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Casos Clínicos

022 - VARICELLA ZOSTER VIRUS MIMICKING GIANT CELL ARTERITIS: THE IMPORTANCE OF ULTRASOUND IN THE DIFFERENTIAL DIAGNOSIS

Martins-Martinho J[1, 2], Inês Pintado Maury[3], Inês Leal[4], Cristina Ponte[1, 2]

[1]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Department of Infectious Diseases, Hospital de Santa Maria, CHLN, Lisbon Academic Medical Centre, Lisboa, Portugal, [4]Serviço de Oftalmologia, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introduction: Giant cell arteritis (GCA) can lead to blindness in up to 20% of cases, most frequently due to arteritic anterior ischemic optic neuropathy (AION). Ultrasound of the temporal (TA) and axillary (AX) arteries is recommended as the first imaging modality in cases of suspected GCA given its high diagnostic sensitivity and specificity. Other differential diagnoses should be considered when the ultrasound is negative, including other causes of anterior neuropathy, such as infectious or non-arteritic ischemic.

Case Report: An 81-year-old woman presented to the Emergency Department with a 3-day painless and sudden vision loss in the right eye (RE). She reported having had an episode of acute anterior uveitis in the 6 months prior, treated with topical glucocorticoids (GCs) and cycloplegics for 3 months with resolution. She denied headaches, jaw claudication, weight loss and polymyalgia rheumatica symptoms. Her background history included arterial hypertension, dyslipidemia and bilateral cataract surgery at 79 years old. On ophthalmic examination, the patient's best corrected visual acuity (BCVA) was counting fingers at 50cm in the RE and 1.0 in the left eye (LE), with relative afferent pupillary defect in the RE. The intraocular pressure (IOP) was increased (26mmHg) in the RE and normal (13mmHg) in the LE. On biomicroscopy, the RE presented mild cilioconjuntival hyperemia, mild corneal edema, and granulomatous keratic precipitates in the lower third of the cornea, with no cells in the anterior chamber. The RE fundus examination showed a paled edematous optic disc. These findings suggested AION overlapping with earlier anterior uveitis with papillitis. The remaining physical examination was unremarkable. The ESR was 64mm/hr and C-reactive protein was 1.1mg/dL. Orbits and cranial computed tomography showed ischemic microangiopathic leukoencephalopathy. The patient was started on 60mg/day of oral prednisolone, valacyclovir 1g tid, sulfamethoxazole and trimethoprim bid, and topical treatment for the RE with tropicamide, dexamethasone, levofloxacin and lowering-IOP drops. An ultrasound of TA and AX arteries was performed and only identified hyperechoic wall thickening of the TA suggestive of atherosclerosis. Thus, a TA biopsy was requested. On further enquiry the patient reported having had an episode of painful blisters on the skin with spontaneous resolution in the 2 months prior. The dose of valacyclovir was therefore increased to 2g tds and the GCs tapered 5mg every 5 days. The optic disc edema improved but the BCVA remained at counting fingers. Serology results were positive for type 2 herpes simplex virus (IgM and IgG) and negative for HIV, toxoplasma, HBV, HCV and VDRL. IGRA and ANCA were negative and ACE was normal. Aqueous humor analysis revealed positive DNA for varicella-zoster virus (VZV) and the TA biopsy was negative for GCA. The final diagnosis was VZV optic neuritis. After completing treatment, prophylaxis was maintained. The patient stopped prophylaxis after 1 year and, after 3 months, relapsed with LE optic neuropathy. Repeated TA and AX ultrasound was negative for GCA. The diagnosis of sequential VZV optic neuropathy was established and

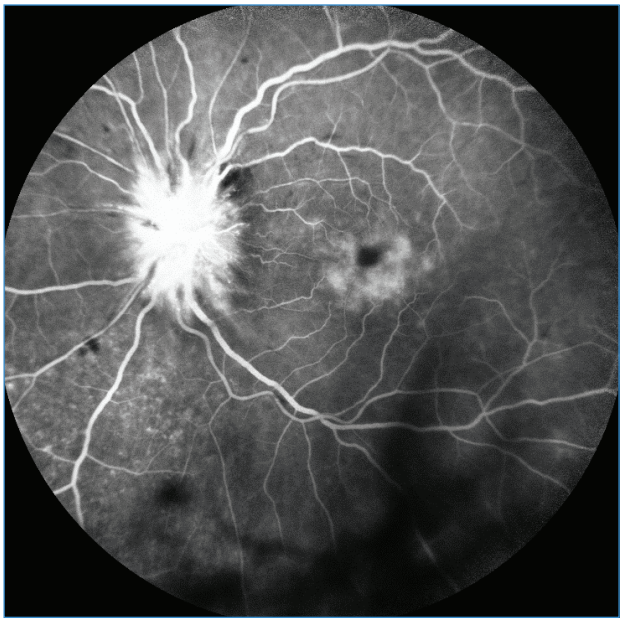

Figure 1. LE fluorescein angiography with contrast diffusion from the optic disc and macular region

valacyclovir restarted ad aeternum.

Conclusion: This case highlights the importance of ultrasound in assessing a suspected GCA. Although GCA was initially considered the most probable diagnosis, the negative ultrasound prompted a more profound investigation that ultimately led to the identification of the correct diagnosis and prevented diagnostic delay and unnecessary GC toxicity.

029 - ANTIPHOSPHOLIPID ANTIBODIES-ASSOCIATED DIFFUSE ALVEOLAR HEMORRHAGE

Sara Paiva Dinis [1], Ana Sofia Pinto[2], Filipe Cunha Santos[1], Duarte Augusto[1], Filipa Fernandes [3], Nathalie Madeira[1], Cláudia Vaz[1], Joana Fonseca Ferreira[1]

[1]Serviço de Reumatologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar Vila Nova de Gaia/ Espinho, Gaia, Portugal, [3]Serviço de Pneumologia, Unidade Local de Saúde da Guarda - Hospital Sousa Martins, Guarda, Portugal

Primary antiphospholipid syndrome (PAPS) is an acquired prothrombotic condition characterized by vascular thrombosis and/or obstetric morbidity in the presence of antiphospholipid antibodies (aPL) (1). While thrombosis is the most common cause of lung injury in PAPS, there are a growing number of case reports describing lung pathology in the absence of thrombosis, including diffuse alveolar hemorrhage (DAH) due to pulmonary capillaritis, suggesting that aPL can lead to tissue injury through means other than thrombosis (1,2). We describe a case of a 45-year-old, previously healthy, nonsmoker female who presented to the emergency department complaining of hemoptysis, dyspnea and chest pain for 1 month, worsening in the last few days. On presentation she was polypneic with a decreased breath sound and had a room air oxygenation of 83% with hypoxemic respiratory failure. She had microcytic anemia, normal platelets, white blood cell counts and urinalysis. PTT, TP and INR were normal. Chest radiographs (fig. A) and computed tomography (CT) (fig. B) showed diffuse bilateral alveolar infiltrates in a ground-glass pattern. Bronchoscopy showed DAH and no evidence of infection. She was diagnosed with DAH, and further investigation showed an IgG anticardiolipin antibody (ACA) titer of 233.8 units (U) (normal<20U), an IgM ACA of 14.4U (<3.5U), titers of IgG and IgM antibodies against β-2-glycoprotein I (β2GPI) elevated at greater than 20U (<20U) and a positive lupus anticoagulant. She had a positive antinuclear antibody titer (1:160) but remaining immunology panel negative, namely anti-double stranded DNA, anti-neutrophil cytoplasmic, anti-glomerular basement membrane and anti-citrullinated peptide antibodies. Rheumatoid factor was negative and complement levels C3 and C4 were normal. She was treated with methylprednisolone 1000mg daily for 3 days, followed by high-dose prednisone (PDN). Within 5 days she had clinical improvement and near-complete resolution of radiographic findings and mycophenolate mofetil (MMF) was started and up-titrated to 1,5 g. Two weeks after hospital discharge, she had recurrence of dyspnea and hemoptysis and worsening of radiographic findings. Pulsed high-dose methylprednisolone was repeated for 3 days with good response and the patient was discharged with MMF 3 g daily and PDN 60 mg daily. When PDN was lowered to 40 mg daily, the patient had recurrence of HAD and it was decided to start intravenous rituximab 1 g infusion, which was repeated 2 weeks later, with clinical and radiographic improvement. One month later, she was transitioned to a tapering PDN dose and was on maintenance treatment with MMF 3 g daily and hydroxychloroquine 400mg daily, with no recurrence of DAH. We describe a case of DAH in the setting of high titers of aPLs and no evidence of acute thrombosis or other etiologies of DAH. While DAH from PAPS or aPL appears to be unusual, physicians need to be aware of this entity and its associated morbidity. Further elucidation of a possible nonthrombotic mechanism of aPL-mediated pathology and of the real importance of a positive aPL in clinical situations as DAH is needed to guide future therapies for this unusual manifestation of PAPS.

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030 - DIAGNOSTIC DILEMMA – VERTEBRAL FRACTURES IN A "BAMBOO SPINE": CASE REPORT

Mariana Rocha Sebastião[1], Luís Maurício Santos[1], Teresa Novoa[1]

[1]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal

Introduction: Ankylosing spondylitis (AS) is associated with an increased risk of vertebral fracture (VF), that aggravates every year after the diagnosis. Low energy trauma is the most common cause, in relation to the high prevalence of osteoporosis in AS. These fractures are often unstable and associated with clinically relevant spinal cord injury. The majority of VF are

not identified in plain radiographs, which contributes to delayed diagnosis and may ultimately aggravate the neurologic complications.

Case Report: A 61-year-old man with a 30-year history of AS presented to the emergency department (ED) with acute low back pain after a fall from his own height, 2 days prior to the visit. Dorsal and lumbar spine radiographs showed severe vertebral body fusion and spinal ligament ossification – "bamboo spine". No fractures were found, and in the absence of neurological symptoms, he was discharged. 5 days later he developed bilateral leg weakness, while still maintaining gait ability. 11 days after the fall he was readmitted in the ED with complete loss of motor and sensory functions of both legs and acute urinary retention. On examination a vesical globe was palpated, and patellar and Achilles tendon reflexes were absent. After consultation with his Rheumatologist, a dorso-lumbar CT was performed, revealing fracture of the D9-D10 longitudinal anterior ligament and posterior column (bilateral lamina and pars interarticularis, and right transverse process). An MRI then revealed an epidural hematoma (EH) from D8 to D10 compressing the medulla. The patient underwent percutaneous fixation, laminectomy and EH drainage with success. At discharge he presented an ASIA B classification and was transferred to an inpatient rehabilitation centre.

Conclusion: AS patients have a fourfold VF risk when compared to the general population. The ossification of spinal ligaments and surrounding tissues, and vertebral body fusion lead to biomechanical changes of the vertebral column. For instance, it may turn the spine into "long lever arms" on which forces can act during trauma, potentially aggravating it's resulting injuries. There is a markedly higher prevalence of VF associated neurologic complications in AS vs unaffected individuals. Late diagnosis, unprotected transfers and development of epidural hematoma can result in secondary motor or sensory deterioration. The pathologic osseous changes in AS make spinal radiographs difficult to interpret and these may very well not reveal a non-displaced fracture. In addition, patients frequently present multiple noncontiguous lesions. For these reasons several authors recommend the use of CT and/or MRI of the entire column.

Images:

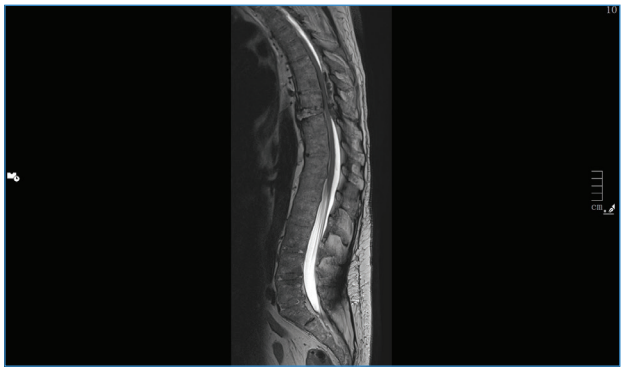

Figure 1. Spinal MRI

035 - SYSTEMIC VASCULITIS EXPOSED BY A NEW (NOT SO SECRET) AGENT

Frederico Rajão Martins[1], Rodrigo Rei[1], Vítor Teixeira[1]
[1]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, Faro, Portugal

Introduction: Dupilumab is an anti-IL-4 and anti-IL13 receptor monoclonal antibody with several indications in allergology and dermatology, such as moderate to severe asthma (isolated or in association with severe atopic dermatitis), eosinophilic esophagitis, prurigo nodularis and rhinosinusitis with nasal polyposis. Recently cases of new onset ANCA-positive eosinophilic granulomatosis with polyangiitis (EGPA) in patients under dupilumab were published (1,2,3).

Clinical case: A 45-year-old man with a history of adult-onset asthma, rhinitis and nasal polyposis with multiple surgical excisions was treated with long-term steroids at a minimum dose of 4 to 8mg q.i.d. methylprednisolone. He received treatment with dupilumab with significant clinical improvement of asthma and rhinitis and consequently steroid withdrawal. Three weeks after that, he presented with lower limb palpable purpuric lesions on the anterior aspect of the lower limbs, popliteal fossae and waist, associated with one night of fever-associated symptoms. No known cardiac issues, neurologic symptoms or urine alterations were reported. Previous studies included a paranasal sinuses CT scan showing multiple bilateral polyps and sinus mucosal thickening of probable inflammatory aetiology. Pulmonary function tests showed mild obstructive changes. Blood tests showed mild increase of eosinophil count (1320 cells/µL, 13.7%), slight elevation of CRP 12.5 mg/L (<5 mg/L) with normal ESR, no abnormalities regarding ANA, RF, ANCA. Lower limb electromyography revealed no sensorimotor neuropathy.

Considering palpable purpura as a surrogate of vasculitis, this patient could fill ACR/EULAR 2022 classification criteria for EGPA. The patient was reinitiated on methylprednisolone 4 mg with complete resolution of skin lesions. Treatment switch to mepolizumab, which is approved for the treatment of EGPA, was not yet considered due to good response to dupilumab and because the vasculitis flare was considered minor and responded to low-dose steroids.

Conclusion: Whether or not dupilumab had a directly effect on the development of purpura in our patient

or whether it unmasked an EGPA by allowing steroid withdrawal is not yet known. In the past, drugs used for asthma such as montelukast were thought to cause EGPA, but the occurrence of EGPA in such patients appears to be related to unmasking of an underlying vasculitic syndrome that was initially clinically recognized as just a steroid dependent moderate to severe asthma. We believe that the same principle may apply to dupilumab.

In fact, other more severe cases of EGPA following dupilumab initiation have been described, with variable courses described after treatment discontinuation.

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053 - LARGE GRANULAR LYMPHOCYTIC LEUKEMIA AND RHEUMATOID ARTHRITIS: EXPERIENCE OF A SINGLE CENTER REGARDING THREE CLINICAL CASES

Mariana Diz-Lopes [1, 2], Carlos Marques Gomes[1, 2], Inês Santos[3], Teresa Martins-Rocha[1, 2], Miguel Bernardes[1, 2], José Pinto[1], Lúcia Costa[1]

[1]Rheumatology Department, Centro Hospitalar Universitário de São João, Porto, Portugal, [2]Department of Medicine, Faculty of Medicine, University of Porto, Porto, Portugal, [3]Rheumatology Unit, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Introduction: Large Granular Lymphocytic Leukemia (LGLL) is a rare lymphoproliferative disorder with a peculiar association with rheumatoid arthritis (RA). The most common feature is neutropenia and patients can have splenomegaly, resembling Felty's Syndrome. These diseases have similar clinical and laboratory abnormalities, but the diagnosis of T-cell LGLL (T-LGLL) requires evidence of clonality. We present three cases of LGLL associated with RA from a single center in the last 10 years.

Case 1: A 82-year-old man had a diagnosis of RA for 12 years, negative for rheumatoid factor (RF) and positive for anti-cyclic citrullinated peptides (ACCP). He was on methotrexate (MTX) 12,5mg/week. He was admitted in the rheumatology department for investigation of newly developed thrombocytopenia and leucopenia, with severe neutropenia (270/mm3). Stopping methotrexate did not improve the full blood count. Flow cytometric immunophenotyping of a bone marrow aspirate was performed, identifying a population of aberrant cytotoxic T-lymphocytes (Table 1). The T-cell receptor (TCR) rearrangement analysis confirmed monoclonality. He started cyclosporine but the treatment was stopped 1 month later due to renal toxicity. The follow-up had multiple infectious intercurrences, and he required chronic treatment with granulocyte colony stimulating factor (GCS-F). He died 7 years later with a severe pneumonia.

Case 2: A 51-year-old woman had a three-year history of polyarthritis of small joints and was diagnosed with RA in 2017. She was RF positive and ACCP negative. MTX was started but had to be later stopped due to hepatic toxicity with concomitant treatment with isoniazid for latent tuberculosis (LT). Six months after, she developed leucopenia with neutropenia (490/mm3). Toxic etiology was discarded after stopping rifampicin she was then taking for LT treatment. She had no fever and no history of recent infections. T-LGL count in peripheral blood smear was 0,59x10^9/L. Lymphocyte surface markers on flow cytometry reflected a constitutively activated T-cell phenotype. Flow cytometry analysis of the TCR variable region allowed for the presumption of monoclonality. She was started on MTX 20mg/week with complete normalization of blood count after four months. Two years later, increased RA disease activity required starting Rituximab, achieving remission.

Case 3: A 55-year-old man with a diagnosis of seropositive RA for 10 years was admitted in the rheumatology department due to newly identified pancytopenia. Blood analysis on admission showed anemia, leucopenia with neutropenia (630/mm3), thrombocytopenia and elevation of c-reactive protein. He was taking leflunomide 10mg/day and the wash out with cholestyramine had no improvement in full blood count. Abdominal ultrasound revealed splenomegaly. Peripheral blood smear showed a T-LGL count of 0.53x10^9/L with an immunophenotype analysis compatible with T-LGLL. Flow cytometry analysis of the TCR favored monoclonality. Starting of MTX was delayed due to several infections, and he required treatment with GCS-F.

Discussion: RA usually precedes the development of T-LGLL and patients are commonly seropositive and have a long-standing disease. In these three RA patients, the development of neutropenia prompted exclusion of infectious and toxic etiologies, and the diagnosis of T-LGLL was made with consistent clinical and hematologic features, immunophenotypic markers and evidence of monoclonality. T-LGLL associated

TABLE 1. RELEVANT CLINICAL AND ANALYTICAL DATA FROM THE THREE PATIENTS.

	Case 1	Case 2	Case 3
Duration of RA (years)	12	3	10
RF positive RF titer (IU/mL)	No	Yes 300	Yes 320
ACCP positive ACCP titer (U/mL)	Yes 3540	No	Yes 351
Erosive disease	No	No	No
Splenomegaly	No	No	Yes
Hematologic features			
Hemoglobin (g/dL) REF: 13.0-18.0 g/dL	14,6	13,2	11,4
Platelets (x10^9/L) REF: 150-400x10^9/L	122	150	124
Leucocytes (x10^9/L) REF: 4.00-11.00 x10^9/L	5,3	2,1	1,5
Lymphocytes (x10^9/L)	4,2	1,2	0,9
Neutrophils (x10^9/L)	0,27	0,49	0,12
T-LGL (x10^9/L)	2,28	0,59	0,53
Immunophenotypic characteristics of cytotoxic (CD3 + CD8 +) T-lymphocytes	Bone marrow aspirate: CD57+, CD5+ diminished, T-Cell Receptor alpha/beta + CD28-, CD45R0-, CD16-	Peripheral blood smear: CD57+, CD5+ diminished, CD45 R0+, T-Cell Receptor alpha/beta + CD28-, CD16-	Peripheral blood smear: CD57+, CD5+ diminished, T- Cell Receptor alpha/beta + CD28-, CD45R0-, CD16-
Evidence of monoclonality	TCR genes PCR rearrangement analysis	Flow cytometry analysis of TCR variable region	Flow cytometry analysis of TCR variable region TCR genes PCR rearrangement analysis – in process
Treatment of LGL-T leukemia	Cyclosporin A	Methotrexate	Methotrexate
Response to treatment	No response - Death	Hematologic Complete Response*	Not evaluated**

RA: Rheumatoid Arthritis; RF: Rheumatoid Factor; ACCP: anti-cyclic-citrullinated peptides; REF: reference values; T-LGL: T-Cell Large Granular Lymphocytes; TCR: T-Cell Receptor; PCR: polymerase chain reaction.
*Hematologic Complete Response is defined as the complete normalization of blood counts (hemoglobin > 12g/dL; platelets > 150 x 109/L; absolute neutrophil count > 1.5x109/L and lymphocytosis < 4 x 109/L) and circulating LGL in the normal range (0,25x109/L).
** The response is evaluated after 4 months of starting therapy.

with RA should be treated with immunosuppression. Disease-related death is most commonly due to severe infections.

063 - PATHERGY IN DISSEMINATED GONOCOCCAL INFECTION

Roberto Costa[1, 2], Matilde Bandeira[1, 2], Tomás Fontes[1, 3], Eduardo Dourado[1, 2]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal

Introduction: Pathergy is a skin condition in which a minor trauma leads to a skin lesion. A pathergy test consists of an intradermal prick and subsequent observation of the local reaction after 48 hours to check for a raised lesion. The test is positive when a local reaction leads to an indurated erythematous papule. The mechanism that causes this reaction is not well established, but it is postulated that there is a polymorphonuclear-mediated mixed inflammatory response. Despite pathergy being reported in several diseases, it is classically used to diagnose Behçet's disease. The positivity

rate of the pathergy test varies according to the population (higher in the Middle East and East Asia) and has been decreasing since the decade of 1980. Disseminated gonococcal infection (DGI) is an acute infectious disease with Neisseria gonorrhoeae bacteriemia that can manifest as arthritis, tenosynovitis and cutaneous lesions.

Case: A 30-year-old male with no known medical history presented to the Emergency Department due to a four-day history of persistent right ankle and left foot pain associated with local swelling. He also presented fever (maximum axillary temperature of 39.4ºC), myalgia and headache. Upon questioning, he reported unprotected sexual contact the previous week. On physical examination, he presented oligoarthritis affecting the right tibiotarsal and left intertarsal joints, tenosynovitis of the left wrist extensors and hemorrhagic pustules in one finger of the hand and on the radial border of the left wrist. Acute phase reactants were elevated (leucocytosis 16100/µL, neutrophil count 12460/µL, erythrocyte sedimentation rate 56mm/h, C-reactive protein 9.76mg/dL).

The patient was admitted to the Rheumatology ward and started on ceftriaxone (2g 24/24h), doxycycline (100mg 12/12h), and a non-steroidal anti-inflammatory drug due to suspected DGI.

During the first four days of hospital stay, he developed arthritis of the right elbow and left wrist, accompanied by increased C-reactive protein (maximum 18.7mg/dL). The initial incomplete response to therapy and a local indurated papular reaction after a venous puncture raised suspicion for Behçet's disease, so a pathergy test was performed on the eleventh day after admission, which was positive after 24 and 48 hours (Figure 1A). He was also submitted to an arthrocentesis of the elbow, with negative cultural exams.

More than a week after admission, Neisseria gonorrhoeae growth was confirmed on blood cultures that had been drawn upon admission, so a diagnosis of DGI was assumed. The patient completed 14 days of intravenous ceftriaxone, after which all symptoms and signs resolved, and was discharged with oral cefixime for an additional week.

The pathergy test was repeated four months after the resolution of DGI, and it was negative at the 48 hours reading (Figure 1B).

Conclusions: We present a case of DGI in which the pathergy test was positive but resolved after cefalosporin treatment. There are no such cases described in the literature. This finding highlights that a positive pathergy test may be due to active infection, not only Behçet's disease, which impacts its value in the differential diagnosis of oligoarthritis. Considering that active bacterial infection leads to neutrophil activation, this finding may be interpreted in the context of the polymorphonuclear-mediated hypothesis postulated to be behind the pathergy phenomenon.

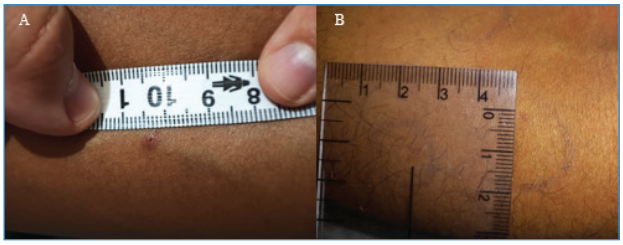

Figure 1. Result of pathergy test (A) during the acute phase of DGI and (B) four months after resolution of DGI

In conclusion, multiple diagnostic hypotheses must be considered in the differential diagnosis of oligoarthritis and a positive pathergy test.

078 - LESÕES NAS MÃOS E LÁBIOS SECOS EM CONTEXTO DE ANSIEDADE OU ALGO MAIS?

Diana Marinha Cruz[1], José M. Seara[1], Flávio Silva[1]
[1]USF Arquis Nova, Unidade Local de Saúde do Alto Minho, Viana do Castelo, Portugal

Enquadramento: As alterações dermatológicas e reumatológicas e a ansiedade, apesar de entidades distintas, podem coexistir e são muito comuns na prática clínica. Fatores psicossociais, incluindo eventos de vida adversos e ansiedade, podem influenciar a gravidade das manifestações clínicas.

Descrição do caso: Mulher de 48 anos, casada com dois filhos e ensino superior. Antecedentes pessoais de ansiedade generalizada, distimia com episódio depressivo e ataques de pânico associados a conflitos laborais. Sem outros antecedentes pessoais ou familiares patológicos.

Recorre a consulta aberta por queixas de lábios e mãos secas, medicada com emoliente por suspeita de eczema disidrótico, com alguma melhoria.

Quatro meses depois, volta ao médico de família, por surgimento de lesões cutâneas no dorso das mãos de coloração eritematosa, por vezes violácea, sem prurido associado, e lesão aftosa com 2mm na região interna do lábio inferior. Referência a agravamento com cremes hidratantes e calor. Tinha recorrido a dermatologista, medicada com nifedipna e tacrolimus em pomada por suspeita de eritema pérnio, sem melhoria.

