankles. Laboratory investigations showed elevated inflammatory markers, and imaging revealed joint erosions consistent with PsA AR-like.

Given the temporal association with pembrolizumab administration and the clinical features, a diagnosis of pembrolizumab-induced PsA was made. The patient was started on systemic corticosteroids (prednisolone 10mg/day), resulting in significant improvement in joint symptoms. Patient refused further treatment and lost follow-up.

Conclusion: This case underscores the importance of recognizing pembrolizumab-induced PsA, particularly in the elderly. Early identification and prompt treatment of irAEs are crucial to balancing effective cancer therapy and immunosuppression. A multidisciplinary approach between oncology and rheumatology is es-



121 – Figure 1. A: erosion of bilateral styloid process, reduction of the radiocarpaljoint space, bilateral carpal erosion, densification of soft tissues at the bilateral carpal level, reduction of the joint space in MCP, PIP, and DIP Joints and erosion and bone formation in PIP and DIP Joints; B: reduction of joint space and osteophyte formation in talonavicular, calcaneocuboid, and tibiotalar Joints

sential to ensure adequate outcomes for both cancer control and irAEs management.

122 - THE HIDDEN ITCH: URTICARIA AND UVEITIS ON THE PATH TO MOTHERHOOD

Tiago Beirão[1], Liliana Dias[2], Inês Cunha[2], Rita Amorim[3], Catarina Rua[1], Catarina Silva[1], Beatriz Samões[1], Taciana Videira[1], Tiago Meirinhos[1]
[1]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [2]immunoallergology department, Unidade Local de Saude Gaia e Espinho, Vila Nova de Gaia, Portugal, [3]Anatomical Pathology, Unidade Local de Saude Gaia e Espinho, Vila Nova de Gaia, Portugal

Introduction: Hypocomplementemic urticarial vasculitis (HUV) is a rare autoimmune condition characterized by chronic urticarial eruptions, systemic inflammation, and low serum complement levels, particularly C1q. HUV can also involve various organs, including the eyes, where it may present as uveitis. This case report discusses the diagnostic and therapeutic challenges in managing HUV with uveitis in a 39-year-old woman who is planning a pregnancy.
Clinical Case: A 39-year-old female presented with a two-year history of recurrent urticarial lesions that were resistant to conventional antihistamine treatment (Figure 1A). She also reported episodes of ocular discomfort, including redness, photophobia, and decreased vision. After ophthalmology evaluation, a diagnosis of uveitis was made. Comprehensive laboratory investigations revealed hypocomplementemia, with significantly reduced levels of C1q, and a skin biopsy confirmed leukocytoclastic vasculitis (Figure 1B). The clinical findings led to a diagnosis of hypocomplementemic urticarial vasculitis with secondary uveitis. Given her desire to conceive, after a multidisclinary approach with immunoallergology, rheumatology, ophthalmology and obstetrics, a combination of corticosteroids and azathioprine, in order to control systemic and ocular inflammation, with minimal teratogenic risk.
Conclusion: This case highlights the complexities of diagnosing and managing hypocomplementemic urticarial vasculitis with uveitis in a patient planning pregnancy. A multidisciplinary approach is crucial to address the systemic and ocular manifestations while considering the reproductive implications. Effective treatment requires a balance between disease control and minimizing potential risks to both mother and child.

135 - POLIARTRITE SIMÉTRICA- UM CASO RARO DE RETICULO HISTIOCITOSE MULTICÊNTRICA

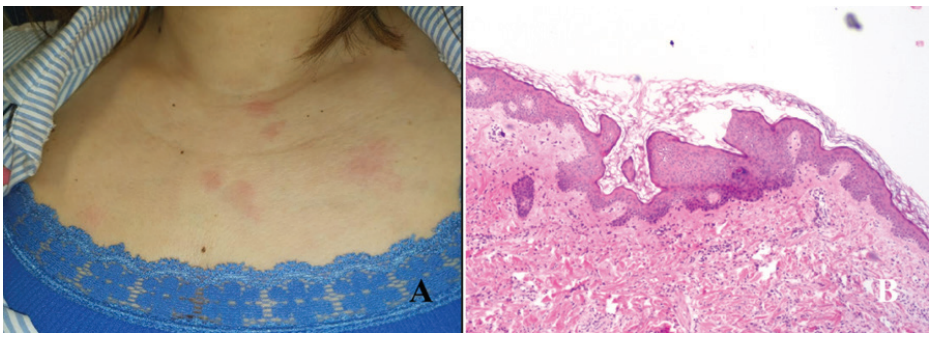
Susana Matias[1], Catarina Abreu[1], Vanessa Fraga[1], Ana Cordeiro[1], Maria José Santos[1]
[1]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal

Introdução: A Reticulo Histiocitose Multicêntrica (RHM) é uma doença sistémica rara, inflamatória, de causa desconhecida, atualmente sem orientações específicas de tratamento. Ocorre preferencialmente em mulheres entre os 40 e 50 anos e carateriza-se por uma ser uma doença granulomatosa que tipicamente envolve a pele e mucosas –lesões pápulo-nodulares que contêm proliferação de histiócitos (macrófagos)- e o sistema músculo-esquelético- poliartrite. No entanto, pode afetar praticamente qualquer órgão. Em cerca de 1/4 dos casos, pode-se associar a neoplasia.

Na RHM o envolvimento articular tende a ser simétrico, poliarticular e erosivo com afeção das pequenas e grandes articulações. Assim sendo, em alguns casos com artrite exuberante e lesões cutâneas pouco específicas, pode ser erroneamente diagnosticada como artrite reumatoide (AR).

O diagnóstico é sempre baseado na biópsia dos nódulos onde se observam células gigantes multinucleadas (CD68+) com citoplasma eosinofílico e aspeto em vidro despolido.

Em seguida, apresenta-se um caso de reticulo histiocitose multicêntrica.

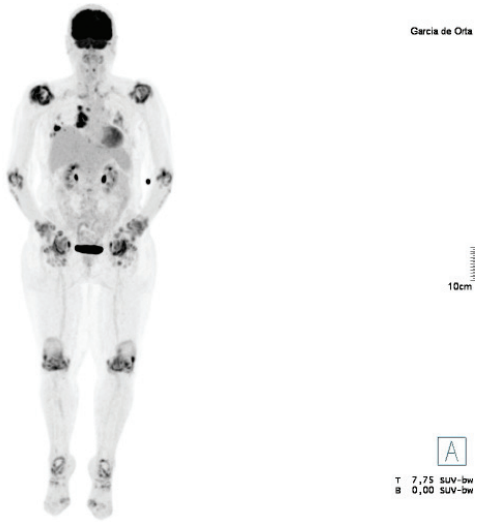


122 – Figure 1.

Descrição do caso: Doente do sexo feminino, 78 anos, com antecedentes pessoais de tuberculose na infância e dislipidemia. Apresentava quadro com um ano de evolução de poliartralgias dos joelhos, tibio-társicas, ombros e punhos associados a eritema da face e região do decote e a lesões pápulo-nodulares nas articulações interfalângicas, cotovelos e pavilhões auriculares. Adicionalmente, anorexia e perda ponderal de 8kg em 2 meses. Ao exame objetivo apresentava poliartrite simétrica (joelhos, punhos e ombros), sem deformações. Da investigação complementar salienta-se normalidade dos parâmetros inflamatórios, fator reumatoide, anticorpos anti-péptidos citrulinados e anticorpos anti-nucleares (ANA) negativos. Biópsia cutânea dos nódulos das mãos com numerosas células gigantes multinucleadas com citoplasma intensamente eosinofílico e aspeto em vidro despolido, confirmando o diagnóstico de RHM. Realizou tomografia computorizada torácica que mostrou múltiplas nodularidades pulmonares suspeitas e PET scan com suspeita de malignidade pulmonar direita e ganglionar mediastino-hilar direita e evidência de patologia inflamatória ativa em múltiplas articulações. A citologia aspirativa de vários nódulos paratraqueais guiada por ultrassonografia endobrônquica foi negativa para tecido neoplásico, evidenciando fragmentos de tecido ganglionar linfático permeado por numerosos macrófagos (CD68+) mono e multinucleados, com abundante citoplasma eosinófilo, por vezes em vidro despolido. Assumida RHM com envolvimento articular, mucocutâneo e pulmonar.

Em termos de tratamento, sem resposta articular a corticoterapia intra-articular ou sistémica, pelo que iniciou metotrexato.

Conclusão: Este caso clínico destaca-se pela raridade da doença apresentada e alerta-nos para a necessidade de diagnóstico diferencial com AR dado a frequente apresentação com poliartrite simétrica. Levanta também questões sobre que terapêutica instituir.



135 – Figure 1. PET scan FDG com evidência de patologia inflamatória activa em várias articulações

137 - HIPERTENSÃO ARTERIAL PULMONAR, UMA COMPLICAÇÃO INESPERADA

Susana Matias[1], Catarina Abreu[1], Maria Margarida Cunha[1], Alice Castro[1], Maria José Santos[1]
[1]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal

Introdução: A hipertensão pulmonar (HTP) é uma síndrome rara, complexa, mas que se associa a elevada morbilidade e mortalidade. A hipertensão arterial pulmonar (HAP) – grupo 1 da HP- é uma complicação vascular bem conhecida dos doentes com esclerose sistémica, no entanto, pode estar presente noutras doenças reumáticas sistémicas (DRS), como na doença mista do tecido conjuntivo e no lúpus eritematoso sistémico. Os sintomas típicos da HAP, como cansaço, intolerância ao exercício, tosse seca, dispneia, podem estar associados às DRS e às suas comorbilidades, sendo muitas vezes desvalorizados pelos doentes, levando ao atraso no diagnóstico de HAP e consequente tratamento.

Apresenta-se um caso clínico de hipertensão arterial pulmonar associada a síndrome de sobreposição Sjögren/lúpus eritematoso sistémico.

Descrição do caso: Doente do sexo feminino, de 32 anos de idade, com diagnóstico de síndrome de sobreposição Sjögren/lúpus eritematoso sistémico desde 2015, com envolvimento predominantemente articular e vascular - fenómeno de Raynaud.

Apresentava quadro com um mês de evolução de cansaço, dispneia e toracalgia anterior para grandes esforços, sem irradiação e sem outros sintomas acompanhantes. Ao exame objetivo havia apenas a destacar uma taquicardia sinusal em repouso. Do estudo efetuado, em termos analíticos, a frisar um aumento dos parâmetros inflamatórios (velocidade de sedimentação de 120mm na 1ªhora e proteína C reativa de 5.46mg/dL, N<0.20) e do NT-proBNP(2917 pg/mL, N<125); Na radiografia do tórax com cardiomegalia; Na cintigrafia pulmonar de ventilação/perfusão sem evidência de tromboembolismo pulmonar; No ecocardiograma documentava-se uma dilatação das cavidades direitas e uma pressão sistólica na artéria pulmonar de 81mmHg. Foi submetida a cateterismo cardíaco direito com evidência de hipertensão pulmonar pré-capilar (Pressão arterial pulmonar média de 49mmHg, Resistência vascular periféria de 8.26 UW, Pressão capilar pulmonar de 11mmHg), sem critérios de gravidade. Iniciou ter-

apêutica com tadalafil 20mg/dia e ambrisentano 5mg/dia, com boa tolerância.

Conclusão: Este caso clínico destaca-se pela raridade do aparecimento de HAP em doentes com uma síndrome de sobreposição e alerta-nos para a necessidade de valorizar queixas inespecíficas como o cansaço. Levanta também questões sobre a necessidade de fazer rastreio de HP (mesmo na ausência de sintomas sugestivos) nestes doentes, a fim de a diagnosticar e tratar atempadamente.

144 - MIOPATIA INFLAMATÓRIA COM POSITIVIDADE PARA ANTICN1A, EM DOENTE COM DOENÇA DE SJÖGREN

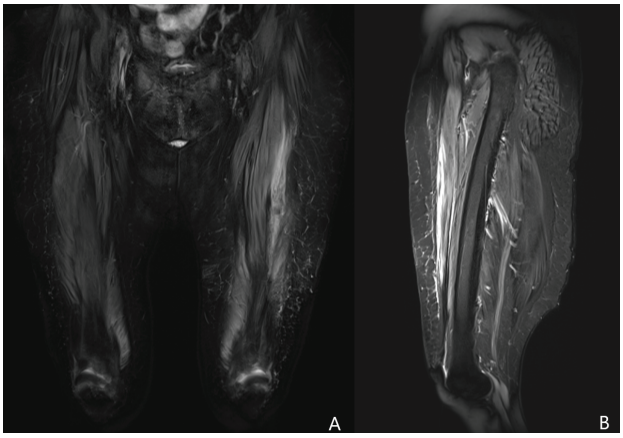
Nuno Delgado[1], Miguel Guerra[1, 2], Rita Pinheiro Torres [1], Ana Filipa Rocha Águeda[1, 2], Joana Ramos Rodrigues[1], Margarida Oliveira[1, 2]

[1]Serviço de Reumatologia, Unidade Local de Saúde da Cova da Beira, Covilhã, Portugal, [2]Faculdade de Ciências da Saúde, Universidade da Beira Interior, Covilhã, Portugal

Introdução: A doença de Sjögren primário (DSp) é um distúrbio sistémico caracterizado por infiltração linfocítica e comprometimento funcional das glândulas exócrinas. No entanto, o processo imuno-mediado pode afetar potencialmente qualquer órgão incluindo os músculos com grande variação na prevalência (0,85–14%) em diferentes series.

Caso Clínico: Doente do sexo feminino, de 70 anos, seguida em consulta de Reumatologia por Doença de Sjögren primária com 10 anos de evolução, medicada com hidroxicloroquina 200mg/dia. Em consulta de rotina, descreveu quadro de mialgias/fraqueza muscular proximal dos membros inferiores, com disfonia e disfagia associadas, com 3 meses de evolução. Ao exame objetivo, apresentava fraqueza muscular proximal dos membros inferiores, dos membros superiores e dos músculos flexores do pescoço, com MMT8 de 115/150. Analiticamente, destacava-se aumento franco da creatina quinase (CK) (1277U/L, N<192) e da mioglobina (873.7ng/mL, N<106), LDH 462U/L, VS 24mm/H, com TSH/T4 livre normais e PCR negativa. Admitiu-se a doente em internamento para estudo adicional e tratamento dirigido, tendo-se suspendido a hidroxicloroquina pelo seu potencial mio-tóxico.

Foi iniciada corticoterapia com 1 pulso endovenoso de 125mg de metilprednisolona, seguido de prednisolona oral 30mg/dia (0.5mg/kg/dia). A ressonância magnética das coxas evidenciou edema muscular centrado ao músculo quadricípite bilateralmente, envolvendo praticamente todos os seus componentes, embora de forma mais severa o recto femoral. Do estudo de realizado, destacava-se positividade para anticorpos antoRo52 e antiCN1A. Realizou-se biópsia muscular



144 – Figure 1. RM das Coxas em corte coronal (A) e sagital esquerdo (B)

no quadricípete esquerdo, que revelou "presença de fibras musculares hipertróficas e atróficas, de morfologia redonda, com significativo infiltrado inflamatório a nível do endomísio, que morfologicamente parece ser de predomínio linfocitário, sugestivo de miopatia inflamatória idiopática, enquadrável em síndrome de sobreposição". Para exclusão de eventual neoplasia oculta associada a miopatia inflamatória, realizou radiografia torácica, ecografia da tiróide, mamária, renal e abdomino-pélvica, sem achados suspeitos.

Foi assim assumido o diagnóstico de miopatia inflamatória associada a Doença de Sjögren. Durante o internamento, verificou-se melhoria clínica e analítica progressivas, com normalização dos valores de CK ao 10º dia e MMT8 de 127/150 à data de alta. Para controlo a longo prazo do quadro, iniciou azatioprina oral 25mg, com boa tolerabilidade.

Discussão e Conclusão: O anticorpo anti-cN1A está tipicamente associado à miopatia de corpos de inclusão, com especificidade de 94-96% e sensibilidade 49-53%. Esta é uma entidade de mau prognóstico, sem resposta à corticoterapia ou outros tratamentos imunossupressores.

No entanto, têm surgido relatos de pacientes com outras condições imuno-mediadas (particularmente doença de Sjögren) com miosite inflamatória com positividade para estes anticorpos e com resposta favorável às terapêuticas instituídas. O caso descrito é um exemplo disso, não se tendo documentado corpos de inclusão na biópsia muscular, e com melhoria franca após introdução de corticoterapia.

152 - OSTEOMALACIA MISDIAGNOSED AS OSTEOPOROSIS - A CASE REPORT

Inês Almeida[1], Liliana Saraiva[1], Couto M[1]

[1]Rheumatology Department, Unidade Local de Saúde de Viseu Dão-Lafões, Viseu, Portugal

Introduction: Osteomalacia is a metabolic bone disease characterized by deficient mineralization of the bone matrix. Crohn's disease (CD) can impair nutrient absorption, either by inflammation of the intestinal mucosa or by resection of segments of the small intestine. This can lead to deficits in phosphate (P), calcium (Ca) and vitamin D (vit D) that predispose to metabolic bone disease.
Case report: A 56-year-old man, diagnosed with CD since the age of 15, with a history of 3 ileocecal resections, high cumulative dose of glucocorticoids and multiple intravenous therapies with ferric carboxymaltose, presented with nonspecific complaints of musculoskeletal pain. Bone scintigraphy revealed multiple foci of dispersed hyperuptake, suggestive of multiple fragility fractures of cervical and dorsal vertebrae, costal arches and lesser trochanter, confirmed by CT. Bone densitometry showed a femoral neck T-score of -2.7 and a lumbar spine T-score of -1.3, and bisphosphonates were started. Due to worsening musculoskeletal pain and muscle weakness, he was referred to our rheumatology department. Clinical laboratory tests revealed P 1.0mg/dL (2.3 – 3.7), Ca 9.2mg/dL (8.7 – 10.4), alkaline phosphatase (ALP) 251IU/L (25 – 100), parathormone (PTH) 193.7pg/mL (18.50 – 88.00), vit D 18.9ng/mL (30.0 – 95.0) and tubular P reabsorption rate 59% (78 – 98%). Ultrasound showed no nodular formations in the parathyroid glands. He started phosphate and calcitriol supplementation and stopped bisphosphonates. After 4 months of therapy, there was clinical and analytical improvement: P 1.4mg/dl; ALP 210IU/L; PTH 204pg/mL; vit D 28.6ng/mL. The supplementation dosage was adjusted.
Discussion: In this case, hypophosphataemia resulted from several factors: intestinal malabsorption; increased renal excretion of P secondary to increased fibroblast growth factor 23 (FGF23) due to frequent therapy with ferric carboxymaltose; vit D deficiency with secondary hyperparathyroidism amplifying renal P excretion.
Conclusion: Fragility fractures in a patient with low bone mineral density are not necessarily synonymous with osteoporosis. A complete study of bone metabolism is essential for an accurate diagnosis and appropriate treatment, especially in the presence of pathologies that compromise the uptake of nutrients.

156 - A RARE CAUSE OF SICCA SYNDROME

Catarina Abreu[1], Vanessa Fraga[1], Susana Matias[1], Ana Catarina Duarte[1], Maria Margarida Cunha[1], Alice Castro[1], Maria José Santos[1, 2]
[1]Rheumatology Department, Unidade Local de Saúde de Almada-Seixal, 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

Sicca syndrome, characterized by dryness of the eyes and mouth, is often a manifestation of Sjögren's syndrome. However, sicca syndrome can also arise from other causes such as medications, radiotherapy and non-lymphocytic gland infiltration due to granulomatous diseases, amyloidosis, malignancies or IgG4 -related disease.

We report a case of a 78-year-old female patient with a medical history of hypercholesterolemia. She presented to the emergency department (ED) with bilateral painful red eyes accompanied by photophobia. She was diagnosed with conjunctivitis and treated with topical antibiotics without improvement. Upon re-evaluation she was diagnosed with keratitis and treated with ocular lubricants. Concomitantly the patient progressively developed anorexia, weight loss (>10% of her baseline weight), xerostomia, rhinorrhea, nasal obstruction and unilateral hearing loss. Due to new-onset odontalgia she returned to the ED and was discharged with the diagnosis of peri-implant infection, for which she started oral antibiotics. Two months after the onset of the symptoms, she developed fever and productive cough. She was observed at the ED and was prescribed amoxicillin-clavulanate and azithromycin for suspected pneumonia. Due to persistent symptoms, she was reevaluated. Blood tests revealed new onset normochromic and normocytic anemia (10.9mg/dL), thrombocytosis (637 000/µL), leukocytosis and elevated C-reactive protein (21mg/dL) and erythrocyte sedimentation rate (120mm in 1st hour). A chest computed tomography (CT) scan showed nodular consolidations, one of which had a central cavitation. She was admitted to the hospital and her antibiotics were switched to piperacillin-tazobactam. Despite treatment, there was no clinical or laboratory improvement. A subsequent chest CT scan showed persistent consolidations with resolution of the cavitated lesion. Microbiological investigations including blood cultures, bronchoalveolar lavage and bronchoscopy cultures were negative, leading to the suspension of antibiotics. Lung biopsy revealed chronic inflammatory infiltrates, without neoplasia or granulomas. Rhinolaryngoscopy examination revealed nasal crusts and a polyp on the nasal fossa and auditory tests confirmed conductive hearing loss, suggesting seromucous otitis secondary to ipsilateral nasal obstruction. A biopsy of the nasal polyp showed inflammatory infiltrates without granulomas. Maxillofacial examination revealed significant signs of dehydration of the oropharyngeal mucosa. Serological tests were positive for anti-neutrophil cytoplasmic an-

tibodies (ANCA) with anti-proteinase-3 antibodies (34 UI/ml).