Realizou estudo analítico com hemograma, leucograma, esfregaço de sangue periférico, coagulação, função hepática, renal e tiroideia com anticorpos, serologias (HIV, VDRL, HVB, HCV), SACE, CK, mioglobina, anti-DS-DNA, IgA, IgG, IgM, normais.

Referenciada a Dermatologia. Sem queixas sugesti-

vas de esclerodermia ou lúpus. Capilaroscopia normal. Apresentava lesões cutâneas papulosas aplanadas nas articulações interfalângicas proximais e metacarpofalângicas. Efetuada biópsia, resultados compatíveis com suspeita clínica de pápulas de Gottron. Pela distrofia cuticular, repetiu capilaroscopia dois meses depois, observando-se capilares trombosados. Referenciada a Reumatologia.

Um mês depois, apresentou dor ligeira nos músculos das coxas e gastrocnémios ao caminhar, sem dor noturna, sem agravamento com a manutenção da marcha ou necessidade de a interromper. No teste dos flexores do pescoço, desistência ao fim de um minuto e meio, com desconforto na região anterior do pescoço, sem dor na região posterior. Sem outras alterações de novo ao exame objetivo. Considerada hipótese diagnóstica de Dermatomiosite.

Efetuados novos exames complementares. Enzimas musculares incluindo aldolase, VS, PCR e ionograma, normais. Anti ENA (RNP, SM, SSA, SSB, Scl70, Jo1), ANAS, Mi2, Ku, PL7 e PL12 negativos; P155/140, RP155 e RP11 positivos. Eletromiografia com alterações moderadas e inespecíficas da fibra muscular, podendo traduzir processo inflamatório/metabólico pouco marcado, restantes estudos da condução dentro da normalidade.

Iniciou hidroxicloroquina, com alguma melhoria inicial, mas suspenso após 9 meses por surgimento de lesões da pele sugestivas de fotossensibilidade e hiperpigmentação.

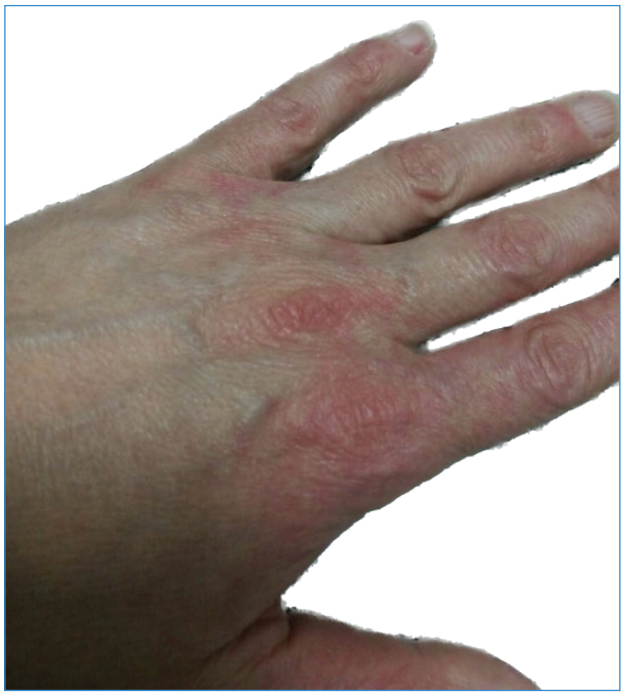

Figure 1. Imagens do caso clínico - manifestações cutâneas das mãos e mucosa labial

Entretanto, mudou de emprego e iniciou psicoterapia. Considera que a ansiedade constante durante vários anos terá contribuído para o desenvolvimento da doença. Atualmente, quatro anos depois, mantem dermatomiosite amiopática, sem medicação além de cuidados de pele e benzodiazepina em SOS.

Discussão e conclusão: A Dermatomiosite é uma miopatia inflamatória rara e as manifestações podem ser inicialmente subtis, constituindo um desafio diagnóstico. A ansiedade, cada vez mais comum, revela-se uma comorbilidade frequente. Estudos mais aprofundados das implicações fisiopatológicas nos mecanismos de doença poderão ser úteis quer para compreender se a mesma poderá ser um trigger, ou se será apenas parte integrante da dolência do doente no seu contexto biopsicossocial.

080 - ARTRALGIAS MECÂNICAS – UM DIAGNÓSTICO PARA ALÉM DO ÓBVIO

Gonçalves, R[1], Oliveira, D[2], Giestas, A[2], Fonseca, R[3]
[1]USF Egas Moniz, Santa Maria da Feira, Portugal, [2]Endocrinology, Centro Hospitalar de Entre o Douro e Vouga, Santa Maria da Feira, Portugal, [3]Rheumatology, Centro Hospitalar Entre o Douro e Vouga, Santa Maria da Feira, Portugal

Introdução: A acromegalia é uma doença rara, caracterizada pela hipersecreção da hormona de crescimento e aumento do seu mediador, o fator de crescimento semelhante à insulina tipo 1 (IGF-1). Uma complicação frequente é a artropatia, que por vezes constitui a manifestação inicial.

Caso clínico: Doente de 63 anos de idade, com história pregressa de uma poliartrite seronegativa diagnosticada aos 50 anos de idade. Na altura, referia queixas de poliartralgias de ritmo misto, descrevia tumefação das mãos e pés, mas não apresentava alterações no estudo analítico. Chegou a estar medicada prednisolona e sulfassalazina com um benefício apenas parcial.

Ao longo do tempo, referiu alteração do padrão de dor para um ritmo puramente mecânico, com envolvimento das mãos, pés, joelhos ancas e coluna lombar associado a macroglossia. Notava ainda alteração gradual da mandíbula. O estudo analítico inicial não revelou alterações. As radiografias mostraram, além de fenómenos degenerativos das articulações descritas, um alargamento do espaço articular e aumento das polpas digitais.

Após nova observação da doente, foi objetivado protuberância frontal, prognatismo da mandíbula, macroglossia, mãos e pés alargados e grosseiros. Contudo, a doente negou alterações do número de calçado ao longo dos anos.

Foi pedido novo estudo analítico que revelou el-

evação do IGF-1 (484 ng/mL). A RMN da hipófise revelou a presença de um macroadenoma da hipófise (de 11.6X11.4 mm) permitindo o diagnóstico de acromegalia.

Conclusão: O caso descrito ilustra a dificuldade diagnóstica e a necessidade de elevada suspeita clínica para o diagnóstico desta patologia.

Na acromegalia, visto que as lesões ocorrem de forma lenta e progressiva e se assemelham às da osteoartrose, o diagnóstico é feito muito tardiamente. Devem levantar suspeita as alterações da fisionomia e, na radiografia, o aumento do espaço articular na presença de osteofitose e aumento dos tecidos moles. Na fase inicial podem surgir queixas semelhantes a uma artrite seronegativa, como foi o caso.

083 - "I CAN NO LONGER HOLD MY BABY" – A CASE OF PREGNANCY – AND LACTATION-ASSOCIATED OSTEOPOROSIS WITH VERTEBRAL FRACTURES

Filipe Araújo[1], Ana Luís[2]

[1]Rheumatology and Osteopororis Unit, Hospital de Sant Ana, Lisboa, Portugal, [2]Neurosurgery Department, Centro Hospitalar de Lisboa Ocidental, EPE, Lisboa, Portugal

Introduction. Pregnancy- and lactation-associated osteoporosis (PLO) is a very rare condition complicating 4-8 pregnancies in each million. Most cases present with back pain during late pregnancy or early postpartum resulting from vertebral fractures. These fractures significantly impact maternal function and quality-of-life and compromise their ability to provide care for the newborns.

Clinical case. An otherwise healthy 33-year old woman was referred to our rheumatology clinic by her spine surgeon for a 1-month history of intense and refractory dorsal pain that started after she picked her 4-month baby up. After several visits to the emergency department and taking different combinations of painkillers, thoracic spine CT scan (Fig.1A) and MRI (Fig.1B T1-weighted, 1C STIR) were ordered confirming vertebral fractures of D5, D7, D8, D9, D10, D12 and L2. She had no known clinical risk factors for osteoporosis, was a non-smoker and non-alcohol drinker. This was her second pregnancy (very early spontaneous abortion in the first one) and at 39 weeks of gestation she had a cesarean delivery. She had been breastfeeding since then. Her mother had also suffered from 2 vertebral fractures but at age 67. She was 156 cm in height and weighed 66 kg (BMI 27.1 kg/m2). Laboratory abnormalities included a mildly insufficient 25-OH-vit. D (25 ng/mL) and elevated N- and C-terminal telopeptide of type I collagen (185 and 0.764 ng/mL, respectively). Dual-energy X-ray absorptiometry (DEXA, GE Healthcare Lunar DPX NT) revealed a bone mineral density (BMD) of 0.777 g/cm2 (Z-score: -3.4) in her lumbar spine and 0.877 g/cm2 (Z-score: -0.8) in the femoral neck. Breastfeeding was stopped and, after shared decision, teriparatide and calcium and vitamin D supplements were started. Teriparatide was done for only 6 months (severe vertigo), but the patient remained on supplementation, physical exercise and clinical monitoring for another 3 years, with pain reduction, no new fractures and BMD improvement (Table 1).

Discussion. Current knowledge on PLO is scarce and arises essentially from case reports and case series. A literature review on 338 cases found a mean age of 35.7 (19-47) years at diagnosis, the majority were primiparous. They presented with back pain, the earliest at the 5th month of pregnancy and the latest 9 months after delivery. A striking feature of PLO is multiple vertebral fractures (average of 4.4 per woman, most at the thoracolumbar transition). Etiology and mechanisms of disease remain unclear. Genetic factors and classical risk factors may play a role, but disruption of normal pregnancy calcium metabolism is likely. Fetal demands for calcium are higher in the third trimester, but maternal adaptation compensates by duplicating intestinal absorption (mediated by increased 1,25-dihydroxyvitamin D). PLO probably arises from significant maternal skeletal resorption during lactation to ensure milk production, mediated by PTH-related peptide (secreted by the placenta and breasts). Treatment remains controversial. Halting breastfeeding and a balanced diet are consensual, as well as calcium and vitamin D supple-

TABLE 1. Bone mineral density (BMD) changes up to 36 months of follow-up. FN: femoral neck; IR: increase rate

	Baseline	12 months	IR (%)	24 months	IR (%)	36 months	IR (%)
L1-L4 BMD (g/cm2)	0.777	0.880	+13.3	0.801	-9.0	0.812	+1.4
L1-L4 Z-score	-3.4	-2.2		-2.2		-2	
FN BMD (g/cm2)	0.877	0.904	+3.1	0.743	-17.8	0.756	+1.8
FN Z-score	-0.8	-0.3		-0.8		-0.6	

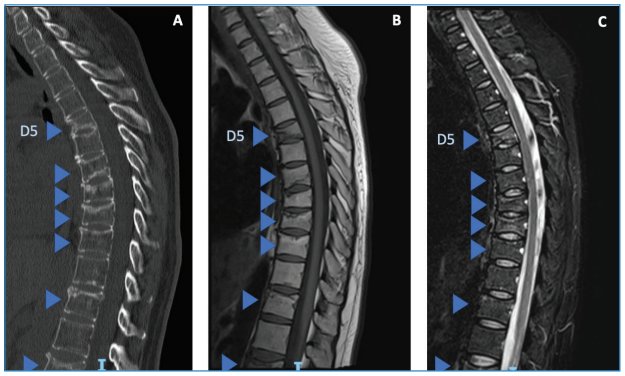

Figure 1. Dorsal spine lateral view of CT-scan (A) and MRI T1 (B) and STIR (C) showing D5, D7-D10, D12 and L2 osteoporotic fractures.

mentation. Although no formal trials exist to support the use of either antiresorptive or anabolic agents in postpartum, dozens of cases have been published in the literature with good results in bone mass recovery and pain reduction. These patients should be advised that the risk of new fractures in subsequent pregnancies can be as high as 25%.

085 - CHRONIC SYNOVITIS DUE TO HYPERSENSITIVITY REACTION TO COBALT AFTER TOTAL KNEE REPLACEMENT: A CASE REPORT

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

Introduction: Total knee replacement is a common surgical procedure that can improve function and reduce pain in patients with advanced knee osteoarthritis. However, complications can occur, including hypersensitivity reactions to metal components of the prosthesis. Cobalt is a common component of modern knee prostheses and can cause chronic synovitis due to a delayed-type hypersensitivity reaction. We report a case of a 70-year-old male with chronic synovitis due to cobalt hypersensitivity after total knee replacement.
Case report: A 70-year-old male underwent total knee replacement with a cobalt-chromium-molybdenum prosthesis. A few months postoperatively, he developed persistent pain, swelling, and limited range of motion in the affected knee (Figure 1A). Imaging studies showed no signs of infection or loosening of the prosthesis (Figure 1B). Synovial fluid culture was negative, and patch testing confirmed cobalt hypersensitivity. The patient underwent revision surgery with a non-cobalt-containing prosthesis.
Discussion: Cobalt hypersensitivity is a rare but important complication of total knee replacement that can cause chronic synovitis and lead to implant failure. The diagnosis is often delayed due to its nonspecific presentation, and high index of suspicion is required in patients with persistent knee pain and swelling. Patch testing is a reliable method for confirming cobalt hypersensitivity, and avoidance of cobalt-containing implants is the mainstay of treatment. Revision surgery with a non-cobalt-containing prosthesis can lead to resolution of symptoms and prevent further complications.
Conclusion: Cobalt hypersensitivity is a rare but important cause of chronic synovitis after total knee replacement. Rheumatologists should be aware of this complication and consider it in the differential diagnosis of patients with persistent knee pain and swelling after total knee replacement.

087 - LÖFGREN SYNDROME: A RARE CAUSE OF ERYTHEMA NODOSUM AND ARTHRITIS IN AN ADULT MALE

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

Introduction: Löfgren's Syndrome is an acute and self-limiting form of sarcoidosis, characterized by oligoarthritis, erythema nodosum and perihilar adenopathies. It is more common in young women. In this case report, we present a 39-year-old man with oligoarthritis of the ankles, erythema nodosum and perihilar adenopathies, who was diagnosed with Löfgren's Syndrome.
Case Report: 39-year-old man, without previous medical history, is referred to the rheumatology department due to oligoarthritis. Patient describes a 2-week history of inflammatory ankle arthralgia and nodular skin lesions on the anterior tibial area. Physical exam showed bilateral ankle arthritis and erythema nodosum on the right leg (Figure 1A). Patient denied gastrointestinal, ocular and other cutaneous lesions. Blood tests showed elevated inflammatory biomarkers (ESR of 79 mmHr and CPR 12.79 mg/dL), with increased angiotensin converting enzyme (42 nmol/mL/min). Chest radiography showed bilateral adenopathies, which was later confirmed on CT scan (Figure 1B). Administration of 15mg/day of prednisolone resulted in complete resolution of symptoms.
Discussion: Löfgren's Syndrome is considered a benign form of sarcoidosis, with good prognosis and spontaneous resolution of symptoms in a few weeks to months. There is no specific treatment for Löfgren's Syndrome, but nonsteroidal anti-inflammatory drugs

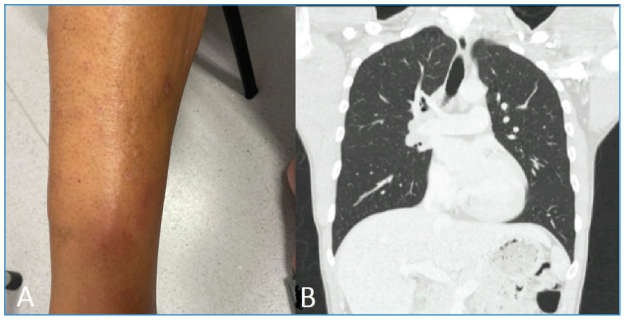

Figure 1.

and corticosteroids are often used to relieve pain and inflammation.

Conclusion: This case report highlights the importance of considering Lofgren syndrome as a differential diagnosis in young adult males presenting with acute lower limb oligoarthritis and erythema nodosum. Early recognition and treatment with glucocorticoids can result in rapid and complete resolution of symptoms.

088 - VENOUS SUBTYPE THORACIC OUTLET SYNDROME: A RARE CAUSE OF UPPER EXTREMITY OEDEMA AND PAIN

Tiago Beirão[1], Sofia Moreira[2], Beatriz Samões[1], Catarina Rua[1], Romana Vieira[1], Joana Abelha-Aleixo[1], P Pinto[1], Flávio Campos Costa[1], Diogo Guimarães da Fonseca[1], Taciana Videira[1]

[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal, [2]Medicina Física e de Reabilitação, Centro de Medicina de Reabilitação da Região Centro – Rovisco Pais, Coimbra, Portugal

Introduction: Thoracic outlet syndrome (TOS) is a group of compression disorders that occur in nerves and/or blood vessels in the thoracic outlet. Venous subtype TOS is characterized by compression of the subclavian vein, leading to upper extremity oedema and pain. We present a case report of a 35-year-old female with venous subtype TOS.

Case Report: The patient presented to the rheumatology clinic with a 2-year history of asymmetric oedema of her right arm, with shoulder pain and limited range of motion. The oedema was godet negative. Increased pain occurred on shoulder abduction. A previous diagnosis of complex regional pain syndrome was made, but no improvement with target treatment was achieved. Electromyography was normal. MRI of the shoulder was also normal. A venous subtype TOS was suspected, and an angioCT scan of the right arm in neutral position and abduction of 120º was ordered. The scan showed diminished blood flow on the subclavian vein in abduction but normal on neutral position (Figure 1A and 1B), confirming the diagnosis of venous subtype TOS. The patient was referred for surgical evaluation, and decompression surgery was performed. Postoperative follow-up showed resolution of the edema and improvement in pain and range of motion.

Discussion: Venous subtype TOS is a rare condition that accounts for approximately 5-10% of all TOS cases. The diagnosis can be challenging, as symptoms are nonspecific and can mimic other conditions such as cervical radiculopathy or rotator cuff injury. Imaging studies, such as angioCT, are essential in making a correct diagnosis. Treatment options for venous subtype TOS include conservative management, such as physical therapy, and surgical intervention, such as decompression surgery. Early recognition and treatment are crucial in preventing long-term complications, such as chronic venous insufficiency.

Conclusion: Venous subtype TOS should be considered in the differential diagnosis of patients presenting with upper extremity oedema and pain. Imaging studies, such as angioCT, can aid in making an accurate diagnosis. Early recognition and treatment can prevent long-term complications, and surgical intervention should be considered when conservative management fails.

089 - MIGRATORY ARTHRALGIA AS AN ATYPICAL PRESENTATION OF WHIPPLE DISEASE: A CASE REPORT

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

[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Introduction: Whipple disease is a rare systemic disorder that affects multiple organs, including the gastrointestinal tract, central nervous system, and joints. Is caused by *Tropheryma whipplei* bacteria, which can evade the immune system and cause chronic infection. Migratory arthralgia is a rare and nonspecific presentation of Whipple disease, causing often delayed diagnosis due to its nonspecific presentation. We report a case of a 62-year-old woman with migratory arthralgia as the first manifestation of Whipple disease.

Case Report: A 62-year-old woman presented with a 4-month history of diarrhea, abdominal pain, and weight loss. She had a 3-year-old history of migratory arthralgia of ankes and wrists. Physical examination showed diffuse abdominal pain. Gastroduodenoscopy showed exuberant duodenal lymphangiectasia (Figure 1A). Capsule endoscopy and duodenal biopsy showed PAS-positive macrophages in the lamina propria containing nonacid-fast Gram-positive bacilli (Figure 1B).

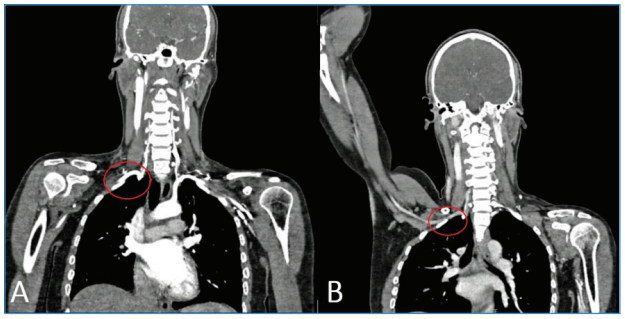

Figure 1.

PCR test for tropheryma whipplei was positive. Intravenous ceftriaxone therapy was prescribed, with resolution of symptoms after 2 weeks, with maintenance therapy with oral co-trimoxazole.

Discussion: Migratory arthralgia is a rare but important manifestation of Whipple disease, and its recognition can lead to earlier diagnosis and treatment. It is thought to be caused by deposition of immune complexes in the joints, leading to inflammation and pain. The diagnosis of Whipple disease can be challenging, as it requires a high index of suspicion and multiple diagnostic modalities. The gold standard for diagnosis is duodenal biopsy, which shows pathognomonic periodic acid-Schiff-positive macrophages containing the bacilli.

Conclusion: Migratory arthralgia and intestinal symptoms can be an atypical presentation of Whipple disease and should prompt investigation for this rare disorder. Early diagnosis and treatment with antibiotics can lead to resolution of symptoms and prevent complications. Rheumatologists should be aware of this rare manifestation and consider Whipple disease in the differential diagnosis.

090 - P-ANCA POSITIVE VASCULITIS INDUCED BY COCAINE WITH SEPTAL DESTRUCTION

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

Introduction: Cocaine abuse is a significant public health concern, with numerous associated complications. One of the rare but severe consequences of cocaine use is the development of vasculitis. We present a case of P-ANCA positive vasculitis induced by cocaine with septal destruction in a 35-year-old male, highlighting the importance of recognizing this unique presentation and its potential implications.

Case Report: A 35-year-old male with a history of chronic cocaine abuse presented with progressive nasal obstruction, epistaxis, and septal deformity. The patient reported a long-standing habit of intranasal cocaine use. Physical examination revealed bilateral nasal crusting, septal perforation, and mucosal ulceration. Facial CT confirmed septal perforation (Figure 1A). Laboratory investigations demonstrated elevated inflammatory markers and a positive perinuclear antineutrophil cytoplasmic antibody (P-ANCA) test, with negative Anti-MPO and Anti-PR3. Nasal biopsy confirmed eosinophilic vasculitis (Figure 1B) with septal destruction (figure 1C) and inflammation (figure 1D). Imaging studies revealed no evidence of systemic vasculitis or underlying connective tissue diseases. The patient refused treatment and lost follow-up.

Discussion: The development of P-ANCA positive vasculitis associated with cocaine abuse is a rare phenomenon. Cocaine, as a potent vasoconstrictor, may lead to ischemic injury and subsequent vasculitis. The involvement of the nasal septum in our case highlights the specific tissue vulnerability associated with cocaine-induced vasculitis. Septal destruction can occur due to direct ischemic injury, subsequent infection, or chronic inflammation caused by the local tissue insult. The presence of P-ANCA antibodies is of particular interest in this case. While P-ANCA positivity is commonly associated with microscopic polyangiitis, cocaine-induced vasculitis with P-ANCA positivity has been reported previously. The exact pathogenesis of this immune response remains unclear, warranting further investigation into the immunological mechanisms involved. Management of cocaine-induced vasculitis primarily revolves around discontinuation of cocaine use and appropriate medical therapy. On most cases, low-

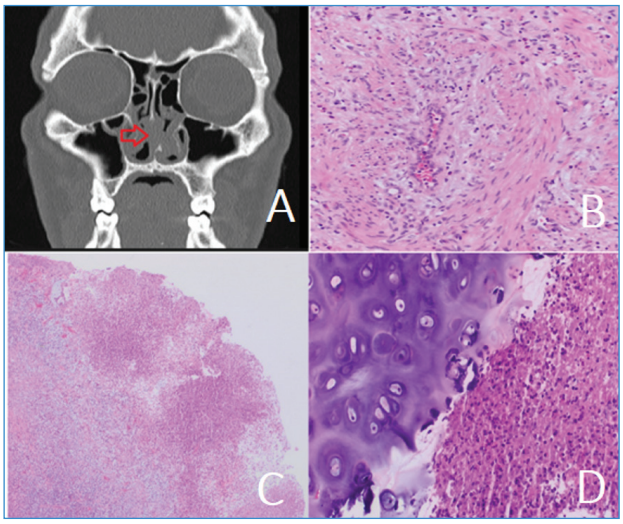

Figure 1.

dose prednisolone, along with wound are measures, are sufficient to promote healing and aiding symptom control. However, the potential for relapse remains significant, emphasizing the importance of addressing the underlying substance abuse disorder.

Conclusion: We present a rare case of P-ANCA positive vasculitis induced by cocaine with septal destruction in a 35-year-old male. This report serves as a reminder to healthcare providers about not only the potential complications associated with cocaine abuse, but also the possible relationship with P-ANCA positivity, therefore being a differential diagnosis of systemic vasculitis.

091 - A CHAIN OF LIGHT EVENTS LEADING TO A LETHAL OUTCOME

Frederico Rajão Martins[1], Teresa Abegão[2], Karolina Aguiar[2], Margarida Lucas Rocha[1], Vítor Teixeira[1]
[1]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, [2]Internal Medicine, Centro Hospitalar Universitário do Algarve, Faro, Portugal

Introduction: Imaging detected hand tenosynovitis has shown to occur in early RA and pre-RA phases. Additionally, it is also noted to be a strong predictor for RA development and undifferentiated arthritis and arthralgia.

We present the case of a patient with marked bilateral tenosynovitis of extensor and flexor tendons of the wrists who was diagnosed with seronegative rheumatoid arthritis (RA), but during follow-up the diagnosis was re-challenged.

Clinical case: A 70-year-old woman with a diagnosis of previous renal carcinoma was referred to Rheumatology in early 2020 due to wrist and finger arthralgias, associated with hand paraesthesia. Marked bilateral tenosynovitis of extensor and flexor tendons of the wrists was evident on ultrasound (US). She was seronegative for rheumatoid factor, anti-CCP, ANA, anti-dsDNA and antiphospholipid antibodies. On blood tests there were no changes on hepatic enzymes, renal function, protein electrophoresis and viral serologies. Urinalysis was normal.

A presumptive diagnosis of non-erosive seronegative rheumatoid arthritis was made. Methotrexate and steroids were ineffective. In the following months she developed periungual erythema and haemorrhages, submandibular gland swelling, dry mouth, severe asthenia and 20kg weight loss, associated with constipation, abdominal distension and sporadic vomiting.

Nailfold capillaroscopy revealed a mixed active/late scleroderma pattern; thoracic-abdominal-pelvic CT scan showed bilateral pleural effusion, pericardial effusion and ascites, dilation of gastric cavity, small intestine loops and transverse colon. Soon after, she was hospitalized due to intestinal ileus with incoercible vomiting. Periorbital ecchymosis and macroglossia was noted during admission.

New blood analysis revealed presence of monoclonal lambda light chain proteins (2420 mg/l) and a kappa/lambda ratio of 0. Bone marrow aspiration evidenced infiltration by 5.8% monoclonal plasma cells, suggestive of a neoplastic process. Gastrointestinal endoscopic studies showed slow gastric motility and wide gastric lumen without obstruction and random biopsies of gastric mucosa and a 12 mm sigmoid polyp were performed. Anatomopathological findings included the presence of amorphous substance, salmon red coloration with Congo red and birefringence under polarized light. Immunohistochemistry was weak positive for amyloid P substance, amyloid A and transthyretin. Echocardiogram revealed a circumferential effusion with apparent fibrin deposits, in the absence of myocardial enhancement on scintigraphy.

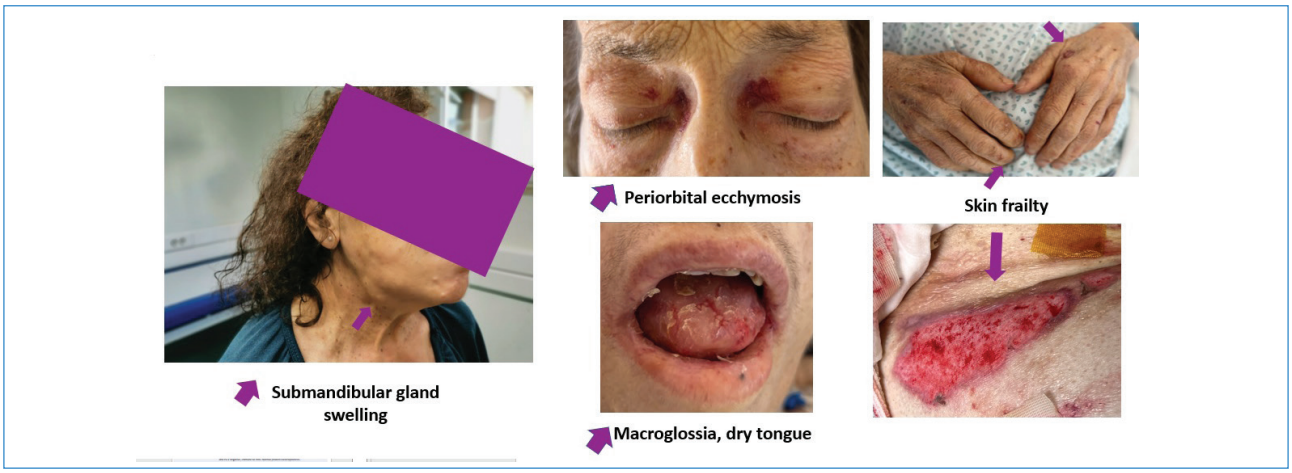

Figure 1. Clinical signs on physical examination

A diagnosis of light chain amyloidosis (lambda) was made, and treatment with VCD adapted protocol: cyclosporine + bortezomib + prednisolone was initiated. Two weeks posteriorly, she died of uroseptic shock.

Conclusion: Seronegative rheumatoid arthritis (RA) remains an imprecise diagnosis, according to some experts. It's not uncommon for a diagnosis of seronegative RA to be re-classified, like in our case.

When the clinical course is atypical, it is important to review diagnosis. Our patient's symptoms were attributed to AL amyloidosis, which is a rare mimic of common rheumatic inflammatory disorders. Association with other plasma cell dyscrasias such as multiple myeloma is frequent. Monoclonal proteins may be missed by routine serum or urine protein electrophoresis, so more sensitive techniques such as immunofixation are recommended.

101 - RS3PE. WHAT ELSE?

Inês Almeida[1], Inês Santos[1], Joana Andrade[2], Paulo Monteiro[1], Couto M[1]
[1]Rheumatology Unit, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [2]Internal Medicine Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Introduction: Remitting seronegative symmetrical synovitis with pitting edema syndrome (RS3PE) is a rare inflammatory arthritis characterized by abrupt onset of symmetrical synovitis of the hands and/or ankles and marked pitting edema of the dorsum of the hands and/or feet, elevated acute phase reactants and negative rheumatoid factor. It affects typically the elderly and is more frequent in men. It has a good response to glucocorticoids in a medium dose (10–20 mg/day), with most of the cases been tapered off within 6 to 18 months of treatment, with long term remission. Ultrasound may evidence tenosynovitis of flexor and extensor tendons at the wrist and extensor tendons of the feet. Blood tests typically show normal white cell count, varying degrees of anemia of chronic disease and elevated acute phase reactants with low or absent ANA titters.

Case report: A 71-year-old women presented to the emergency service with asthenia, fever, edema, pain and heat of the right hand dorsum after peripheral venipuncture in the previous week. Blood tests showed leucocytosis, neutrophilia, macrocytic anemia, normal procalcitonin, elevated CRP and erythrocyte sedimentation rate (ESR): 40,18 mg/dl and 81 mm/h, respectively. She was hospitalized for a suspected cellulitis and vancomycin and meropenem were started. After 17 days of antibiotics, the pitting edema evolved affecting both hands and left tibiotarsal, despite of maintained apyrexia accompanied by reduction of inflammatory markers. Blood and urine cultures were negative. The rheumatology team was asked to collaborate at this point. Radiographies showed no joint erosions. Ultrasound evidenced grade 2 radiocarpal synovitis with grade 2 Doppler sign, exuberant extensor tenosynovitis in both hands and left peroneal tenosynovitis. Her rheumatoid factor, anti-CCP, ANA and ANCA were negative. Protein electrophoresis suggested an inflammatory state, with no other abnormalities. Based on clinic and ultrasound findings, RS3PE was suspected and the patient started prednisolone 20mg/day. Due to unsatisfactory response, a 250mg methylprednisolone pulse was performed and prednisolone was increased to 30mg/day, with benefit. We verified a significant decrease of hand edema and left tibiotarsal edema resolution. On suspicion of a paraneoplastic condition, the patient underwent neck, thoracic, abdominal and pelvic CT scans that did not catch neoplastic suggestive findings. After 17 days of glucocorticoid therapy, CRP decreased to 12,74 mg/dl, ESR to 61 mm/h and she had a meaningful clinical improvement, but still had moderate edema of her right hand. The patient was discharged medicated with subcutaneous methotrexate 20mg/week, folic acid 10mg/week, prednisolone 20mg/day, daily calcium and vitamin D and monthly ibandronic acid. Three months later there is still some remaining edema of the right hand accompanied of ESR decreased to 21 mm/h and CRP to 3,5 mg/dl. Prednisolone is being slowly tappered.

Discussion/Conclusion: RS3PE may be a harbinger of an underlying malignancy. Solid tumors (lung, gastrointestinal, genitourinary) and haematological malignancies have both been reported. Typically, these cases have a more severe presentation, are resistant to medium doses of glucocorticoids and resort after treatment. The prognosis depends on the tumor prognosis. In this case, proper investigation must keep carry on for the exclusion of other malignancies, such as haematological malignancy.

102 - MACROVASCULOPATHY IN SYSTEMIC SCLEROSIS - A LESS DESCRIBED VASCULAR INVOLVEMENT

Inês Almeida[1], Paulo Monteiro[1], Couto M[1]
[1]Rheumatology Unit, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Introduction: Systemic sclerosis (SSc) is a complex and heterogenic disease characterized by vasculopathy and organ fibrosis. Raynaud phenomenon (RF) and digital ulcers (DU) with severe scarring and tissue necrosis are fearsome vasculopathy expressions. Mostly, they are a manifestation of progressive microangiopathy, assessed via nailfold capillaroscopy (NC). Studies point macrovasculopathy, assessed via colour doppler

ultrasonography (CDUS), as a risk factor and a disease severity marker, thus supporting its role in the DU's pathophysiology.

Case report: An eighteen-year-old women with the diagnosis of limited SSc since 2018 was first observed in our rheumatology department on 12/2021. She referred RF and DU for the last 3 years, dyspepsia and epigastric pain. She denied palpitations, dyspnea, chest pain, intestinal symptoms, drugs or tobacco consumption. She was on nifedipine 30mg/day and pentoxifylline 800mg/day. By the time of our first observation, she had deep DU in the pulps of the right 2nd, 3rd and 4th fingers, left 2nd and 3rd fingers and lateral surface of some digits. She didn't have sclerodactyly or skin thickening. She presented telangiectasia on the upper lip and fingers. Her ANA were positive (titre 1/2560), anti-centromere pattern, positive for anti-Ro-52 and anti-nucleosomes. Her lupus anticoagulant was positive, anti-cardiolipin and anti-beta2 glycoprotein I were negative. NC showed a late scleroderma pattern. She was started on iloprost through elastomeric pump, completing 3 cycles, with benefit. During the follow up, since she had multiple episodes of DU under nifedipine 60mg and AAS 100 mg, with the need of multiple perfusions of iloprost, sildenafil was added in dose titration up to 150mg/day. There was substantial improvement in the pre-existing DU, no appearance of new ulcers and a lower frequency of RF until 2/2023. By that time, new multiple DU appeared, so a new cycle of iloprost was performed and she was started on bosentan, titrated up to 125mg twice a day. On 03/2023, after an initial favourable healing, we verified a severe worsening of DU on the left 2nd and 3rd fingers, with signs of infection. She was hospitalized to intravenous prostanoid therapy, antibiotic and wound care. There were no alterations on physical examination, except for the loss of continuity of epithelial coverage on the left 2nd and 3rd distal fingertips, covered by purulent exudate and necrotic tissue. Distal pulses were palpable and symmetric. She maintained iloprost perfusion and nifedipine was optimised to 90mg/day. Opioid based analgesia and statins were also started. Her blood analysis showed Hb 14.1 g/dl, leukocytes 4970/mm3, platelets 126000/mm3, erythrocyte sedimentation rate 14 mm/h, CRP 0,07 mg/dl. Titres of anti-dsDNA, ANCA, cryoglobulins, C3, C4, Ig and SPEP were normal, hematuria and proteinuria were absent. Inherited thrombophilia screening was normal, except for homocysteine concentration that was 20.1 µmol/L (normal <13.9 µmol/L). CDUS revealed left radial artery stenosis. AngioCT scan showed bulky splenomegaly, but no liver enlargement, renal or suprahepatic arteries involvement. She continues the process of a favourable but very slow healing, being on the remodelling stage of wound healing 30 days after hospitalization. The cause for splenomegaly is being studied.

Discussion: SSc is a complex disease and DU are a feared complication. Micro-and macrovasculopathy can both contribute to severe scarring and tissue necrosis and should be evaluated to understand the underlying mechanism and treat accordingly.

111 - DRUG-INDUCED SYSTEMIC LUPUS ERYTHEMATOSUS IN PSORIATIC ARTHRITIS ASSOCIATED WITH SECUKINUMAB

Pedro Ávila Ribeiro[1, 2], Ana Rita Lopes[1, 2], Martins-Martinho J [1, 2], Estela Nogueira[3], José Carlos Romeu[1], Ana Rita Cruz-Machado[1, 2], Elsa Vieira-Sousa[1, 2]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Serviço de Nefrologia e Transplantação Renal, Centro Hospitalar e Universitário Lisboa Norte, Lisboa, Portugal

Introduction: Drug-induced lupus erythematosus (DILE) manifests as cutaneous or systemic features of lupus erythematosus following treatment with a wide range of drugs. Only one case of systemic secukinumab-associated DILE was previously reported.

Case report: A 68-year-old male patient treated with secukinumab in monotherapy for psoriatic arthritis, previously in remission, suspended treatment due to COVID pandemic onset. Three months later hand polyarthritis ensued and secukinumab was reintroduced but remission was not attained and the patient developed anorexia and weight loss. Three months after reintroduction he had an abrupt onset of low-grade fever, fatigue, flu-like symptoms, dyspnea and worsening of widespread inflammatory arthralgias. Laboratory investigations showed anemia, leukopenia with lymphopenia, cytocholestasis, raised erythrocyte sedimentation rate (56 mm/h) and C-reactive protein (2.73 mg/dL), C3 complement consumption (85 mg/dL), proteinuria (1630mg/24h) and active urine sediment. Antinuclear (1:1280) and anti-double-stranded (ds) DNA (212.3 IU/mL) antibodies were found. Anti-histone, anti-extractable nuclear antigens as well as anti-phospholipid antibodies were negative. Chest imaging showed a peripheral pulmonary embolism and pneumonia of the inferior left lobe, and a small bilateral pleural effusion. The patient was admitted to hospital, secukinumab therapy was discontinued and treatment with enoxaparin, antibiotics, enalapril, hydroxychloroquine and prednisolone 0.5mg/Kg qd was started with

remission of symptoms and laboratorial changes after one month except for proteinuria, which decreased to 653mg/24h. Due to concomitant hemophilia A a kidney biopsy was not performed. Assuming significant renal involvement with probable lupus nephritis class III-IV, treatment with mycophenolate mofetil (MMF) was started and complete renal response was achieved and persisted during follow up, without evidence of relapse even when immunosuppression was tapered. Maximum prescribed MMF dose was 1g qd because proteinuria decreased to 302mg/dL two weeks after MMF was started. Complete renal response (haematuria <5 cells/µL, leukocyturia <10 cells/µL, proteinuria 80mg/g creatinine) was achieved six months after discharge. Total follow-up was thirty-three months, prednisolone was slowly tapered and withdrawn after one year in renal remission and MMF tapered to 500mg in the last four months.

Discussion: This is the second reported case of systemic secukinumab-associated DILE, and the first with renal involvement. Acute onset with constitutional symptoms, serositis and hepatic involvement are typical of DILE. Although other features are unusual for DILE, including nephritis, hypocomplementemia, anti-dsDNA positivity, and absence of anti-histone antibodies may rise suspicion of intercurrent idiopathic systemic lupus erythematosus, the temporal association with secukinumab reintroduction, acute presentation in a 68-year old male and rapid resolution after secukinumab discontinuation are strongly suggestive of DILE with secukinumab as a culprit.

117 - UMA CAUSA SURPREENDENTE DE METATARSALGIAS

Diana Capela[1], Ricardo Sampaio[2], Fonseca, R[3]
[1]USF Famílias, Santa Maria da Feira, Portugal, [2]Imagiologia, Hospital Lusíadas Porto, Porto, Portugal, [3]Rheumatology, Centro Hospitalar Entre o Douro e Vouga, Santa Maria da Feira, Portugal

Enquadramento: As metatarsalgias são um sintoma muito comum e a sua etiologia pode ser muito diversa. A história clínica e o exame objetivo nem sempre são suficientes para o diagnóstico pelo que os exames complementares de imagem podem esclarecer a etiologia final que, por vezes, pode ser surpreendente.

Descrição do Caso: Mulher de 42 anos de idade, sem antecedentes médicos e cirúrgicos de relevo, com queixas de metatarsalgias mecânicas à esquerda, sobretudo localizadas sob as primeiras 3 articulações metatarso-falângicas e com extensão até à região plantar do médio pé. A radiografia não mostrou alterações e a ecografia não mostrou neuromas de Morton. Pela persistência das queixas, idade da doente, ausência de traumatismo ou uso de calçado inadequado, realizou Ressonância Magnética que mostrou uma massa de tecidos moles no antepé, no alinhamento do segundo metatarso de contornos lobulados compatível com malformação vascular de baixo débito (venosa) que infiltrava os músculos quadrado plantar e adutor do hallux. Esta lesão media 5,5 cm de diâmetro anteroposterior e 3 cm de diâmetro transversas e 2 cm de espessura dorso-plantar (figura 1).

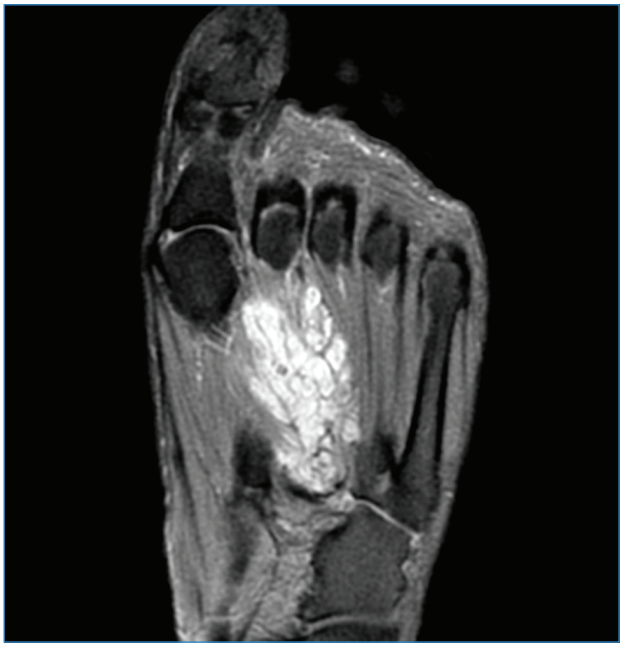

Figura 1. Ressonância Magnética do Pé

Discussão: As malformações vasculares venosas de baixo débito são habitualmente assintomáticas, constituindo uma causa muito rara de metatarsalgias. Em ortostatismo ou esforço físico podem aumentar de volume e causar dor como foi o caso da doente apresentada. O aparecimento de uma lesão de grandes dimensões na região plantar é raro sobretudo não havendo qualquer sinal ao exame objetivo. O tratamento passa pela vigilância estando a escleroterapia reservada para casos selecionados.

118 - SYSTEMIC SCLEROSIS- COLD HANDS, NOT SO WARM HEART- A CASE DESCRIPTION

Sofia Ferreira Azevedo[1, 2], Carolina Mazeda[1, 2], Anabela Barcelos[1, 2]
[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, EPE, Aveiro, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz Health Alliance, Portugal, Aveiro, Portugal

Introduction: Systemic sclerosis (SSc) is an autoim-

mune disease with multisystemic involvement. Primary systemic sclerosis heart involvement (pSHI) is a significant contributor to morbidity and mortality in SSc patients. The clinical spectrum of pSHI is wide-ranging, spanning from asymptomatic perfusion abnormalities to diastolic dysfunction, acute myocarditis, and congestive heart failure.

Case description: We present the case of a 71-year-old female followed in our Rheumatology centre since 2010 with limited cutaneous SSc, presenting with Raynaud phenomena, sclerodactyly, telangiectasis, calcinosis, and oesophageal involvement. Her autoimmunity profile was characterized by anti-nuclear antibodies in a titter of 1/1280, with strong positivity for anti-centromere antibodies. The patient's medical history included atrial fibrillation and reduced left ventricular ejection fraction (LVEF) heart failure. In December 2021, her echocardiogram evidenced a LVEF of 45% and the patient was clinically stable, with a New York Heart Association (NYHA) functional scale of II.

One year later, in December 2022, the patient experienced an acute worsening of symptoms, reaching a NYHA IV. An echocardiogram revealed a significant decline in LVEF (28%) with severe left atrial dilation. The patient had an elevated nT-ProBNP level (17452 pg/ml), while troponin, myoglobin, and creatine-phosphokinase were within normal limits.

Laboratory findings indicated a slightly increased erythrocyte sedimentation rate (ESR) (26 mm/h) and a C-reactive protein (CRP) level of 0.6 mg/dl. Serologies for CMV, EBV, Herpes simplex, and Influenza were negative. The patient was initially admitted to the Cardiology department and later transferred to the Intensive Care Unit due to cardiogenic shock, necessitating inotropic support, diuretics, vasopressors, and ventilatory assistance. Amiodarone was initiated but discontinued promptly due to acute hepatitis resulting from the medication.

To further evaluate possible myocardial tissue damage, cardiac Magnetic Resonance Imaging (MRI) was performed, revealing extensive myocardial oedema, late gadolinium enhancement, and subepicardial fibrosis, suggestive of acute and/or subacute myocarditis.

Cultures, serologies, and PCR testing were conducted to investigate potential causes of myocarditis, but all results were negative. Subsequently, an endomyocardial biopsy was performed, demonstrating interstitial fibrosis and a lymphocytic infiltrate, consistent with the diagnosis of myocarditis, particularly in relation to autoimmune diseases. Further tissue cultures and PCR testing for common viruses associated with myocarditis were negative.

Based on the imaging findings, histological evidence, and exclusion of other potential causes, pSHI in the form of myocarditis was established as the most likely diagnosis.

Treatment was initiated with Prednisolone 20 mg and Mycophenolate mofetil 2g, accompanied by tight control of kidney function and blood pressure. The patient responded favorably, allowing for a gradual tapering of Prednisolone and discharge to a rehabilitation clinic for further care and recovery.

Conclusion: We present a rare case of myocarditis in a patient with limited cutaneous SSc. The occurrence of myocarditis in SSc patients, particularly those with anticentromere positivity, is uncommon. The limited number of reported cases and the absence of clear treatment recommendations emphasize the need for further research and a multidisciplinary approach, improving the prompt diagnosis and individualized treatment strategies for these patients.

121 - DENOSUMAB NO TRATAMENTO DE OSTEOPOROSE SECUNDÁRIA A DISTROFIA MUSCULAR DE DUCHENNE

Beatriz Samões[1], Tiago Beirão[1], Catarina Rua[1], Joana Aleixo[1], Romana Vieira[1]

[1]Rheumatology Department, Centro Hospitalar de Vila Nova de Gaia/Espinho, Vila Nova de Gaia, Portugal

Introdução: A Distrofia Muscular de Duchenne (DMD) é uma doença neuromuscular progressiva grave ligada ao X. A corticoterapia crónica e a imobilização inerentes condicionam um risco substancial de osteoporose e de fraturas ósseas. Ainda há poucos dados sobre o uso de denosumab no tratamento da osteoporose induzida pela DMD.

Casos clínicos: Caso 1: Homem de 22 anos, com antecedentes de DMD (com incapacidade para a marcha há 5 anos) e hipertensão arterial, sob deflazacorte 30 mg/dia, alendronato 70 mg/semana (início há um mês), carbonato de cálcio 1250 mg + colecalciferol 400 UI/dia, lisinopril 5 mg/dia e pantoprazol 20 mg/dia. Negava raquialgia ou fraturas prévias e referia baixo aporte de cálcio na dieta (2 porções de leite ou derivados/dia). Analiticamente com défice de testosterona livre (5,2 pg/mL) e de vitamina D (44 nmol/L). A radiografia dorso-lombar mostrava fraturas de L1-L4. A densitometria óssea apresentava em L1-L4 densidade mineral óssea (DMO) de 0,855 g/cm2, T-score de -3,1 e Z-score de -2,9, no colo do fémur DMO de 0,368 g/cm2, T-score de -5,4 e Z-score de -5,6 e no fémur total DMO de 0.362 g/cm2, T-score de -5.6 e Z-score de -5.7. Foi feito o diagnóstico de osteoporose fraturária multifatorial (DMD, corticoterapia crónica, imobilização prolongada, défice de testosterona e baixo aporte de cálcio) e trocado o tratamento anti-reabsortivo para denosumab 60 mg semestral e calcifediol 0,266 mg/mês. Durante

o follow-up de 44 meses (8 administrações de denosumab), não ocorreram novas fraturas. Aos 24 meses, registou-se uma melhoria na DMO de 24% em L1-L4, de 10% no colo do fémur e de 11% no fémur total. Durante o seguimento, registou-se hipocalcemia ligeira assintomática de 8,2 mg/dL antes da 3ª toma de denosumab, revertida com aumento transitório da suplementação de cálcio e que não contraindicou a manutenção do fármaco.

Caso 2: Homem de 19 anos, com antecedentes de DMD, depressão e escoliose submetida a correção cirúrgica, sob prednisolona 5 mg/dia (desde há 14 anos, inicialmente sob 20 mg/dia), venlafaxina 75mg/dia e alprazolam 2 mg/dia. Referia história de fratura de baixo impacto da tíbia há 8 anos com incapacidade para a marcha desde então. Negava raquialgia, consumo de produtos lácteos ou tratamento anti-osteoporótico prévio. O estudo analítico não mostrou alterações, à exceção de hipovitaminose D (10 nmol/L). A radiografia dorso-lombar revelou uma fratura de D2. A densitometria óssea apresentava no colo do fémur DMO de 0,957 g/cm2, T-score de -0,9 e Z-score de 0,0 (não realizada a nível lombar por presença de material metálico). Foi feito o diagnóstico de osteoporose fraturária multifatorial (DMD, corticoterapia crónica, imobilização prolongada, baixo aporte de cálcio e défice de vitamina D) e iniciado tratamento com denosumab 60 mg semestral, carbonato de cálcio 1000 mg + colecalciferol 880 UI/dia e calcifediol 0,266 mg/mês. Durante o follow-up de 13 meses (3 administrações de denosumab), não foram reportados efeitos adversos nem ocorrência de novas fraturas.

Conclusão: Estes casos sugerem que o denosumab tem benefício no tratamento de osteoporose fraturária, mesmo em doentes sem capacidade de carga, promovendo a formação óssea tanto na coluna vertebral como no fémur. Existem apenas três outros casos publicados sobre o uso deste fármaco na osteoporose secundária a DMD mostrando igualmente eficácia na prevenção de novas fraturas, melhoria da DMO e boa tolerância[1,2]. São necessários estudos de maiores dimensões e com maior follow-up para confirmar estes achados.