Due to the suspected diagnosis of granulomatosis with polyangiitis (GPA), the patient was started on prednisolone 0.5mg/kg/day. Significant clinical, laboratorial and imaging improvements were observed, including sustained apyrexia, resolution of sicca symptoms, respiratory and otolaryngological complaints, anti-PR3 antibodies normalization and clearance of chest CT condensations.

In this case, sicca syndrome, particularly xeropthalmia, was the presenting symptom of GPA. The progressive occurrence of nasal and ear involvement alongside systemic symptoms and specific serological findings were pivotal in distinguishing GPA from other mimickers of Sjögren's syndrome. Recognizing glandular involvement as a manifestation of GPA is crucial for its timely diagnosis and management, which can significantly improve patient outcomes.

157 - INDUÇÃO DE HIPERTIROIDISMO DURANTE O TRATAMENTO COM LEFLUNOMIDA – A PROPÓSITO DE DOIS CASOS CLÍNICOS

Marina Oliveira[1], Bernardo Dias Pereira[2], Carolina Furtado[1], Uladzislau Sushko[1], Mariana Rocha Sebastião[1], Tomás Fontes[1], Filipe Oliveira Pinheiro[1], Luís Maurício Santos[1], Teresa Novoa[1]

[1]Serviço de Reumatologia, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal, [2]Serviço de Endocrinologia e Nutrição, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal

Introdução: A leflunomida é um fármaco antirreumático modificador da doença usado no tratamento da artrite reumatoide (AR). Existe a possibilidade deste medicamento alterar a resposta imunológica, predispondo ao desenvolvimento de doenças imunomediadas da tiroide, nomeadamente hipertiroidismo. Doentes com AR apresentam um risco aumentado de patologia tiroideia, com uma prevalência de hipotiroidismo e de hipertiroidismo variando entre 6% e 34%. A relação entre estas duas condições não é clara, porém sabe-se que a etiologia de ambas é multifatorial, envolvendo componentes ambientais e genéticos. A literatura é limitada a um relato de caso, relativamente a efeitos adversos da leflunomida na função tiroideia.

Descrevemos dois utentes com AR com diagnóstico de hipertiroidismo após tratamento com leflunomida.

Caso clínico 1: Homem de 76 anos, referenciado à consulta de Endocrinologia, após deteção incidental de bócio multinodular em eutiroidismo, com desenvolvimento de tireotoxicose a T4, 18 meses após o início da leflunomida. O painel completo de anticorpos anti-tiroideus foi negativo. A cintigrafia tiroideia não mostrou captação na topografia tiroideia. A leflunomida foi suspensa e o utente permaneceu apenas numa estratégia de vigilância clínica e analítica. Após 21 e 26 meses de interrupção da leflunomida, respetivamente, ocorreu normalização da função tiroideia e captação normal da tiroide na cintigrafia tiroideia.

Caso clínico 2: Mulher de 58 anos, com hipotiroidismo autoimune, controlado com levotiroxina 75 mcg/dia, recorreu à consulta de Endocrinologia com fadiga, palpitações, tremores, sudorese, fotofobia e diplopia, 14 meses após o início de leflunomida. À avaliação clínica, apresentava oftalmopatia, e a avaliação analítica revelou hipertiroidismo por doença de Graves. Iniciou metimazol e propranolol e interrompeu leflunomida e levotiroxina. A ecografia cervical revelou glândula tiroide de tamanho normal, hipervascularizada e difusamente hipoecóica, sem nódulos. A utente desenvolveu novo agravamento do hipertiroidismo, após 15 meses de metimazol, bem como oftalmopatia refratária aos glicocorticoides. Reiniciou metimazol e está, atualmente, a aguardar o início de tocilizumab para oftalmopatia de Graves.

Discussão/Conclusão: Dada a elevada frequência de associação entre AR e doença autoimune tiroideia, o clínico deve considerar o rastreio de doença nodular da tiroide e respetiva autoimunidade em utentes com AR, se equacionar o tratamento destes com leflunomida, uma vez que este fármaco pode induzir diversas etiologias de tireotoxicose. Além disso, a leflunomida, contendo iodopovidona, deve ser evitada em utentes com AR com histórico de doença nodular da tiroide ou autoimunidade tiroideia.

160 - PARVOVÍRUS B19 E VASCULITE CRIOGLOBULINÉMICA: UMA ASSOCIAÇÃO IMPROVÁVEL

Sara Alves Costa[1], Fernando Albuquerque[1], Filipa Canhão André[1], Marcelo Neto[1], Maria João Cadório[1], João Alexandre Oliveira[1], Adriana Carones[1], Beatriz Mendes[1], Sara Serra[1]

[1]Rheumatology Department, Unidade Local de Saúde de Coimbra, Coimbra, Portugal

Introdução: A infeção por Parvovírus B19 apresenta um fenótipo clínico heterogéno, manifestando-se, frequentemente, com eritema infecioso e febre em crianças, e artralgia nos adultos. Podem, contudo, ocorrer outras manifestações mais raras, havendo alguns relatos de caso de vasculite crioglobulinémica associada a esta infeção.

Caso clínico: Doente do sexo feminino, 47 anos, recorre ao Serviço de Urgência por quadro de febre, com temperatura máxima de 38.5 ºC, fadiga, mial-

gia dos membros inferiores e superiores, artralgia inflamatória de punhos, joelhos e tornozelos, e lesões cutâneas dos membros inferiores com cerca de uma semana de evolução.

Os seus antecedentes pessoais incluíam rinossinusite alérgica, taquicardia supraventricular e gastrite, estando medicada com bisoprolol 5 mg/ dia e pantoprazol 20 mg/ dia.

Ao exame objetivo, apresentava sinais de artrite de punhos e tornozelos, com dor à palpação e mobilização ativa e passiva, e lesões maculopapulosas nos membros inferiores, com componente purpúrico palpável, sem desaparecimento à digitopressão, distribuídas bilateralmente, enquadráveis em vasculite cutânea (figura 1).

O estudo analítico revelou uma elevação ligeira da velocidade de sedimentação (24mm/h), um sedimento urinário com 14 eritrócitos por campo e dismorfismo eritrocitário, e diminuição de C4 (0.09 g/L), sem alterações de bioquímica, hemograma e proteína C reativa. Serologias de Hepatite B e C e HIV negativas.

A doente foi medicada com prednisolona 20mg/ dia com resolução das suas queixas, tendo sido reavaliada em consulta de Reumatologia após uma semana, não se evidenciando artrite ou lesões cutâneas.

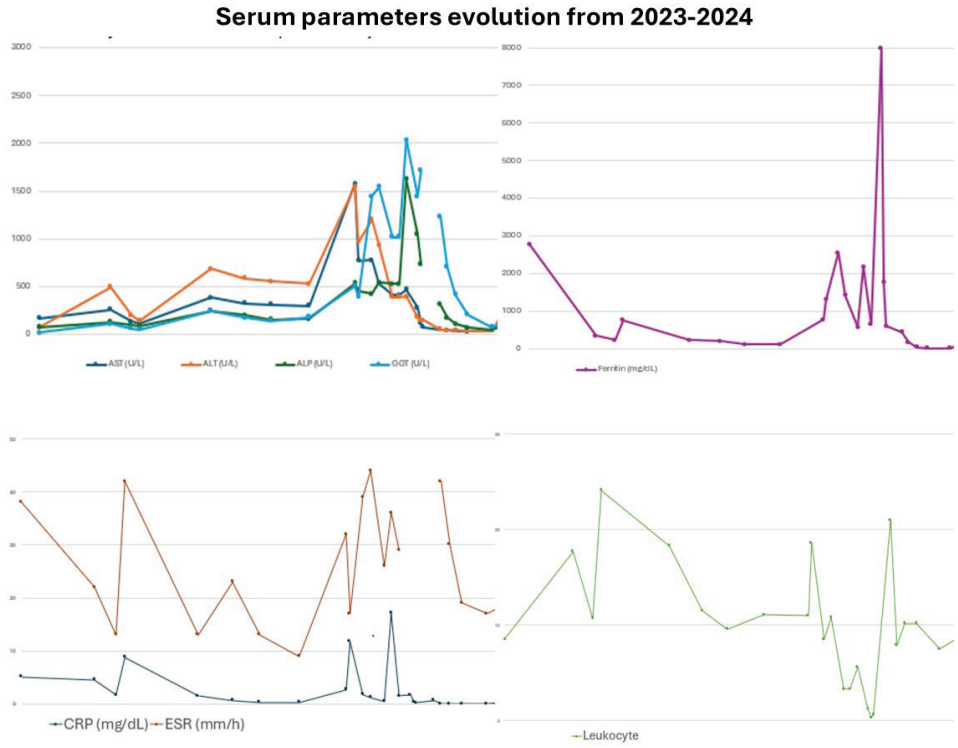
Do estudo analítico complementar, verificou-se normalização de VS, C4 e sedimento urinário. Apresentava, ainda, serologia sugestiva de infeção em evolução por Parvovírus B19 (em duas medições separadas por 3 semanas) e presença de crioglobulinémia, com imunofixação a revelar IgG e IgM policlonais, sugestiva de crioglobulinémia tipo III.

Desta forma, foi assumido como diagnóstico mais provável uma infeção por Parvovírus B19 com vasculite crioglobulinémica tipo lll, estando sob vigilância clínica.

Discussão: A combinação de púrpura, artralgia e fadiga deve levantar a hipótese de vasculite crioglobulinémica, a qual é suportada pela presença de crioglobulinas no soro, estando frequentemente associada a diminuição de C4, presença de Fator Reumatoide e/ ou imunoglobulina M no soro. As causas mais frequentes são infeções, principalmente hepatite C, doenças do tecido conjuntivo e neoplasias hematológicas, pelo que uma avaliação clínica detalhada é fundamental para determinar a sua etiologia.

Na literatura, apenas existem relatos de caso isolados associados à infeção por Parvovírus B19. O achado clínico mais comum da infeção vírica em adultos é a artralgia, também presente neste caso. A coexistência de crioglobulinémia mista é rara e foi apenas descrita para o tipo ll, não existindo registos, até à data, de casos de crioglobulinémia tipo lll, como se apresenta.

Assim, este caso reforça a importância do reconhecimento do Parvovírus B19 como umas das causas de crioglobulinémia mista e a sua pesquisa deve ser incluída no estudo complementar.

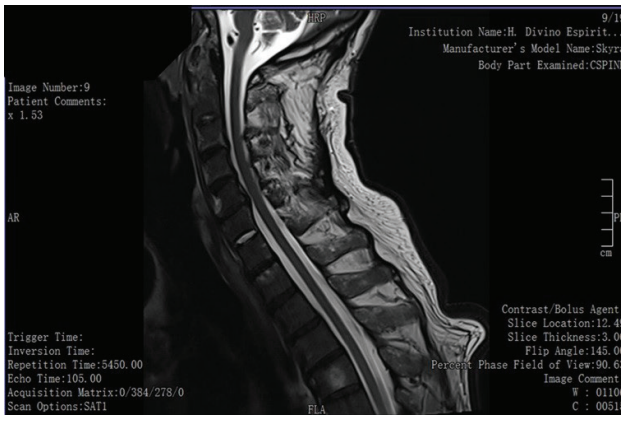


160 – Figure 1. A - Lesões cutâneas sugestivas de vasculite. B- Tumefação da tibiotársica direita.

161 - CERVICALGIA COMO MANIFESTAÇÃO DE ESPONDILODISCITE INFECIOSA NUM DOENTE COM ESPONDILARTRITE – DESAFIO DIAGNÓSTICO

Marina Oliveira[1], Uladzislau Sushko[1], Mariana Rocha Sebastião[1], Tomás Fontes[1], Carolina Furtado[1], Luís Maurício Santos[1], Teresa Novoa[1], Filipe Oliveira Pinheiro[1]
[1]Serviço de Reumatologia, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal

Introdução: A cervicalgia é muito prevalente na prática clínica, sendo a sua etiologia multifatorial. A apresentação clínica da cervicalgia inflamatória e infeciosa é semelhante, tornando o seu diagnóstico diferencial desafiante e essencial, atendendo às implicações terapêuticas associadas.
Caso clínico: Descrevemos o caso de um homem de 69 anos com espondilartrite axial (sacroileíte radiográfica) e periférica, medicado com sulfassalazina 2g/dia desde há sete anos. Recorreu ao Serviço de Urgência por anorexia, astenia, períodos de desorientação e febre, com cerca de dois meses de evolução. Os meios complementares de diagnóstico revelaram uma endocardite da prótese biológica aórtica por Streptococcus gallolyticus, tendo iniciado tratamento com ampicilina dirigida ao isolamento microbiano. Cerca de duas semanas depois, o doente apresentou quadro de cervicalgia constante associada a impotência funcional marcada, com rigidez matinal prolongada e despertares noturnos frequentes, sem alívio significativo ao longo do dia ou posição antálgica. Ao exame físico, objetivou-se limitação das mobilidades ativas e passivas cervicais (< 5 graus de amplitudes articulares), com mobilidades dorsais e lombares mantidas e indolores, não existindo trigger points miofasciais ou alterações ao exame neurológico sumário. Perante os achados de novo, num doente com espondilartrite, com endocardite infeciosa, considerou-se pertinente a exclusão de foco séptico cervical. Nesse sentido, realizou ressonância magnética cervical que mostrou um foco de espondilodiscite contida no disco intervertebral de C6-C7. Atendendo ao contexto clínico do doente, assumiu-se envolvimento infecioso do disco por êmbolo séptico, sem complicações locais, tendo sido prolongada a antibioterapia para um total de oito semanas, que o doente cumpriu com melhoria do quadro clínico.
Discussão: Os utentes com próteses valvulares têm maior risco de endocardite infeciosa, que pode complicar-se com espondilodiscite. A coluna vertebral é um local frequente de focos sépticos secundários, por disseminação hematogénica, devido à sua extensa vascularização. Outros fatores de risco para a espondilodiscite são a doença inflamatória reumática e a imunossupressão. Existem várias causas que podem mimetizar a espondilodiscite, nomeadamente o flare da atividade inflamatória da espondilartrite axial. O diagnóstico diferencial entre ambas pode ser desafiante, tendo em conta o caráter inespecífico e insidioso da cervicalgia de ritmo predominantemente inflamatório.
Conclusão: A espondilodiscite infeciosa deve ser equacionada como diagnóstico diferencial nos doentes com espondilartrite com queixas axiais de novo, sobretudo na presença de fatores de risco para infeção, pela necessidade de tratamento dirigido e célere.



161 – Figure 1. Discite C6-C7

173 - CHALLENGES AND INSIGHTS IN DIAGNOSING AND MANAGING OSTEOPOROSIS IN SYSTEMIC MASTOCYTOSIS: A CASE REPORT AND REVIEW

Miguel Correia Natal[1], Bárbara Fernandes Esteves[1], Lúcia Costa[1], Georgina Terroso[1]
[1]Rheumatology Department, Unidade Local de Saúde de São João, Porto, Portugal

Introduction: Mastocytosis describes a group of rare disorders characterized by the abnormal proliferation of neoplastic mast cells and their infiltration and accumulation in various tissues. This invasion can be confined to the skin (cutaneous mastocytosis) or involve extracutaneous tissues (systemic mastocytosis [SM]), with the bone being the most commonly affected.

The prevalence of osteoporosis (OP) in SM is high. Although the pathogenesis is not fully understood, it is believed to involve a combination of neoplastic infiltration and the release of mediators from mast cells, leading to predominant bone resorption, with significant involvement of the RANK/RANKL pathway.
Case presentation: A 64-year-old woman, followed in an Internal Medicine outpatient clinic for osteoporosis since the age of 37, presented with multiple low-impact fractures, including vertebral fractures, a left intertro-

chanteric femoral fracture (treated with osteosynthesis), and rib fractures, resulting in significant functional limitation. She had been on weekly alendronate therapy for over 20 years.

During this period, osteoporosis was thought to be secondary to primary hyperparathyroidism due to a left lower parathyroid adenoma. She underwent parathyroidectomy, which normalized her parathyroid hormone (PTH) levels. She had no other risk factors for osteoporosis.

However, due to a history of three anaphylactic episodes, two of which occurred during anesthesia induction, she was referred to the Immunoallergology clinic. Initial studies documented a baseline serum tryptase of 380 µg/L (REF < 11.40 µg/L), raising the suspicion of SM. Further investigations revealed normal complete blood count and biochemical studies, a bone marrow biopsy with histological and immunofluorescence findings consistent with SM, and a skin biopsy showing perivascular and interstitial mast cell infiltration. Thus, the diagnosis of SM was established.

Due to complaints of left hip pain, a CT scan was performed, revealing osteonecrosis of the femoral head, which led to the removal of osteosynthesis material and placement of a total hip prosthesis.

As part of the SM work-up, bone densitometry was conducted, showing T-scores of -2.6 and -3.3 for the lumbar spine and femur, respectively. She was referred to the Rheumatology clinic, where it was concluded that her long-standing osteoporosis was secondary to SM. As such, she was started on biannual denosumab therapy, which she is currently maintaining.

Discussion: Although SM is a rare cause of secondary OP, it should always be considered in patients with unexplained osteoporosis, particularly in the presence of symptoms related to mast cell release. In the absence of cutaneous manifestations and triggers for anaphylaxis, fragility fractures may be the only clinical manifestation of SM. Tryptase measurement is the initial test, and the diagnosis can be definitively established with a bone marrow biopsy.

In this patient's case, the diagnosis was delayed due to the presence of hyperparathyroidism as a potential cause of OP. However, even after bisphosphonate treatment and PTH normalization, there was no improvement in bone mineral density, suggesting a significant contribution of SM to the condition.

The apparent predominance of osteoclasts and bone resorption in this disease makes anti-resorptive agents the treatment of choice and the most studied option. Bisphosphonates are effective, although denosumab has been suggested as preferable due to its mechanism of action, which appears to have specific effects on the likely pathogenesis of the disease.

174 - POST-BARIATRIC SURGERY REGIONAL MIGRATORY OSTEOPOROSIS: A RARE CASE OF RAPID ONSET AND BILATERAL HIP INVOLVEMENT

Miguel Correia Natal[1], Bárbara Fernandes Esteves[1], Lúcia Costa[1], Georgina Terroso[1]

[1]Rheumatology Department, Unidade Local de Saúde de São João, Porto, Portugal

Case presentation: a 36-year-old woman with no significant medical history was seen for disabling left hip pain requiring crutches, 4 days after an uncomplicated bariatric surgery (Roux-en-Y anastomosis) for morbid obesity. She had no other complaints. Magnetic resonance imaging (MRI) was performed, revealing marked bone marrow edema consistent with transient osteoporosis of the hip (TOH), although early-stage osteonecrosis of the femoral head (ONFH) could not be ruled out. Bone densitometry (DEXA) yielded T-scores of -0.5 in the lumbar spine and -2.2 in the femoral neck. A conservative approach based on limited weight bearing and analgesia was adopted.

Over the next 6 months, the patient reported gradual symptom improvement. However, as the pain on the left side was resolving, she developed similar, equally limiting pain in the right hip. Plain radiographs showed no significant changes. A subsequent MRI demonstrated signs of bilateral TOH, showing regression of findings in the left hip and the emergence of new ones in the right. Comprehensive laboratory tests were normal. Thus, the diagnosis of regional migratory osteoporosis (RMO) was established.

The recommendation for non-weight bearing of the limb was maintained, and calcium supplementation was initiated. A follow-up MRI performed 5 months later showed resolution of the bone marrow edema in both hips. Around this time, the joint complaints also disappeared, leading to a full recovery. The patient is currently asymptomatic.

Discussion: RMO is a rare condition most commonly presenting as focal, migrating arthralgia in weight-bearing joints of the lower limbs, often accompanied by joint effusion. Although self-limiting, symptom duration and time between primary and secondary joint involvement may vary widely. It mainly affects men in their fifth and sixth decades of life.

Biochemical and serological markers are typically normal, and radiographs may show no changes at symptom onset. Therefore, MRI is crucial for diagnosis, showing bone marrow edema that normally resolves within a year. DEXA has been suggested to play a role in confirming the diagnosis and monitoring therapeutic response.

Since the most common primarily affected joint is

the hip and its imaging findings are indistinguishable from those of TOH, a diagnosis cannot be made until migratory symptoms have appeared. Many authors consider these two entities, together with ONFH, as different stages of the same disease, often referred to as "bone marrow edema syndrome". Many case reports point out that RMO has probably been underrepresented in the literature due to this uncertainty in terminology and lack of classical features.

Conservative treatment with limited weight bearing, analgesia, and repletion of Vitamin D and calcium levels is an appropriate and effective regime. Comparison between therapeutic options is difficult since it is a very rare and self-limiting disease.