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122 - ARTRITE, PSORÍASE E HLA-B27 – UM DIAGNÓSTICO INESPERADO

Diana Capela[1], Mariana Sá[1], Fonseca, R[2]
[1]USF Famílias, Santa Maria da Feira, Portugal, [2]Rheumatology, Centro Hospitalar Entre o Douro e Vouga, Santa Maria da Feira, Portugal

Introdução: A sífilis é uma doença sexualmente transmissível causada pelo *Treponema pallidum*. As manifestações clínicas podem ser muito variadas ou mesmo assintomáticos, dependendo da fase de infeção. A infeção pode ser precoce, nos 12 meses iniciais, que compreende a sífilis primária, secundária e latente precoce ou se não tratada pode evoluir para infeção tardia que inclui a sífilis latente tardia e a terciária. As manifestações musculoesqueléticas surgem habitualmente na sífilis secundária e podem confundir-se com outras patologias osteoarticulares inflamatórias.

Caso clínico: Homem de 37 anos, solteiro, mecânico, fumador, sem antecedentes médicos de relevo ou medicação crónica. Na família tinha avó com doença reumatológica e uma irmã com artrite psoriática. Foi encaminhado para consulta de reumatologia por quadro de 4 meses de evolução de lombalgia intensa, de características inflamatórias, que agravavam durante a noite e que estavam associadas a rigidez matinal, assim como artralgias migratórias com tumefação dos punhos e tornozelos. Não tinha obtido melhoria com anti-inflamatórios, mas sim com prednisolona oral na dose de 5 mg e agravamento quando tentava suspender. Ao exame objetivo, não apresentava sinovite e as manobras das articulações sacroilíacas não despertavam dor. Analiticamente apresentava aumento da VS (73mm/1ªh) e da PCR (59,8 mg/L), ANA e anti-dsDNA negativos, Ra teste negativo, HLA-B27 positivo, VDRL positiva (1/32) e sem outras alterações. O doente afirmava ter tido apenas 2 parceiras sexuais nos últimos 6 meses, negava úlceras genitais, contudo referiu o surgimento de um exantema há cerca de 1 ano, com resolução após medicação prescrita por dermatologista. O doente repetiu o estudo e mantinha a VS (71 mm/1ªh) e PCR (13,5 mg/L) elevadas e VDRL positivo, tinha os anticorpos Anti-*Treponema pallidum* IgM e IgG positivos e as serologias dos HIV, HVC e HVB estavam negativas. Nesta consulta, optou-se por encaminhar o doente para consulta de Infeciologia e medicou-se com Penicilina G benzatina 2,4 milhões UI, IM, dose única. Após 2 dias o doente ficou totalmente assintomático. Voltou a ser avaliado em consulta após 2 meses, onde se encontrava sem medicação e analiticamente tinha apenas ligeiro aumento da VS (16 mm/1ªh) e a PCR negativa. Portanto, foi assumido quadro de artrite em provável contexto de sífilis.

Discussão: A poliartralgia é dos sintomas mais frequente na nossa prática clínica, podendo ter múltiplas causas desde patologias autolimitadas a doenças incapacitantes, podendo ser o primeiro sintoma de uma doença sistémica. Dada a história familiar, história clínica e positividade do HLA-B27 a primeira hipótese de diagnóstico era de uma artrite psoriática ou espondilartrite. Contudo, a anamnese, os exames e evolução clínica permitiu o diagnóstico atempado e definitivo de sífi-

lis, assim como o seu tratamento dirigido.
Conclusão: Os sintomas musculoesqueléticos da sífilis secundária são manifestações pouco comuns, porém muito confundidores. Este caso clínico veio realçar a importância deste diagnóstico diferencial na avaliação de doentes com patologia osteoarticular inflamatória.

123 - LIKE WALKING ON PEBBLES - A SERIES OF THREE CASES OF PLANTAR FIBROMATOSIS

Marcelo Neto[1], Filipa Canhão André[1], André Saraiva[1], Margarida Coutinho[1]
[1]Serviço de Reumatologia, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

Background: Plantar fibromatosis (PF) is a rare disease characterized by disordered fibrous tissue proliferation and subsequent formation of nodules on the plantar aponeurosis – usually on the central or medial bands. Symptoms range from painless nodules to tender lesions that can affect the ability to bear weight. The presence of single or multiple nodules along the plantar aponeurosis is pathognomonic, but ultrasound and MRI are both acceptable imaging modalities to aid in the diagnosis. Compared with Dupuytren's disease, the upper extremity analogue of PF, relatively little has been published on this disease. This work aims to illustrate this pathology by the description of the following 3 cases:
Case 1: 50-year-old woman concerned with left plantar pain, especially on weight-bearing, and the presence of an ipsilateral plantar nodule. Physical examination revealed a left painless plantar nodule (Figure 1 – A) and ultrasonography confirmed the diagnosis by showing a nodule of approximately 1cm in diameter on the medial band of the plantar aponeurosis (Figure 1 – B). There was moderate improvement with soft insoles.

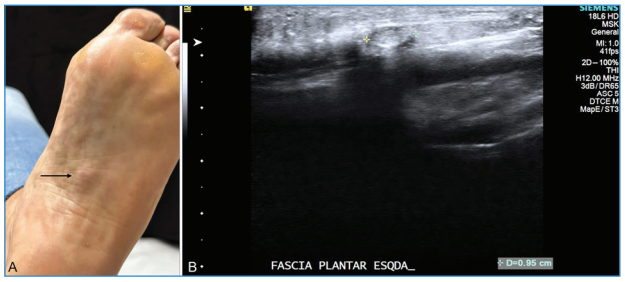

Figure 1. (Case 1): A - nodular lesion in the plantar surface of the left foot (arrow). B - musculoskeletal ultrasound of the left plantar aponeurosis (Siemens Acusion S-2000MT device, 18 MHZ transducer, longitudinal view): presence of an iso-hypoechoic lesion with well-defined borders adjacent to the plantar aponeurosis, with 0.95 cm of longitudinal diameter, without power doppler signal, suggestive of a plantar fibroma.

Case 2: 56-year-old woman, presenting with mild bilateral plantar pain triggered by long-distance walking. Multiple, palpable and mildly tender nodules on the right plantar aponeurosis were found on physical examination. The diagnosis was confirmed by ultrasonography showing exuberant focal thickening of the central plantar aponeurosis. The symptoms remain stable after 2 years of follow-up under conservative management.
Case 3: 72-year-old woman complaining of painful bilateral plantar nodules, particularly while bearing weight. Physical examination showed one slightly tender plantar nodule on the central part of each plantar aponeurosis. Ultrasonography was performed solely on the right foot, showing 2 nodules (14x5mm and 11x4mm) adherent to the plantar aponeurosis. After 1 year of follow-up, there was moderate clinical benefit with the use of soft insoles and a stretching program.
Discussion: Plantar fibromatosis is a scarcely studied condition, and even though its clinical presentation is diverse, it can lead to significant impact on daily activities. Treatment is mainly conservative owing to its benign nature and lack of quality evidence. Suggested therapies include steroid injections, verapamil, radiation therapy, extracorporeal shock wave therapy, and surgery in selected cases.

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127 - CLINICAL CASE: HEPATITIS B INFECTION - WHEN MUSCULOSKELETAL SYMPTOMS ARE INTERPRETED AS RHEUMATOID ARTHRITIS

Duarte Augusto[1], Sara Paiva Dinis [1], Filipe Cunha Santos[1], Ana Sofia Pinto[1], Cláudia Vaz[1, 2], Joana Fonseca Ferreira[1, 2], Catarina Quinaz[3], Nathalie Madeira[1]
[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: Hepatitis B (HBV) infection is related to a broad range of rheumatic symptoms, both in the acute and chronic phases. The most frequently reported HBV-associated rheumatic manifestations are serum sickness-like syndrome, arthritis and polyarteritis nodosa. A quarter of patients with chronic HBV complain of joint symptoms but severe joint damage is uncommon. Arthritis occurs as the result of formation

and deposition of immune complexes containing viral antigens.

Case report: We describe the case of a 67-year-old hypertensive female who, at first observation in our Rheumatology Department, provided us medical reports from her previous 2-year follow-up at another European Rheumatology Center that supported that she had been diagnosed with Rheumatoid Arthritis (RA). She described an insidious onset of inflammatory pain involving the metacarpophalangeal and proximal interphalangeal joints of the hands and swelling of the right hand that persisted for more than 6 weeks. At that time, hands MRI described 2nd, 3rd and 4th flexors rays' tenosynovitis of the right hand and mild 3rd and 4th flexors rays' tenosynovitis of the left hand; blood tests from the onset of symptoms with rheumatoid factor (RF) titer of 40 UI/mL (normal range, NR < 20), anti-cyclic citrullinated peptide (CCP) and antinuclear antibody (ANA) both negative, HBsAg and HBcAb both positive. Pain and swelling improved with 40 mg of prednisolone that was promptly reduced to 20 mg followed by tapering until suspension and introduction of 7.5 mg of weekly injectable methotrexate. The patient was unaware of the history of HBV and no references to therapy for this infection were mentioned in the medical reports.

During our observation, we noticed normal blood pressure, presence of 3 tender and 1 swollen joints, and absence of symptoms suggestive of neuropathy and of mucocutaneous lesions. Further investigation in our Center revealed hands and feet X-rays with degenerative changes without erosions and blood tests with positive HBsAg (1149.44 S/CO, NR < 1.0), positive HBsAb (12.76 mUI/mL NR < 10.0), positive HBcAb (7.28, NR <1.0), positive HBeAb (0.01, <1 positive), ANA titer of 1:160, positive IgM RF (21.0 UI/mL, NR < 5), positive HBV viral load (Genotype D, 677.4 UI/mL, NR < 3.8), C3, C4 liver function and renal tests in NR, abdominal ultrasound without changes.

Methotrexate was discontinued and observation by an Infectious Disease specialist was requested. In this evaluation, the patient confirmed family history of hepatitis B (several siblings and possibly also the mother), lack of appetite (but not significative weight loss), nocturnal sweating and fatigue. After discussion, probable musculoskeletal (MSK) involvement of HBV was considered and entecavir 0.5 mg per day was introduced, with consequent reduction of viral load until its non-detection. After 4 months of treatment, total resolution of MSK symptoms was verified and has remained so after one year of follow-up.

Discussion: The positivity of RF should be interpreted with caution in patients with MSK symptoms as it may be present in a variety of clinical conditions such as acute and chronic infections, including HBV. In addition to the positivity of HBsAg and HBcAb, the asymmetry of hand swelling, flexor tenosynovitis without extensor involvement and the negativity of anti-CCP could promptly have raised the suspicion of another diagnosis besides RA.

130 - EYE FOR SARCOIDOSIS: A CASE REPORT

Filipa Canhão André[1], André Saraiva[1], Sara Alves Costa[1], Margarida Coutinho[1]

[1]Rheumatology Department, Centro Hospitalar Universitário de Coimbra, Coimbra, Portugal

Background: Virtually all of the systemic inflammatory diseases that require rheumatologic care affect the eye or its surrounding structures. Diagnosis may be challenging, as many of the ocular manifestations are not specific, requiring a very thorough anamnesis.

Case report: Here in we report a case of a 66-year-old heavy smoker male, with the diagnosis of Rheumatoid arthritis (RA), treated with methotrexate and etarnecept. After being in sustained clinical remission for a long period of time, the patient began experiencing a progressive but sustained decrease in visual acuity on the right eye. This prompted an ophthalmologic evaluation that came to show a posterior chronic granulomatous uveitis. After a thorough assessment, the patient also admitted having persistent productive cough and right pleuritic chest pain. Infectious causes, like tuberculosis, were excluded and a high-resolution chest CT scan was performed. This showed mediastinal adenomegaly and discrete areas of ground glass densification. A 18F-fluorodeoxyglucose-positron emission tomography (18F-FDG-PET) was then used to assess extra-pulmonary involvement and it ended up confirming the lung changes and showing mediastinal-hilar and supraclavicular ganglion involvement, with increased uptake of 18F-FDG, suggestive of sarcoidosis. A tissue biopsy (lymph node) confirmed the diagnosis. As the patient developed an uveitis under etanercept, treatment was switched to adalimumab, with good response.

Discussion: This case had many confounding factors that hindered the diagnosis of sarcoidosis. The patient already had a rheumatic disease that could present with uveitis, even though scleritis is more common in RA. He was also a heavy smoker and was on methotrexate, both of which could be responsible for the patient complaints and pulmonary involvement. The CT scan showing hilar adenomegaly and the tissue biopsy with granulomatous involvement were key for diagnosis.

Conclusion: Sarcoidosis is an inflammatory disease that involves the eyes in 10-55% of cases, sometimes without systemic involvement. All eye structures can be

affected, but uveitis is the most common ocular manifestation and a worrying cause of vision loss.

Of note, etarnecept is the least effective anti-TNF for uveitis, which prompted the change to adalimumab.

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131 - RAYNAUD PHENOMENON AS AN ISOLATED MANIFESTATION OF AUTOIMMUNE/INFLAMMATORY SYNDROME INDUCED BY ADJUVANTS (ASIA)

Augusto Silva[1, 2], Elsa Vieira-Sousa[1, 2], Ana Rita Cruz-Machado[1, 2]

[1]Rheumatology and Bone Metabolic Diseases Department, Centro Hospitalar Lisboa Norte, EPE - Hospital de Santa Maria, Lisbon Academic Medical Centre, Lisboa, Portugal, [2]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introduction: In genetically predisposed individuals, some adjuvants, such as silicone widely used in the manufacture of medical implants, may trigger a set of symptoms, described as autoimmune/inflammatory syndrome induced by adjuvants (ASIA), also known as Shoenfeld's syndrome[1]. It may present a wide range of manifestations, including rheumatic and autoimmune-related symptoms such as arthralgia, myalgias, sicca syndrome, and Raynaud's phenomenon (RP). The presence of autoantibodies has also been reported(1,2).

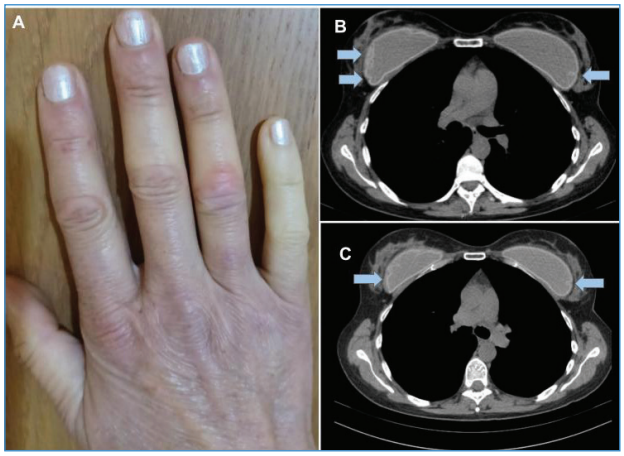

Figure 1. A: Raynaud's Phenomenon. Fig. 1B, 1C: HRCT revealing retro-mammary prostheses with bilateral intraprosthetic rupture

Several studies reported an increased incidence of RP in women with breast implants (BI), in most cases secondary to a systemic rheumatic disease or accompanied by other symptoms[1-3]. In neither of these cases, RP appeared as the only symptom or there was a spontaneous resolution, even after BI removal[2-7].

Case-report: A 51-year-old female, followed at our Rheumatology outpatient clinic over the last 15 years for spondyloarthritis, presented with a new onset biphasic RP on both hands and feet (Figure 1), manifested by exposure to cold and not associated with digital ulcers, for the last six months. She reported dry cough for more than one year, with no other de novo symptoms or traumatic injuries. One and a half years before, she received silicone BI for aesthetic reasons. She had a background medical history of smoking, chronic gastritis and periarticular shoulder pathology. She was taking sulfassalazine, acemetacin, esomeprazole and cyclobenzaprine. Symptomatic therapy for RP was not initiated considering its benign course.

Blood results were unremarkable. Antinuclear and extractable nuclear antigen antibodies were negative, however on the Systemic sclerosis (SSc) immunoblot panel, anti-RNA-polymerase II was transiently weak positive, and ten months later anti-Th/To also tested weak positive. Capillaroscopy and lung function tests were normal. High-resolution chest computed tomography revealed diffuse centrilobular emphysema and the presence of retro-mammary prostheses with bilateral pleating, possibly corresponding to intraprosthetic rupture (figure 1). Between appointments, she removed her both BI and two weeks later she reported a spontaneous resolution of RP, that has not relapsed over the last year.

Discussion: Silicone in BI is usually composed by various sized water-insoluble straight chains of polydimethylsiloxanes, many of them smaller than the pores of the shell. Leakage can occur with or without the shell rupture. This phenomenon can trigger an immune response(8) and subsequent ASIA.

The vast majority of cases of RP reported after silicone BI implantation, are considered secondary to systemic rheumatic diseases, usually SSc (1-4,6). Following BI removal, the disease frequently persists, although cases of improvement have been also reported. In most instances, regardless of whether BI are removed or not, clinical benefit was only noted after the initiation of vasodilating, antiplatelet or immunosuppressive therapy[1,5,7].

This case is unique because RP appeared as an isolated symptom and totally resolved after the removal of BI, meeting ASIA syndrome 2 major criteria, in

line with the diagnostic criteria for ASIA developed by Shoenfeld et al[9].

Conclusion: To the best of our knowledge, this is the first description of RP as an isolated manifestation of ASIA syndrome that totally reverted after the removal of silicone BI. We hope our report contributes to raise awareness of similar manifestations of Schoenfeld's syndrome that may benefit, when feasible, from the removal of the inciting agent.

133 - SUSPICIOUS LYMPHADENOPATHIES: A RARE DIAGNOSIS AMONG A MYRIAD OF POSSIBILITIES

Maria João Cadório[1], João Gama[2], João Alexandre Oliveira[1], Fernando Albuquerque[1], Marcelo Neto[1], Adriana Carones[1], Beatriz Mendes[1], Ana Isabel Maduro[1], André Saraiva[1], Cátia Duarte[1, 3]

[1]Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal, [2]Anatomical Pathology Department, Coimbra, Portugal - Centro Hospitalar Universitário de Coimbra, Coimbra, Portugal, [3]Coimbra Institute for Clinical and Biomedical Research (iCBR) - Faculty of Medicine, University of Coimbra, Coimbra, Portugal

Introduction: Kikuchi-Fujimoto Disease (KFD), also known as Kikuchi histiocytic necrotizing lymphadenitis, is an extremely rare and benign condition characterized by lymph node involvement and low-grade fever. KFD has been reported in association with Systemic Lupus Erythematosus, and, to a lesser extent, other immunomodulated inflammatory rheumatic diseases, such as Sjögren syndrome.

Clinical Case: We report the case of a 66-year-old woman currently followed in a rheumatology clinic, with primary Sjögren syndrome diagnosed at the age of 45, and diffuse large B cell lymphoma (DLBCL) in remission after treatment with R-CHOP chemotherapy. She presented with recent constitutional symptoms, namely fever, night sweats, weight loss and anorexia, associated with multiple tender lymphadenopathies. These findings raised the hypothesis of lymphoproliferative disease relapse or active Sjögren syndrome. Lab results showed normal cell blood count, acute phase reactants, renal function and urine analyses, with a polyclonal hypergammaglobulinemia (IgG and IgM), hypocomplementemia (C3 and C4), high levels of rheumatoid factor and positive ANAs, with positive anti-SSA and -SSB antibodies. PET scan revealed supra and infra-diaphragmatic hypermetabolic lymphadenopathies, concerning of lymphoma involvement. However, histopathology of a cervical ganglia obtained via excisional biopsy revealed lymphadenitis with monocytoid hyperplasia and necrosis (Figure 1), findings suggestive

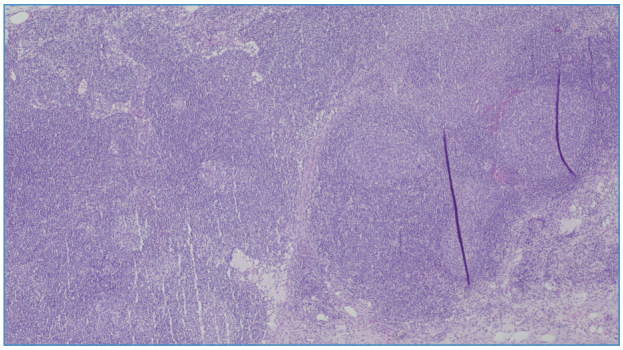

Figure 1. Cervical lymph node on microscopy

of Kikuchi-Fujimoto Disease. The patient was started on prednisolone 0,5 mg/kg/day on a slow tapering regimen, with resolution of her symptoms through follow-up.

Discussion: KFD, also known as Kikuchi histiocytic necrotizing lymphadenitis, is an extremely rare and benign condition that mostly affects young women. It is characterized by lymph node involvement with a predilection for the cervical region, commonly presenting with tender lymphadenopathy and low-grade fever. The diagnosis requires lymph node biopsy. KFD mostly self-resolves within a few weeks to months, and only some patients require symptomatic relieve with NSAIDs or corticosteroids. A minority of cases develop recurrent episodes of the disease, and it has been repeatedly reported in association with SLE, and, to a lesser extent, other immunomediated inflammatory rheumatic diseases, such as Sjogren syndrome, whose clinical presentation may include lymphadenopathy. For these reasons, KFD patients must undergo a regular follow-up for several years.

Conclusion: With this case report, the authors aim to raise awareness to a rare diagnosis that, although benign, can mimic serious conditions that must be excluded from the differential, which, in this patient, was especially difficult given her complex past medical history.

136 - IT'S MORE THAN LUPUS

Beatriz de Carvalho Mendonça[1], Susana Fernandes[1], Helena Santos[1]

[1]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal

Systemic Lupus Erythematosus (SLE) is an immune-mediated, multisystemic disease with a variable course and prognosis. It's associated with several comorbidities, including certain malignancies. According to a 2021 meta-analysis, it was identified an 18% increased risk of cancer among patients with SLE compared with the general population. In these patients, Hodgkin's and Non-Hodgkin's lymphomas have the highest risk.

In addition to these, there is also an increased risk of monoclonal gammopathies, such as monoclonal gammopathy of undetermined significance (MGUS) which is a premalignant condition of multiple myeloma (MM). Despite this, the association between SLE and MM is extremely rare. A case is presented of a patient who developed MM 6 months after SLE diagnosis.

A 67-year-old healthy woman, with no usual medication or drug allergies, presented to the Rheumatology consultation for inflammatory, symmetrical and additive polyarthralgias of the shoulders, wrists, metacarpophalangeal (MCF) and tibiotarsal joint, associated with nocturnal awakenings with 2 weeks of evolution. She denied other systemic signs and symptoms. On physical examination it was noted symmetrical polyarthritis of wrists and MCFs. Analytically with ANA + 1/640; FR +; Anti dsDNA +; Anti SSA +++ Anti SSB + and hypovitaminosis D, she was diagnosed with SLE according to EULAR/ACR 2019 criteria. She was treated with prednisolone 10mg/day, methotrexate 15mg/week, folic acid 10mg/week and vitamin D and calcium replacement, with remission of polyarthritis in 4 months. Simultaneously to the remission, she presented acute incapacitating low back pain, without irradiation and without associated trauma. An X-ray and MRI of the spine revealed new vertebral fractures in D12, L1 and L2. To clarify these fractures, a thoracic-abdominal-pelvic CT was carried out, as well as bone scintigraphy, analyses of phospho-calcium metabolism, serum and urinary immunofixation and Bence Jones proteinuria, which results were negative with the exception of hypovitaminosis D (9.7 [30-100 ng/mL]). As her back pain remained refractory to analgesia and spinal orthosis, she underwent vertebroplasty. The biopsy of the surgical specimen revealed medullary infiltration by CD138+ cells, very suggestive of MM. A myelogram and bone marrow biopsy confirmed the diagnosis of non-secretory MM. She discontinued all rheumatologic medication to begin a cycle of chemotherapy with cyclophosphamide, bortezomib, and dexamethasone. After the end of chemotherapy and zoledronic acid, the patient remained with controlled pain, no arthritis and no evidence of lupus activity.

This clinical case is a diagnostic challenge, since besides the coexistence of these two diagnoses being rare, non-secretory MM associated with SLE is even rarer. Non-secretory MM is defined by the presence of plasmacytoma or by ≥10% plasma cells in the myelogram, and electrophoresis and immunofixation are negative. The development of MM in SLE patients most often occurs years after the diagnosis of SLE, as opposed to the clinical case described. The mechanisms explaining this coexistence are unclear, but the following theories have been proposed: (a) B-cell hyperactivity and defective immune surveillance may help B-cell clones escape the immune system; (b) increased resistance to cell apoptosis; (c) SLE immunosuppressive therapy may increases the risk of malignancies. In conclusion, in SLE patients presenting with atypical clinical features and/or a monoclonal peak on electrophoresis, monoclonal gammopathies should be suspected.

139 - (NÃO SÓ) UMA POLIARTRITE PÓS COVID-19 - UM CASO DE SÍNDROME ANTISSINTETASE

Inês Santos[1], Maria Inês Seixas[1], Ana Viegas[2], Inês Almeida[1], Telma Santos[3], Paulo Monteiro[1], Couto M[1]
[1]Unidade de Reumatologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [2]Serviço de Medicina Interna, Centro Hospitalar Tondela-Viseu, Viseu, Portugal, [3]Serviço de Neurologia, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Introdução: O diagnóstico de várias doenças imunomediadas, entre as quais miopatias inflamatórias idiopáticas (MII), após infeção ou vacinação COVID-19 tem sido reportado na literatura. A síndrome antissintetase é um subtipo de MII caracterizado por doença intersticial pulmonar e/ou miosite e positividade para anticorpos antissintetase (autoanticorpos específicos de miosite), sendo o mais frequente o anti-Jo1. Outros sintomas incluem artrite, "mãos de mecânico", fenómeno de Raynaud e febre. Diferentes autoanticorpos associam-se a subgrupos fenotipicamente distintos. O anticorpo anti-Jo1 está associado a um envolvimento muscular mais frequente e mais grave. Os autoanticorpos associados a miosite podem estar presentes, sendo que o anti-Ro52 se relaciona com artrite mais precoce e "mãos de mecânico".
Caso clínico: Doente de 45 anos, caucasiano, com quadro de poliartrite envolvendo punhos e pequenas articulações das mãos, com cerca de 7 meses de evolução e que teve início 1 semana após doença ligeira por infeção SARS-CoV-2. Veio a cumprir duas doses da vacina contra a COVID-19 aos 6 e aos 13 meses após o quadro infecioso. Sem outros antecedentes pessoais relevantes. Negou sintomas constitucionais e outras queixas de órgãos ou sistemas. A radiografia das mãos e punhos não demonstrou alterações de relevo. Analiticamente destacava-se hiperuricemia 7,3 mg/dL e hiperferritinemia 371 ng/mL; sem alterações do hemograma, parâmetros inflamatórios, função renal, provas hepáticas, enzimas musculares, função tiroideia ou proteinograma eletroforético; ANA, FR e aCCP negativos; serologias VHB, VHC e VIH negativas. Iniciou metilprednisolona na dose máxima de 8 mg id e metotrexato titulado até 20 mg/semana subcutâneo. Após 7 meses de terapêutica, iniciou queixas de mialgias e noção de fraqueza muscular proximal com agravamento progressivo, sem

sintomas respiratórios. Ao exame objetivo apresentava paraparésia proximal simétrica grau 4 e reflexos osteotendinosos ligeiramente diminuídos nos membros inferiores, bem como fissuração e descamação ao nível das polpas digitais ("mãos de mecânico"); sem alterações sensitivas ou outras lesões cutâneas. Analiticamente destacava-se elevação das enzimas de citólise e musculares (ALT 219 UI/L, AST 172 UI/L, LDH 523 UI/L, CK 3422 UI/L), pelo que o metotrexato foi suspenso para estudo complementar. A ecografia abdominal e renovesical não revelou alterações. O estudo imunológico revelou ANA negativo, anti-Jo1 +++ e anti-Ro52 ++. O eletromiograma foi compatível com MII e a biópsia muscular confirmou o diagnóstico. Foi iniciada terapêutica com prednisolona 60 mg id. A TCAR torácica revelou achados ligeiros de pneumonite intersticial não específica e as PFR, DLCO e gasimetria arterial eram normais. Cumpriu 3 pulsos de metilprednisolona 1 g e iniciou terapêutica com rituximab, apresentando melhoria clínica e analítica progressivas, com normalização da CK aos 3 meses. Veio a apresentar reagravamento muscular aos 6 meses após o 1º ciclo de rituximab, à data sob micofenolato de mofetil 2 g id e prednisolona 7,5 mg id. Cumpriu 3 pulsos de metilprednisolona 500 mg, aumento da dose de prednisolona oral para 60 mg id e retratamento com rituximab.

Discussão: As formas incompletas de síndrome antissintetase requerem um alto índice de suspeição para assegurar um diagnóstico precoce. Os clínicos devem manter-se vigilantes sobre um potencial envolvimento articular e/ou muscular após infeção ou vacinação COVID-19, ainda que sejam necessários estudos adicionais acerca das suas associações a MII.

146 - LIPOMA ARBORESCENS – DON'T MISS THE DIAGNOSIS!

Inês Almeida[1], Maria Inês Seixas[1], Inês Santos[1], José António Gomes[2], Luísa Negrão[2], Paulo Monteiro[1], Couto M[1]
[1]Rheumatology Department, Centro Hospitalar Tondela -Viseu, Viseu, Portugal, [2]Orthopaedics Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Introduction: Lipoma arborescens (LA) is a rare benign lesion characterized by villous proliferation of the synovium, with replacement of the sub-synovial tissue by mature adipocytes. It consists of a non-specific reactive response to chronic synovial irritation due to mechanical or inflammatory insults. It most commonly involves the suprapatellar pouch of the knee, and the majority of cases are monoarticular, with progressive joint swelling, which may be associated with effusion and pain. Magnetic resonance imaging (MRI) is the gold standard for the diagnosis of LA and open or arthroscopic synovectomy is the treatment of choice which could delay osteoarthritis.

Case Report: A 38-year-old female presented with relapsing bilateral knee swelling and moderate pain over the past 24 years, without involvement of other joints. Multiple arthrocentesis were performed, in urgency departments, without synovial fluid analysis. She denied fever, bleeding tendencies, loss of weight or trauma. On examination, both knees were enlarged, tender and felt warmer than the surrounding skin, with a positive patellar tap test indicating a moderate knee effusion. There was no tenderness at tendon insertion. Laboratorial study revealed normal blood count, ESR 8.0 mm/hour, CRP 0.55 mg/dL, uric acid level was normal, positive ANA 1/320, homogeneous pattern), anti-ds-DNA, ENA screening, RF, anti-CCP and HLA-B27 were negative. Hepatitis B, C and HIV serologies were also negative. Knee radiographs were normal. Ultrasound of the left knee revealed villous projections of synovial membrane and a large effusion in the suprapatellar recess. Synovial fluid analysis showed 102 leukocytes/mm3 with a mononuclear cells predominance, without microbial development on culture. MRI showed bilateral effusion, more expressive on the left knee, containing suspended material with a micronodular appearance, without clear identification of hemosiderin deposits and lacking the typical picture of pigmented villonodular synovitis. Admitting the diagnosis of undifferentiated inflammatory oligoarthritis, a conservative treatment was the initial approach with oral prednisolone at a maximum dose of 10 mg/day and methotrexate at a maximum dose of 17,5 mg/week, with no improvement. Arthrocentesis

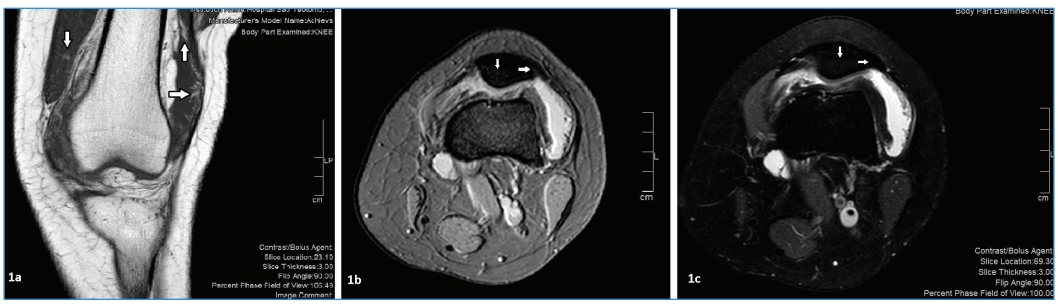

Figure 1. MR images

and intra-articular injection with triamcinolone hexacetonide had only brief relief. The case was discussed with an orthopaedic surgeon, and we decided to proceed to arthroscopic synovial membrane biopsy, which was consistent with moderate chronic synovitis with extensive lipomatosis and an arthroscopic synovectomy was proposed.

Discussion: This case highlights LA as a rare condition with specific imaging features, particularly on MRI, that are important for early recognition and to avoid misinterpretation with other intra-articular masses (non-infectious synovial proliferative lesion, infectious granulomatous lesion, depositional joint disease, neoplastic lesion or vascular malformation) that require a different approach. A high grade of suspicion is needed, particularly in the face of chronic mono or oligoarticular swelling with recurrent joint effusion episodes refractory to conservative management. The treatment of choice is either open or arthroscopic synovectomy, with uncommon recurrence and delay of secondary degenerative changes.

148 - ARTRITE REUMATÓIDE - O DESAFIO MULTIDISCIPLINAR

Alice Neves[1], Catarina S Cortesão[1], Catarina Artilheiro[2], Cláudia Miguel[1], Helena Santos[1]

[1]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal, [2]Internal Medicine, Hospital Garcia de Orta, Almada, Portugal

Introdução: A Artrite Reumatóide é uma patologia inflamatória crónica que afeta principalmente as pequenas articulações e estruturas periarticulares e tem adicionalmente um grande número de manifestações extra-articulares. Para além disso, as opções terapêuticas disponíveis têm efeitos adversos que exigem uma adequada monitorização.

Assim, os pacientes com Artrite Reumatóide necessitam de uma abordagem multidisciplinar, que engloba Reumatologia e várias outras especialidades. Descreve-se o caso de uma doente com necessidade de abordagem de patologias graves em vários domínios da medicina - Infecciologia, Gastroenterologia, Medicina Interna, Neurologia, Cardiologia.

Caso clínico: Sexo feminino, 63 anos, seguida no Instituto Português de Reumatologia desde 2006 por Artrite Reumatóide, com fator reumatóide e anticorpo anti-proteínas citrulinadas positivos. Outros antecedentes pessoais conhecidos: hipertensão arterial, dislipidemia, osteoartrose e tuberculose pulmonar tratada na infância.

Medicada com Metotrexato e Deflazacorte, por manutenção da atividade da doença, iniciou tratamento com Adalimumab em 2009. Previamente, foi observada em consulta de Pneumologia e realizou terapêutica profilática com Isoniazida. Após 7 meses de Adalimumab, atingiu baixa atividade da doença.

Em 2019 iniciou quadro de amigdalites purulentas de repetição, que resolviam com a administração de antibioterapia de primeira linha, mas recorriam posteriormente, obrigando a múltiplas paragens da administração do biotecnológico. Por este motivo, iniciou terapêutica com Penicilina intramuscular mensal em 2020 que mantém até à atualidade, sem novas recorrências de amigdalite, o que permitiu retomar regularmente o tratamento com Adalimumab, com ajuste da dose.

Em 2022 realizou colonoscopia que identificou uma lesão infiltrativa, ulcerada com envolvimento da válvula ileocecal e mucosa subjacente cuja biópsia confirmou tratar-se de tuberculose intestinal. Foi excluído o envolvimento de outros órgãos.

Iniciou terapêutica antibacilar quádrupla e suspendeu terapêutica com Adalimumab, o que levou a um agravamento da artrite e necessidade de reintrodução de corticoterapia sistémica com Prednisolona.

Em Setembro de 2022 em ECG de rotina, foi diagnosticada uma fibrilhação auricular e iniciou anticoagulação oral.

Em Fevereiro de 2023 iniciou um quadro súbito de perda de força muscular e desvio da comissura labial à esquerda. Realizou estudo por TC-CE que confirmou a oclusão do segmento M1 da artéria cerebral média direita. Por não apresentar contraindicações e se encontrar dentro da janela terapêutica realizou trombectomia aspirativa. O estudo etiológico realizado era sugestivo de causa cardioembólica. A doente recuperou totalmente, não se registando défices neurológicos sequelares.

Atualmente aguarda colonoscopia de reavaliação, para confirmação da cura da tuberculose intestinal e ponderação de reinício de terapêutica biotecnológica. O score DAS-28 atual é de 2.8.

Discussão: Apresenta-se este caso pela complexidade da abordagem de uma artrite reumatóide grave, com necessidade de terapêutica biotecnológica, que se tornou um desafio pelas complicações infecciosas - amigdalites purulentas e tuberculose intestinal, mas também pelas comorbilidades cardiovasculares - fibrilhação auricular, acidente vascular cerebral - que, apesar dos tratamentos bem sucedidos evidenciam o elevado risco destas comorbilidades nestes doentes.

153 - MIOPATIA NECROTIZANTE IMUNO-MEDIADA (MNIM) APÓS INFEÇÃO POR SARS-COV-2: A IMPORTÂNCIA DA DETEÇÃO PRECOCE

Carlos Marques-Gomes[1, 2], Mariana Diz-Lopes [1, 2], Miguel Bernardes[1, 2], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar de São João, Porto, Portugal, [2]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal

Caso clínico: Sexo feminino, 41 anos, admitida a internamento de Reumatologia em julho de 2020 para estudo de provável doença do tecido conjuntivo inaugural. Apresentava história de poliartralgias de ritmo inflamatório (punhos, metacarpofalângicas e interfalângicas proximais, bilateralmente) associadas a sensação de edema difuso das mãos, com 6 meses de evolução, descrevendo, nas mãos, um fenómeno de Raynaud de novo, desde há 2 meses. Ao exame objetivo, destacava-se hipertensão arterial de novo, puffy hands e esclerodactilia bilaterais, cianose do 1º ao 4º dedos de ambas as mãos e incapacidade de preensão palmar (sobretudo, à direita). A videocapilaroscopia do leito ungueal mostrou, sobretudo, capilares desorganizados e muito arborizados e abundantes megacapilares. Dos antecedentes pessoais patológicos, salientava-se infeção assintomática por SARS-CoV-2, detetada pelo método de reverse transcription polymerase chain reaction (RT-PCR), há 3 meses atrás. Analiticamente, apresentava elevação dos parâmetros inflamatórios (VS 78 mm/1ªh, PCR 7,2 mg/L), das enzimas musculares (CK 1642 U/L, aldolase 26,3 U/L, CK-MB 35 ng/mL, mioglobina 303 U/L), proteinúria de novo (1 g/24h; função renal normal e ecografia reno-vesical sem alterações), ANA 1/1000 (padrão mosqueado), anticorpos anti-RNP e anti-SSA positivos, anti-Ro52 positivo fraco (restantes auto-anticorpos negativos, incluindo os dos painéis de esclerose sistémica e miosites). O estudo infecioso foi negativo. Realizou ressonância magnética das coxas que identificou sinais de edema muscular nas junções miotendinosas dos músculos isquiotibiais, sartórios e adutores, bilateralmente. A biópsia muscular revelou características de miopatia necrotizante imuno-mediada (MNIM). Foi iniciado tratamento com prednisolona (7,5mg/dia), hidroxicloroquina (HCQ) (400mg/dia), metotrexato (MTX) (17,5 mg/sem, via oral) e fármacos antihipertensores. Ao fim de 2 anos de follow-up, a doente manteve-se sob imunossupressão com HCQ e MTX, já sem corticoterapia, sem evidência de défice de força muscular e com normalização das enzimas musculares.

Discussão: As infeções virais respiratórias sintomáticas são, muitas vezes, acompanhadas de envolvimento muscular agudo que pode originar rabdomiólise aquando ou logo após o período de sintomatologia respiratória e, nem sempre, evoluir para miosite. A MNIM é uma patologia rara (10 a 20% de todas as miopatias inflamatórias) que se caracteriza pela existência de necrose de fibras musculares com pouca ou nenhuma inflamação na biópsia muscular. Apesar de ser frequentemente idiopática, existem raros casos descritos de MNIN após a infeção por SARS-CoV-2 (embora a fisiopatologia desta associação não seja bem conhecida), cuja apresentação clínica foi em tudo semelhante à de uma miosite, com fraqueza muscular e elevação marcada das enzimas musculares.

Apresentamos um caso de doença mista do tecido conjuntivo com miopatia necrotizante imuno-mediada subclínica objetivada 2 meses após infeção por SARS-CoV-2. Embora a doente não apresentasse manifestações clínicas exuberantes, foi necessário o início de imunossupressão para o controlo da doença, de forma a impedir a progressão para um quadro de miosite grave (cujo tratamento, na maioria dos casos, passa por imunossupressão mais agressiva). Este caso clínico enfatiza a importância do reconhecimento de fenótipos subclínicos, mais atípicos, da MNIM num contexto de pós-infeção viral, reforçando o facto de que o início precoce de tratamento adequado pode ser suficiente para uma evolução favorável da doença.

156 - A RARE CASE OF CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY (CIDP) ASSOCIATED WITH ACTIVE SYSTEMIC LUPUS ERYTHEMATOUS (SLE): ANIFROLUMAB AS A SUCCESSFUL THERAPEUTIC ASSET

Carlos Marques-Gomes[1, 2], Mariana Diz-Lopes[1, 2], Ana Martins[1, 2], Daniela Oliveira[1, 2], Rafaela Nicolau[3], Inês Santos[3], Luís Braz[4], Miguel Bernardes[1, 2], José Pinto[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]Rheumatology Unit, Centro Hospitalar Tondela -Viseu, Viseu, Portugal, [4]Serviço de Neurologia, Centro Hospitalar de São João, Porto, Portugal

Introduction: Chronic inflammatory demyelinating polyneuropathy (CIDP), rarely reported as being associated with systemic lupus erythematous (SLE), is the most common form of acquired autoimmune peripheric neuropathy. This condition manifests as progressive muscle weakness, with or without sensory loss, sometimes coinciding with a lupus flare. The treatment strategy should include SLE disease activity control; intravenous immunoglobulins (IVIg) and immunosuppressants are mostly used. Anifrolumab, a newly authorized monoclonal antibody, has demonstrated efficacy in treating moderate to severe SLE.

Presentation: A 53-years-old woman was diagnosed with SLE [positive antinuclear (ANA) and anti-(double stranded)-DNA (anti-dsDNA) antibodies (atbs)] and secondary Sjögren's syndrome (SS) at the age of 26. She had articular (polyarthritis), mucocutaneous (malar rash, photosensitivity, recurrent oral ulcers), hematologic (anemia, leukopenia) and lung (UIP) involvement, Raynaud's phenomenon, subacute cutaneous lupus and

cutaneous vasculitis in the hands. Sicca symptoms, positive anti-SSA/anti-SSB atbs and minor salivary gland biopsy confirmed the association with SS. During the disease course, several lupus flares occurred, and she was treated with prednisolone, hydroxychloroquine 400 mg/day (since the diagnosis), azathioprine 150 mg/day (from 2005 to 2019) and rituximab (2 cycles of 1g each). In 2016, she developed persistent hypogammaglobulinemia (unrelated to rituximab usage) and suffered from recurrent respiratory infections, requiring IVIg infusions. Because of allergic reaction to IVIg, the treatment was interrupted for approximately one year, but it was restarted in 2022 and the patient tolerated successive infusions (with premedication protocol) until July 2022, when she missed medical appointment.

In September 2022, a new lupus flare has occurred and treatment with anifrolumab was approved. Additionally, she was admitted with pyelonephritis successfully treated. On admission, in October 2022, she presented with a two months history of polyarthritis and distal muscular weakness witch evolved to loss of proximal muscle strength. On physical examination, tetraparesis and hypoesthesia of the hands and feet was present. Pleural effusion, leukopenia, elevated anti-dsDNA atbs and low complement levels confirmed SLE disease activity. Nerve conduction studies revealed a diffuse and symmetrical, sensory-motor demyelinating neuropathy, with signs of axonal damage, compatible with CIDP. Treatment with methylprednisolone (500 mg/5 days), IVIg (2 mg/Kg/day) once every 3 weeks and anifrolumab (300 mg/28 days) was started. The patient was admitted to a rehabilitation center. After 2 anifrolumab infusions and 5 IVIg cycles, there were no signs of disease activity and she started to partially perform some of her daily routine with the help of others.

Discussion: This long-standing SLE patient had poor disease control (despite the use of several immunosuppressants, including rituximab) and developed CIPD, concurrently with a lupus flare, even after years of IVIg treatment (albeit at the lowest dose used in hypogammaglobulinemia). With anifrolumab and IVIg therapy there was clinical and immunological improvement. The case highlights the need to achieve disease control in order to treat and prevent such rare associated complications that must be promptly identified. We described the first Portuguese case of CIPD occurring together with SLE in which we observed a clear effectiveness of anifrolumab in managing the complications arising from active severe SLE.

160 - VASCULITE ISOLADA DA ARTÉRIA HEPÁTICA

Susana Matias[1], Maria José Santos[1], Ana Cordeiro[1], Filipe Vinagre[1], Raquel Freitas[1], Inês Ferreira[2]
[1]Serviço de Reumatologia, Hospital Garcia de Orta, Almada, Portugal, [2]Nuclear Medicine, Hospital Garcia de Orta, Almada, Portugal

Introdução: As vasculites são um grupo heterogéneo de doenças sistémicas caracterizadas por inflamação dos vasos sanguíneos. O atingimento de órgão único (single organ vasculites-SOV) coloca particular desafio no diagnóstico e seguimento. São raros os casos descritos de SOV do trato gastrointestinal, afetando predominantemente a vesícula biliar, o apêndice e o intestino delgado. A apresentação clínica pode incluir dor abdominal, náuseas, vómitos, hemorragias, mas alguns casos são diagnosticados acidentalmente em exames de imagem ou em peças cirúrgicas. Abaixo, descrevemos um caso clínico de vasculite isolada da artéria hepática, sendo que na literatura encontrámos apenas outros três casos publicados.

Caso Clínico: Doente do sexo masculino, 54 anos, sem antecedentes pessoais relevantes. Admitido no serviço de urgência por quadro com 10 dias de evolução de dor abdominal na região epigástrica, intensidade 5/10 com irradiação para os quadrantes inferiores, náuseas e anorexia não seletiva. Adicionalmente, referia obstipação desde há 5 dias. Objetivamente apenas a salientar hipertensão arterial (180/109mmHg), sendo a palpação abdominal indolor. Analiticamente apresentava ligeiro aumento da proteína C reativa (1.03mg/dL) e VS 39 mm na 1ªhora, sem leucocitose, anemia ou trombocitose. A radiografia do abdómen em pé não

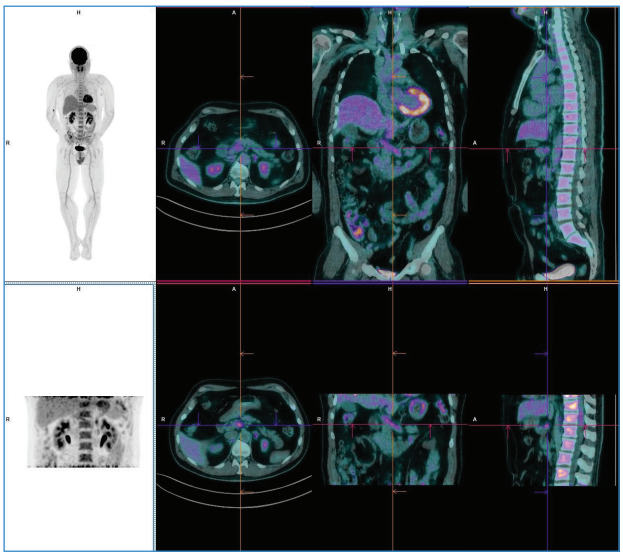

Figura 1. PET scan com FDG mostra aumento difuso moderado e relativamente homogéneo da captação do radiofármaco em todo o trajeto da artéria hepática comum (ver ponto focalizado), tanto no estudo inicial (painéis superiores) como nas imagens tardias, (painéis inferiores).

mostrava alterações. Por fraca resposta a analgesia e ausência de restabelecimento do trânsito com enemas, solicitou-se tomografia computorizada (TC) abdomino-pélvica que documentou edema da gordura que envolve o tronco celíaco. Para melhor esclarecimento, realizou angio- TC, que revelou uma hipodensidade mais expressiva ao nível da artéria hepática comum, que parecia traduzir um aumento da espessura parietal do vaso, estando o lúmen filiforme, tendo-se colocado a hipótese diagnóstica de vasculite. Tal diagnóstico, foi confirmado por angio-RM e PET Scan (Figura 1), sem evidência de envolvimento de outros territórios vasculares. O doente apresentava imunologia negativa, tendo sido excluídas causas infecciosas virais, bacterianas e parasitológicas. Iniciou corticoterapia sistémica com prednisolona 1mg/kg/dia com resolução das queixas e melhoria das alterações laboratoriais. Aos seis meses de terapêutica e atualmente sob 10mg/dia de prednisolona, o doente está assintomático, apresentando uma PCR de 0.28mg/dL, VS de 27 e sem evidência de envolvimento de outros órgãos.

Conclusão: Este caso clínico destaca-se pela raridade de vasculite isolada da artéria hepática e paucidade na apresentação clínica ,reforçando a importância da suspeição diagnóstica e a mais valia dos exames imagiológicos. Levanta também questões sobre a terapêutica mais apropriada e como se deve fazer a monitorização da doença.

164 - PARANEOPLASTIC RAYNAUD'S PHENOMENON: CASE REPORT

Mariana Rocha Sebastião[1], Luís Maurício Santos[1], Carolina Furtado[1], Teresa Novoa[1]
[1]Rheumatology Department, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal

Introduction: Raynaud's phenomenon (RP) is a vascular acrosyndrome characterised by the classic triphasic colour change of the extremities. According to its association with an underlying systemic disease, it can be referred to as primary or secondary RP. Paraneoplastic RP (PRP) is a rare complication of a variety of malignancies: it can appear at any stage of the disease and has been reported as an initial manifestation.

Case Report: A 56-year-old man presented with a 3-week history of painful cyanosis of the upper extremities and hardened scaly skin of the fingertips. Other than a 60 pack-year smoking habit, he had no relevant medical or occupational history, nor presented any other symptom. On examination he revealed RP of all 10 fingers, with severe allodynia and hardened skin. All upper limb pulses were normal, which was confirmed by continuous wave Doppler. Initial etiological investigations excluded diffuse connective tissue disease, vasculitis, antiphospholipid syndrome or embolic source. Treatment with Alprostadil, Sildenafil and Bosentan during hospital admissions, and with Nifedipine in ambulatory care, led to an initial mild subjective and objective improvement, but didn't prevent ischemic complications: ulceration and necrosis of several fingers and toes, with concomitant infections. These resulted in the recurrent need for surgery: partial amputation of the second finger of both hands and Chopart amputation of the left foot. Chest-abdomen-pelvic angio-CT showed right paratracheal lymph nodes enlargement. Biopsy performed via mediastinoscopy revealed Diffuse large B cell lymphoma. After 4 months of combination chemotherapy there was complete remission of the RP, and no other ischemic events occurred. At the 6-month follow-up PET-scan showed no evidence of malignancy, and no vascular symptoms recurred after 1 year.

Conclusion: Although PRP has been reported in many types of cancer, it is more frequent in lung, ovarian and gastric carcinomas, as well as in lymphomas and leukaemia. Secondary RP more often presents after 50 years of age with asymmetrical distribution, associated with pain and ischemic complications (ex. ulceration, gangrene). PRP should be suspected when there is poor response to the usual initial treatment, rapid progression to gangrene, and when other more common etiologies have been excluded. PRP's clinical course is similar to that of the underlying neoplasm: it usually resolves with the malignancy's effective treatment and its recrudescence should raise suspicion of cancer recurrence.