Its pathogenesis remains unclear, but some risk factors have been suggested, such as a history of smoking, low calcium intake, and the presence of systemic osteoporosis or osteopenia. Very few cases of TOH after bariatric surgery have been reported. To the best of our knowledge, there have been no previous reports of RMO after bariatric surgery in the literature, nor cases of TOH cases occurring such a short time after the procedure. This clinical case highlights the importance of early disease identification to educate the patient, commence appropriate treatment, and avoid unnecessary invasive interventions.

175 - DENOSUMAB IN PAGET'S DISEASE OF BONE: A NEW THERAPEUTIC OPTION?

Bárbara Fernandes Esteves[1], Miguel Correia Natal[1], Lúcia Costa[1], Raquel Miriam Ferreira[1]

[1]Rheumatology Department, Unidade Local de Saúde de São João, Porto, Portugal

Introduction: Paget's disease of bone (PDB) is the second most prevalent metabolic bone disease, surpassed only by osteoporosis. The clinical spectrum varies from asymptomatic to bone pain, deformities, or fractures. The most commonly used marker for diagnosis and monitoring is alkaline phosphatase (ALP). First-line therapy is bisphosphonates, particularly a single dose of 5 mg zoledronate. Bisphosphonates are contraindicated in patients with a glomerular filtration rate (GFR) below 30 mL/min. Even though few cases are described in the literature, denosumab appears to be an option for these patients. The following case is notable for involving a patient with polyostotic PDB with reactivation after zoledronate therapy, but with a favorable response to denosumab.

Case Report: We present a case of a 70-year-old man with a polyostotic PDB diagnosed in 2000, with involvement of the sacrum, iliacs, D11 and L1. He received treatment with pamidronate in September 2000 and July 2003 and zoledronate in 2009, 2011, and 2015. He also had history of crohn's disease, microcrystalline arthropathy involving the knees, dyslipidemia, hypertension, chronic alcoholic liver disease and chronic kidney disease that progressed slowly over the years to a stage 4 by 2021. In March 2022, he developed a disabling low back pain that some times radiated to the right leg, without response to analgesic treatment. A lumbar spine computed tomography (CT) showed changes suggestive of PDB in L1 and L5 and radicular involvement between L4-L5 and L5-S1. Nuclear bone scan showed heterogeneous radiopharmaceutical uptake consistent with reactivation of PDB. Analytically, he presented an increase in bone-specific ALP (101µg/L, normal <20,1µg/L), serum ALP (346U/L, normal 30-120U/L), beta-crosslaps (1.38ng/mL, normal <0.30ng/mL) and osteocalcin (123.4ng/mL, normal <26.3ng/mL). Due to low renal clearance (GFR 24-32mL/min), he started treatment with denosumab 60 mg every 6 months with complete resolution of pain after about 2 weeks. He showed decrease in bone-specific ALP (24.5µg/L), serum ALP (154U/L), beta-crosslaps (1.15ng/mL) and osteocalcin (60.0ng/mL) over 1 year. Repeat nuclear bone scan after 6 months of therapy was comparable to the previous one. The patient showed rapid laboratory response, however, the ALP tended to increase once the next administration approached. These laboratory findings may suggest a rebound effect similar to the one observed in osteoporosis.

Discussion: The pathophysiology of PDB is not yet fully understood. Zoledronate is the first-line treatment. When there is intolerance or contraindication to bisphosphonates, the indications are less clear. Denosumab is a safe alternative for patients with renal impairment. To the authors' knowledge, there are no comparative studies between zoledronate and denosumab in PDB. Another question remains, how long and frequently should these patients be treated. In the literature, there are no studies indicating the frequency or duration of denosumab in PDB.

182 - CLINICAL EFFICACY OF ZOLEDRONATE IN THE TREATMENT OF GARRÉ'S OSTEOMYELITIS: A CASE REPORT

Augusto Silva[1], João André correia[2, 3], Francisco Salvado[2, 3], José Carlos Romeu[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 Estomatologia, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Clínica Universitária de Estomatologia, Faculdade de Medicina da Universidade de Lisboa, Lisboa, Portugal

Introduction: Garrè's osteomyelitis (GOM) is a rare inflammatory disease with an unclear etiology. It mainly affects the region of the mandible in young children and adults under 30 years. The goal of the treatment is to control symptoms, however the most appropriate management remains unclear. There are reports of off-label use of antiresorptive drugs, namely bisphosphonates, in patients with refractory disease, with positive results.

Case Report: A 20-year-old woman was referred due to pain and swelling of the right jaw over the last 5 years. In 2016, after an endodontic treatment of tooth 4.6 (mandibular right first molar), the patient started experiencing pain and swelling of the right mandible region (fig A). The tooth was extracted, but an inflammatory process persisted and she did several cycles of antibiotic therapy and analgesics.

In 2018 she underwent a second molar extraction (fig B-C). Intraoperatively, dense bone was observed with areas of osteoid material and periosteal reaction and biopsy revealed a chronic inflammatory process. She started high dose prednisolone (PDN, initially 40mg per day, then reduced to 10mg) that was kept for 3 years. A CT of the jaw in 2019 revealed a periosteal reaction with progressive ossification of the right mandibular body (fig D-E).

After PDN dose reduction she had a recurrence of symptoms at a dose inferior to 7.5mg per day. She underwent hyperbaric chamber treatments without improvement. A CT in 2021 revealed a layered bone sclerosis, resulting in a double contour of the mandible and facial asymmetry, which confirmed the diagnosis of GOM (fig F-G).

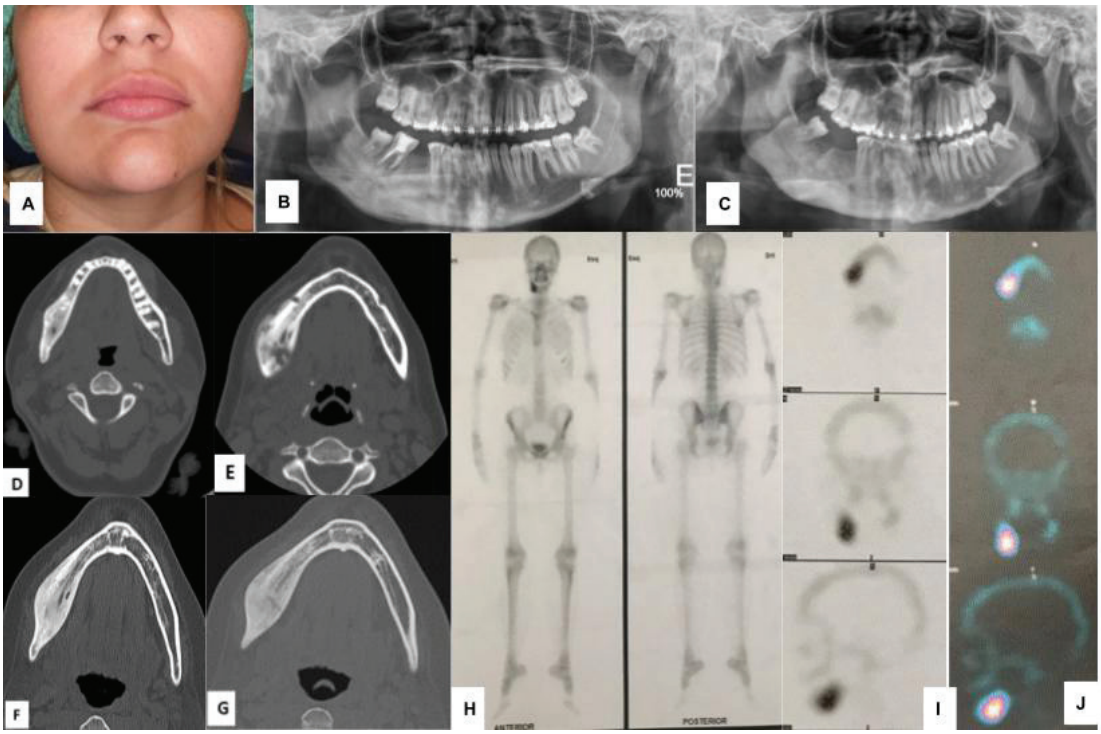
During follow-up the patient progressively reduced the PDN dose over 6 months, until its discontinuation in September 2021. By this time, laboratory tests showed elevation of the inflammatory parameters (ESR 50 mm; CRP 2.17 mg/dl). Whole-body bone scintigraphy with head SPECT/CT showed focal hyperfixation in the right jaw (figures H-J).

Since the patient maintained recurrent complaints of pain, at least one crisis per month, she accepted treatment with zoledronate, single IV administration, with symptoms resolution and improvement of inflammatory parameters (ESR 29 mm; CRP 1.09 mg/dl).

Discussion: GOM is a rare chronic inflammatory disease characterized by an insidious onset, localized pain with episodic nonprogressive nature, facial asymmetry and trismus, without suppuration. The duration is variable and may persist for several months or years. Between crises, most patients are assymptomatic.

The etiology is unclear and usually the diagnosis is made based on the patient's history and conventional radiographic methods or CT images.

The most appropriate treatment remains unclear. Reported modalities include antimicrobial therapy, glucocorticoids, occlusal splint and physiotherapy, hyperbaric oxygen therapy, antiresorptive medica-



182 – Figure 1. Osteomyelitis of Garré

tions, targeted biologic therapy or surgical resection. The most accepted is antibiotic administration and extraction of teeth with inflammatory lesions or decay, which leads to lesion regression and bone remodeling over a period of 6 to 8 months. However, in most cases these treatments fail in achieving a long-lasting reduction of the recurrent pain, trismus and swelling. In these cases, bisphosphonates can be useful to achieve symptom control.

Conclusion: We present a case of a 20-year-old woman with GOM for at least 5 years, refractory to conventional medication, who was treated effectively with zoledronate. The use of bisphosphonates should be considered for refractory cases of GOM.

190 - SUCCESSFUL TREATMENT OF REFRACTORY ADULT-ONSET STILL'S DISEASE WITH THE ASSOCIATION OF IL-1 INHIBITOR ANAKINRA AND JAK INHIBITOR BARICITINIB: A CASE REPORT

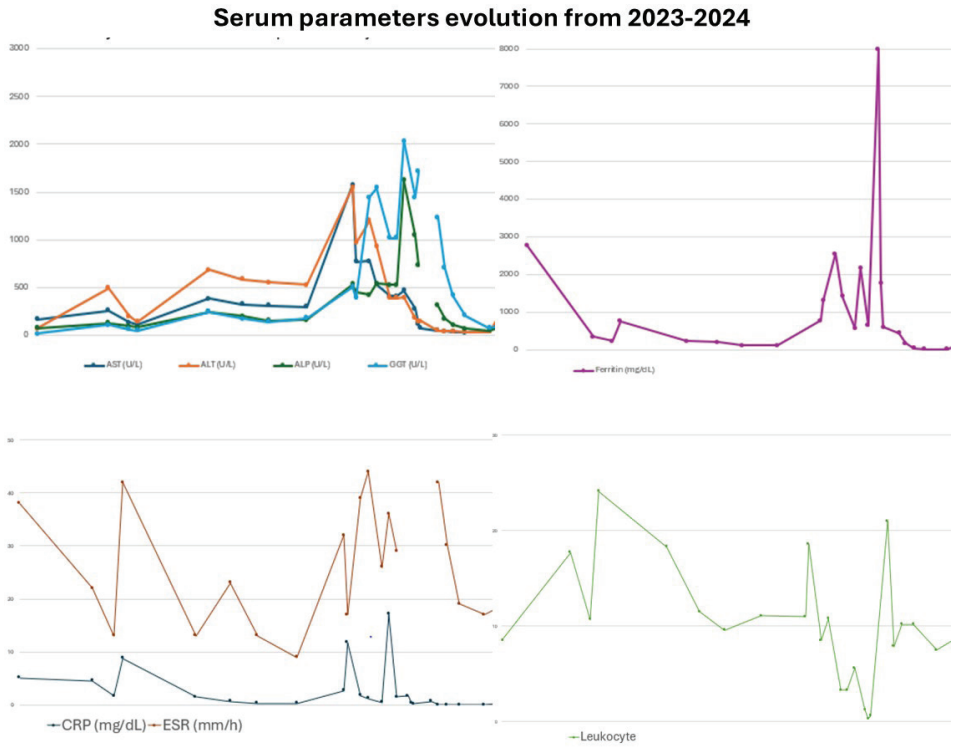
Sofia Ferreira Azevedo[1, 2], Susana P. Silva[1, 2], C. Pinto Oliveira[1, 2], Carolina Vilafanha[1, 2], Pedro Miguel Teixeira[1, 2], Eduardo Dourado[1, 2], Ana Rita Prata[1, 2], Gisela Eugénio[1, 2], Inês Cunha[1, 2], Anabela Barcelos[1, 2, 3, 4], Carolina Mazeda[1, 2, 3]
[1]Rheumatology Department, Unidade Local de Saúde da Região de Aveiro, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz Health Alliance, Portugal, Aveiro, Portugal, [3]EpiDoc Research Unit, CEDOC, NOVA Medical School, Lisboa, Portugal, [4]Comprehensive Health Research Center, NOVA University School, Lisboa, Portugal

We present the case of a 23-year-old female that presented in 2020 with fever, evanescent rash, lymphadenopathy, hepatosplenomegaly, hepatic involvement, and elevated inflammatory markers. She progressed to macrophage activation syndrome (MAS), requiring intensive care admission due to multi-systemic dysfunction. She was treated with IV methylprednisolone (MPDN) pulses and IV immunoglobulin (IVIg) and, after an exhaustive study a diagnosis of adult-onset Still disease was made. Following initial prednisolone (PDN) therapy, methotrexate was initiated and later combined with anakinra (100 mg/day SC), achieving a good response until April 2023 when a new episode of MAS ensued. She was treated with IV MPDN pulses followed by high-dose oral PDN and methotrexate was

190 – TABLE 1. Analytical evolution from april 2023 to june 2024

	AST (U/L)	ALT (U/L)	ALP (U/L)	GGT (U/L)	Ferritin (mg/dL)	Leukocyte u/L	CRP (mg/dL)	ESR (mm/h)
17/04/2023	167	66	76	15	2787	8.5	5.08	38
18/07/2023	108	138	80	45	759	24.1	8.8	42
21/06/2023	260	493	127	109	353	17.7	4.57	22
10/07/2023	135	196	94	59	229	10.7	1.72	13
18/08/2024	923	1448	375	337	1060	12.6	1.36	25
21/09/2023	383	684	241	245	233	18.3	1.53	13
22/10/2023	324	587	201	172	205	11.5	0.63	23
15/11/2023	312	552	152	137	123	9.6	0.25	13
20/12/2023	298	529	162	181	112	11.1	0.26	9
31/01/2024	1577	1550	536	507	773	11.0	2.7	32
03/02/2024	772	968	456	389	1300	18.6	11.73	17
15/02/2024	776	1201	422	1442	2546	8,5	1,76	39
05/03/2024	410	387	528	1021	580	3,3	0,5	26
11/03/2024	410	387	528	1021	2174	3,3	17.10	36
18/03/2024	465	395	1622	2033	648	5.6	1.5	29
31/03/2024	116	147	728	1717	1784	0.3	0.36	120
18/04/2024	46	50	311	1233	441	21.00	0.7	71
24/04/2024	38	44	174	704	182	7.9	0.05	42
02/05/2024	32	38	107	415	52	10.2	0.05	30
13/05/2024	29	32	65	208	22	10.2	0.05	19
04/06/2024	38	34	47	76	12	7.5	0.05	17

ALT: alanine transaminase; AST: aspartate transaminase ALP: alkaline phosphatase ; CRP: C- reactive protein; ESR: Erythrocyte sedimentation rate; GGT: gamma-glutamyl transferase



190 – Figure 1. Serum parameters evolution from 2023 to 2024

switched to cyclosporine (discontinued a few weeks later due to suspected liver toxicity). By August 2023, while under treatment with anakinra 100 mg/day SC and PDN 35 mg/day, she was readmitted with fever, elevated inflammatory markers, liver enzyme abnormalities, and hyperferritinemia (Table 1, Figure 1). Liver histology suggested disease activity/anakinra toxicity, prompting a switch to canakinumab 4 mg/kg/month. Despite the normalization of inflammatory markers, serum liver enzymes remained high. In February 2024, she presented with acute hepatitis (jaundice, dark urine, pale stools, aggravated cytocholestasis) and high inflammatory markers, with progressive worsening during admission and onset of fever (39°C) and evanescent salmon-colored rash. A thorough evaluation to exclude infectious, neoplastic, and other immune-mediated etiologies indicated the likely contribution of disease activity and amoxicillin-clavulanic acid toxicity (administered in the previous 2 weeks) to the clinical picture. Due to a lack of liver enzyme improvement after anakinra suspension and a poor response to canakinumab, anakinra was reintroduced and titrated up to 400 mg/day, resulting in an initially favorable clinical and laboratory response. However, one-week post-discharge, the patient developed a daily fever (38.5-39°C) and a new increase in inflammatory markers. Blood cultures identified Listeria monocytogenes bacteremia, for which she completed 21 days of IV ampicillin with clinical and laboratory improvement. However, the liver enzyme profile worsened during this period, likely due to ampicillin toxicity. Further contributing to the case complexity, the patient had been submitted to a tooth extraction without antibiotic prophylaxis days before readmission and experienced delayed healing, with signs of dry alveolitis and a high risk of progressing to osteomyelitis, requiring regular Stomatology care and IV metronidazole treatment. After an initial one-week apyrexia period, fever recurred, with an increase in ferritin, triglycerides, and CRP, along with new-onset severe leukopenia and neutropenia. Infectious causes were excluded, and a third MAS episode was assumed. Treatment with IV MDPN pulses and IVIg reduced inflammatory markers, but severe leukopenia (nadir 0.3/µL) and neutropenia (nadir 0/µL) persisted. Bone marrow aspirate excluded primary hematologic malignancy, so anakinra toxicity was assumed, and this treatment was temporarily suspended. This resulted in a recovery of leukocyte and neutrophil counts to normal values. Ten days later, anakinra was reintroduced at a lower dose (200 mg/day SC) and combined with baricitinib, leading to clinical and serological improvement, with normalization of liver enzymes and decrease in inflammatory markers, allowing for progressive PDN tapering.

192 - ORGANIZING PNEUMONIA AFTER MORE THAN 10 YEARS OF METHOTREXATE TREATMENT IN A RHEUMATOID ARTHRITIS PATIENT

Maria Pontes Ferreira[1], Anita Cunha[1], Catarina Soares[1], Susana Almeida[1], José Tavares-Costa[1]
[1]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal

Introduction: Organizing pneumonia (OP) is a rare entity, that can be secondary to lung injury such as infections, drug toxicity, radiotherapy, among other causes, or it can be idiopathic, also known as cryptogenic1. We present a case of methotrexate (MTX) induced OP.
Clinical case: A 53 years old female patient is followed in our centre since 2003 due to rheumatoid arthritis (RA), with positive rheumatoid factor and anti-citrullinated protein antibodies. She was previously treated with MTX alone, MTX plus ADA, with secondary failure and etanercept, with which the patient developed erythema nodosum requiring its discontinuation. For this reason, rituximab (RTX) was introduced in association with MTX in 2010, making it possible to reach clinical remission.

In September of 2023, about 2 months after the last RTX perfusion, the patient began to have low-grade fever, anorexia and fatigue and she was hospitalized for etiological study. Her blood work showed a low grade normocytic normochromic anaemia (haemoglobin 12.1g/dL) and elevated erythrocyte sedimentation rate (ESR) and c-reactive protein (CRP), 47 mm and 15.47 mg/dL, respectively, and her respiratory virus panel was negative. A computer tomography (CT) scan was performed, revealing densification in the right lower lobe (RLL), with inverted halo sign, ground-glass and peribronchial densifications in the upper left lobe (LL). Given the suspicion of pulmonary infection, blood cultures were collected and intravenous antibiotics were started, namely amoxicillin+clavulanic acid. A bronchoscopy was performed, showing no significative changes, and no isolation of any microorganisms in bronchoalveolar lavage. Due to lack of response, the therapy was escalated to piperacillin-tazobactam and then cotrimoxazole was added. The thoracic CT scan was repeated, showing resolution of densification with inverted halo in the RLL, and peripheral densifications were present in the LL, with migratory pattern, consistent with OP. Thus, the patient begun prednisolone 40mg/day and was discharged with a referral to Pneumology consult, where the was reevaluated 2 months later. Since the fever persisted, the prednisolone dose was increased to 1mg/Kg/day.

Despite high doses of prednisolone, the fever and migratory pulmonary lesions persisted and other causes of OP were investigated and excluded, such as indolent and atypical bacterial like Pneumocystis jirovecii or tuberculosis, and fungi.

Since no other cause was discovered, the MTX induced OP was hypothesised and so MTX was discontinued in February 2024, with consequent sustained apyrexia and normalization of ESR and CRP.
Discussion: OP is a rare acute lung injury, with its incidence ranging between 1.97-7 cases per 100,000 people1. Although pulmonary involvement of RA can sporadically present with OP1, in this case the patient was in clinical remission for years, making this hypothesis less likely. Granting the clinical response to MTX suspension, this patient developed OP after more than 10 years of MTX treatment. To our knowledge, there are few cases described of MTX induced OP, and most of patients developed OP weeks (and not years) after its initiation2, so this case provides a new perspective on methotrexate-associated OP.

REFERENCES

1. Baque-Juston M, Pellegrin A, Leroy S, Marquette CH, Padovani B. Organizing pneumonia: what is it? A conceptual approach and pictorial review. Diagn Interv Imaging. 2014
2. Faisal M, Roslan A, Nik Abeed NN, Ban Yu-Lin A. Disease-modifying antirheumatic drugs and organising pneumonia. BMJ Case Rep. 2021

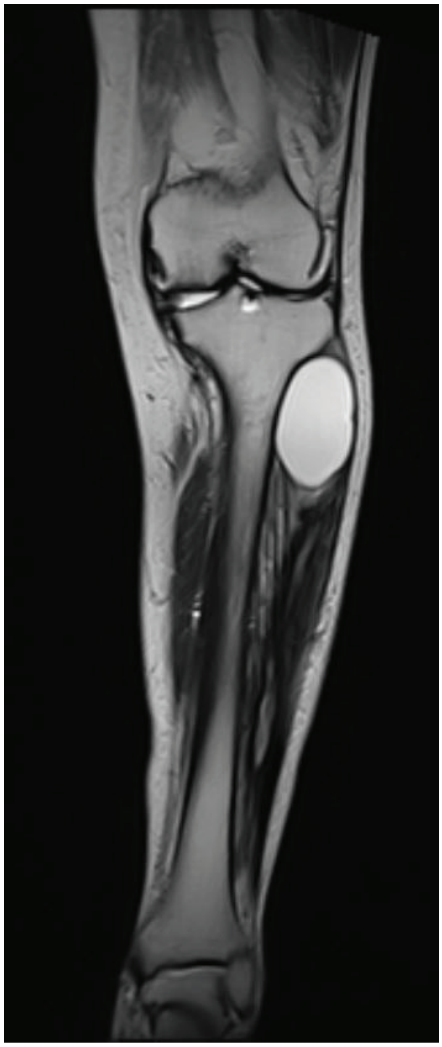
199 - PÉ PENDENTE - COMPRESSÃO DO NERVO PERONEAL POR QUISTO GANGLIÓNICO

Carla Campinho Ferreira[1], Paulo Pereira[1], Ana Margarida Correia[1], Joana Leite Silva[1], Ana Ribeiro[1]
[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

As neuropatias compressivas são comuns, podendo afetar qualquer nervo periférico, sendo que a compressão pode ser de causa extrínseca ou intrínseca ao nervo. A clínica vai depender do nervo afetado e do local de compressão, caracterizando-se por défice de força e/ou atrofia muscular, se atingimento de um nervo motor ou parestesias e dor neuropática se atingimento de um nervo sensitivo.

Doente do sexo feminino, 64 anos, seguida na consulta de Reumatologia por Artrite Reumatóide em remissão há vários anos, medicada com leflunomida 20 mg/d e prednisolona 5 mg/d. Como antecedentes pessoais, destaca-se hipertensão arterial e dislipidemia, ambas medicadas e controladas, gonartrose bilateral, espondilartrose e tendinopatia da coifa dos rotadores à direita. Em consulta de seguimento habitual, referiu quadro com 2 meses de evolução de sinais inflamatórios na perna esquerda, tendo recorrido ao SU por 3 vezes e medicada com 3 ciclos de antibioterapia por

presumível erisipela com melhoria após o último antibiótico. Há 1 semana, reportava défice de força no pé esquerdo, com dificuldade na marcha. Ao exame físico destacava-se força grau 0 na dorsiflexão do pé esquerdo e na extensão do hálux esquerdo e hipostesia no território do peroneal profundo. Não foram objetivados sinais de atividade inflamatória da artrite reumatóide nem outros défices de força e o estudo analítico não revelou alterações. Para estudo deste quadro de pé pendente foi solicitada eletromiografia (EMG) dos membros inferiores que revelou lesão axonal do nervo peroneal profundo esquerdo, sem outras alterações, nomeadamente sinais sugestivos de sofrimento radicular lombossagrado motor bilateral. Iniciou tratamento fisiátrico, com melhoria da força na extensão do hálux, mantendo o défice de dorsiflexão do pé. Cerca de 10 meses após o início do quadro, refere aparecimento de tumefação dura, na região proximal da face antero-lateral da perna, sem trauma associado. Foi solicitada ressonância magnética (RM) da perna esquerda, que, além de alterações degenerativas evoluídas nas articulações femuro-tibial e femuro-patelar, revelou a presença de quistos gangliónicos/sinoviais proeminentes, justa-articulares, inferiormente à articulação do joelho, ocupando a parte proximal da perna, na vertente postero-lateral, com 7,2 cm de maior eixo transversal e 4,2 cm de diâmetro anteroposterior, ocupando a zona de cruzamento do nervo peroneal comum e dos seus ramos, causando provável neuropatia compressiva (figura 1). Além disso, foi objetivada a presença de acentuada atrofia e involução adiposa dos músculos do compartimento anterolateral por provável desnervação, a favor de etiologia compressiva. Neste contexto, foi encaminhada para a consulta de Ortopedia, tendo sido submetida a exérese do referido quisto e neurólise do nervo peroneal comum. Na última avaliação, 3 meses após a cirurgia, referia melhoria das queixas, nomeadamente na dorsiflexão do pé esquerdo.



199 – Figure 1. RM a mostar a presença de quisto gangliónico justa-articular ocupando a zona de cruzamento do nervo peroneal comum.

A neuropatia do nervo peroneal é uma causa comum de pé pendente com múltiplos diagnósticos diferenciais, sendo que as causas mais comuns são a compressão extrínseca ou o trauma. O recurso a meios complementares de diagnóstico como e EMG e a RM são cruciais na marcha diagnóstica das neuropatias. Neste caso, inicialmente não havia história de trauma nem outra causa evidente para a compressão do nervo, provavelmente pelas reduzidas dimensões do quisto na fase inicial. A identificação atempada dos quistos gangliónicos que causam neuropatias compressivas é essencial para um tratamento precoce, de forma a evitar o dano neurológico permanente.

203 - MASSA TUMORAL DA ÓRBITA EM DOENTE COM SÍNDROME DE SJÖGREN

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

Introdução: A síndrome de Sjögren (SSj) é uma doença crónica autoimune com tropismo particular para as glândulas exócrinas nomeadamente as lacrimais/salivares, que origina a síndrome sicca, manifestação cardinal nestes doentes. Apesar disso, esta doença pode afetar virtualmente qualquer órgão, pelo que a sua severidade vai desde uma forma muito "leve" com queixas secas, até uma apresentação bastante grave, com atingimento glandular e extra-glandular, cujas complicações podem ser fatais.

Entre estas complicações encontra-se a doença linfoproliferativa, cuja incidência está aumentada de forma global nas doenças autoimunes, mas parece haver uma tendência especialmente marcada na SSj.

Caso Clínico: A respeito desta complicação, apresenta-se o caso de uma mulher de 53 anos, com overlap de SSj e Lúpus Eritematoso Sistémico (LES), desde 1997, com atingimento vascular (Raynaud), articular, cutâneo, polisserosite, hematológico e síndrome sicca. Apresenta doença estável nos últimos anos, em remissão, com terapêutica otimizada de Metotrexato e Hidroxicloroquina, sem flares nem intercorrências major registadas.

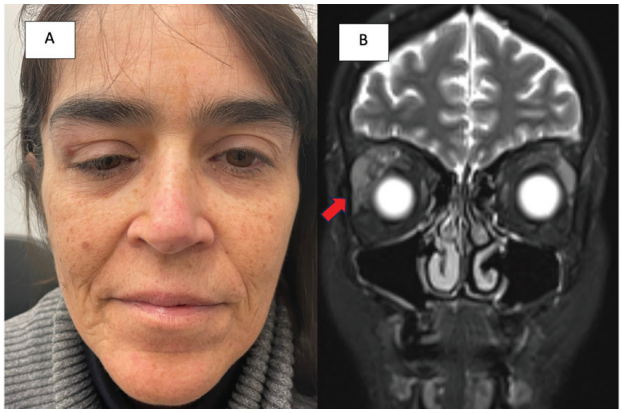
Desde meados de 2023, doente descreve noção de tumefação da pálpebra direita (inicialmente impercetível a olho nú) em agravamento, associada também a perda ponderal involuntária significativa (6 kg no período decorrido), astenia e adinamia. De forma progressiva, desenvolveu também quadro de cefaleia hemicraneana direita, que focalizava na região retro ocular direita, com noção de ptose direita progressiva, agora constatada ao exame objetivo (figura 1A).

Tendo em conta a clínica da doente e sua patologia de base, colocada hipótese de complicação neoplásica, tendo-se avançado com estudo aprofundado. Fez RMN CE (figura 1B) que revelou aumento marcado da glândula lacrimal direita e lesão expansiva anexa ao rego olfativo direito, sem outras lesões expansivas nem outras alterações de relevo. Apesar de os achados imagiológicos favorecerem aumento glandular de etiologia inflamatória, optou-se por biópsia puncional da lesão, que revelou massa compatível com tumor amilóide. Foi realizada PET que excluiu existência de outras lesões sugestivas de neoplasia, bem como atividade metabólica da lesão biopsada.

Analiticamente, apresenta eletroforese de proteínas séricas com pico monoclonal IgG/lambda e aumento das cadeias leves séricas, com rácio de K/L aumentado (1.15). Após investigação do seu contexto familiar, constatou-se existência de PAF em vários familiares, tendo-se assumido, até conclusão do estudo, diagnóstico presuntivo de Amiloidose TTR, aguardando a doente, neste momento, conclusão de estudo e estratificação da doença.

Conclusão: A história natural de um SSj compreende a sua evolução de envolvimento estritamente glandular, até aparecimento de manifestações extraglandulares e, eventualmente, surgimento de linfoma, que constitui a complicação mais grave e com pior sobrevida. Este conceito, associado ao contexto clínico descrito, torna imperativo que se exclua doença linfoproliferativa nesta doente, bem como um follow-up mais apertado.

Importa também referir que se trata, caso se venha a confirmar o diagnóstico, de manifestação inaugural de Amiloide ATTR, e que esta constitui uma atipia no tipo e localização da manifestação.



203 – Figure 1. A: ptose causada pela massa tumoral; B: massa tumoral em RMN

Por fim, constitui também uma complicação que pode ter implicações no seguimento, prognóstico e orientação terapêutica desta doente, dai o interesse na partilha deste caso.

206 - NEOPLASIA PULMONAR – UM RARO DIAGNÓSTICO DIFERENCIAL DE DOENÇA POR DEPOSIÇÃO DE IGG4?

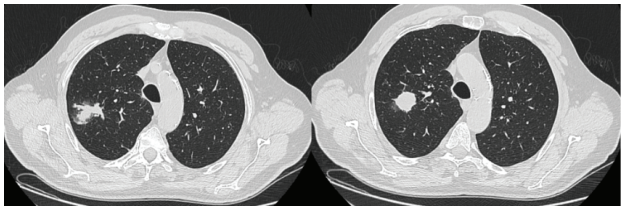
Paulo Jorge Pereira[1], Carla Campinho Ferreira[1], Ana Margarida Correia[1], Emanuel Costa[1], Diogo Esperança Almeida[1], Joana Leite Silva[1], Marcos Cerqueira[1], José Redondo[1], Ana Ribeiro[1], Joana Sousa-Neves[1]
[1]Serviço de Reumatologia, Hospital de Braga, Braga, Portugal

Introdução: A doença de IgG4 é uma doença crónica imunomediada, fibroinflamatória, que ocorre por deposição de IgG4, podendo afetar praticamente qualquer sistema de órgãos, pelo que a sua apresentação é muito variável, de caso para caso.

No global, as manifestações mais típicas passam pelo atingimento da via hepato-biliar, glândulas exócrinas (nomeadamente as salivares/lacrimais), rim e fibrose retroperitoneal.

Numa pequena parte dos doentes, esta doença pode atingir o parênquima e vias aéreas pulmonares, bem como simular algum tipo de atingimento constitucional, vascular (como aortite) e adenopático. Em casos como estes, com apresentações consideradas atípicas, o diagnóstico diferencial é vasto e essencial, tendo em conta a elevada morbimortalidade dessas mesmas condições.

Exemplo disso é o caso de um doente avaliado recentemente, com diagnóstico histológico de Doença por Deposição de IgG4, sem manifestações clínicas sugestivas.



206 – Figure 1. Nódulo pulmonar descrito em corte transversal de TC de Tórax.

Caso Clínico: Apresenta-se o caso de um homem de 69 anos, com antecedentes de dislipidemia e DPOC com vários anos de evolução, como consequência de um historial pesado de tabagismo (cerca de 50-60 UMA), sem antecedentes familiares de relevo. Até meados de 2023, sem queixas de relevo, exceto tosse e expetoração crónicas, estáveis, correlacionadas inicialmente com os seus antecedentes. Desde então, este doente descreve padrão de dispneia de novo para grandes esforços, bem como agravamento do seu padrão habitual de tosse e expetoração, em intensidade e quantidade. Em simultâneo, referia também xerostomia sobretudo matutina, sem xeroftalmia nem noção de alteração/tumefação das glândulas salivares. Na altura, sem quaisquer outras queixas de relevo, nomeadamente clínica constitucional ou focalizadora de determinada etiologia.

Durante o estudo etiológico inicial, foi requisitada uma radiografia do tórax que demonstrou presença de formação nodular relativamente regular, com polo espiculado, no hemitórax direito, que motivou TC Pulmonar (figura 1) e biópsia para avaliação histológica, uma vez que, neste contexto clínico, a hipótese de Neoplasia Pulmonar seria a mais coerente.

Desse estudo resultou a deteção de uma doença de IgG4, confirmada histologicamente, motivo pelo qual o doente foi referenciado a consulta de Reumatologia, onde mantém seguimento. Para estratificação de atingimento de órgão alvo, foi requisitado TC Abdominopélvico, que excluiu, até ao momento, outro tipo de atingimento pela doença, bem como PET, no sentido de excluir doença neoplásica ativa, dadas as características imagiológicas altamente sugestivas de neoplasia, deste nódulo.

Atualmente, encontra-se sem sintomas sistémicos nem terapêutica dirigida.

Conclusão: O caso supracitado reveste-se de elevada importância por vários motivos, desde o diagnóstico diferencial colocado, até ao follow-up necessário nestes doentes. De facto, a apresentação sob forma de nódulo pulmonar é pouco típica desta entidade, sobretudo se considerarmos que se trata da única manifestação até ao momento. Por outro lado, coloca vários desafios no seu seguimento, como a decisão relativamente ao tratamento, uma vez que não existem sintomas específicos deste achado neste doente, bem como a necessidade de manter índice de suspeição elevado, uma vez que existe possibilidade, ainda que diminuta, de erro amostral na obtenção do material histológico, pelo que a hipótese de neoplasia não pode ser completamente afastada.

Serve este caso para alertar para uma doença pouco comum, com uma manifestação atípica, cujo tratamento e follow-up não está ainda preconizado.

208 - DIFFUSE CALCINOSIS – A CAUSE OF RECURRENT LOWER LIMB ULCERS

Rita Silva-Vieira[1], Alice Neves[1], Beatriz de Carvalho Mendonça[1], Ana Bispo Leão[1], Leonor Reynolds[1], Bárbara Lobão[1], Joana Borges[1], Helena Madeira[1], Helena Santos[1, 2]
[1]Instituto Português de Reumatologia, Lisboa, Portugal, [2]Comprehensive Health Research Center (CHRC), NOVA Medical School, University of Lisbon, Lisboa, Portugal

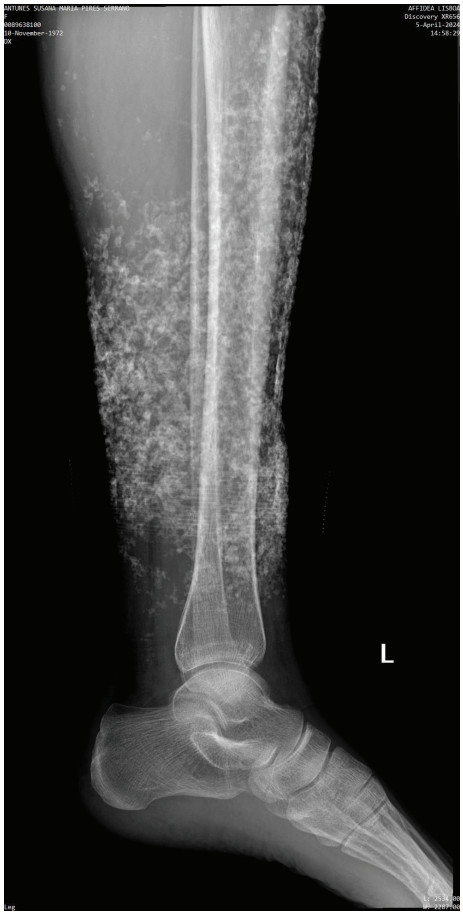
Introduction: Calcinosis within the extracellular matrix of the dermis and subcutaneous tissue, is a possible manifestation of systemic rheumatic diseases, such as systemic sclerosis (SS), dermatomyositis, systemic lupus erythematosus (SLE), and mixed connective tissue disease. It is due to chronic inflammation, ischemia and/or recurrent trauma. The prevalence varies depending of the disease. Calcinosis may be complicated by skin ulceration complicated by recurrent episodes of local inflammation and infection.

Clinical Case: We present the case of a 51-year-old woman diagnosed with overlap syndrome (SLE, SS and Sjögren's syndrome), with 27 years of evolution. From an immunological standpoint, she presents positive ANA (titre 1/2560), anti-dsDNA, anti-Sm, anti-RNP, anti-SSA, anti-SSB, rheumatoid factor and low complement levels. Genetically, HLA DR3 was positive.

The patient currently presents with Raynaud's phenomenon without ulceration, pulmonary hypertension, chronic venous insufficiency of the lower limbs and portal hypertension in the context of chronic liver disease (regenerative nodular hepatic hyperplasia).

At the end of 2022, she began experiencing extensive subcutaneous nodules and plaques accompanied by painful recurrent ulcers, compatible with diffuse dystrophic calcinosis. According to Plastic Surgery, the patient didn't have surgical indication given the extent of the lesions, so she underwent local treatment with sodium thiosulfate, with little benefit.

Imaging and therapeutic reassessment were performed in 2024. X-ray revealed a diffuse pattern of soft tissue calcifications along the entire length of the legs, but with anterior and distal predominance, which followed the location of the ulcers. Ultrasound confirmed multiple calcification foci with 2-3mm depth, without



208 – Figure 1. Difuse Calcinosis - Lower Limb X-ray

Doppler sign. The assistant clinicians proposed treatment with pamidronate in 12-week intervals, and the first administration was completed without complications.

Conclusion: Diffuse calcinosis has a major impact on patients' quality of life, as there is no standard therapy. Control of the underlying disease, individualized care and a multidisciplinary approach with Rheumatology and Plastic Surgery are essential.

211 - ILIAC FRACTURE AS A SIMULATOR OF SACROILIITIS: A CASE REPORT

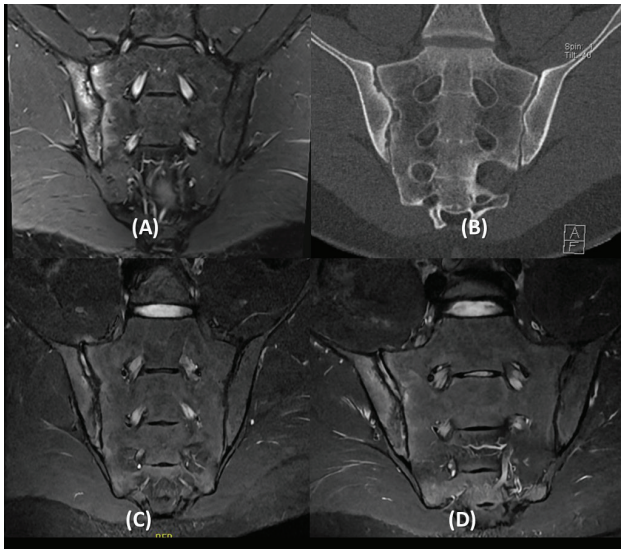
Sara Dias Rodrigues[1], Mónica Jorge[2], Ana Catarina Moniz[1], Daniel Melim[1, 3], Mariana Emília Santos[1, 4], Tiago Saldanha[5], Jaime C. Branco[1, 4], Carina Lopes[1, 4]

[1]Rheumatology Department, Unidade Local de Saúde de Lisboa Ocidental, Hospital Egas Moniz, Lisboa, Portugal, [2]Serviço de Reabilitação de Adultos (SRA1), Centro de Medicina de Reabilitação de Alcoitão, Cascais, Portugal, [3]Rheumatology Department, Centro Hospitalar do Funchal, SESARAM, Funchal, Portugal, [4]Universidade Nova de Lisboa, Nova Medical School, Lisboa, Portugal, [5]Radiology department, Unidade Local de Saúde de Lisboa Ocidental – Hospital Egas Moniz, Lisboa, Portugal

Introduction: Sacroiliitis consists of inflammation of the sacroiliac joints (SIJ) and can manifest as unilateral or bilateral. When the presentation is bilateral and symmetrical, the most likely diagnosis is axial spondyloarthritis. However, in cases of unilateral presentation, the differential diagnosis may be complex. In fact, we must consider inflammatory etiology (mainly psoriatic arthritis and reactive arthritis), osteoarthritis, infectious sacroiliitis, stress fracture, among others.