166 - LIFE-THREATENING HYPEREOSINOPHILIC SYNDROME IN A PATIENT WITH RHEUMATOID ARTHRITIS: A CASE REPORT

Mariana Emília Santos[1, 2], Maria João Gonçalves[1], Ana Ramalhal Jorge[3], Joana Vasconcelos[4], Maria Manuela Costa[1], Jaime C. Branco[1, 2], Alexandre Sepriano[1, 2]
[1]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [2]CEDOC, NOVA Medical School, Lisboa, Portugal, [3]Clinical Hematology Department, Centro Hospitalar Lisboa Ocidental - EPE, Hospital São Francisco Xavier, Lisboa, Portugal, [4]Infectious Diseases Department, Centro Hospitalar Lisboa Ocidental EPE, Hospital de Egas Moniz, Lisboa, Portugal

Introduction: Hypereosinophilia is unusual in rheumatoid arthritis (RA), but can occur in severe long-lasting disease, especially in patients with extra-articular manifestations and high titers of rheumatoid factor (RF). The hypereosinophilic syndrome (HES) occurs when hypereosinophilia leads to organ dysfunction.

There are only a few case reports of RA associated to eosinophilic pneumonia and myocarditis (Loeffler's endomyocardial fibrosis), and the possible association of RA and HES remains yet poorly known. Here, we present a case of HES with life-threatening multiorgan involvement in a patient with known RA.

Clinical case: A 46 year-old female patient presented to the emergency department (ED) in March 2022 with acute oppressive chest pain. She also reported one week of shortness of breath and inflammatory arthralgias of the metacarpophalangeal and interphalangeal joints of both hands. The patient was diagnosed with RA by a rheumatologist when she was 33 years old and remained for several years in remission with methotrexate and low-dose prednisolone, without known extra-articular involvement. She had lost follow-up between 2017 and 2022 but remained asymptomatic without medication during that period. Physical examination revealed both reduced respiratory murmur in pulmonary bases and heart sounds, pruritic erythematous skin papules and symmetric polyarthritis of hand joints. While in the ED, she had 2 episodes of cardiopulmonary arrest, responsive to resuscitation. The urgent cardiopulmonary study revealed a constrictive pericarditis with cardiac tamponade and bilateral pleural effusion. She was submitted to urgent pericardial drainage and partial pericardiotomy and admitted on cardiology intensive care unit. Laboratory analyses showed highly elevated inflammatory parameters - leukocytosis (22300/cm3) due to hypereosinophilia (7000/cm3, 20%), thrombocytosis (1027000/cm3), elevated CRP (25.4 mg/dL) and ESR (46 mm/h). She also had elevated IgE (3740 UI/mL). The histological study of the pericardium showed thickened pericardial wall, with fibrosed areas and abundant eosinophilic infiltration, with no granulomas. A diagnosis of HES was assumed, and the patient was started on prednisolone (0,25 mg/Kg/day), with a significant clinical and analytical improvement. In the first Rheumatology visit, she still had eosinophilia (1560/cm3) and mildly elevated inflammatory parameters, with arthritis of 5 joints but no skin lesions. Hands and feet radiographs revealed typical erosions of RA, and the immunological study identified high titers of RF (1130 UI/mL) and ACPA (305 UA/mL). She was treated with methotrexate 15mg/week, maintaining 20 mg/day of prednisolone initially, which resulted in completed clinical and analytical remission in 12 weeks. After 24 weeks, steroids were completely tappered, and she had normal blood eosinophils count, showing no signs of pleural or pericardial effusion and no irreversible cardiac lesions, or other suspected organ involvement. After an intensive study, ruling out other potential causes (e.g., parasitic infections, malignancies, drug-induced, other rheumatic diseases) of HES, and considering the presence of some characteristics that raise the suspicion of reactive HES instead of idiopathic HES (elevation of inflammatory parameters, e.g.), the diagnosis of HES associated with RA was made.

Conclusion: Clinicians should be aware that HES (including severe life-threatening cases) can occur in patients with RA, especially in long-lasting disease and if high titters of RF and extra-articular features are present.

168 - GIANT CELL ARTERITIS AFTER RADIATION THERAPY: CAUSE OR COINCIDENCE?

Tomás Fontes[1, 2], Pedro Ávila Ribeiro[1, 3], Martins-Martinho J [1, 3], Tiago Tavares Abreu[4], Ana Amado[5], Cristina Ponte[1, 3]

[1]Serviço de Reumatologia e Doenças Ósseas Metabólicas, Centro Hospitalar e Universitário de Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Serviço de Reumatologia, Hospital do Divino Espírito Santo, Ponta Delgada, Portugal, [3]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [4]Unidade de Técnicas Invasivas Pneumológicas, Pneumologia II, Hospital Pulido Valente, Centro Hospitalar Universitário Lisboa Norte, CAML, Lisboa, Portugal, [5]Serviço de Radioterapia, Hospital de Santa Maria, CHLN, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Background: Giant cell arteritis (GCA) is the most common form of primary systemic vasculitis in the elderly. It predominantly involves the temporal arteries but can also affect extracranial arteries such as the aorta and its main branches. Radiation-induced arteritis (RIA) may occur as a result of radiation therapy (RT) and consists of a localized vasculitic phenomenon with rapid arterial inflammation that can lead to wall thickening and stenosis, mimicking GCA. Moreover, a potential association between malignancy and GCA has also been reported. We describe a case of a patient who developed GCA following RT for a suspected lung adenocarcinoma.

Case report: An 86-year-old woman was referred to our rheumatology clinic with a six-month history of weight loss (10kg), asthenia, and bilateral scapular and pelvic girdle pain with morning stiffness. She had a background history of asthma, dyslipidaemia, coronary artery disease and a slow-growing lung mass in the right superior lobe, identified four years prior, which on fluorodeoxyglucose (FDG)-positron emission tomography (PET) assessment showed an increase in its standardized uptake value (SUV) from 1.5 to 3.5 units over the course of three years. Due to the suspicion

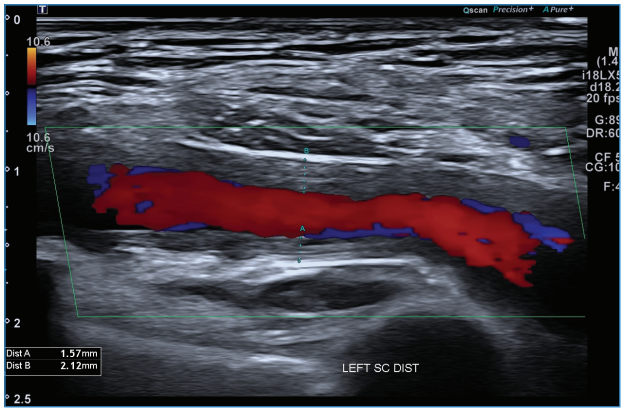

Figure 1. Ultrasound image with colour Doppler flow of the left subclavian artery, depicting a massive increase of the intima-m

of indolent lung adenocarcinoma but difficult access to obtain histology samples confirming the diagnosis, treatment with stereotactic body RT was chosen over lobectomy. She received a total of 55 Gray/units over nine days, with subsequent regression in the lung mass size (from 4.3 to 1.6 cm in diameter). Approximately one month after finishing RT, the patient began feeling unwell with the abovementioned symptoms. On physical examination, she struggled to walk and had bilateral impairment of shoulder abduction. Systolic blood pressure was asymmetrical on the upper limbs (<10mmHg on the left). Blood tests showed normocytic/normochromic anaemia (Hb 8.3 g/dL) and increased acute phase reactants (APR; ESR 107 mm/h and CRP 1.7 mg/dL). A follow-up FDG-PET showed no activity in the pulmonary lesion but identified an increased uptake in both axillary arteries (AX) with a maximum SUV of 3.4 units. Ultrasound of the temporal, AX, subclavian (SC) and common carotid (CC) arteries was performed showing a halo sign in both AX, CC and left SC (Fig.1), compatible with the diagnosis of GCA with exclusive large vessel involvement. Treatment with prednisolone 1mg/Kg/day was started resulting in significant improvement of symptoms. Laboratory evaluation after four months showed normal APR, and imaging assessment with FDG-PET after six months did not depict metabolic activity in the large vessels or lungs.

Discussion/Conclusion: RIA is a rare complication of RT that tends to be limited to the radiated site but may also affect the surrounding healthy tissues. To the best of our knowledge, no cases of RIA with GCA/polymyalgia rheumatica symptoms and concomitant increase in APR have been previously described. Although this patient presented with elevated APR and vasculitic changes in the left SC and AX and the radiated lung lesion was on the right, the temporal coincidence between events and the relative proximity between structures does not allow exclusion of RIA as the underlying cause for this patient's condition. Moreover, paraneoplastic GCA in this case is a less likely possibility given the improvement in the lung lesion documented by PET when the symptoms started and no arterial FDG uptake in pre-RT PET scans. Nevertheless, this case draws attention to RIA and paraneoplastic syndrome in the differential diagnoses of GCA.

172 - CATASTROPHIC ANTIPHOSPHOLIPID SYNDROME AS THE INITIAL PRESENTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS

Laura Gago[1,2], Mariana Emília Santos[1,2], Ana Catarina Moniz[1,2], Maria João Gonçalves[1,2], Carina Lopes[1,2], Inês Silva[1,2], Ana Filipa Mourão[1,2], Maria Manuela Costa[1,2], Jaime C. Branco[1,2]

[1]Rheumatology Department, Centro Hospitalar de Lisboa Ocidental, Hospital Egas Moniz EPE, Lisboa, Portugal, [2]2Comprehensive Health Research Centre (CHRC), NOVA Medical School | Faculdade de Ciências Médicas, NMS|FCM, Universidade Nova de Lisboa, Lisboa, Portugal

Introduction: Catastrophic antiphospholipid syndrome (CAPS) is a rare, life-threatening disease characterised by multiple vascular occlusive events, presenting over a short period of time, in patients with APLA.

A 32-years-old women with no relevant medical history was admitted in the emergency department with a one-week history of fever(39.0ºC), dizziness, myalgia, cough and arthralgia. She also noted the development of small red lesions on her face and ecchymosis in the feet. She denied hemoptysis or epistaxis.

On objective examination, she was febrile (TT 38.7ºC), with palpable purpura on the face, trunk, and lower limbs, and presented several cervical adenopathies.

Bloodwork showedmicrocytic hypochromic anemia (hemoglobine 6.9 g/dL; VGM 63 fL; HGM 20.0 pg), leucopenya (3500 u/L), thrombocytopenia with 13000 u/L platelets. She also presented elevated ferritin (898 ng/mL), lactate dehydrogenase(LDH) (653 U/L), haptoglobin (402 mg/dL), and C-Reactive Protein(CRP)(14.8 mg/dL). Presented positive direct Coombs test, lupus anticoagulant (negative anti-cardiopilin and anti-B2 glycoprotein1 antibodies) and antinuclear antibodies (1/160), negative anti-dsDNA antibody, C3 consumption (57 mg/dL), normal C4 level. She developed renal disfunction with creatinine of 1.27 mg/dL (GFR 60 mL/min/1.73m2), with proteins, hemoglobin and some granular cylinders in urinalysis. She had no changes in liver function or changesin coagulation times. Theperipheral blood smear showedveryfew schistocytes. Viral

and bacterial infections were also excluded.

A myelogram was performed and showed increased number of megakaryocytes, but no dysmorphisms. Bone marrow immunophenotypingshowed only reactive changes. She presented positive antiplatelet antibody and moderately decreased ADAMTS13activity (although it is not valued since it was collected after platelet transfusion).

She underwent several bloodtransfusions and platelet pool with no improvement. Anticoagulation was not started since the patient had platelets around 3000 u/L. Due tosuspicion of APS secondary to autoimmune pathology, we started high doses of methylprednisolone, with no improvement.

The admission computed tomography(CT)scan showed cervical adenopathies with necrotic tissue and signs of splenic infarction with hepatomegaly. A cranial CT was also performed where cerebral venous thrombosis was evident in the transverse and sigmoid sinuses.

Two days after being admitted to hospital, the patient developed left lower limb edema associated with flushing of the gemellar region. EcoDoppler was compatible with deepvein thrombosis. Due to complaints of photopsia, ophthalmologic evaluation was requested and she was diagnosed with ocular microangiopathy.

Considering the analytical and imaging results, and the worsening of the purpuric lesions mainly on the toes, CAPS secondary to Systemic Lupus Erythematosus was assumed as the most probable diagnosis. Plasmapheresis was performed for 2 days and later treatment with IV Immunoglobulin 2g/kg and Rituximab1000 mg. There was a clear improvement in hematology, renal function, and clinical symptoms. She also started anticoagulation initially with unfractionated heparin with later switch to warfarin.

Now the patient is asymptomatic with no new thrombotic events.

Conclusion: CAPS is associated with high morbidity and mortality. Therefore, an aggressive multidisciplinary treatment strategy is indicated. Anticoagulation, immunosuppression, plasma exchange, intravenous immunoglobulins, and anti-platelet agents, used in various combinations, have resulted in improved patient outcome.

178 - ALOPÉCIA AREATA - REAÇÃO ADVERSA RARA DOS INIBIDORES DO TNF-ALFA TRATADA COM TOFACITINIB

Catarina S Cortesão[1], Barbara Lobão[1], Alice Neves[1], Eugénia Simões[1], Cláudia Miguel[1], Cândida Silva[1], Helena Santos[1]
[1]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal

Introdução: Os inibidores do TNF-alfa (iTNF-alfa) alteraram o paradigma da evolução de várias doenças imuno-mediadas tornando-se indispensáveis no controlo e remissão da sua atividade. O seu modo de ação passa pela interferência nas vias de sinalização mediadas pelo TNF-alfa ou pelo seu recetor, facilitando a regulação e dinâmica de citoquinas e mediadores inflamatórios.

Raramente, esta interferência tem como efeito acionar mediadores de novas patologias imuno-mediadas. A alopécia areata (AA) é uma patologia auto-imune mediada por células T citotóxicas direcionada aos folículos pilosos inibindo o seu crescimento e regeneração, tendo como característica principal a reversibilidade do quadro clínico após o controlo do ataque imuno-mediado.

Aqui apresentamos o caso clínico de um homem com diagnóstico de artrite psoriática controlada com certolizumab que desenvolveu uma reação cutânea rara.

Caso clínico: Homem com 28 anos, iniciou em 2017 um quadro agudo de artrite aditiva e assimétrica de predomínio distal, atingindo pés e tornozelos, associado a lombalgia de ritmo inflamatório e dactilite do 2º dedo do pé. Tem HLA-B27 positivo, antecedentes de "eczema" atópico, sem outro antecedente familiar conhecido. Foi admitido o diagnóstico de artrite psoriásica.

Iniciou metotrexato (MTX) com escalada progressiva de dose. A doença manteve-se ativa com artrite das MTF e interfalângicas dos 2º e 3º dedos, entesite aquiliana e fasceíte plantar direita, com lesões cutâneas descamativas e lesões ungueais. Iniciou certolizumab 200mg/quinzenal associado a MTX 15mg em novembro de 2018, com melhoria clínica significativa após 3 meses, permitindo a redução da dose de MTX, com suspensão deste fármaco.

Em dezembro de 2020 apresentou peladas no couro cabeludo e corpo com cerca de 1cm de diâmetro, que evoluíram com progressivo aumento em número e diâmetro. Em janeiro de 2021 suspendeu o certolizumab. Na consulta de dermatologia foi diagnosticada AA e iniciou terapêutica dirigida com corticóide tópico e intralesional, sem eficácia. Iniciou prednisolona na dose de 60mg/dia com redução da perda de cabelo e pêlo mas sem reversão das lesões.

Em junho de 2021 iniciou tofacitinib 5mg bid, com melhoria significativa, segundo critérios ASDAS, e repovoamento das lesões cutâneas. Mantém tofacitinib 5mg bid em monoterapia, com doença reumática inativa e praticamente sem lesões de AA.

Discussão: Numa revisão da literatura, as reações cutâneas ao certolizumab são raras e correspondem 1,9% das reações cutâneas totais sob iTNF-alfa[1], sendo a AA considerada muito rara. Neste doente, o aparecimento e a evolução rápida das lesões de AA, levaram à suspensão do certolizumab. Dado não haver consenso

relativo à manutenção da terapêutica neste contexto[2], a decisão, partilhada com o doente, teve em conta o impacto das lesões cutâneas na sua qualidade de vida e a necessidade de manter o controlo da atividade inflamatória. A introdução de tofacitinib teve em conta, também, os estudos que já decorriam na AA e que demonstravam bons resultados, com bom perfil de segurança[3].

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182 - DOENÇA DE CROHN – APRESENTAÇÃO ATÍPICA E MULTIFACETADA.

Mariana Sá[1], Rute Cerqueira[2], Fonseca, R[3]
[1]USF Famílias, Santa Maria da Feira, Portugal, [2]Gastroenterology, Centro Hospitalar Entre o Douro e Vouga, Santa Maria da Feira, Portugal, [3]Rheumatology, Centro Hospitalar Entre o Douro e Vouga, Santa Maria da Feira, Portugal

Introdução: A presença de artralgias de ritmo inflamatório não implica necessariamente a presença de uma doença reumatológica subjacente. Contudo, quando associadas a manifestações extra-articulares atípicas, como a uveíte ou eritema nodoso, devem levar a suspeita de doença inflamatória com fenótipo multivariado.

Descrição do caso: Doente do sexo feminino, de 38 anos de idade, com história prévia de uveíte anterior granulomatosa bilateral, foi referenciada à consulta de Reumatologia por poliartralgias. Na consulta, referia artralgias dos ombros, tornozelos e lombossacralgia de ritmo misto com rigidez matinal de 1 hora. Ao exame objetivo não apresentava alterações. Analiticamente, verificou-se uma discreta elevação da velocidade de sedimentação (29 mm/1ªh) e da proteína-c-reativa (21,6 mg/L) sem outras alterações. O HLAB27, fator reumatoide, anticorpos anti-nucleares e HLAB51 eram negativos. O IGRA, teste direto e cultural para mycobacterium tuberculosis foram também negativos. A enzima de conversão da angiotensina era borderline, tendo sido efetuado TAC de tórax que não mostrou adenopatias. A radiografia e ressonância magnética das articulações sacroilíacas não mostraram sinais de sacroileíte. A doente manteve a vigilância na consulta e, cerca de 1 ano mais tarde, apresentou episódio de eritema nodoso que foi tratado com prednisolona. Mais tarde, durante o seguimento, iniciou queixas de diarreia alternada com obstipação tendo sido referenciada à consulta de Gastroenterologia. A calprotectina fecal era elevada (141 mg/kg). A colonoscopia mostrou áreas com mucosa eritematosa com erosões/úlceras aftóides e a biópsia foi compatível com Doença de Crohn.

Atualmente, a doente encontra-se medicada com mesalazina, sem recidiva das queixas gastrointestinais, artralgias, uveíte ou eritema nodoso.

Conclusão: A Doença de Crohn é uma doença inflamatória intestinal que pode ser precedida em meses/anos por manifestações extra-intestinais. As queixas coloproctológicas e/ou perda de peso em doentes com patologia reumatológica ou uveíte/episclerite deve ser rapidamente investigadas para pesquisa de doença inflamatória intestinal (Doença de Crohn ou Colite Ulcerosa). A singularidade deste caso deve-se aos sintomas inaugurais que além de serem extra-intestinais envolvem 2 sistemas – musculo-esquelético e ocular.

185 - EXPLORING NUCLEAR IMAGING TECHNIQUES IN CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS (CRMO): PET AND SCINTIGRAPHY

Vanessa Fraga[1], Sandra Sousa[1], Maria José Santos[1, 2]
[1]Rheumatology Department, Hospital Garcia de Orta, Almada, Portugal, [2]Unidade de Investigação em Reumatologia, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introduction: CRMO is a rare bone disorder affecting children and adolescents. Diagnosing and monitoring CRMO requires advanced imaging techniques, with magnetic resonance imaging (MRI) being the gold standard. However, due to limited availability, nuclear medicine techniques like bone scintigraphy and positron emission tomography (PET) can also be used.

Case presentation: We present two cases of children with CRMO. The first case involves a 6-year-old boy with a limping and swollen ankle presenting an erosive lesion in the third cuneiform bone in MRI. The second case is a 7-year-old girl who presented with a refractory torticollis caused by an erosive atlas lesion documented in a cervical-CT. Both cases underwent extensive diagnostic investigations, including bone biopsies, to exclude malignancy and infection and to identify the underlying cause. Given the suspicion of CRMO and the unavailability of total body MRI, alternative imaging techniques were requested, including PET-scan and bone scintigraphy.

In the boy's case, bone scintigraphy revealed an inflammatory/infectious process in the 1st e 3rd cuneiform bones, accompanied by oedema of soft tissues of the ipsilateral foot and ankle.

In the girl's case, PET-scan played a crucial role in identifying multiple sites of abnormal metabolic activity, suggesting ongoing inflammatory/infectious processes. The atlas, iliac bone, coccyx, tibial junctions, and distal

2nd metatarsal were among the areas affected.

Both patients were treated with intravenous bisphosphonates with clinical improvement.

Conclusion: In both cases, the use of PET-scan and scintigraphy proved valuable in localizing areas of active inflammation, assessing disease activity and extent, aiding in the diagnosis and subsequent management. PET-scan provided functional information by detecting metabolic activity, while scintigraphy demonstrated tracer uptake in affected areas, supporting the diagnosis. This highlights the importance of considering alternative imaging techniques when total body MRI is unavailable or contraindicated.

190 - POLIMORFISMO DA SARCOIDOSE E SUAS COMPLICAÇÕES

Nuno Delgado[1], Miguel Guerra[1], Ana Filipa Rocha Águeda[1], José Marona[1], Joana Ramos Rodrigues[1], Margarida Oliveira[1]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário Cova da Beira, Covilhã, Portugal

Introdução: A Sarcoidose constitui um desafio diagnóstico na prática clínica da Reumatologia devido ao seu polimorfismo clínico, com uma panóplia de manifestações extrapulmonares heterogéneas, nomeadamente no que ao envolvimento cutâneo se refere. Pode cursar com manifestações ligeiras ou apresentações mais graves, como a hipercalcémia maligna, que comprometem o prognóstico vital do paciente.

Caso Clínico: Doente do sexo feminino, de 57 anos, com zonas de tumefação induradas e indolores no antebraço e no 2º dedo esquerdos, com 3 semanas de evolução, sem outras manifestações clínicas. A ecografia de partes moles descrevia ``áreas de tecido celular subcutâneo com aumento da refletividade e edema associado a processo inflamatório´´, confirmado posteriormente com RM. O estudo anatomopatológico foi compatível com paniculite granulomatosa, a sugerir sarcoidose. Prosseguiu-se com estudo complementar, com deteção de adenopatias hilares, peri-hilares, cervicais e mediastínicas na TC do tórax e hipermetabólicas na PET-TC. Analiticamente com VS 23mm/h, ECA 77.1 U/L (N < 52 U/L), PCR 1.5mg/dL e Cálcio 9.7mg/dL. A doente foi submetida a broncofibroscopia e biópsia brônquica guiada por eco-endoscopia. No lavado alveolar, a relação CD4/8 foi de 2.83; o estudo histológico das adenopatias revelou linfadenite granulomatosa não necrotizante. Considerando a ausência de achados compatíveis com micobacteriose, assumiu-se o diagnóstico de Sarcoidose. Inicialmente, protelou-se a corticoterapia, face à melhoria espontânea das lesões cutâneas e em virtude das comorbilidades da doente, nomeadamente, diabetes mellitus descompensada. Em posterior reavaliação, foi detetada hipercalcémia grave de novo

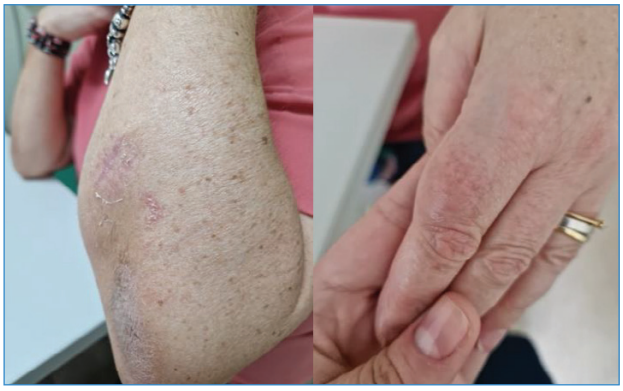

Figura 1. Tumefações compatíveis com paniculite observados no antebraço e 2º dedo esquerdos

(cálcio ionizado 8.35mg/dL), lesão renal aguda (creatinina 2.5mg/dl; TFG 25ml/min), aumento da ECA para 182 U/L, associado a quadro de lentificação psicomotora, desorientação temporo-espacial e desequilíbrio. A doente foi internada e realizou fluidoterapia ev, tratamento com metilpredisolona ev (500mg) e zoledronato ev (5mg). Apesar da melhoria analítica, a doente desenvolveu um quadro de alucinações e síndrome confusional agudo, interpretado como psicose secundária a corticoterapia (excluída neurosarcoidose por RM), pelo que suspendeu corticoterapia endovenosa e iniciou tratamento com deflazacorte (30mg id), anti-psicóticos e metotrexato oral (15mg/semana). A doente evoluiu favoravelmente em termos clínicos e analíticos, encontrando-se, de momento, assintomática.

Conclusão: O caso descrito reflete o desafio diagnóstico e terapêutico que a sarcoidose constitui na prática clínica de um Reumatologista. As lesões cutâneas, em particular, apresentam marcada variabilidade morfológica, incluindo pápulas, nódulos, placas e infiltração de cicatrizes. Devido a este polimorfismo, o seu diagnóstico baseia-se na conjugação de critérios clínicos, radiográficos, laboratoriais e histopatológicos. Por outro lado, a hipercalcémia grave e as manifestações clinicas a ela associada constituem uma manifestação rara desta doença, sendo que a grande maioria dos doentes apresenta valores de cálcio normais ou ligeiramente elevados, sem sintomatologia clinica associada.