Clinical case report: We report a case of a 19-year-old male patient, roller hockey goalkeeper, with no relevant personal history, but with significant paternal family history of axial spondyloarthritis (axSpA) (father, brother, uncle, cousin, grandmother). He was referred to our Rheumatology department due to an intermittent mixed rhythm right glutalgia with 1 year of evolution that was only partially relieved by non-steroidal anti-inflammatory drugs (NSAIDs), analgesics and physiotherapy. He denied all possible clinical SpA features and significant trauma. Physical examination was normal except for positive sacroiliac maneuvers on the right side. The laboratory analysis revealed a positive HLA-B27. A SIJ-MRI (Figure 1.A) was performed and reviewed in a multidisciplinary meeting with an expert radiologist. The images revealed a linear hypointensity on the right iliac bone parallel to the SIJ contour surrounded by peripheral bone edema; the left side was unremarkable. A pelvic computerized tomography (CT) scan was carried out (Figure 1.B) revealing SIJ erosions in the postero-inferior region of the iliac aspect of the joint with well-defined sclerosis. The CT and MRI results together were suggestive of a stress fracture. As such, an extensive workup was performed to exclude all possible causes of a secondary stress fracture, such as osteoporosis, neoplasm, and infectious diseases. After a complete rest period of 6 weeks, the patient showed symptomatic improvement. He repeated the SIJ-MRI (Figure 1.C) showing an evident improvement of the fracture trace and edema, however there was new evidence of SIJ synovitis. He resumed his usual activities, maintaining physical exercise restriction. 6 weeks later, a new SIJ-MRI (Figure 1.D) was performed keeping signs of synovitis (without fracture recurrence). Therefore, the overlapping diagnoses of stress fracture and inflammatory sacroiliitis were assumed. The patient gradually restarted physical exercise and maintained follow-up.

Conclusion. Despite some undeniable SpA features, the atypical clinical presentation for axSpA and the initial results of the SIJ-MRI were suggestive of a stress iliac fracture. However, his high-impact sport activity as-



211 – Figure 1. (A) First SIJ-MRI. (B) Pelvic CT. (C) Second SIJ-MRI. (D) Third SIJ-MRI.

sociated with the clinical and radiological evolution led to the suspicion of a stress fracture superimposed on an inflammatory sacroiliitis. This case highlights the importance of keeping in mind the differential diagnoses of axSpA. Additionally, it embodies the already described bone fragility associated with inflammatory joint process, predisposing to stress fractures, particularly in patients with high-impact sports.

212 - PSEUDOTUMOR DO TENSOR DA FÁSCIA LATA – RELATO DE CASO CLÍNICO

Susana Almeida[1], Anita Cunha[1], Catarina Soares[1], Maria Pontes Ferreira[1], Filipa Teixeira[1], José Tavares-Costa[1], Diogo Roriz[2], Soraia Azevedo[1]

[1]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [2]Serviço de Radiologia, Unidade Local de Saúde do Alto Minho, Viana do Castelo, Portugal

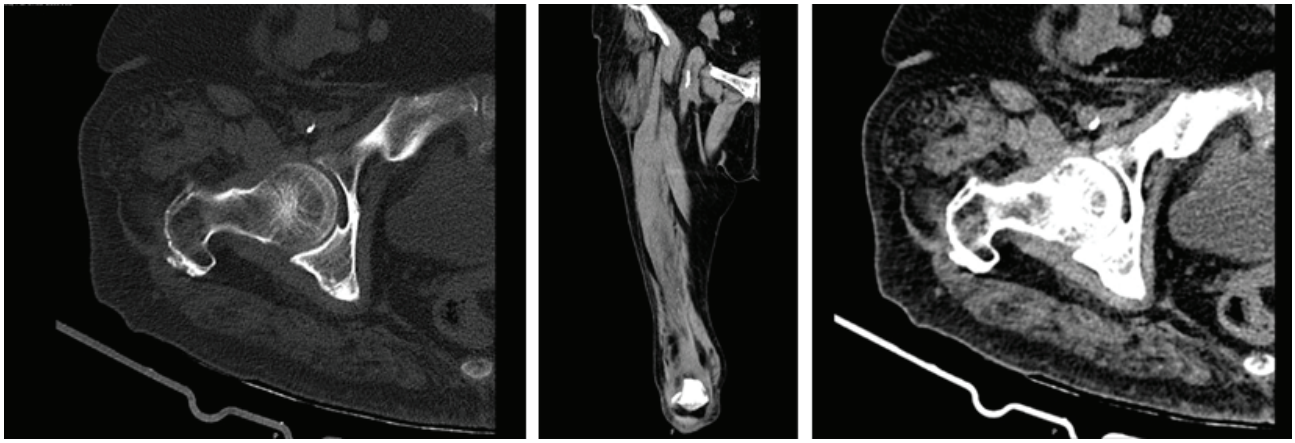
Introdução: O tensor da fáscia lata (TFL) é um músculo localizado na região anterolateral da coxa, que se insere na margem lateral da espinha ilíaca anterosuperior e desce lateralmente ao longo da coxa até se inserir no tubérculo de Gerdy no côndilo lateral da tíbia. O TFL ajuda a estabilizar a pélvis durante a marcha, participa na abdução e rotação interna da anca, na flexão e estabilização do joelho durante a extensão.1 O pseudotumor do TFL consiste numa massa/hipertrofia rara e benigna na face anterolateral da coxa proximal, que frequentemente mimetiza um tumor maligno.2 O diagnóstico pode ser feito por imagem.

Caso clínico: Mulher de 85 anos, com antecedentes de fatores de risco cardiovasculares (hipertensão arterial, dislipidemia e hiperuricemia), seguida na consulta de reumatologia por polimialgia reumática. Numa consulta de vigilância, referiu dor mecânica e noção de tumefação na face lateral da anca direita, com cerca de 2 meses de evolução, sem história de traumatismo ou cirurgia. Ao exame físico, apresentava uma tumefação duro-elástica móvel na face lateral da crista ilíaca direita. A ecografia revelou edema do músculo TFL, tendo-se solicitado TC para esclarecimento diagnóstico, que mostrou pseudohipertrofia do TFL direito, por infiltração adiposa, e ligeira atrofia do glúteo médio direito sem sinais de lesões ósseas ou da aponevrose, compatível com pseudotumor do TFL. Tendo em conta a benignidade da lesão, optou-se por uma atitude de vigilância na consulta, verificando-se estabilidade da lesão após 1 ano de seguimento.

Discussão: O diagnóstico diferencial de uma massa palpável na face anterolateral da coxa inclui neoplasias benignas e malignas, depósitos focais de gordura, hematomas, abscessos, anomalias da fáscia ou dos grupos musculares presentes naquela área. Uma hipertrofia muscular é caracterizada pelo aumento do volume muscular devido ao aumento do número e tamanho das fibras musculares. Em contrapartida, na pseudohipertrofia, o aumento de volume ocorre por infiltração de gordura e tecido conjuntivo no músculo. A hipertrofia do músculo TFL é uma patologia rara, com poucos casos publicados na literatura.1 Os doentes podem apresentar dor e tumefação na região anterolateral da anca ou sintomas que mimetizam uma radiculopatia lombossagrada.2 Relativamente à etiologia existem duas hipóteses: hipertrofia mecânica e hipertrofia por desnervação. A causa mais comum é a sobrecarga seletiva deste músculo como consequência da alteração da biomecânica com redistribuição da carga. A desnervação por lesão da raiz ou do plexo nervoso, embora se manifeste principalmente na forma de atrofia muscular, também pode levar à hipertrofia compensatória ou pseudohipertrofia do músculo afetado.1,2 O tratamento consiste no fortalecimento dos músculos abdutores da coxa e, em alguns casos, na administração de toxina botulínica.2 A RM é o exame gold standard para o diagnóstico da hipertrofia do TFL, pela melhor diferenciação nos tecidos moles. No entanto, o TC e a ecografia também desempenham um papel importante no diagnóstico. A eletromiografia pode auxiliar no estudo etiológico.2,3

Neste caso clínico, a doente apresenta um pseudotumor do TFL de etiologia não esclarecida.

Conclusão: O pseudotumor do TFL é uma condição benigna rara que pode mimetizar situações mais graves. A suspeita diagnóstica e o apoio da imagiologia podem permitir o diagnóstico, evitando procedimentos invasivos.



212 – Figure 1. TC da coxa direita com pseudotumor do tensor da fáscia lata

214 - UM DIGNÓSTICO DIFERENCIAL DESAFIANTE DE ESPONDILARTRITE AXIAL

Inês Guimarães[1], Diana Rosa-Gonçalves[2]
[1]USF Laços, ULS Região de Aveiro, Aveiro, Portugal, [2]Serviço de Reumatologia, Unidade Local de Saúde de Entre Douro e Vouga, Santa Maria da Feira, Portugal

Introdução: A Síndrome de Quebra-Nozes é uma condição vascular rara causada pela compressão da veia renal esquerda entre a artéria mesentérica superior e a aorta abdominal. Manifesta-se com sintomas pouco específicos, resultando frequentemente no diagnóstico tardio ou incorreto. As queixas mais frequentes são lombalgia, dor abdominal/flanco esquerdo e hematúria. A ecografia com doppler é o exame de primeira linha.
Caso Clínico: Mulher, 42 anos. Sem antecedentes médicos de relevo. Referenciada à consulta de Reumatologia por lombossacralgia esquerda praticamente constante e irradiação ocasional para a região inguinal ipsilateral com 2 anos de evolução. EO destacava-se palpação da região trocantérica e sacroilíaca (SI) esquerda dolorosas com manobras positivas para sacroileíte, avaliação das articulações coxofemorais normais e sinal de lasegue negativo. O estudo analítico mostrou VS 13 mm/hr (≤20 mm/hr), PCR normal, FR e HLA-B27 negativos, exame sumário de urina sem alterações. As radiografias de coluna lombar e bacia não apresentavam alterações de relevo. A RM das SI demonstrou edema medular ósseo subcondral na vertente anterior da articulação sacroilíaca esquerda, com ligeiro hipersinal quer da vertente ilíaca quer da vertente sagrada com esclerose e hipossinal em T1 com uma ligeira irregularidade da interlinha articular numa extensão que é sugestiva já de processo inflamatório traduzindo alterações sugestivas de sacroileíte. Efetuou vários ciclos de, pelo menos, dois anti-inflamatórios diferentes sem qualquer benefício. Iniciou Adalimumab 40mg quinzenal ao qual houve falência primária e fez switch para Secucinumab 150mg mensal. Por ausência de melhoria das queixas, apesar de aumento de dose para 300mg/mês, foi interrompido passado 6 meses do início do tratamento e solicitou-se reavaliação imagiológica. Na RM SI observou-se na vertente ântero-inferior do compartimento sinovial das articulações SI discreta irregularidade das placas ósseas subcondrais, traduzindo prováveis alterações erosivas, bem como ligeira esclerose e edema da medula óssea subarticular, aspetos sugestivos de sacroileíte, mais evidente à esquerda, salientando-se que o edema medular ósseo é menos significativo do que o observado na RM anterior. Iniciou Etanercept 50mg semanal que suspendeu após 3 meses também por falência terapêutica. Ao longo do seguimento, apresentou micro-hematúria isolada e intermitente no exame sumário de urina e, por este motivo, foi solicitada TC Abdomino-Pélvico que demonstrou redução do ângulo aortomesentérico (16º) com compressão da veia renal esquerda, num padrão compatível com "Síndrome de Quebra-Nozes", demonstrou ainda sinais de congestão pélvica com ectasia e tortuosidade das estruturas venosas peri-uterinas, de predomínio à esquerda. Desta forma, foi referenciado à consulta de Urologia onde realizou eco com Doppler de veias renais que confirmou o diagnóstico, aguardando orientação.
Conclusão: A pertinência deste caso prende-se com a dificuldade na abordagem diagnóstica e terapêutica que nos confrontamos muitas vezes na pratica clínica. Mesmo na presença de clínica e de imagem sugestivas de determinada doença não devemos excluir outras opções principalmente quando há falência a múltiplas terapêuticas sucessivas.

215 - SACROILIAC CYST: AN UNUSUAL SUSPECT

Rita Silva-Vieira[1], Beatriz de Carvalho Mendonça[1], Bárbara Lobão[1], Leonor Reynolds[1], Ana Bispo Leão[1], Joana Borges[1], Helena Santos[1, 2]

[1]Instituto Português de Reumatologia, Lisboa, Portugal, [2]Comprehensive Health Research Center (CHRC), NOVA Medical School, University of Lisbon, Lisboa, Portugal

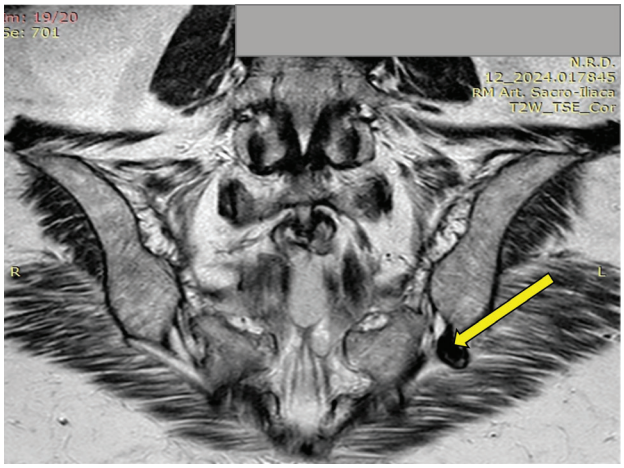
Introduction: Synovial cysts typically develop as a result of degenerative processes. They may develop in any synovial joint and have been commonly described arising from the knee and hip joints. Symptoms may be absent or, when present, can derive from the mass effect on the surrounding structures.

Sacroiliac synovial cyst descriptions are rare in the literature, but they may enhance low back pain.

Clinical case: We present the case of a 60 year-old woman, HLA B27 positive, with a 5-year evolution of low back pain with mixed rhythm and irradiation to the left gluteus region, associated with decreased proximal muscle strength of this limb. She also referred mechanical bilateral artralgias of the hands, knees and feet, without inflammatory signs. Symptoms partially improve with NSAIDs.

Physical examination showed a slight proximal decrease in muscle strength of the left lower limb (degree 4+/5 on hip flexion), ipsilateral positive Lasègue sign and allodynia. Rotulian reflexes were normal, sacroiliac maneuvers were negative and no signs of arthritis were found. Blood analysis revealed normal CK, ESR and CRP levels and negative auto-antibodies (ANA, ds-DNA, RF). X-ray showed undefined sclerosis of the lower end of left sacroiliac joint. CT scan evidenced L5-S1 spondylodiscarthrosis with left parasagittal diffuse herniation with evidence of possible nerve compression. Sacroiliac MRI showed a synovial cyst in the lower end of left sacroiliac joint, with evidence of gluteus minimums focal compression, and no inflammatory local features.

In multidisciplinary discussion, the presence of the cyst was considered as a possible contributor to the low back pain, in consequence of the local mass effect and muscle compression.



215 – Figure 1. Sacroiliac cyst in MRI (T2)

Conclusion: Although low back pain is frequently multifactorial, with degenerative processes such as spondylosis, spinal stenosis, herniation and spondylolisthesis being the most common causes, it is important to consider less frequent etiologies and contributors, particularly in atypical presentations.

217 - ESTATINA, O FÁRMACO BOM E MAU DA FITA? – DOIS CASOS DE MIOPATIA NECROTIZANTE IMUNOMEDIADA

Susana Matias[1], Catarina Abreu[1], Vanessa Fraga[1], Maria Margarida Cunha[1], Sandra Sousa[1], Maria José Santos[1]
[1]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal

Introdução: A miopatia necrotizante imunomediada é uma doença rara que faz parte do grupo das miopatias inflamatórias e se carateriza por fraqueza muscular proximal, valores elevados de creatinoquinase (CK) e positividade para os anticorpos anti-SRP ou anti-HMG-CR. No entanto cerca de 20% dos doentes são seronegativos. É a mais grave das miopatias inflamatórias em termos de dano muscular, com frequente atrofia deste, substituição por gordura e incapacidade.

As miopatias anti-HMGCR positivas estão frequentemente associadas ao uso prévio de estatinas (entre 15-65% dos doentes expostos, dependendo da origem geográfica e da idade), colocando-se a hipótese de a doença ser um efeito adverso raro e grave da exposição a estas. Difere da miopatia tóxica comum induzida por estatinas por não resolver com a suspensão do fármaco.

Apresentam-se dois casos de miopatia necrotizante imunomediada anti-HMGCR positiva.

Descrição do caso: Caso 1 – Doente do sexo feminino, caucasiana, 54 anos, com antecedentes pessoais de vasculite de grandes vasos diagnosticada em 2019 sob tocilizumab, hipertensão arterial, dislipidemia, obesidade e cardiopatia isquémica. Medicada com atorvastatina 40mg desde 2017.

Apresentava desde final de agosto de 2023 diminuição progressiva da força nos membros inferiores, inicialmente com dificuldade em subir escadas e mais tarde com dificuldade em levantar-se de cadeiras e da sanita. Ao exame objetivo com força muscular proximal grau IV dos membros superiores e inferiores. Analiticamente com elevação das enzimas musculares (CK 14774 UI/L, mioglobina 2285 ng/mL, AST 193 UI/L, ALT 290 UI/L). Foi suspensa a atorvastatina sem melhoria dos sintomas. Da marcha diagnóstica, a destacar anti-HMGCR IgG positivo (176 UA, VR< 20), eletromiografia (EMG) com sinais de irritabilidade do sar-

colema em localização crural e proximal e ressonância magnética (RM) a mostrar edema difuso de todos os grupos musculares das cinturas escapular e pélvica. Iniciou prednisolona 0.5mg/kg/dia e metotrexato 10mg oral e substituiu a atorvastatina por evolucumab.
Caso 2: Doente do sexo feminino, caucasiana, 70 anos, com antecedentes pessoais de diabetes mellitus tipo 2, hipertensão arterial e enfarte agudo do miocárdio. Inicialmente medicada com atorvastatina, mas mudança para pitavastatina em abril de 2023 por ligeiro aumento da CK.

Em outubro de 2023 iniciou cansaço, edema dos membros inferiores e sensação de diminuição da força muscular destes. Recorreu a consulta não programada de cardiologia, tendo ficado internada por suspeita de descompensação cardíaca e toxicidade à estatina dado valor elevado de CK (5791 U/L). Descontinuou a pitavastatina no internamento, mas manteve queixas de fraqueza muscular. À observação por reumatologia, apresentava força muscular proximal grau IV dos membros superiores e grau III dos inferiores. Analiticamente com elevação não só da CK, mas de outras enzimas musculares (ALT 165 UI/L, AST 201 UI/L) e anti-HMGCR IgG positivo (167 UA, VR< 20). No EMG com sinais de irritabilidade do sarcolema e na RM com edema difuso de vários grupos musculares ao nível dos braços e coxas. Iniciou prednisolona 0.5mg/kg/dia, metotrexato 15mg subcutâneo e manteve suspensa pitavastatina, ficando apenas com ezetimibe.
Conclusão: Estes dois casos clínicos destacam-se pela raridade das miopatias necrotizantes imunomediadas anti-HMGCR positivas, reforçando a importância da suspeição diagnóstica, principalmente nos doentes expostos a estatinas. Levanta também questões sobre que terapêutica instituir e sobre a possibilidade de um rechallenge com estatina.

219 - RECURRENT UNILATERAL PERIPHERAL ULCERATIVE KERATITIS IN A PATIENT POSITIVE FOR ANTI-CENTROMERE

Sara Dias Rodrigues[1], João Romana[2], Ana Catarina Moniz[1], Daniel Melim[1, 3], Mariana Emília Santos[1, 4], Miguel Leitão[2], Carina Lopes[1, 4], Jaime C. Branco[1, 4], Maria Manuela Costa[1]
[1]Rheumatology Department, Unidade Local de Saúde de Lisboa Ocidental, Hospital Egas Moniz, Lisboa, Portugal, [2]Ophtalmology Department, Unidade Local de Saúde de Lisboa Ocidental, Hospital Egas Moniz, Lisboa, Portugal, [3]Rheumatology Department, Centro Hospitalar do Funchal, SESARAM, Funchal, Portugal, [4]Universidade Nova de Lisboa, Nova Medical School, Lisboa, Portugal

Introduction. Systemic sclerosis is a connective tissue disease that can affect multiple organs, in particular skin, vessels, lungs, joints, gastrointestinal tract, kidneys and other organs. However, the eye involvement is uncommon. When present, ocular manifestations can appear as a consequence of thickening of the eyelid skin, ocular dryness, changes in the iris epithelium and retinal pathology, with corneal affection being rare. Peripheral Ulcerative Keratitis (PUK) is a group of inflammatory diseases whose final common pathway is peripheral corneal thinning. Patients may present with pain (worse at night), redness, tearing, photophobia, and decreased vision. PUK has been associated with many autoimmune disorders, frequently rheumatoid arthritis (34 – 42% of PUK patients), but also other systemic rheumatic diseases. As far as we know, this is the first case report of PUK associated with anti-centromere positivity.
Clinical case report. A 39-year-old woman with no relevant personal history was referred to a Rheumatology consultation due to recurrent PUK with no established etiology. She denied experiencing arthralgia, myalgia or muscle weakness, Raynaud's phenomenon, skin lesions, alopecia, photosensitivity, dyspnea, cough, dysphagia, heartburn, mouth ulcers, dryness, or other symptoms of connective tissue diseases. Physical exam was unremarkable except for the eye. Extensive workup was carried out, including the study of infectious, neoplastic and immunological causes. The laboratory investigation unveiled positive antinuclear antibodies (in a titer of 1/1280) and positive anti-centromere (CENP-B). The capillaroscopy showed nonspecific changes in microcirculation. The chest computerized tomography (CT) and echocardiogram were normal. A diagnosis of PUK was suspected, and treatment was initiated with intensive ocular lubrication, topical ciclosporin, oral doxycycline and a combination of topical and oral steroids. The ulcer exhibited favorable progression initially; however, deterioration was noted 2 weeks after the patient decided to stop all therapy. There was no clinical improvement upon reinstitution of the medical therapy previously described. Hence, the patient underwent surgical intervention and was treated with prednisolone 1 mg/kg/day and methotrexate 20 mg/week. After this, there was a gradual significant improvement until the ulcer resolved. During follow-up at the Rheumatology appointment, no new signs or symptoms of connective tissue diseases, particularly systemic sclerosis, appeared.
Conclusion. Eye involvement in rheumatic diseases is common and may have various presentations. PUK is an urgent situation and a quick referral for corneal consultation is necessary. Having excluded other etiologies and after double laboratory confirmation of positivity for anti-centromere antibodies, this was the only analytical and immunological change present in this

case. Despite not having signs of systemic rheumatic disease, systemic immunosuppressants are sometimes necessary, like in this case. The patient will maintain follow-up and surveillance in a Rheumatology appointment. This case is especially interesting as it highlights the rare association between PUK and anti-centromere antibodies positivity. In the future, the patient's temporal and clinical evolution may help us draw new conclusions.

222 - PHANTOM SCAPULA - A CASE OF GORHAM'S DISEASE

Rita Silva-Vieira[1], Ana Bispo Leão[1], Beatriz de Carvalho Mendonça[1], Leonor Reynolds[1], Bárbara Lobão[1], Joana Borges[1], Manuela Parente[1], Helena Santos[1]
[1]Instituto Português de Reumatologia, Lisboa, Portugal

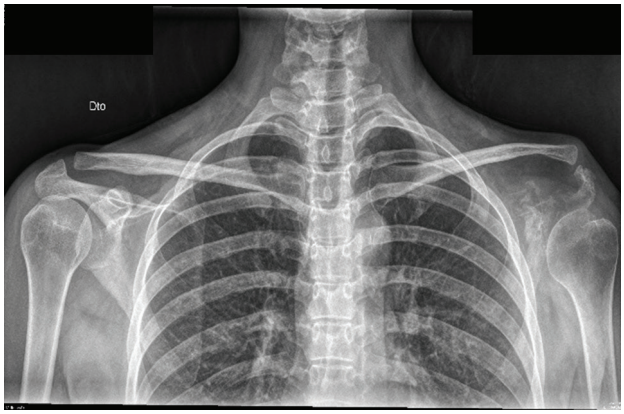
Introduction: Gorham's disease is a rare disorder characterized by proliferation of thin-walled vascular or lymphatic channels within bone, which leads to resorption and replacement of bone with angiomas and/or fibrosis. In some cases, progression may be almost painless until a fracture occurs. Etiology is unknown, but in some cases it is preceded by a minor trauma such as a fall.

Clinical case: We present the case of a 47-year-old woman, previously healthy, who in 2015 fell on her back and used her left arm to support the impact of the fall. This movement triggered intense pain in the scapular region. She was evaluated in the emergency department (ED), where a fracture was ruled out and she was discharged with brachial suspension and analgesia. After 8 weeks, she maintained intense pain in the scapula despite having maintained active mobility, so she underwent X-ray, CT and MRI that were reported as normal. After 9 months, she reports a sudden episode of intense pain and functional impairment of the left arm, so she was re-evaluated in the ED, where acute injuries were again ruled out.

At the beginning of 2016, 10 months after the fall, a CT scan revealed a possible fracture of the body of the scapula, an MRI showed changes in the morphology of the acromion/scapula's spine transition, and a scintigraphy excluded valuable hyperperfusion images. She underwent 6 months of physiotherapy with full mobility recovery.

In 2017, after an episode of acute neuropathic pain in the scapula non-responsive to analgesia, triggered after left arm elevation, she performed an EMG that showed a lesion in the upper part of the left brachial plexus, however, MRI of the plexus was normal.

Between 2017 and 2022, disease progressed with periods of intense pain lasting around 2 days accompanied by great functional limitation and impact on daily life activities, followed by periods of mechanical pain and progressively less response to physiotherapy. Due to the symptoms, she adapted her daily tasks: driving an automatic car, performing self-care with limited use of the left arm.



222 – Figure 1. Gorham X-Ray

At the end of 2022, due to a new disabling and therapy refractory crisis, a repeat CT study revealed osteolysis of the shoulder blade with bone destruction and deformation, as well as osteolysis at the level of the acromion at its base. At this time, scintigraphy showed hyperfixation on the scapula, coracoid process and glenoid.

In 2023, a bone biopsy was performed which revealed an intraosseous hemagioma, the adjacent muscle tissue was normal. Evaluation by a Rheumatologist was recommended and, in 2024, her physical examination showed clear asymmetry of the scapulas and limited mobility, with maximum 30º of elevation and abduction, 40º of adduction, without changes in the flexion of the forearm. An X-ray showed a new slight erosion of the external region of the acromion, phospho-calcium metabolism investigation revealed hypovitaminosis D (19.50ng/mL) with normal calcemia, phosphatemia, PTH, osteocalcin, CTX, alkaline phosphatase, protein electrophoresis and DEXA.

Zoledronic acid was started, and she is being followed up, with controlled pain symptoms.

Conclusion: Gorham's disease is poorly understood, with an unpredictable progression and prognosis. Treatment should be individualized, and an early diagnosis offers a window of opportunity to try to prevent irreversible bone damage.

223 - ISOLATED MUSCLE INVOLVEMENT IN ANCA-ASSOCIATED VASCULITIS - A CASE REPORT

Anita Cunha[1], Maria Pontes Ferreira[1], Catarina Soares[1], Susana Almeida[1], Francisca Guimarães[1], Soraia Azevedo[1], Daniela Santos-Faria[1], Ana Nascimento[2], Miguel Pinto[3],

José Tavares-Costa[1], Daniela Peixoto[1]
[1]Serviço de Reumatologia, Unidade Local de Saúde do Alto Minho, Ponte de Lima, Portugal, [2]Internal Medicine Department, Unidade Local de Saúde do Alto Minho, Viana do Castelo, Portugal, [3]Department of Neuropathology, Centro Hospitalar Universitário de Santo António, Porto, Portugal

Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) is a rare disease characterized by diverse clinical manifestations. Muscles are seldom affected by systemic vasculitis (1). We report a case of AAV presenting with isolated muscle involvement, emphasizing the diagnostic complexities in this infrequent presentation.

We describe a case of a 79-year-old female who, starting in August 2023, developed profound asthenia and increasing dependence in daily activities. Initially, the hypothesis of an infectious complication, specifically a urinary tract infection(UTI) was considered, given her history of recurrent UTIs and findings of leukoerythrocyturia. However, there was no response to the instituted antibiotic therapy, prompting her admission to Internal Medicine department in December 2023 for further evaluation.

On examination, she had generalized muscle weakness, complicated by her recumbent posture and somnolence, which confounded assessment. She had no fever and was hemodynamically stable. Initial laboratory findings showed new-onset anaemia, leucocytosis, elevated inflammatory markers and normal muscle enzyme levels. Investigations included protein electrophoresis (indicating monoclonal IgG/K peak) and a PET scan revealing diffuse 18F-FDG uptake in axial and proximal appendicular skeleton, suggesting marrow reactivity, and a suspicious thyroid nodule.

Haematology consultation, following bone marrow biopsy and medullary study, suggested that secondary hematologic disorders were less likely. Thyroid nodule biopsy indicated benign changes. Additional imaging, including temporal and axillary artery ultrasounds, transthoracic echocardiogram, and thoracoabdominopelvic and spine CT scans, found no significant abnormalities.

Immunologic workup post-exclusion of neoplastic and infectious causes revealed positive ANCA (MPO+). Electromyography showed subacute myopathic changes and whole-body MRI confirmed bilateral, symmetrical muscle oedema in the scapular girdle, forearm and all compartments of the legs. The patient was subsequently referred to Rheumatology for further evaluation. A muscle biopsy was performed, revealing type 2 fiber atrophy accompanied by inflammatory changes consistent with vasculitis, and predominance of IgG4 plasmocytes, which can be seen in AVV, and in this case, less likely linked to IgG4-Related Disease, given the clinical context.

This led to the diagnosis of MPO AAV with isolated muscle involvement. The patient began treatment with high-dose prednisolone, resulting in gradual clinical improvement, muscle strength recovery, and reduced inflammatory markers. Considering age, comorbidities, and infection risk, methotrexate (MTX) was initiated before considering biologic disease-modifying anti-rheumatic drugs. Additionally, she started rehabilitation treatment at a specialized rehabilitation unit.

By June 2024, her health significantly improved with assisted ambulation and partial independence in daily activities, supported by stable haemoglobin and normalized inflammatory markers. She continued on methotrexate with a tapering dose of prednisolone.

This case underscores diagnostic and therapeutic complexities in rare AAV manifestations, emphasizing glucocorticoids and conventional immunosuppressants' efficacy in this severe presentation. Further research is crucial for optimizing treatment strategies, in managing localized forms of vasculitis.

REFERENCES

1. E. Conticini et al. Inflammatory muscle involvement in systemic vasculitis: A systematic review Autoimmunity Reviews 2022 Mar;21(3):103029

225 - FEVER AND EOSINOPHILIA FOLLOWING INTRODUCTION OF ADALIMUMAB

Diana Belchior Raimundo[1], Nuno Pina Gonçalves[1], Sandra Falcao[1, 2]
[1]Serviço de Reumatologia, Hospital Beatriz Ângelo, Loures, Portugal, [2]NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

Background: Adalimumab (ADA) is an anti-tumour necrosis factor α (TNFα) agent widely used for the treatment of several inflammatory rheumatic diseases. Common side effects include increased risk of infections, hypersensibility reactions, transient headache and increased serum lipids. Eosinophilia is a rare secondary effect and is estimated to occur in 0.02% of patients (eHealthMe). Here we present the case of a patient with psoriatic arthritis who developed moderate eosinophilia after starting ADA.

Case Report: A 41-year-old man, whose father had an ankylosing spondylitis diagnosis, was diagnosed with psoriasis in 2004 and four years later developed polyarthritis (wrists and several metacarpal-phalangeal (MCF) joints) and a diagnosis of psoriatic arthritis was made. The patient was treated with indomethacin, with significant improvement.

After ten years of adequate disease control with in-

domethacin on-demand, he was referred to our department due to recurrence of polyarthritis (several MCF and proximal inter-phalangeal (PIP) joints) and was prescribed s.c. methotrexate (MTX) 15mg/week. Due to insufficient response, MTX was progressively increased until 25mg/week and finally leflunomide 10mg was added to the regimen with clinical improvement in 2019.

The patient was then lost to follow-up until April 2023 when he was again referred to our department for polyarthritis of the small joints of the hands after stopping the medication for several years. He was again started on subcutaneous MTX 15mg/week (which was titrated up to 25mg/week) and prednisolone 15mg/day which was tapered to 5mg/day. Laboratory tests evidenced elevated erythrocyte sedimentation rate of 25mm/h (<11) and c-reactive protein 2.02mg/dL (<0.05); pelvic x-ray showed grade III sacroiliitis on the right. Considering insufficient response after optimized MTX dose, the patient was started on ADA 40mg every other week in February 2024.

After the first month of treatment, the patient reported a self-limited fever (38-39ºC), one week after each ADA administration (up to 3-5 days). No other accompanying symptoms were present, including rashes, weight loss, night sweats, diarrhoea or enlarged lymph nodes. Blood work evidenced moderate eosinophilia (2030/uL) which was not present before starting the medication (610/uL). ADA was suspended, with remittance of the fever and progressive decrease of the eosinophile count (710/uL). Other causes of eosinophilia were ruled out and, while no other red flags were present, the patient is currently waiting for a Haematology consultation to evaluate the possibility of a lymphoproliferative disorder.

Conclusion: This patient developed fever and eosinophilia shortly after starting treatment with ADA, which resolved after suspension of the drug. Given the temporal association and clinical and analytical response to the withdrawal it seems very likely that these symptoms are due to an adverse reaction to ADA . There are reported cases of eosinophilia secondary to treatment with ADA described since 2012, one case with associated eosinophilic fasciitis. Other eosinophilic disorders have been associated with other anti-TNFα agents (e.g asthma). Although very rare, clinicians should be aware of this potential side effect for patients treated with anti-TNFα drugs as eosinophilia is usually asymptomatic, but it can lead to important organ damage mediated by release of toxic granule proteins.

230 - PARVOVIRUS B19 INFECTION PRESENTING AS POLYARTHRITIS: CASE SERIES OF A SINGLE CENTER

Sara Dias Rodrigues[1], Daniel Melim[1, 2], Ana Catarina Moniz[1], Mariana Emília Santos[1, 3], Jaime C. Branco[1, 3], Carina Lopes[1, 3]

[1]Rheumatology Department, Unidade Local de Saúde de Lisboa Ocidental, Hospital Egas Moniz, Lisboa, Portugal, [2]Rheumatology Department, Centro Hospitalar do Funchal, SESARAM, Funchal, Portugal, [3]Universidade Nova de Lisboa, Nova Medical School, Lisboa, Portugal

Introduction: Viral arthritis is responsible for 1% of acute polyarthritis cases and can be caused by various agents, such as parvovirus B19 (B19V), Epstein-Barr virus, and hepatitis B and C viruses. B19V infection usually presents with a facial rash and fever and around 60% of patients have joint disease. Although B19V polyarthritis usually has an acute onset with spontaneous resolution within a few weeks, in rare cases it can evolve to a chronic and persistent form. The diagnosis is made based on typical clinical findings in the presence of B19V antibodies and after exclusion of other infectious and non-infectious aetiologies. Since March 2024, different European countries have reported increased detections of B19V in the general population.

Clinical case report: We present a case series of 5 patients with acute polyarthritis due to B19V. Patient and disease characteristics are presented in Table 1. All patients were women of childbearing age (39-48 years, mean age 44 years), with children and/or jobs with a significant infectious epidemiological risk. Cases were detected during springtime (March - May 2024). All cases presented with acute, additive, symmetric, inflammatory polyarthralgias, with progressive worsening over a week. Only one patient mentioned fever and none had a typical rash. Other symptoms suggestive of connective tissue diseases were denied. Physical examination revealed symmetric polyarthritis affecting large joints (knees, ankles, elbows) and small joints of the hands and feet. A complete workup was carried out to search for other etiologies. At baseline, laboratory findings included anemia (n=2), elevation of transaminases (n=3), increased C-reactive protein (n=2) and Erythrocyte sedimentation rate (n=3). The immunological workup revealed low-titer antinuclear antibody positivity in three patients, rheumatoid factor in one (titer of 25UI/mL) and no patient had anti–citrullinated protein antibody. All patients had positive Immunoglobulin M (4.44-150 UI/mL, reference value ≥1.1) and Immunoglobulin G (4.4-44.5 UI/mL, reference value >2.5) B19V antibodies. Patients were refractory to non-steroidal anti-inflammatory drugs and therefore were prescribed with prednisolone 10-15mg/day (one patient did a pulse of methylprednisolone 125mg due to severe polyarthritis). At 1-month follow-up, three out of five patients presented complete resolution of joint

230 – TABLE 1. Patient and disease characteristics of the five patients with Parvovirus B19 infection

Total cases (n)	**5**
Sex (n)	
Woman	5
Man	0
Age (in years)	
Minimum	39
Maximum	48
Clinical presentation (n)	
Fever	1
Rash	0
Joint pain	5
Joint swelling	5
Joint stiffness	5
Laboratory findings (n)	
Anemia [rv < 12.0]	2
Raised AST [rv > 33]	3
Raised ALT [rv > 32]	3
Elevated CRP [rv > 0.5]	2
Elevated ESR [rv > 20]	3
Immunology (n)	
Parvovirus IgM [rv ≥1.1]	5
Parvovirus IgG [rv: > 2.5]	5
Positive ANA	3
Positive CCP [rv > 5]	0
Positive RF [rv > 15]	1

ANA – antinuclear antibody. ALT - Alanine transaminase. AST - Aspartate transaminase. CCP - Anti–citrullinated protein antibody. CRP – C-reactive protein. ESR - erythrocyte sedimentation rate. Ig G - Immunoglobulin G. IgM - Immunoglobulin M. n – number of patients. RF - Rheumatoid factor. RV – reference value.

symptoms and laboratory findings, starting successful weaning of corticosteroid therapy. The remaining two patients experienced recurrence of joint complaints after corticosteroid weaning, suggesting an evolution to a chronic disease state.

Conclusion. Although some case reports have been published in the literature, to our knowledge, this is the first case series of polyarthritis associated with B19V. Despite the well-described possible joint involvement by B19V, it is unusual to detect such a high number of cases in such a short period of time. Furthermore, we highlight that, of the five cases presented, two appear to have progressed to a chronic disease state, which is even rarer.

232 - ACUTE MONOARTHRITIS - THE SAME OLD DILEMMA

Diana Belchior Raimundo[1], Nuno Pina Gonçalves[1], Sandra Falcao[1, 2]

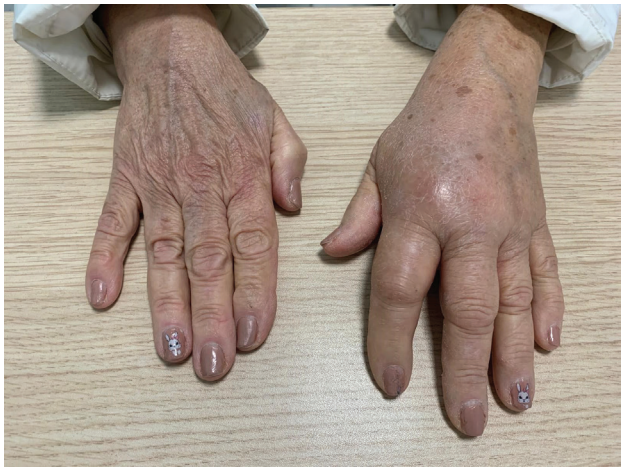
[1]Serviço de Reumatologia, Hospital Beatriz Ângelo, Loures, Portugal, [2]NOVA Medical School, Faculdade de Ciências Médicas, Lisboa, Portugal

Background: Acute monarthritis is a frequent motive for Rheumatology consultation. The differential diagnosis often is based on the distinction between an infectious and an inflammatory cause which can often be rather challenging.

Case Report: A 76-year old woman with diagnosis of hypertension, recent ischaemic stroke, stage V chronic kidney disease of undetermined cause and gout, with an episode of arthritis of the first left metatarsal-phalangeal joint five years ago, was admitted through the Emergency Department in septic shock from a urinary tract infection which warranted urgent induction of dialysis and antibiotic therapy (piperacillin-tazobactam). The patient also reported pain on the second left finger for three weeks with no improvement after being treated with prednisolone 20mg for several days. A strain of multisusceptible Enterococcus faecalis was isolated in the urine culture and the inflammatory markers slowly decreased with therapy: leukocytes 15.730 » 10.280 » 8.170 cells/mm3, c-reactive protein 47.7 » 45.5 » 37.3mg/dL. However, the patient maintained low grade fever and a worsening of the pain in the second left finger, so a request for observation was made to the Rheumatology Department. The physical examination showed redness and diffuse swelling on the second left metacarpal-phalangeal (MCP) and proximal inter-phalangeal (PIP) joints, with important tenderness, most evident on the palmar side, and limitation of flexion and extension of the second digit (Figure 1).

An ultrasound was performed and evidenced significant active synovitis of the second left MCP with multiple small (<1mm) intra-articular hyperechoic foci, a large erosion of the metacarpal bone and intense tenosynovitis of the flexor tendons with severe subcutaneous tissue oedema and numerous hyperechogenic foci – these findings can be suggestive of a microcrystalline disease but don't exclude a septic component. Magnetic resonance imaging of this finger was obtained which documented extensive bone marrow oedema of the second PIP and almost all of the second metacarpal bone, marked second MCP active synovitis, intense tenosynovitis of the flexor tendons of the second finger and oedema of the subcutaneous tissue and superficial muscles.

These exuberant clinical and imagiological findings prompted the possibility of septic arthritis/osteomyelitis, and the patient underwent surgical debridement by the Orthopaedics team who reported an abundant macroscopic presence of devitalised tissue, pus and substantial articular destruction of the MCP joint,



232 – Figure 1. Patient´s hands at admission with intense swelling of the second metacapal-phalangeal and proximal interphalangeal joints

highly suggestive of an infectious process. No agent was identified in the cultures of the material collected during the surgery. Antibiotic therapy was extended for four more weeks during which the pain and inflammation markers quickly subsided. In the follow-up the patient had no pain or inflammatory signs although there was some limitation of flexion of the finger.