197 - ERASMUS SYNDROME: A CASE REPORT OF A RARE ASSOCIATION

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

Background: Silica exposure is considered a risk factor for a number of autoimmune disorders. The particular association between silica exposure and the development of systemic sclerosis is a rare occurrence and often referred to as Erasmus syndrome.
Case report: We present the case of a 46-year-old Romanian man, living in Portugal for 20 years, working as a car painter with known exposure to silica through sandblasting for 15 years, without using adequate protective equipment, and recent ex-smoker of 26 pack-years, who was referred to our department for a three-month history of bilateral hand swelling, Raynaud's phenomenon and digital ulcers in multiple fingers. The physical exam showed puffy hands and digital ulcers in the 1st to 3rd left fingers and 3rd right finger, with loss of substance in the 2nd left and 3rd right fingers. Blood work evidenced non-elevated inflammatory markers, positive antinuclear antibodies in high titre (1: 1280) and a strongly positive anti-Scl70 antibody; anti-double strand DNA, rheumatoid factor and anti-citrullinated-peptide antibodies were negative. A pulmonary CT-scan revealed enlarged pre-tracheal and hilar nodes and diffuse opacities in the right upper lobe and both lung bases, with a ground-glass pattern. The diagnosis of systemic sclerosis was established and the patient was started on prednisolone 10 mg daily, methotrexate 20 mg weekly, nifedipine 20 mg twice daily, pentoxifylline 400 mg twice daily and acetylsalicylic acid 150 mg daily, as well as scheduled for iloprost treatment. Oesophageal manometry was performed following complaints of dyspepsia, and documented dysmotility compatible with early oesophageal involvement of systemic sclerosis. The echocardiographic assessment revealed mild biatrial enlargement with slight tricuspid regurgitation, estimated pulmonary artery systolic pressure of 38 mmHg and a minimal pericardial effusion. Lung function tests showed a restrictive disease pattern (forced vital capacity of 62%, forced expiratory volume in the first second of 61% and total lung capacity of 64%), with decreased diffusing capacity for carbon monoxide (69%). Endobronchial ultrasound revealed no obvious abnormalities and the biological material collected shoewed no evidence of neoplastic cells, granulomas, organic or inorganic fibre deposits or bacterial infection. With these findings, as well as the clinical background of the patient, the diagnosis of Erasmus syndrome was made, with a clinical, functional and imagiologic pulmonary re-evaluation planned in six months, to consider treatment with an antifibrotic agent.
Conclusion: Systemic sclerosis is a systemic and progressive autoimmune disease in which pulmonary involvement represents a significant concern for the management of these patients, as the leading causes of death in this group of individuals include pulmonary fibrosis and pulmonary arterial hypertension. Erasmus syndrome in particular has worse pulmonary outcomes and, as such, these patients should be more closely monitored and more aggressively treated.

198 - SARCOID REACTION INDUCED BY ANTI-TNF THERAPY

Ana Catarina Moniz[1], Sónia Mota Faria[2], Madalena Silveira Machado[3], Manuela Zita Veiga[4]
[1]Rheumatology Department, Hospital Egas Moniz (CHLO), Lisboa, Portugal, [2]Serviço de Patologia Clínica, Hospital Prof. Doutor Fernando Fonseca, Amadora, Portugal, [3]Instituto Português de Oncologia de Lisboa Francisco Gentil, Lisboa, Portugal, [4]Medicina Interna, Centro Hospitalar de Lisboa Ocidental EPE, Hospital de Egas Moniz, Lisboa, Portugal

Paradoxical reactions (PR) are exacerbations or new onset of non-infectious inflammatory signs that appear due to a substance that is in principle therapeutically effective. Those can occur with any drug group but are more frequent with biologics and particularly anti-TNF drugs.

We describe a case of a 52-year-old man treated with Golimumab for Axial Spondylarthritis, presenting with nephrolithiasis, hypercalcemia, progressively decreased renal function and multiple thoracic and abdominal enlarged lymph nodes. Detailed laboratory studies revealed normal Parathyroid Hormone (PTH), normal 25-hydroxyvitamin D, negative PTH-related protein, elevated Erythrocyte Sedimentation Rate, elevated Angiotensin-converting enzyme and elevated 1,25-dihydroxyvitamin D. A Endobronchial ultrasound guided biopsy of a mediastinal lymph node was negative for bacteria, mycobacteria, lymphoma, and other cancer cells. Diagnosis of sarcoid reaction associated with anti-TNF drug was made. Patient suspended Golimumab and eventually developed an uveitis, for which he was treated with Prednisolone for a short period. On follow-up, the enlarged lymph nodes were reduced to normal, as well as serum calcium levels, supporting the presumptive diagnosis.

Drug-induced sarcoid reaction (DISR) is a systemic granulomatous tissue reaction that is indistinguishable from sarcoidosis and occurs in a temporal relationship with initiation of an offending drug. Diagnosis of DISR, as well as sarcoidosis, is one of exclusion. It usually resolves with discontinuation of the offending drug but may sometimes require corticosteroids and further sarcoidosis treatment. We present a case of a sarcoid reaction manifested as mediastinal lymphadenopathy and altered calcium and phosphorus metabolism, which resulted from treatment with Golimumab and resolved spontaneously without treatment.

199 - SCHNITZLER'S SYNDROME: A CASE CROSSING MEDICAL SPECIALTIES

Rodrigo Rei[1], Frederico Rajão Martins[1], Maria João Paes[2], Catalina Gómez[3], Catarina Tenazinha[1]
[1]Rheumatology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, Faro, Portugal, [2]Immunoallergology Department, Centro Hospitalar Universitário do Algarve, Faro, Portugal, [3]Hematologia Clínica, Hospital da Luz, Lisboa, Portugal

Introduction: Schnitzler's Syndrome is a rare disease, likely underdiagnosed, with fewer than 400 literature reports. It is acquired, of unknown etiology, but it does have inflammatory clinical and physiopathological profiles and established diagnostic criteria, and is classically associated to a monoclonal gammopathy.

Chronic idiopathic urticaria and Adult Onset Still's Disease (AOSD) are the main differential diagnosis and atypical presentations are not infrequent in the latter. The M gradient, not routinely determined in patients with suspected AOSD, as well as response to therapy, may help in diagnosis.

Clinical report: An 84 year-old male with a known history of rheumatoid arthritis, characterized by recurrent oligoarthritis of the wrists refractory to conventional disease modifying anti-rheumatic drugs, presented with 1 year duration complaints of a spontaneous urticarial rash, mildly pruritic, little evanescent and unresponsive to antihistamine regimens at supratherapeutic doses, with anorexia and weight loss of approximately 9 Kg since the onset of the complaints. He has history of hypertension and dyslipidemia. Physical examination showed bilateral inguinal adenopathies and reduced wrist mobility. Wrist X-rays showed a significant diffuse decrease in the radiocarpal, intercarpal and carpometacarpal joint lines. Blood tests identified anemia of chronic disease, leukocytosis with neutrophilia, persistent elevation of erythrocyte sedimentation rate and C-reactive protein, moderate elevation of serum ferritin, and monoclonal gammopathy of undetermined significance (MGUS) to IgM. On skin biopsy, there were findings of urticarial dermatitis with neutrophilic infiltrate, and the inguinal lymph node biopsy showed dermatopathic lymphadenopathy. Myelogram and bone biopsy excluded the presence of lymphoproliferative syndrome and plasma cell neoplasia.

The diagnosis of Schnitzler's Syndrome was admitted, and the patient was started on oral corticosteroid therapy at a moderate dose, verifying complete resolution of the complaints. Considering the high probability of recurrence of the complaints after corticosteroid therapy discontinuation, the patient is a current candidate for treatment with anakinra.

Discussion: If, on one hand, monoclonal hypergammaglobulinemias and chronic idiopathic urticaria are common in the elderly, on the other hand, manifestations of SchnS previously not attributable to the disease have been described, mainly by retrospective evaluation which results in diagnostic rectification. Arthritis of the wrist with joint damage is a classic finding of chronic forms of AOSD, but it is not described in SchnS, and its presence might be misleading.

It is estimated that 10-20% of SchnS cases develop lymphoproliferative disorders about 10 years after diagnosis. Additionally, untreated patients have a high probability of developing systemic AA amyloidosis, which requires adequate surveillance and treatment. IL-1 inhibitors are the only truly effective treatments known to date.

Identification and dissemination of rare clinical entities is critical to improve current knowledge, with potential impact on clinical practice, namely promoting earlier detection of underdiagnosed pathologies, with consequent impact on prognosis.

201 - DUPILUMAB ASSOCIATED ACHILLES TENDINOPATHY: A NOVEL MECHANISM AND ADVERSE EFFECT

Pedro Miguel Teixeira[1, 2], Carolina Vilafanha[1, 2], Diogo Valente Dias[2, 3], Gisela Eugénio[1, 2], Anabela Barcelos[1, 2]
[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz Health Alliance, Portugal, Aveiro, Portugal, [3]Otolaryngology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal

Introduction: Dupilumab is a monoclonal antibody that blocks the interleukin (IL)-4 receptor thus blocking Th2 cytokines IL-4 and IL-13. It is approved for atopic dermatitis and more recently for asthma and chronic rhinosinusitis with nasal polyps (CRSwNP). Although dupilumab is generally safe, musculoskeletal adverse effects that were not described in clinical trials, have been reported. We report a case of tendinopathy occurring during dupilumab therapy for CRSwNP.

Clinical case: We present the case of a 55-year-old woman with asthma and CRSwNP who was referred to a Rheumatology consultation after experiencing bilateral Achilles tenderness following the initiation of dupilumab treatment. Approximately one month after initiating dupilumab therapy, the patient reported a significant improvement in upper respiratory symptoms. However, she began to develop pain and swelling around both Achilles' tendons, which gradually worsened and became incapacitating. The patient experienced only

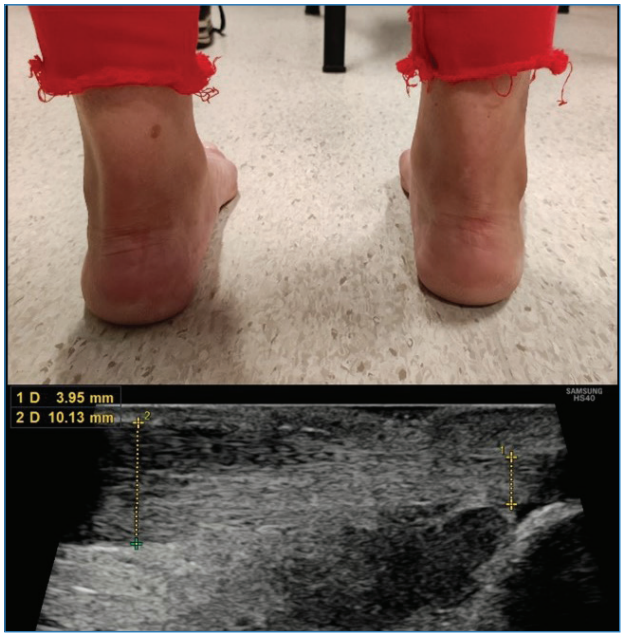

Figure 1. Bilateral Achilles' swelling and ultrassound of the left Achiles tendon

mild relief with the administration of etoricoxib 60mg/day. There was no history of musculoskeletal diseases, rheumatic conditions, or preceding infections, trauma, or surgery. Upon physical examination, diffuse tenderness and swelling were observed in both Achilles' tendons. Laboratory investigations yielded normal results, including erythrocyte sedimentation rate and C-reactive protein. Rheumatoid factor, anti-cyclic citrullinated peptide antibody, antinuclear antibodies, and HLA-B27 were negative. Musculoskeletal ultrasonography revealed bilateral thickening and hypoechogenic areas consistent with Achilles tendinopathy (Figure 1). Following a multidisciplinary discussion, the decision was made to discontinue dupilumab and initiate treatment with etoricoxib 90mg/day. The patient experienced a dramatic improvement in symptoms within three weeks of this treatment modification.

Conclusion: Enthesitis and/or inflammatory arthritis associated with the use of dupilumab in atopic dermatitis has been reported in the literature only in a limited number of cases. It has been hypothesized that the blocking of IL-4/IL-13 by dupilumab may result in heightened activation of the IL-23/IL-17 axis, potentially leading to spondyloarthritis-related conditions such as enthesitis/tendinitis, arthritis, uveitis, and psoriasis. In most instances, treatment with oral non-steroidal anti-inflammatory drugs and/or discontinuation of dupilumab appears to be effective. Further research is necessary to establish a causal relationship, comprehend the process of pathogenesis and determine the proper treatment in each case.

204 - ERITEMA MULTIFORME – UM IMPORTANTE DIAGNÓSTICO DIFERENCIAL NA REUMATOLOGIA

Susana P. Silva[1, 2], Sofia Ferreira Azevedo[1, 2], C. Pinto Oliveira[1, 2], Manuela Loureiro[2, 3], Luís Santiago[2, 3], Gisela Eugénio[1, 2], Anabela Barcelos[1, 2]

[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [2]Centro Académico Clínico Egas Moniz Health Alliance, Portugal, Aveiro, Portugal, [3]Dermatologia, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal

Apresentamos o caso de um doente do sexo masculino, de 31 anos, sem antecedentes pessoais de relevo, observado na consulta de reumatologia por aftose oral recorrente, com 5 anos de evolução. Não existia história sugestiva de artrite, lesões cutâneas, alterações gastrointestinais, vasculares ou oculares. Recorreu à consulta por quadro agudo com 3 semanas de evolução de febre (temperatura máxima 39ºC), perda de peso de 8kg, lesões ulceradas orais, odinofagia, e lesões cutâneas nas pernas. Ao exame objetivo apresentava lesões ulceradas e erosões na mucosa bucal (Fig. 1A), queilite exuberante com crostas hemorrágicas aderentes (Fig. 1B), erosão no orifício da narina direita e lesões papulares eritematosas nos membros superiores e em placas e bolhas com descolamento a nível do dorso das mãos (Fig. 1C), joelhos e pernas (Fig. 1D). Analiticamente, destacava-se proteína C-reativa de 3.19mg/dL, anticorpos IgG para o vírus do Herpes Simplex 1 (VHS) e do Epstein Barr (VEB), com restantes serologias e autoimunidade negativas. Sem alterações no raio-X tórax. A biopsia cutânea revelou acantose e infiltrado linfomonocitário pericapilar intersticial dérmico superficial, não se evidenciando imagens de dermatose neutrofílica. Apesar da aftose oral recorrente ser uma manifestação clássica da Síndrome de Behçet, a presença de lesões orais generalizadas (inclusive na região externa dos lábios) e as lesões cutâneas em alvo não são comuns nesta patologia. Trata-se de quadro típico de eritema multiforme. Após avaliação multidisciplinar (Dermatologia- Reumatologia), o doente iniciou 60 mg de prednisolona oral em desmame com melhoria completa das lesões e da dor.

O Eritema Multiforme é uma reação mucocutânea de hipersensibilidade de etiologia variável. Apresenta-se caracteristicamente por lesões papulares, bulhosas e necróticas. Precipitantes comuns incluem fármacos, VHS, VEB, Mycoplasma pneumoniae e histoplasmose. A maioria dos casos são ligeiros, contudo os casos mais severos podem ser ameaçadores de vida. O tratamento é dirigido à causa subjacente, embora a corticoterapia seja utilizada frequente como primeira linha de tratamento, apesar da escassa evidência. A recorrência não

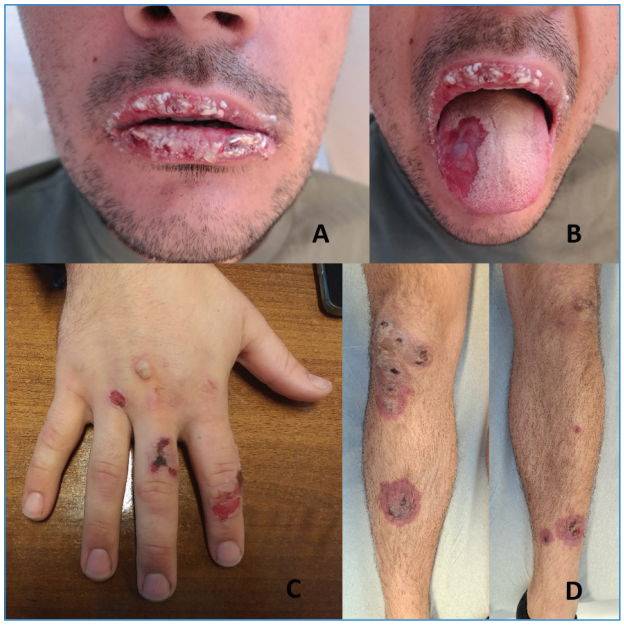

Figura 1. Eritema Multiforme. A - lesões ulceradas e erosões na mucosa bucal; B - queilite exuberante; C e D - lesões cutâneas.

é infrequente sobretudo nos casos relacionados com o VHS. Este caso destaca-se pela importância no diagnóstico diferencial, dado também existirem casos descritos de lesões tipo eritema multiforme associadas à Síndrome de Behçet.

214 - DIGITAL ULCERS AND SHORTENING OF THE SECOND AND THIRD FINGERS IN AN ELDERLY MAN

Catarina Dantas Soares[1], Hugo Parente[1], Maria Pontes Ferreira[1], Anita Cunha[1], Vera Teixeira[2], Daniela Santos-Faria[1]
[1]Rheumatology Department, Hospital Conde de Bertiandos (ULSAM), Ponte de Lima, Portugal, [2]Dermatology, Unidade Local de Saúde do Alto Minho, Viana do Castelo, Portugal

Introduction: The median nerve is a motor and sensory nerve that contains fibres from roots of C5-T1. The median nerve provides motor innervation to the muscles of the thenar eminence and radial the lateral two lumbricals. The sensory distribution of the median nerve supplies the palmar aspect of the lateral palm and the palmar and distal dorsal aspects of the lateral three and a half digits.

Carpal tunnel syndrome (CTS) is the most common compressive neuropathy resulting from median nerve entrapment. Its incidence tends to increase with age, and it is more prevalent among females.

Case Report: A 83-year-old man presented with recurrent digital ulcers with purulent inflammation on the second and third fingers. These lesions had started several years ago and were initially oriented as a bacterial cellulitis and treated with different antibiotics, however without relevant improvement. He also had paresthesia of the hands, without pain and had no systemic symptoms. He had no complaints suggestive of Raynaud's phenomenon. The patient was taking levothyroxine for hypothyroidism and an antihypertensive agent. There was no relevant family history. Physical examination findings revealed skin thickening, nail dystrophy, shortening of the second and third fingers and presented a global atrophy of the hand's muscles (Fig. 1). There were no similar skin lesions in other locations nor evidence of arthritis.

Histopathologic examination of prior wound biopsy showed granulation tissue with inflammatory infiltrate. Blood test results indicated slightly elevated erythrocyte sedimentation rate and C-reactive protein and all other laboratory findings were normal. Hands X-ray showed degenerative changes of small joints and acroosteolysis of distal phalanx of second fingers bilateral and third finger of left hand. Electrophysiological findings reveal severe bilateral median nerve sensory and motor neuropathy. The patient was redirected to an orthopedic consult and a surgical decompression was performed. A few months after surgery, there was a complete resolution of paresthesia and digital ulcers were practically healed.

Discussion: The ulcerative and mutilating clinical variant of CTS is rare and a marker of a severe neural lesion. Usually presents in advanced stages in patients who have suffered from its symptoms for years. This variant is more frequent in males in the sixth decades who had been manual workers, and it is bilateral in 25% of cases.

Symptoms may include hypoesthesia, tingling, with or without pain and, beyond that, this variant is characterized by skin and nail lesions. It includes ulcers in the second and third fingers, sometimes with pus-filled inflammation, cutaneous hyperkeratosis and onychodystrophy. If the median nerve compression further progresses acro-osteolysis may occur. An accurate history is crucial for diagnosis. Neurologic examination

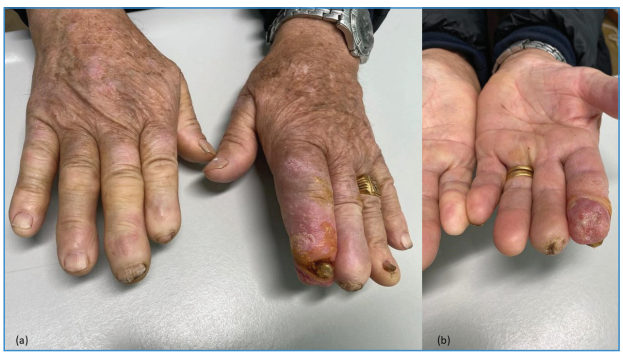

Figure 1. Nail dystrophy, skin erosions and shortening of the second and third fingers are visible on the hands of a 83-year-old

may show hypesthesia and dysesthesia in the sensory distribution of the median nerve and atrophy of the thenar muscles. Electromyography allows precise localization and severity of nerve injury and hand radiography, showing bone reabsorption of the distal phalange of the affected fingers, can support the diagnosis.

Surgical decompression is recommended and the definitive treatment for this severe form of the disease, leading to a considerable improvement in majority.

218 - ARTHRITIS AS THE PRESENTING MANIFESTATION OF CHILDHOOD LEUKEMIA

Vanessa Fraga[1], Catarina Abreu[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: Childhood leukemia can present with musculoskeletal manifestations, such as joint pain, swelling and limping, mimicking Juvenile Idiopathic Arthritis (JIA)1. Distinguishing between the two conditions can be challenging and requires high clinical suspicion.

Objective: The objective of this study was to characterize children who presented with arthritis as the first manifestation of acute leukemia.

Methods: We performed a retrospective study of children referred to the pediatric rheumatology outpatient clinic over the past 15 years with arthritis and who were subsequently diagnosed with leukemia. Demographic characteristics, clinical manifestations, initial laboratory findings and outcomes are reported.

Results: A total of 3 patients were identified, two girls and one boy. The mean age at clinical presentation was 6± 4.3 years and the mean interval between joint symptoms and leukemia diagnosis was of 5 ± 1 months.

All patients presented with limb pain, limping and arthritis, with a polyarticular pattern in 2 of them. The most common joints involved were the knee, wrist and the ankle.

Other symptoms such as weight loss, anorexia or skin rash were present in 2 patients.

Table 1 - Demographic characteristics, clinical features, laboratory results and outcomes

	Patient 1	Patient 2	Patient 3
Sex	Female	Male	Female
Age at clinical presentation (years)	11	3	4
Time until diagnosis (months)	4	6	5
Musculoskeletal involvement	Arthritis and limping	Arthritis and limping	Arthritis and limping
Articular pattern	Polyarticular	Oligoarticular	Polyarticular
Joints involved	Wrist, tibiotarsal and PIP	Hip and knee	Wrist, elbow, knee and ankle
Other symptoms	Weight loss		Anorexia and maculo-papular skin rash
Hemoglobin (g/dL)	7.8	9.7	11.5
Leukocyte count (10^9/L)	4.0	6.3*	7.5
Platelet count (10^9/L)	98	371	248
Peripheral blast cells (%)	Yes (20%)	No	No
ESR (mm/h)	120	89	113
CRP (mg/dL)	3.25	2.1	0.2
Peripheral Blood Smear	Blast cells with elevated N:C	Lymphocytes with moderate N:C	Normal
RF	Negative	N/A	Positive (borderline)
ANA	Negative	N/A	Negative
Leukemia type	B-ALL	B-ALL	AML
Outcome/follow-up	Ongoing chemotherapy	Remission	Death

* There was inversion of the leukocyte formula. ANA: Antinuclear Antibodies, AML: Acute Myeloid Leukemia, B-ALL: B-cell Acute Lymphocytic Leukemia, CRP: C-reactive protein, ESR: Erythrocyte Sedimentation Rate. N/A: Not Available, N:C Nucleus-to-cytoplasm ratio, RF: Rheumatoid Factor

Regarding laboratory results, cytopenias were observed in 2 patients and only 1 presented peripheral blast cells. One patient had no hematologic abnormalities in the initial investigation. Elevated inflammatory markers were present in all patients and the mean ESR was 107±16 mm/h. The patients' demographic characteristics, clinical features, laboratory results and outcomes are presented in Table 1.

The most frequent diagnosis was B-cell Acute Lymphocytic Leukemia (B-ALL) (n=2) and one patient was diagnosed with Acute Myeloid Leukemia (AML) through skin lesion biopsy. Regarding clinical outcomes, one patient is in remission, one is currently receiving chemotherapy and one died.

Discussion: Arthritis and limb claudication can be the initial manifestations of childhood leukemia. Large joints were mostly affected and more frequently in a polyarticular pattern. Hematologic abnormalities are not always present, especially peripheral blast cells.

To conclude, the presence of persistent constitutional symptoms, sudden onset of bone or joint pain and abnormal laboratory results should raise suspicion for childhood leukemia over JIA, particularly B-ALL, which has been more frequently associated with musculoskeletal involvement[1].

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219 - PAQUIDERMODACTILIA: UM DIAGNÓSTICO A EXCLUIR

Daniel Melim[1], Daniel Carvalho[1], Margarida Faria[1], Lídia Teixeira[1], Ricardo Figueira[1], Mário Rodrigues[1]
[1]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal

A paquidermodactilia é uma forma rara de fibromatose localizada nas articulações interfalângicas proximais (IFP). Caracteriza-se pelo espessamento dos tecidos moles das IFP, de forma progressiva, indolor e assimétrica, afetando sobretudo jovens do sexo masculino, reportando-se cerca de 150 casos na literatura. Apesar de ser definida como uma forma de fibromatose benigna e idiopática, fatores de stress mecânico, como tiques ou traumatismos, são encontrados em até 42% dos casos.