Conclusions: This case displays the importance of a high index of suspicion for infectious causes regarding the differential diagnosis of acute monoarthritis. In this patient, even though she had prior history of gout, the location, the exuberant local inflammatory signs, absence of response to glucocorticoids and the recent occurrence of septic shock, were red flags to consider an infection as the cause for arthritis in this patient.

238 - A CASE REPORT OF FOCAL MYOSITIS

Beatriz de Carvalho Mendonça[1], Rita Silva-Vieira[1], Bárbara Lobão[1], Susana Fernandes[1], Helena Santos[1]
[1]Instituto Português de Reumatologia, Lisboa, Portugal

Focal myositis (FM) is a rare inflammatory disease of the skeletal muscle with a benign course, often involving just one muscle in one of the lower limbs. Clinically it presents with acute swelling and myalgia, without muscle weakness. The aetiology remains unknown. It is associated with radiculopathies, muscle trauma, viral infections, immune-mediated diseases, neoplasia and drugs. Creatine kinase level (CK) is normal. Immune checkpoint inhibitors (ICI) are monoclonal antibodies that increase the efficiency of the immune system in destroying neoplastic cells. However, altering the regulation of the immune system can lead to the appearance of immune-mediated adverse events, particularly rheumatic ones such as arthritis or myositis. A case of a patient with FM is presented.

A 63-year-old woman with a history of occult neoplasia diagnosed by diffuse thoracic and bone lymph node metastasis, whose histology was compatible with lung adenocarcinoma. She started immunotherapy with pembrolizumab, which was suspended due to therapeutic failure and alternatively started chemotherapy with paclitaxel. She was also on denosumab 120mg/month, gabapentin 300mg/day, fentanyl 25mg/72h and morphine 2mg/ml in SOS. She presented to the rheumatology consultation, six months after stopping pembrolizumab, for intense pain in the anterolateral aspect of the left thigh with inability to walk, with sudden onset and with 1 week of evolution. There was no history of trauma, muscle weakness or systemic signs and symptoms. On physical examination, she had a supported gait, swelling and intense pain on palpation of the anterolateral aspect of the left thigh, as well as pain on mobilisation of the ipsilateral thigh and knee. There were no changes in the contralateral lower limb. Muscle strength was difficult to assess due to disabling pain. Analytically with slightly elevated inflammation parameters, normal CK and normal thyroid function, negative ANA and negative myositis-specific antibodies. MRI of the left thigh showed hyperfixation in STIR and T2 of the vastus externus, suggestive of myositis of this muscle. She began treatment with acemetacin 180 mg/day and became asymptomatic after three weeks. The diagnosis of FM was assumed.

According to the literature, the FM in this clinical case has several possible aetiologies: paraneoplastic and pharmacological (paclitaxel or pembrolizumab). The active and progressing neoplasia stands out in favour of paraneoplastic. For paclitaxel there are no reported cases, but there are published cases of systemic myositis induced by another taxane (docetaxel). As for pembrolizumab, dozens of cases of systemic myositis have been described, even after discontinuation of the drug and according to some authors, for patients who have undergone ICI in the last 2 years, this aetiology should not be excluded. The recommended treatment for FM is the use of NSAIDs, as they reduce muscle oedema and resolve the clinical condition, as was the case here. A muscle biopsy was not carried out because the patient became asymptomatic, but it would have been useful for a definitive diagnosis.

In short, drug-induced myopathy should be part of the differential diagnosis of non-traumatic myalgia. Oncological therapy, such as ICIs and taxanes, is associated with rheumatic immune-mediated adverse effects and therefore requires a close look from both rheumatologists and oncologists.

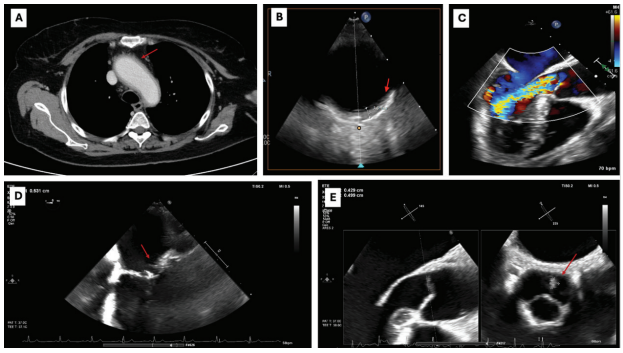
255 - RARE COEXISTENCE OF LARGE VESSEL VASCULITIS AND NONBACTERIAL THROMBOTIC ENDOCARDITIS: A CASE REPORT

Ana Rita Lopes[1, 2], Roberto Pereira da Costa[1, 2], Filipa Marques Costa[1, 2], Bianca Paulo Correia[1, 2], Luís Rosário[3], Cristina Ponte[1, 2]

[1]Serviço de Reumatologia, Unidade Local de Saúde Santa Maria, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Unidade Local de Saúde Santa Maria, Centro Académico de Medicina de Lisboa, Lisboa, Portugal (Serviço de Cardiologia), Lisboa, Portugal

Introduction: Large-vessel vasculitis (LVV) involves inflammation of the large arteries, often leading to vessel occlusion or aneurysm formation. Nonbacterial thrombotic endocarditis (NBTE) is a rare condition primarily associated with hypercoagulable states and malignant diseases, characterized by sterile vegetations on previously undamaged cardiac valves. Here, we present a patient diagnosed with LVV, who also had echocardiographically confirmed vegetations on the mitral and aortic valves, suggestive of NBTE.

Case report: A 61-year-old woman presented to the emergency department with a one-week history of episodes of syncope preceded by generalized weakness, sweating, and dizziness upon standing. Over the past three months, she reported fatigue on minimal exertion, unintentional weight loss (6 kg; 8% of total body weight), intermittent claudication of the lower limbs and tinnitus in the left frontal region. She denied fever, headache, visual symptoms, jaw claudication, or arthralgia. Her background history consisted of long-standing arterial hypertension and an ischemic stroke in the prior three months with minor sequelae (left hemiparesis). On physical examination, her temperature was 38°C, blood pressure 167/67 mmHg on the right and 188/67 mmHg on the left arm, and cardiac auscultation revealed rhythmic S1 and S2 sounds with a diastolic murmur at the aortic focus (grade III/VI) and a left basal systolic murmur (grade III/VI) radiating to the axilla. Laboratory tests showed microcytic/hypochromic anaemia (Hb 12.9 g/dL), minor leukocytosis (11.60x10^9/L), elevated ESR (84 mm/h) and CRP (16.6 mg/dL), and cholestasis (GGT 239 U/L). Anti-nuclear, anti-neutrophil cytoplasmic and anti-phospholipid antibodies and blood cultures were negative. Ultrasound of the temporal and axillary arteries showed no halo sign. Thoracic-abdominal-pelvic CT revealed wall-thickening of the aorta (ascending, aortic arch and proximal descending thoracic) and carotid and vertebral arteries indicative of vasculitis (Figure 1) Transthoracic and transoesophageal echocardiogram revealed multiple hypoechoic masses attached to the aortic and mitral valves, with severe aortic and minimal mitral regurgitation (Figure 1). The diagnosis of LVV with NBTE was established. The patient was started on anticoagulation therapy with enoxaparin 80 mg/day and pulses of methylprednisolone (1g/day for three days), followed by 60 mg/day of oral prednisolone (with gradual tapering) and methotrexate 12.5 mg/weekly. Her symptoms resolved rapidly, and after five months of follow-up, the echocardiogram showed mild aortic regurgitation and no evidence of valve vegetation. Treatment with tocilizumab 162 mg/weekly was initiated after nine months due to glucocorticoid-related adverse events. Presently, at two years of follow-up, the patient is in clinical remission with occasional mild asthenia and decreasing lower limb claudication, managed with prednisolone 5mg/day, methotrexate and tocilizumab.



255 – Figure 1. Imaging of the aorta demonstrating wall thickening, severe regurgitation, and valve vegetation. A) Thoracic computed tomography slice showing wall-thickening of the aorta (arrow); B) Transoesophageal echocardiogram images showing thickening of the aortic wall (arrow), suggesting an involvement of large-vessel vasculitis (LVV); C) Transoesophageal echocardiogram showing severe aortic regurgitation; D) and E) Transoesophageal echocardiographic views of the vegetation at the level of the aortic valve (arrows).

Discussion: NBTE has been linked with various immune-mediated conditions, such as systemic lupus erythematosus, but its association with LVV is exceedingly rare. To the best of our knowledge, only four cases involving these concurrent conditions have been reported to date, all in patients with giant cell arteritis and older than our patient. Additionally, this report marks the first case of NBTE with extensive large vessel vasculitic involvement documented through imaging. Our findings highlight the need for heightened awareness and personalized management strategies in these complex clinical scenarios.

256 - FENÓMENO DE RAYNAUD UNILATERAL COMO SINAL DE ALARME PARA ATEROSCLEROSE GRAVE – UM CASO CLÍNICO

Guilherme Santos Luís[1], Alexandra Daniel[1]
[1]Serviço de Reumatologia, Centro Hospitalar de Leiria, Leiria, Portugal

Doente do sexo masculino com 63 anos, ex-vidreiro, fumador (carga tabágica de 50 UMA), encaminhado à consulta de Reumatologia por fenómeno de Raynaud unilateral da mão direita com 6 meses de evolução, associada a omalgia direita, de ritmo mal definido, com o mesmo tempo de evolução.

Ao exame objetivo, apresentava palidez dos dedos da mão direita, reversível, sem úlceras digitais, pitting scars ou espessamento cutâneo associado. Também se constatou limitação dolorosa da abdução ativa do ombro direito (<90º). Iniciou nifedipina 30mg/dia e tramadol 75mg + paracetamol 650mg (até duas vezes por dia) e pediu-se estudo complementar diagnóstico.

Um mês depois, o doente é novamente observado em consulta sem melhoria com a terapêutica instituída e com agravamento das queixas. Apresentava agora cianose persistente de ambas as mãos e queixas compatíveis com claudicação de ambos os membros superiores. Foi medida a tensão arterial em ambos os membros superiores, apresentando 62/58mmHg no braço esquerdo e imensurável no braço direito (após várias tentativas). Analiticamente, apresentava hemograma sem alterações, parâmetros inflamatórios normais, doseamento de ANAs negativo e hipercolesterolémia com hipertrigliceridémia. Foi realizada capilaroscopia, que não evidenciou padrão esclerodérmico.

Foi realizada Angio-TC dos membros superiores que revelou extensa ateromatose, com placas fibrocalcificadas dispersas a determinar estenose da artéria braquiocefálica, carótida comum, subclávia (com ausência de opacificação ao longo de todo o seu trajecto) e axilar esquerdas e estenose da artéria subclávia direita, ao qual se associa ateromatose coronária e aórtica calcificada.

A PET/CT não revelou sinais de vasculite.

O doente foi diagnosticado com Doença Arterial Periférica dos membros superiores e orientado de forma urgente para consulta de Cirurgia Vascular. Foi também orientado para consulta de desabituação tabágica e medicado com aspirina 100mg id e rosuvastatina 20mg id.

Este caso ilustra como o fenómeno de Raynaud unilateral e de instalação tardia constitui um sinal de alarme clínico que deve sempre conduzir a realização de estudo complementar etiológico. Dentro da multiplicidade de doenças e condições associadas, a doença arterial periférica dos membros superiores é responsável por menos de 5% dos casos.

264 - VEXAS SYNDROME: THE NEW GREAT MIMICKER?

Sofia Ferreira Azevedo[1, 2], Gisela Eugénio[1, 2], Anabela Barcelos[1, 2, 3, 4], Carolina Mazeda[1, 2, 4]
[1]Rheumatology Department, Unidade Local de Saúde da Região de Aveiro, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz Health Alliance, Portugal, Aveiro, Portugal, [3]Comprehensive Health Research Center, NOVA University School, Lisboa, Portugal, [4]EpiDoC Unit, CEDOC, NOVA Medical School, NOVA University, Lisboa, Portugal

Background: VEXAS syndrome is a recently described rare autoinflammatory disorder, caused by mutations in the UBA1 gene on the X chromosome. It is marked by a broad spectrum of manifestations, often mimicking other inflammatory conditions.
Case presentation: A 77-year-old male presented with nodular erythematous limb lesions and asthenia for two months, followed by recurrent fever. His past medical history included a pancreatic neuroendocrine neoplasm submitted to splenopancreatectomy in 2020. Blood analysis revealed elevated inflammatory markers, normocytic anemia, and low-titer ANAs. ANCA and cryoglobulins were negative. Infectious and neoplastic etiologies were excluded. A skin biopsy revealed erythema nodosum and small vessel vasculitis. A few weeks later he experienced frontal headaches and was admitted for further study. Cranial and perinasal CT showed the presence of sinusitis and antibiotics were initiated, with initial clinical improvement. Nevertheless, shortly after being discharged, the patient developed wrist arthritis and bilateral auricular chondritis. Methotrexate (initially 15 mg oral, with the subsequent need to increase to 20 mg subcutaneous) and prednisolone (PDN) (0.5 mg/kg/day) were initiated assuming granulomatosis with polyangiitis/ relapsing polychondritis as the most likely diagnoses. The regimen led to a favorable response, with clinical and analytical improvement, allowing progressive corticosteroid tapering. However, 9 months later (with PDN at 15 mg/day), the patient experienced new episodes of fever, auricular chondritis, and elevated inflammatory markers. The hypothesis of VEXAS syndrome was considered and later confirmed by molecular testing, identifying the pathogenic mutation UBA1 p.Met41Thr. Considering the patient was under treatment with methotrexate and there was no robust evidence for treatment options in this condition, the methotrexate dose was increased to 25 mg subcutaneously. Nevertheless, no clinical or analytical responses were obtained, with maintenance

of elevated inflammatory markers and with recurrence of wrist synovitis. After a multidisciplinary discussion with the Endocrinology and Oncology, weighing the risk-benefit profile and after patient and family explanation and consent, treatment with jak inhibitor Ruloxitinib was initiated.

Conclusion: This case underscores the importance of maintaining high clinical suspicion for VEXAS in patients with symptoms of chronic inflammation, responsive only to moderate-high corticosteroid doses. The recent discovery of VEXAS adds to the complexity of its management, given the absence of established treatment guidelines or robust evidence supporting specific therapeutic interventions. In this case, the choice of ruloxitinib was supported on the wide range of available doses, allowing for dose adjustment according to the clinical response, as well as its demonstrated efficacy (from case reports and case series) compared to other JAK inhibitors. That may result from its specificity for JAK1/JAK2 and strong inhibition of TYK2 activity.

265 - WHEN YOUR FINGERS POINT TOWARDS YOUR CHEST: A DIAGNOSTIC ODYSSEY

Fernando Albuquerque[1], Marcelo Neto[1], Filipa Canhão André[1], Sara Alves Costa[1], Ana Isabel Maduro[1], André Saraiva[1], Cátia Duarte[1, 2]

[1]Serviço de Reumatologia, Unidade Local de Saúde de Coimbra, Coimbra, Portugal, [2]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal

Introduction: Hypertrophic osteoarthropathy (HOA), a syndrome characterized by digital clubbing and periosteal new bone formation, frequently serves as a harbinger of an underlying systemic disease, namely intrathoracic malignancy and, less commonly, inflammatory conditions.

Clinical Case: We report the case of a 29-year-old man presenting with bilateral heel pain and inflammatory arthralgia affecting the wrists, knees and ankles for approximately 4 months. He also reported a significant weight loss. His medical history was unremarkable except for a maternal history of Crohn's disease.

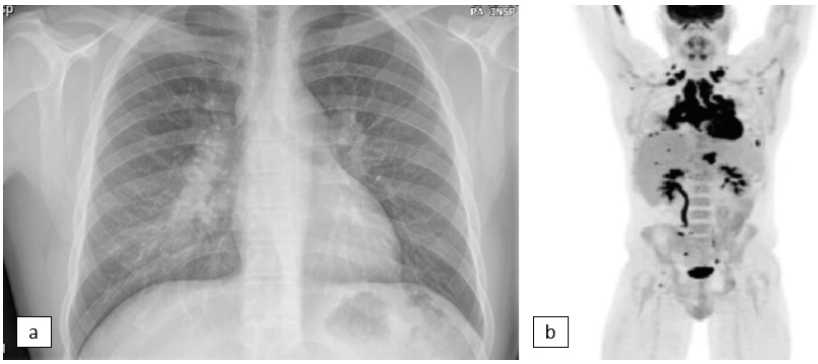
Physical examination revealed mild finger clubbing and bilateral plantar fasciitis. No swollen joints were observed, and the range of motion was normal. Additionally, poorly defined erythematous and warm lesions were present over the anterior surface of the legs.

Blood tests showed mild microcytic/hypochromic anaemia (Haemoglobin: 11,5 g/dL), thrombocytosis (Platelets: 604 G/L), and a high level of ESR (39 mm/h), CRP (8.98 mg/dL), and ferritin (625.2 ng/mL). HLA-B27, anti-CCP, rheumatoid factor, and anti-nuclear antibodies were negative, and the angiotensin-converting enzyme was normal. Bacterial and viral serologies were negative. A thoracic radiography indicated a significant bilateral hilar enlargement (Figure 1a). These findings, along with the presence of signs of HOA raised suspicion of a possible neoplastic condition.

A PET scan was performed, demonstrating multiple extra and intrathoracic adenopathies with bilateral broncho-hilar conglomerates that were intensely hypermetabolic (Figure 1b). There were also areas of lung parenchyma densification in the perihilar region with heterogeneously increased uptake of FDG-F18. These results suggested the diagnosis of sarcoidosis, although a lymphoproliferative disease could not be ruled out.

As a result, the patient underwent an excisional biopsy of cervical adenopathies. Histological examination revealed non-necrotizing epithelioid granulomas with extensive hyaline fibrosis and small lymphocytes without cytological atypia, consistent with Sarcoidosis.

Discussion and Conclusion: This case exemplifies the diagnostic challenges posed by sarcoidosis, since our patient musculoskeletal complaints resembled another inflammatory condition such as spondylarthri-



265 – Figure 1. Chest X-ray and PET scan. Image (a) reveals bilateral hilar enlargment and image (b) shows hypermetabolic adenopathies

tis. However, the presence of subtle digital clubbing, a hallmark of HOA, raised suspicion for an underlying intra-thoracic process. Furthermore, the erythematous lesions on the patient's legs, initially presumed to be nonspecific, probably represented panniculitis lesions, which frequently occurs in conjunction with sarcoidosis as erythema nodosum, especially in the acute presentation of Löfgren syndrome.

This case underscores the importance of recognizing subtle clinical clues, such as HOA and erythema nodosum, in patients presenting with musculoskeletal complaints. A high index of suspicion for underlying systemic disorders, particularly sarcoidosis, is warranted in such cases. Prompt diagnosis and initiation of appropriate therapy are crucial for improving outcomes and preventing long-term complications in patients with sarcoidosis.

267 - SWITCHING TRACKS: RESOLVING EOSINOPHILIA IN ANKYLOSING SPONDYLITIS BY TRANSITIONING FROM ADALIMUMAB TO SECUKINUMAB

Tiago Beirão[1], Catarina Rua[1], Catarina Silva[1], Tiago Meirinhos[1], Taciana Videira[1], Diogo Guimarães da Fonseca[1]
[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Introduction: Eosinophilia may be a rare but important adverse reaction to biologic therapies used in rheumatologic diseases. Ankylosing spondylitis (AS) is a chronic inflammatory disease affecting the axial skeleton. Tumor necrosis factor-alpha (TNF-α) inhibitors, like adalimumab, are commonly prescribed for managing AS. However, these agents can occasionally trigger hypersensitivity reactions, including eosinophilia, which can causes organ damage and hypercoagulability. This report presents a case of a 34-year-old female with AS who developed eosinophilia following treatment with adalimumab, which resolved upon switching to seccukinumab, an interleukin-17A (IL-17A) inhibitor.
Clinical Case: A 34-year-old female with a 2-year history of ankylosing spondylitis presented with worsening back pain and stiffness. Her treatment regimen included full-dose nonsteroidal anti-inflammatory drugs with suboptimal control. She was started on adalimumab, 40 mg subcutaneously every two weeks. After four months, routine blood tests revealed a progressive rise in eosinophil counts, reaching 21680/uL from a baseline of 150 cells/µL, without symptoms of allergic reactions or parasitic infections. Further investigations, including a throughout investigation by hematology and immunoallergology excluded common secondary causes of eosinophilia. Given the temporal relationship between adalimumab administration and eosinophilia, the drug was suspected as the causative agent, with a Naranjo algorithm resulted in "probable". Adalimumab was discontinued, and the patient was transitioned to seccukinumab, 150 mg subcutaneously every four weeks. Following the switch, the patient continued to show good control of AS symptoms. Serial blood tests over the subsequent three months demonstrated a decline in eosinophil count, normalizing to 250 cells/µL, without any recurrence of eosinophilia.
Conclusion: This case highlights a rare instance of adalimumab-induced eosinophilia in a patient with ankylosing spondylitis. Switching to seccukinumab effectively managed the patient's AS while resolving the eosinophilia. Clinicians should be aware of eosinophilia as a potential adverse effect of TNF-α inhibitors and consider alternative therapies, such as interleukin-17A (IL-17A) inhibitor.

269 - EYES AND EARS: COGAN SYNDROME IN A PATIENT WITH RHEUMATOID ARTHRITIS

João Alexandre Oliveira[1], Maria João Cadório[1], Pablo Castro[2], JAP da Silva[1], Mariana Luis[1]
[1]Rheumatology Department, Unidade Local de Saúde de Coimbra, Coimbra, Portugal, [2]Rheumatology Department, Complexo Hospitalario Universitario de Santiago de Compostela,, Santiago de Compostela, Spain

Background: Cogan syndrome (CS) is a very rare chronic inflammatory disorder, characterized by interstitial keratitis and sensorineural hearing loss. Associations with systemic vasculitis and rheumatoid arthritis (RA) have been documented in the literature[1]. Here, we report a case of Cogan syndrome in a patient with long-standing RA.
Case Report: We present the case of a 57-year-old caucasian woman with long-standing, erosive, seropositive rheumatoid arthritis (both rheumatoid factor and anti-citrullinated antibodies) and rheumatoid nodules. She was currently being treated with subcutaneous methotrexate 15mg weekly in monotherapy, in sustained remission within the last year.

In October 2023, she began experiencing acute episodes of red, painful eyes, without visual loss or diplopia, initially affecting the left eye and later the right eye. These symptoms subsided with topical corticosteroids prescribed by an Ophthalmologist but recurred upon treatment discontinuation. She was diagnosed with peripheral ulcerative keratitis in the context of RA (in a specialised Ophthalmology consultation) and received short courses of high-dose oral methylprednisolone and eye lubricants.

In April 2024, she reported rotational vertigo. She

was evaluated in the Emergency Department, underwent a computerised tomography (CT) scan of the head, which showed no abnormalities, and was treated with antihistamines and metoclopramide, without benefit.

In June 2024, she presented with acute and total bilateral hearing loss, first in the left ear and then in the right ear and was admitted to the Otorhinolaryngology ward for further investigation and treatment.

On clinical examination, her vital signs were within normal range. She had difficulty walking due to vertigo and exhibited nystagmus in the left eye. Rinne and Weber tests were consistent with bilateral sensorineural hearing loss. Rheumatological examination showed RA-like deformities but no active synovitis. Ocular examination revealed perilimbal infiltrates, but fundoscopy and neurological examination were normal. Magnetic resonance imaging of the inner ear showed inflammatory signs of the membranous labyrinth bilaterally. Blood tests revealed elevated C-reactive protein at 2,7 mg/dL, erythrocyte sedimentation rate at 60 mm/1 st hour, and mild normocytic anaemia (haemoglobin 11,2 g/dL). Tests for syphilis, angiotensin-converting enzyme, and antineutrophil cytoplasmic antibody were all negative. A diagnosis of Cogan syndrome was made within a week of the hearing losss, and the patient underwent a 3-day course of daily 125 mg methylprednisolone followed by 80 mg/ daily of methylprednisolone. Ocular symptoms showed marked improvement, but unfortunately, there was no improvement in hearing loss.

Discussion: Sudden ocular and audiovestibular manifestations in a patient with long-standing RA (even in sustained remission) should raise the suspicion of associated Cogan syndrome. There is no diagnosis test for Cogan syndome, however recurring cases of keratitis and vestibular symptoms without response to first line therapy are red flags for this condition. Prognosis highly depends on prompt diagnosis and treatment. In this case, treatment delay led to irreversible hearing loss, reinforcing the need to swift action in these circumstances.

REFERENCES

1. Lee L. Y., et al. Interstitial keratitis and sensorineural hearing loss as a manifestation of rheumatoid arthritis: clinical lessons from a rare complication. BMJ Case Reports. 2012;2012

270 - A CASE OF SEVERE GENERALIZED MORPHEA ASSOCIATED WITH NON-CIRRHOTIC PORTAL HYPERTENSION

Carolina Ochôa Matos[1, 2], Cláudia Brazão[3], Francisco Capinha[4], Pedro de Vasconcelos[5], Luís Soares de Almeida[3, 5], Gonçalo Boleto[1, 2], Nikita Khmelinskii[1, 2]

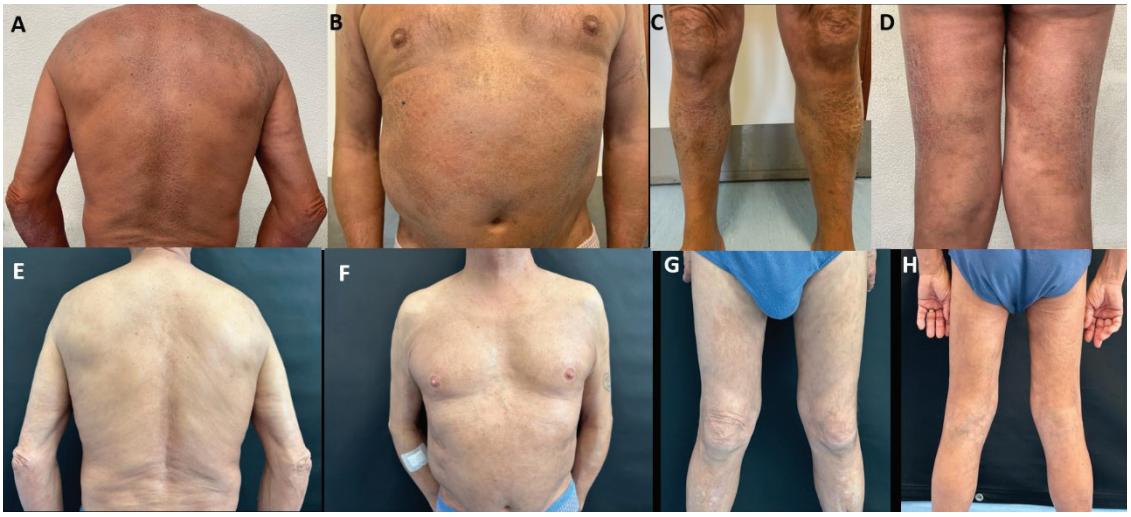
[1]Serviço de Reumatologia, Unidade Local de Saúde Santa Maria, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Serviço de Dermatologia, Unidade Local de Saúde Santa Maria, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [4]Serviço de Gastrenterologia e Hepatologia, Unidade Local de Saúde Santa Maria, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [5]Laboratório de Dermatopatologia, Serviço de Dermatologia, Unidade Local de Saúde Santa Maria, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introduction: Generalized morphea is a rare inflammatory connective tissue disorder, characterized by at least four inflammatory patches and/or bands of thickened skin, more than 3cm in diameter, affecting two or more anatomic sites. Non-cirrhotic portal hypertension (NCPH) is characterized by elevated portal pressures without cirrhosis and it is often a manifestation of an underlying systemic disease. We report a case of severe, refractory, generalized morphea associated with NCPH.

Clinical vignette: A 72-year-old-man presented to the emergency department (ED) with a four-month history of multiple, coalescent, indurated plaques, with overlying erythema and scaling, that began on the upper limbs with subsequent fast progression to the trunk and legs, in a symmetrical distribution, sparing the face, hands and feet (figure 1 A-D). These lesions caused important functional disability. He had no history of Raynaud's phenomenon, constitutional, respiratory or gastrointestinal symptoms.

Laboratory test results were notable for elevated erythrocyte sedimentation rate (103 mm/h) and C-reactive protein (5.21 mg/dL). Antinuclear antibodies and systemic sclerosis-specific autoantibodies were negative. The body computed tomography scan, echocardiogram, upper and lower gastrointestinal endoscopies findings were unremarkable. Nailfold capillaroscopy showed non-specific abnormalities. The skin biopsy corroborated the clinical diagnosis of generalized morphea. At the time of diagnosis, the Localized Scleroderma Skin Activity Index (LoSAI) was 73/162, the Localized Scleroderma Skin Damage Index (LoSDI) 60/216, Physician's Global Assessment of disease activity (PGA-A) 90/100 and of damage (PGA-D) 75/100.

Treatment was started with topical corticosteroids, prednisolone 60mg/day and methotrexate (MTX), titrated up to 25mg/week subcutaneously. Following mild improvement, after 4 months of treatment and tapering of prednisolone to 20mg/day, the disease relapsed. MTX was switched to mycophenolate mofetil (MMF) 2g/day and bridge therapy with intravenous immunoglobulin (IVIg) 2g/kg monthly, given glucor-



270 – Figure 1. A-D: Morphea lesions in the trunk and limbs before treatment; E-H: Improvement after a year of treatment with IVIg.

ticoid induced toxicity. MMF was suspended after 6 months due to new-onset moderate thrombocytopenia. Two months after its suspension, the patient was admitted to the ED with upper digestive hemorrhage from esophageal variceal rupture. After extensive investigation, NCPH with moderate splenomegaly was diagnosed. Transjugular liver biopsy was compatible with porto-sinusoidal vascular disease and common secondary causes were excluded. He initiated treatment with endoscopic variceal ligation and a beta-blocker (carvedilol), with clinical stability, but maintained thrombocytopenia (90-100 x 10 9 /L), which we considered to be due to splenomegaly and not MMF-induced.

Currently, he maintains treatment with IVIg every 6 weeks and low-dose prednisolone (5mg/day), with remarkable improvement, and sustained remission after a year (figure 1 E-H). The current LoSAI is 0/162, LoSDI 19/216, PGA-A is 0/100 and PGA-D is 20/100, maintaining only residual hyperpigmentation and skin thickness in the trunk, forearms and legs.

Conclusion: To the best of our knowledge, we present the first case of generalized morphea and NCPH. While no direct correlation can be definitely established, all other known secondary causes were excluded. Additionally, our report also highlights significant and sustained skin improvement with IVIg monotherapy, with a favourable risk profile, adding to the limited evidence of alternative treatments against morphea.

272 - COMPLEX REGIONAL PAIN SYNDROME: NAVIGATING A WORLD OF UNCERTAINTY

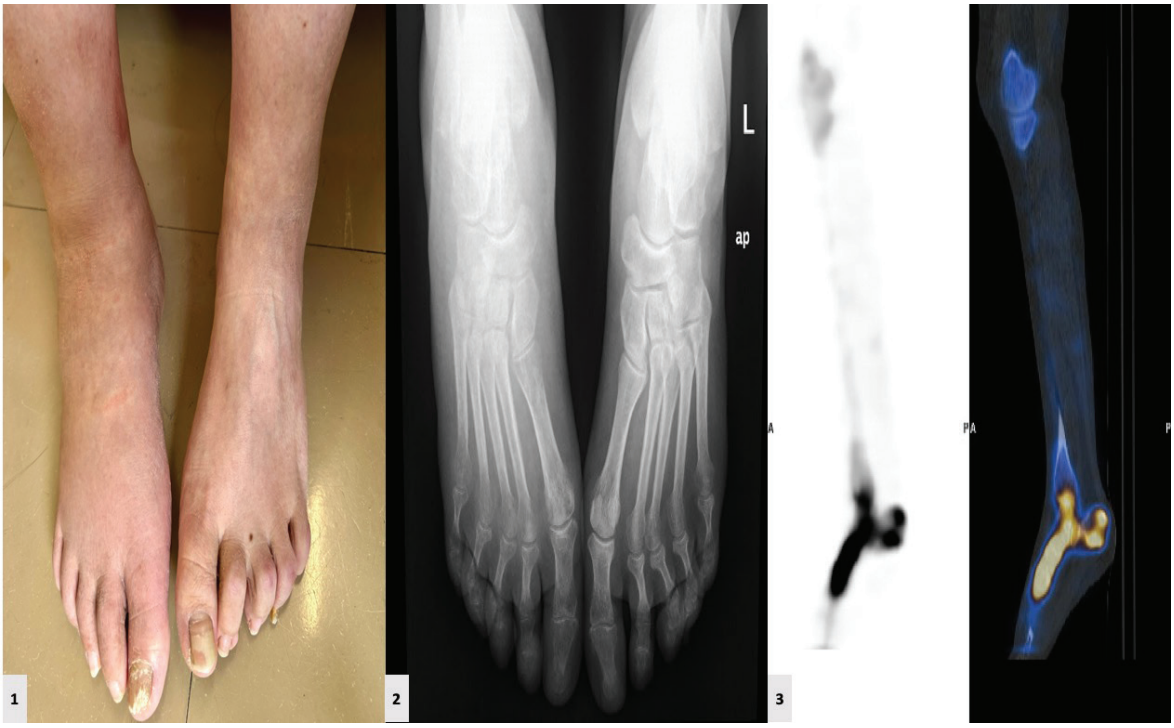
Filipa Canhão André[1], Fernando Albuquerque[1], Sara Alves Costa[1], Marcelo Neto[1], Adriana Carones[1], Beatriz Mendes[1], Ana Isabel Maduro[1], André Saraiva[1], Margarida Coutinho[1, 2], Tânia Santiago[1, 2], Maria João Salvador[1, 2]

[1]Serviço de Reumatologia, Unidade Local de Saúde de Coimbra, Coimbra, Portugal, [2]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal

Introduction: Complex Regional Pain Syndrome (CRPS) is a debilitating chronic pain condition, typically affecting a limb, characterized by diverse autonomic, inflammatory, sensory and vasomotor symptoms. It is classified into two types: CRPS type I, occurring without identifiable nerve injury and CRPS type II, associated with peripheral nerve injury. We present a case illustrating the challenges of diagnosing and managing CRPS type II.

Clinical Case: A 60-year-old woman presented with six months of escalating pain in her right forefoot, described as electric shocks and warmth, accompanied by fluctuating swelling and color changes. Symptoms progressed to the entire foot and ankle, leading to numerous misdiagnoses and ineffective treatments. Notably, the patient had a history of right foot drop due to sciatic nerve injury during a prior total hip arthroplasty. Examination revealed marked hyperesthesia in the dorsum of the foot. A bone scintigram showed increased uptake in all foot and ankle bones and ultrasound revealed soft tissue edema with synovitis of the tibiotalar and other tarsal joints. These findings supported the diagnosis of CRPS.

Pharmacological therapy, including antidepressants, neuroleptics, corticosteroids (1 mg/kg/day) and weak opioids, was optimized but provided minimal relief. A single intravenous dose of pamidronate and rehabili-



272 – Figure 1. 1 – Red and swollen right foot; 2 - Radiograph with soft tissue; 3 - Scintigram with increased bone metabolism

tation were initiated, but the pain persisted, hindering patient adherence to therapy. In consultation with the pain clinic, a peripheral nerve block was offered as a last resort, resulting in significant symptom improvement and increased functional capacity.

Conclusion: CRPS presents a complex diagnostic and therapeutic challenge, with poorly understood pathophysiology and limited effective treatment options. This case emphasizes the importance of early recognition and multidisciplinary collaboration involving rheumatology, physiatry and anesthesiology. It also highlights the potential for iatrogenic nerve injuries that require patient education and informed consent and the potential benefits of peripheral nerve blocks in refractory cases. Further research is crucial to elucidate the mechanisms of CRPS and to develop targeted therapies.

274 - STRIKING THE BALANCE: RISK OF MACROPHAGE ACTIVATION SYNDROME FLARE IN STILL'S DISEASE AFTER TREATMENT SUSPENSION

Filipa Canhão André[1], Fernando Albuquerque[1], Sara Alves Costa[1], Maria João Cadório[1], João Alexandre Oliveira[1], Ana Isabel Maduro[1], André Saraiva[1], Tânia Santiago[1, 2], Maria João Salvador[1, 2], Margarida Coutinho[1, 2]

[1]Serviço de Reumatologia, Unidade Local de Saúde de Coimbra, Coimbra, Portugal, [2]Faculdade de Medicina, Universidade de Coimbra, Coimbra, Portugal

Introduction: Still's disease (SD) often requires lifelong treatment, even in patients in remission. However, adjustments or temporary cessation of therapy may be required during infections or cancer treatment. The balance that leads to remission can be easily disrupted, leading to severe complications. We present a case of macrophage activation syndrome (MAS) in a patient with SD following the interruption of anakinra due to influenza A infection.

Clinical Case: An 18-year-old female with SD in clinical remission on anakinra for at least 6 months, presented with fever, cough and generalized myalgia. Influenza A was diagnosed, prompting the temporary discontinuation of anakinra. One week later the fever persisted and the patient deteriorated, developing multi-organ dysfunction and requiring intensive care admission. Laboratory findings revealed elevated ferritin (60238 ng/mL) and D-dimer (94170 ng/mL), anemia (hemoglobin 7.5 g/dL), hypofibrinogenemia and hypertriglyceridemia. The clinical picture, along with a high HScore (probability >99%), supported the diagnosis of MAS. Prompt initiation of methylprednisolone pulses, followed by prednisolone (1 mg/kg/day) and a higher dose of anakinra (200 mg/daily), resulted in a favorable response.

Conclusion: This case raises concerns about the need for treatment discontinuation during infections in SD patients. While anakinra's rapid onset and short half-

life may be advantageous, the risk of worsening infection must be balanced against the potential for disease flare. As there is a lack of clear guidance on this matter, a case-by-case approach is recommended, with expert consultation. Further research is needed to guide the management of acute infections in patients with SD treated with biological therapy. In the meantime, vigilant monitoring is crucial to prevent and manage potential complications.

280 - HEPATITE LÚPICA: CASO CLÍNICO

Catarina Silva[1], Edgar Afecto[2], Tiago Beirão[1], Catarina Rua[1], Tiago Meirinhos[1], Taciana Videira[1], Romana Vieira[1], Joana Abelha-Aleixo[1], Flávio Costa[1], Diogo Fonseca[1], Ana Sofia Pinto[1], Beatriz Samões[1], Patricia Pinto[1]

[1]Serviço de Reumatologia, Unidade Local de Saúde de Gaia e Espinho, Vila Nova de Gaia, Portugal, [2]Serviço de Gastroenterologia, Unidade Local de Saúde de Gaia e Espinho, Vila Nova de Gaia, Portugal

Introdução: Alterações bioquímicas da função hepática são comuns no Lúpus Eritematoso Sistémico (LES), sendo encontradas em até 50% dos doentes em algum momento do curso da doença. A hepatite lúpica (HL) é considerada uma entidade distinta descrita em cerca de 3 a 8% dos doentes com LES que se caracteriza por uma disfunção hepática subclínica, mais frequentemente associada a uma elevação ligeira a moderada das transaminases. O diagnóstico de HL é amplamente considerado um diagnóstico de exclusão.
Descrição do caso: Doente do sexo feminino, 34 anos. Encaminhada para a consulta de Reumatologia em 2013 para seguimento de um LES conhecido desde os 15 anos, que se encontrava em remissão. Em consulta de seguimento nesse mesmo ano, descreve quadro de fadiga e desconforto abdominal, com documentação de elevação ligeira das transaminases (AST/ALT 54/65U/L), da gama-glutamil transpeptidase (GGT 121U/L) e das fosfatase alcalina (FA 148U/L). Melhoria clínica após introdução de corticoide, contudo sem melhoria analítica, pelo que realiza ecografia abdominal, que não evidencia alterações de relevo. Inicia azatioprina 100mg por agravamento articular, com normalização do perfil hepático, à exceção da GGT.

Do ponto de vista analítico, manteve elevações flutuantes das enzimas hepáticas de padrão misto, com períodos de normalização. Os valores mais elevados foram documentados em 2020, altura em que repete ecografia hepática, que evidencia apenas esteatose hepática de grau moderado. Suspende temporariamente azatioprina, sem melhoria da disfunção hepática. Estudo imunológico com padrão flutuante de IgG e positividade para ANA, anticorpos anti-centrómero B, anti-RNP e anti-Sm, anti-SMA/F-actina. Foram excluídas infeções víricas.

Referenciada a consulta de Gastroenterologia em 2023 por suspeita de quadro de HL ou hepatite autoimune (HAI). Biopsia hepática evidencia inflamação peri-portal ligeira, necrose hepatocitária com esboço de septos, fibrose portal e peri-portal ligeira e esteatose focal. Inicia tratamento com 20mg de cortisona, tendo sido atingida remissão bioquímica ao fim de 3 semanas, sem possibilidade de redução progressiva da dose de corticoide por episódio de agravamento articular e cutâneo da doença.
Discussão: O diagnóstico diferencial de HL deve considerar outras causas de disfunção hepática nos doentes com LES, como fármacos, álcool, vírus, metabólicas, vasculares ou autoimunes. A HL e a HAI são duas condições imunológicas que envolvem o fígado que se podem apresentar de forma semelhante. A apresentação clínica é geralmente insidiosa, com sintomas inespecíficos, como fadiga, anorexia, desconforto abdominal e náuseas. A nível laboratorial, apesar de ambas poderem cursar com elevações de IgG e ANA positivos, o anticorpo anti-proteína P ribossomal é considerado um marcador de HL, enquanto a positividade para os anticorpos anti-SMA/F-actina, anti-LMK-1, anti-LC1 e anti-SLA/LP apoia o diagnóstico de HAI. A histologia pode ser útil na sua distinção: são considerados achados típicos de HAI a presença de hepatite de interface com infiltrado predominantemente linfoplasmocitário, emperipolese e rosetas hepatocitárias, enquanto a HL apresenta geralmente infiltrado lobular ou ocasionalmente periportal com apenas alguns linfócitos. Um diagnóstico preciso é fundamental, uma vez que a HL é uma condição benigna que geralmente responde a corticoides sem complicações e a HAI está associada a um mau prognóstico se não tratada, taxas mais altas de progressão e necessidade de uma abordagem terapêutica mais agressiva.