Reportamos o caso de um doente do sexo masculino, 12 anos de idade, com antecedentes pessoais e familiares irrelevantes, que foi referenciado à consulta de Reumatologia por tumefação dos dedos da mão direita com 3 meses de evolução. Negava dor, rigidez matinal e limitação funcional associada. Não se apurou trauma recorrente dos dedos, história sugestiva de perturbação obsessivo-compulsiva ou outras perturbações psiquiátricas. Ao exame objetivo identificou-se tumefação fusiforme ao nível das IFP dos 2º, 3º e 4º dedos da mão direita, indolor à palpação ou mobilização. O restante exame objetivo não apresentou alterações de relevo. O estudo analítico também não revelou alterações significativas, nomeadamente, com parâmetros de fase aguda negativos, fator reumatoide, anticorpo anti-CCP e HLA B27 negativos. Foi solicitado estudo radiográfico das mãos onde se realça aparente espessamento dos tecidos moles. Realizou-se ecografia que corroborou o espessamento de tecidos moles, não se identificando sinovite, alterações tendinosas, musculares ou ósseas. Optou-se por uma atitude expectante, permanecendo após 3 anos de seguimento, sem qualquer limitação funcional.

Os autores pretendem com este caso realçar a existência desta patologia rara e que, em consonância com a literatura, tem de ser diferenciada da artrite idiopática juvenil, segundo os achados clínicos, laboratoriais e imagiológicos. O diagnóstico desta entidade é essencialmente clínico, podendo ser confirmado com recurso a biópsia cutânea. O seu reconhecimento e diagnóstico precoce é essencial, não só para tranquilizar o doente atendendo à benignidade da doença, como também, para evitar a realização de exames invasivos ou tratamentos desnecessários.

220 - A RARE CASE OF ACANTHOSIS NIGRICANS ASSOCIATED WITH DIFFUSE CUTANEOUS SYSTEMIC SCLEROSIS

Carolina Ochôa Matos[1, 2], Eduardo Dourado[1, 2], Cláudia Brazão[3], Pedro de Vasconcelos[4], Luís Soares de Almeida[3, 4], Nikita Khmelinskii[1, 2], Sofia C Barreira[1, 2]
[1]Serviço de Reumatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [2]Unidade de Investigação em Reumatologia - JE Fonseca Lab, Instituto de Medicina Molecular João Lobo Antunes, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [3]Serviço de Dermatologia, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal, [4]Laboratório de Dermatopatologia, Serviço de Dermatologia, Hospital de Santa Maria, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisboa, Portugal

Introduction: Acanthosis nigricans (AN) is a dermatosis characterized by velvety, papillomatous, dark-brown, hyperkeratotic plaques, distributed symmetrically on the neck, axillae, antecubital and popliteal fossae, and inguinal area. AN is often linked with endocrine disorders or can appear as a paraneoplastic sign. It is rarely found in autoimmune diseases, usually associated with anti-insulin receptor antibodies (IRAb). We

report a case of AN in a patient with diffuse cutaneous systemic sclerosis (dcSSc), showing no signs of insulin resistance or neoplasia.

Clinical vignette: A 55-year-old man of Indian descent presented to the Rheumatology outpatient clinic with a four-month history of abrupt-onset Raynaud phenomenon, nausea, vomiting, abdominal distension, diarrhoea, recurrent and profuse night sweats, occasional fever spikes (>38ºC), and weight loss of 27 Kg. He also reported experiencing inflammatory polyarthralgia accompanied by joint swelling and 30-minute morning stiffness, as well as darkening and thickening of the skin.

During the examination, hyperpigmented velvety plaques were observed on the patient's face (sparing the malar region), axillae (Figure 1-A), lower abdominal and inguinal area, as well as the ankles. These findings were consistent with AN. Additionally, there was mild sclerodactyly extending to the metacarpophalangeal joints, with a modified Rodnan skin score (mRSS) of 2/51.

Laboratory testing results were notable for microcytic, hypochromic anaemia (Hb 12.5 g/dL), elevated erythrocyte sedimentation rate (77 mm/h) and C-reactive protein (1.21 mg/dL), and positivity for antinuclear antibodies (titre 1/160, AC-4 pattern), anti-Ro52, and anti-PL-7. Blood glucose and fasting serum insulin levels were normal.

The body computed tomography (CT) scan showed a non-specific interstitial pneumonia pattern and axillary, mediastinal, and hilar lymphadenopathies. Upper endoscopy revealed chronic non-atrophic antral gastritis, positive for Helicobacter pylori. A biopsy of the axillary lymph nodes and positron emission tomography/CT did not reveal any evidence of malignancy. Lung function tests showed a restrictive lung pattern. Oesophageal manometry showed absent oesophagal contractility. Capillaroscopy revealed a "scleroderma-like" pattern. The colonoscopic and echocardiographic findings were unremarkable.

In the first months of follow-up, the skin thickening progressed (maximum mRSS of 23/51) and dcSSc was diagnosed. Mycophenolate mofetil (MMF), hydroxychloroquine, and vasodilators were started with clinical improvement. Nine months after starting MMF, dark skin pigmentation was markedly fading, especially on the axillae (Figure 1-B), and skin thickening improved (mRSS of 16/51). A skin biopsy confirmed AN (Figure 1-C).

Conclusion: The association of AN and SSc has been seldomely reported in the literature. Two previous reports described AN in SSc patients with metastatic gastric adenocarcinoma and diabetes mellitus with IRAb. Two additional reports described AN as the initial manifestation of SSc without evidence of malignancy or metabolic diseases, as in the present case. Furthermore, as seen in cases with IRAb, AN improved with immunosupressive treatment.

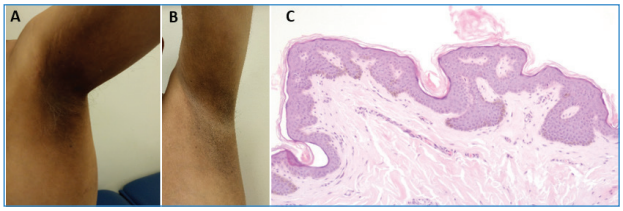

Figure 1. Before(A)and after(B)treatment;C:Epidermal papilomatosis,acanthosis,hyperkeratosis and hyperpigmentation of basal keratinocytes.

In summary, AN commonly signifies an underlying systemic condition, including autoimmune disorders. This case highlights the importance of thorough investigation, especially to exclude malignancy and insulin resistance.

225 - INFEÇÃO POR CITOMEGALOVÍRUS EM DOENTE COM ARTRITE REUMATOIDE SOB BARICITINIB

Daniel Melim[1], Daniel Carvalho[1], Margarida Faria[1], Lídia Teixeira[1], Ricardo Figueira[1], Alberto Quintal[1], Mário Rodrigues[1]

[1]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal

Os doentes com Artrite Reumatoide (AR) têm um risco de infeção aumentado, quer pela imunopatologia inerente à doença, comorbilidades associadas ou uso de imunossupressores como modalidade de tratamento. O Baricitinib é um inibidor seletivo e reversível da Janus Quinase (JAK) 1 e JAK2, sendo uma das opções terapêuticas na AR. Os inibidores da JAK têm sido associados a infeções virais, como o Herpes Zoster e, menos frequentemente, na infeção por outro herpesvírus, o Citomegalovírus (CMV).

Relatamos o caso de uma doente do sexo feminino, 55 anos, com AR seropositiva com 20 anos de evolução, atualmente medicada com Baricitinib, Hidroxicloroquina e Prednisolona. Trata-se de um caso de difícil controlo da atividade da doença, tendo realizado múltiplos fármacos antirreumáticos modificadores da doença, nomeadamente, Metotrexato, Sulfassalazina, Etanercept de 2010 até 2020, Tocilizumab em 2020, Rituximab em 2020 e Baricitinib desde dezembro de 2021. Recorreu à consulta em março de 2023 por quadro de febre, cefaleia, náuseas, anorexia e desconforto abdominal generalizado com uma semana de evolução. Ao exame objetivo encontrava-se febril, com punhos, cotovelos

e joelhos tumefactos e dolorosos, e dor à palpação do hipocôndrio direito. Analiticamente a destacar uma linfopénia de 0.9 (10^3/µL) com 15% linfócitos atípicos, ALT 584 (U/L), AST 575 (U/L), LDH 826 (U/L) e proteína C reativa 50.3 (mg/L). Foram pedidas serologias virais, identificando-se CMV IgM positivo, CMV IgG positivo com baixa avidez e carga viral de 61388 (cópias/mL). Diagnosticada infeção por CMV, pelo que suspendeu a toma de Baricitinib e cumpriu Valganciclovir durante 21 dias, mantendo vigilância semanal em consulta. Verificou-se melhoria clínica e analítica, com carga viral indetetável após o tratamento. Optou-se por não reiniciar terapêutica com Baricitinib.

Os autores pretendem com este caso sensibilizar para a ocorrência de infeções oportunistas em doentes medicados com inibidores da JAK, nomeadamente a infeção por CMV. Em doentes imunocompetentes, a infeção por CMV poderá ser assintomática, mas em estados de imunossupressão as manifestações podem ser graves e variadas, apelando-se à monitorização de envolvimento ocular, gastrointestinal, pulmonar e do sistema nervoso central. A literatura sugere que o risco de infeção por herpesvírus é superior nos doentes sob corticoterapia sistémica concomitante, como no caso exposto. O mecanismo pelo qual a adição de um inibidor da JAK aumenta o risco de infeção por herpesvírus não está totalmente esclarecida, admitindo-se como mais provável fator causal, a inibição de sinal mediado por interferão.

São necessários mais dados a fim de apurar a segurança da reintrodução de um inibidor da JAK após uma infeção por CMV.

227 - HIPOCALCÉMIA GRAVE SECUNDÁRIA AO DENOSUMAB EM DOENTE COM DOENÇA RENAL CRÓNICA

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

Introdução: O denosumab é um anticorpo monoclonal humanizado IgG2 utilizado no tratamento da osteoporose, que atua através da sua ligação ao RANKL, inibindo a reabsorção óssea e, consequentemente, reduzindo o risco de fratura. A hipocalcemia é um reconhecido efeito adverso do denosumab, especialmente em doentes com Doença Renal Crónica (DRC), com uma incidência estimada de 14-15%.

Caso Clínico: Mulher, 63 anos, seguida em consulta de Reumatologia por Artrite Reumatóide com amiloidose AA secundária, a condicionar DRC estadio 5, sob terapêutica com tocilizumab (TCZ) 8 mg/kg endovenoso (EV) mensal, sulfassalazina 1g/dia e prednisolona 5 mg/dia. Apresenta concomitantemente diagnóstico de osteoporose não fraturária sob tratamento com denosumab 60 mg subcutâneo a cada 6 meses, iniciado há cerca de 2 anos.

Na sessão de Hospital de Dia para perfusão de TCZ, 5 semanas após a última administração do denosumab, a doente reportou fadiga fácil, parestesias nos membros superiores e inferiores, assim como agravamento das queixas de artralgias de ritmo misto em ombros com evolução nas últimas semanas. Ao exame físico, destacava-se apenas a presença de artrite dos cotovelos e de pequenas articulações das mãos, sem outros dados de relevo. Salienta-se do estudo analítico realizado, hipocalcemia grave (5.2 mg/dL), hiperfosfatemia (8.5 mg/dL), elevação da PTH (1188.57 pg/mL), anemia normocítica normocrómica (hemoglobina 11.3 mg/dL), creatinina estável face ao basal (3.4 mg/dL) e elevação da ureia (258 mg/dL) e da velocidade de sedimentação (34 mm/h). O eletrocardiograma (ECG) demostrou prolongamento do QT corrigido (507 ms). Assim, a doente foi admitida na Unidade de Cuidados Intermédios por hipocalcemia grave sintomática, admitindo-se como provável etiologia a administração recente de denosumab. Foi iniciada terapêutica de reposição com cálcio EV e oral, tendo a doente reportado uma resolução da fadiga e das parestesias com a normalização da calcémia. À data de alta do internamento, 6 dias após a admissão, a doente apresentava cálcio corrigido de 8.9 mg/dL. Foi suspensa a terapêutica com denosumab e mantida a suplementação com carbonato de cálcio 1500 mg/dia e colecalciferol 22400 UI/mês.

Dois meses após a alta, a doente apresentou novamente hipocalcemia grave (6.2 mg/dL) apesar da suplementação com cálcio oral diária, assintomática, mas com prolongamento do QT no ECG (489 ms). Por recusa de novo internamento por parte da doente, foi realizada a correção iónica com perfusão de gluconato de cálcio no Hospital de Dia e ajustada a suplementação com carbonato de cálcio oral para 1500 mg 3 vezes/dia e iniciada suplementação com alfacalcidol 0,25 µg 2 vezes/dia, tendo-se verificado uma normalização da calcémia (8.7 mg/dL).

Conclusão: Nos doentes com DRC, o tratamento da osteoporose é sempre um desafio, não só pelo limitado leque de opções terapêuticas, mas também pelo maior risco de complicações associadas aos fármacos. O caso clínico apresentado pretende alertar para o reconhecimento da hipocalcemia como efeito adverso do tratamento com denosumab, particularmente em doentes com DRC. Esta pode apresentar-se de forma grave com alterações eletrocardiográficas, sendo a sua identificação e tratamento precoces fundamentais.

229 - MENINGOCOCCÉMIA EM DOENTE COM LUPUS ERITEMATOSO SISTÉMICO JUVENIL

Daniel Melim[1], Daniel Carvalho[1], Margarida Faria[1], Lídia Teixeira[1], Ricardo Figueira[1], Alberto Quintal[1], Mário Rodrigues[1]
[1]Serviço de Reumatologia, Hospital Central do Funchal, Funchal, Portugal

O Lupus Eritematoso Sistémico (LES) é uma doença reumática sistémica crónica que pode afetar qualquer órgão ou sistema, caracterizando-se pela produção e eliminação anormal de autoanticorpos. As infeções mantêm-se como uma das principais causas de morbilidade e mortalidade nos doentes com LES. A Neisseria meningitidis coloniza assintomaticamente a nasofaringe de 40% da população, sendo que em casos de asplenia anatómica ou funcional, imunodeficiência ou défice de complemento pode causar doença invasiva. Os doentes com LES têm comumente níveis de circulantes de C3 e C4 diminuídos, no entanto a associação com meningococcémia na literatura é rara.

Reportamos um caso de uma doente do sexo feminino, caucasiana, de 18 anos, seguida em consulta de Reumatologia por LES diagnosticado em 2013, com envolvimento cutâneo, articular, renal, hematológico e neurológico. Antecedentes relevantes de Sépsis meningocócica em 2011, isolando-se N. meningitidis serogrupo Y em hemoculturas. Atualmente medicada com Micofenolato de Mofetil, Prednisolona, Hidroxicloroquina e Lisinopril. A doente tem um seguimento errático nas consultas, com períodos prolongados de ausência e suspensão de medicação por iniciativa própria. Ausentou-se da consulta durante um ano, reiniciando seguimento e terapêutica em outubro de 2022. Em novembro de 2022, foi enviada ao Serviço de Urgência por quadro de febre, prostração, rash cutâneo, mialgias e artralgias com 3 dias de evolução. Ao exame objetivo apresentava febre, oligartrite com envolvimento das tibiotársicas, joelho e punho direito, rash não pruriginoso envolvendo a face, tronco, membros superiores e lesões petequiais nas mãos e pés. O restante exame objetivo não apresentava alterações de relevo. Analiticamente identificou-se uma leucocitose de 15.2 (10^3/µL), 93% neutrófilos, velocidade de sedimentação de 66 (mm), proteína C reativa de 98 (mg/L) e défice de C4. Atendendo aos achados clínicos e laboratoriais, ficou internada para estudo complementar. As hemoculturas revelaram positividade para N. meningitidis. Foi realizada punção lombar com exame bioquímico e cultural negativos para o agente. Realizou antibioterapia dirigida com Ceftriaxona, apresentando melhoria clínica e analítica progressiva.

A incidência de doença invasiva meningocócica (DIM) em Portugal apresenta uma tendência decrescente nas últimas décadas, que em muito se deve às políticas de vacinação implementadas. Segundo os dados do relatório de vigilância epidemiológica, a incidência global de DIM em Portugal foi de 0,39 casos por 100000 habitantes em 2020. Reportam-se ainda taxas de mortalidade elevadas, na ordem dos 7,1%. Os doentes com LES ativo encontram-se geralmente num estado de hipocomplementemia, pelo que se torna intuitivo admitir que tenham risco aumentado para infeções por microrganismos capsulados, como a N. meningitidis. No entanto, são escassos os casos reportados na literatura, sendo mais frequentemente associada a infeção pelo serogrupo Y nos doentes com LES.

Desde 2020 que o Programa Nacional de Vacinação contempla a administração da vacina contra o a N. meningitidis do serogrupo B e C, reservando-se a imunização contra outros serogrupos para grupos de risco. Questiona-se deste modo a pertinência e, atendendo à atual baixa incidência de DIM, a custo-efetividade de alargar as indicações para imunização com as vacinas quadrivalentes contra os serogrupos ACWY aos doentes com LES.

234 - SPLENOMEGALY AS THE INITIAL PRESENTATION OF SARCOIDOSIS

João Paulo Vilas-Boas[1], João Pacheco[2]
[1]Rheumatology Department, Centro Hospitalar do Baixo Vouga, Aveiro, Portugal, [2]Anatomia-Patológica, Centro Hospitalar e Universitário de São João, Porto, Portugal

Clinical Case: A 58-year-old female patient was referred to a general surgery consultation due to abdominal pain and a feeling of postprandial fullness with progressive worsening after one year of evolution. Her past medical-surgical history included essential arterial hypertension, non-insulin-dependent diabetes mellitus and multi-nodular goiter. At the first evaluation in the hospital context, she mentioned anorexia and weight loss. The patient denied respiratory, genitourinary, musculoskeletal, mucocutaneous symptoms in the previous year. On physical examination it was noted diffuse abdominal pain on palpation, accompanied by a decrease in bowel sounds and dullness on percussion in the left hypochondrium and flank. There were no other alterations on objective cardiopulmonary or musculoskeletal examination. Analytically, she also had previous records of persistent elevation of aminotransferases (about 2 times the normal limit) and sustained elevation of gamma glutamyl transferase and alkaline phosphatase. Her blood count was normal.

Abdominal CT was requested due to suspicion of hepatosplenomegaly, which demonstrated an enlarged spleen (15.5x9x5.8 mm), heterogeneous, with multiple dispersed pericentimetric hypodense nodules (image 1), as well as adenomegaly in the splenic hilum and multiple ganglionic formations along the inter-aortic chain.

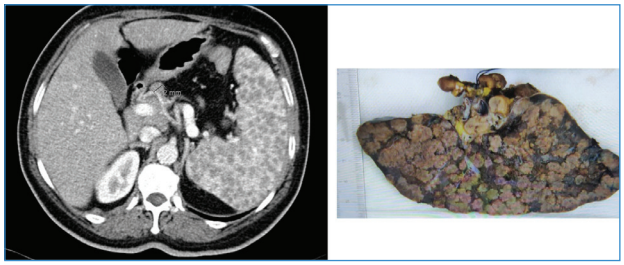

Figure 1. Sarcoidose esplénica (múltiplos nódulos sarcóticos observáveis em TC e em peça de esplenectomia)

The findings described raised the suspicion of a lymphoproliferative or granulomatous disease. Splenectomy was performed with subsequent anatomopathological study, which revealed numerous epithelioid granulomas compatible with a granulomatous inflammatory process of the sarcoidotic type, with extensive involvement of the spleen, lymph nodes and epiploon. Given the suspicion of sarcoidosis, a high-resolution chest CT scan, bone scintigraphy and ophthalmological evaluation were requested in order to exclude involvement of other organs and systems. Chest CT showed multiple peribronchovascular nodules and mediastinal ganglia that were larger than usual, a process compatible with stage II thoracic sarcoidosis. The ophthalmologic evaluation and the scintigraphy showed no alterations. In view of all these data, a diagnosis of hepatic, splenic, and thoracic sarcoidosis was made.

Discussion: This case represents a case of multisystemic sarcoidosis with splenic involvement as the initial presentation. Approximately 6% of patients have splenomegaly initially or during the course of the disease, which is considered an infrequent finding. Complications of splenomegaly include abdominal pain, anorexia, portal hypertension, anemia, leukopenia and thrombocytopenia. Previous studies have shown splenic nodules in 50% of the autopsies of patients with sarcoidosis, which suggests that the involvement may be mostly subclinical. Sarcoidosis should therefore be considered as a differential diagnosis in the presence of splenomegaly, with pulmonary, cutaneous and ophthalmological evaluation being pertinent to exclude involvement of other organs and systems. Other differential diagnoses include infectious diseases and lymphoproliferative disorders.

235 - MIELITE TRANSVERSA LONGITUDINALMENTE EXTENSA NUM DOENTE COM LUPUS ERITEMATOSO SISTÉMICO: EFICÁCIA DO RITUXIMAB NUM CASO DE DOENÇA DO ESPECTRO DA NEUROMIELITE ÓPTICA

Carlos Marques-Gomes[1,2], Mariana Diz-Lopes[1,2], Bruno Miguel Fernandes[1,2], Ricardo Reis[3,4,5,6], Miguel Bernardes[1,2,7,8], Filipe Brandão[7,8], Joana Guimarães[3,4,5,9], Lúcia Costa[1]

[1]Serviço de Reumatologia, Centro Hospitalar de São João, Porto, Portugal, [2]Departamento de Medicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal, [3]Serviço de Neurologia, Centro Hospitalar de São João, Porto, Portugal, [4]Departamento de Neurociências Clínicas e Saúde Mental, Faculdade de Medicina da Universidade do Porto, Porto, Portugal, [5]Departamento de Biomedicina, Faculdade de Medicina da Universidade do Porto, Porto, Portugal, [6]i3S - Instituto de Investigação e Inovação em Saúde, Universidade do Porto, Porto, Portugal, [7]Unidade de Reumatologia, Hospital CUF, Porto, Portugal, [8]Unidade de Reumatologia, Instituto CUF, Porto, Portugal, [9]Center for Drug Discovery and Innovative Medicines (MedInUP), Universidade do Porto, Porto, Portugal

Introdução: A Neuromielite Óptica (NMO) é uma doença desmielinizante inflamatória rara do sistema nervoso central (SNC), que afeta a medula espinhal e os nervos óticos. O envolvimento do SNC no Lúpus Eritematoso Sistémico (LES) é causa de morbimortalidade aumentada, podendo sobrepor-se a doenças primárias do SNC.

Caso Clínico: Homem de 41 anos, caucasiano, diagnosticado com LES desde 2013 [sob hidroxicloroquina (HCQ)], apresentou-se, em agosto de 2018, com espasmos apendiculares tónicos dolorosos, involuntários e autolimitados, sinal de Lhermitte e parestesias simétricas nas extremidades dos membros inferiores (MIs). Um mês depois, referia diminuição da força muscular no membro superior esquerdo e dificuldade no controlo dos esfíncteres. Ao exame físico, apresentava diminuição da força muscular no braço esquerdo (grau 4/5) e hipostesia nos dedos dos pés. Do estudo complementar salientavam-se: eletromiografia normal dos MIs; ressonância magnética (RM) do neuroeixo com múltiplas lesões focais medulares periféricas, hiperintensas em T2 e com captação de contraste, na medula cervico-dorsal (C2, C7, D1-D3, D6-D8 e D11). Com diagnóstico provável de neuro-lúpus, foi internado para realizar punção lombar (PL); após pulsos de metilprednisolona (MP) (1g/dia e.v., 3 dias), verificou-se melhoria neurológica parcial, tendo alta sob HCQ e prednisolona (PDN) (60mg/dia). Os resultados da PL foram inconclusivos.

Em outubro de 2018, por aumento dos episódios tónicos dolorosos, foi reinternado. A RM medular mostrou esbatimento das lesões medulares prévias e desaparecimento das de C7 e D11 e o estudo imunológico mostrou elevação dos anticorpos (ac) antidsDNA e consumo de C4. Perante a hipótese de doença desmielinizante, foi re-

alizada colheita para atcs anti-aquaporina4 (AQP4) e anti-glicoproteína da mielina do oligodendrócito (MOG), tendo alta medicado com HCQ, pregabalina e esquema de desmame da PDN oral até 20mg/dia.

A positividade para atcs anti-AQP4 confirmou o diagnóstico de NMO e, em novembro de 2018, iniciou azatioprina (AZA) com aumento gradual de dose até 150mg/dia. Um mês depois, apresentou-se melhorado, repetiu RM medular que mostrou redução das lesões prévias e ausência de hipersinal medular, iniciando em janeiro de 2019 o desmame da PDN oral (até 5 mg/dia) e redução na dose da AZA.

Em fevereiro de 2019, por atividade do LES (presença de úlceras orais persistentes e de pneumonite NSIP, aumento dos ac antidsDNA, consumo de C4 e agravamento das parestesias), retomou PDN 10 mg/dia e aumentou a AZA para 150 mg/dia. Contudo, por agravamento clínico contínuo, com aumento da frequência e intensidade das disestesias dos MIs e recorrência dos episódios tónicos dolorosos, realizou 9 novos pulsos de MP (fevereiro de 2020). Verificou-se apenas um alívio parcial da sintomatologia, mantendo sinais de atividade do LES. A RM do neuroeixo revelou lesões focais com hipersinal medular em T2 na região dorsal (D6-D9), iniciando terapêutica com rituximab.

Atualmente, encontra-se em remissão e a última RM, após 4 ciclos de rituximab, não apresentou lesões inflamatórias.

Discussão: Estudos recentes sugerem que metade das mielites no contexto do LES apresentam positividade para os ac anti-AQP4. Contudo, desconhece-se o verdadeiro impacto da associação NMO/LES no prognóstico destes doentes a longo prazo e qual a melhor estratégia terapêutica.

Este caso clínico acrescenta mais evidência neste campo, reforçando a utilidade do rastreio dos atcs anti-AQP4 nos doentes com LES e mielite e demonstrando uma boa resposta clínica ao rituximab após falência de resposta à azatioprina.

238 - ARTERITE DE TAKAYASU – T2T: CASO CLÍNICO

Catarina S Cortesão[1], José Melo-Gomes[1]
[1]Rheumatology Department, Instituto Português de Reumatologia, Lisboa, Portugal

Introdução: Arterite de Takayasu (ATK) é uma vasculite rara de grandes vasos, crónica, inflamatória e imunomediada cuja etiologia se desconhece. A apresentação clínica é inespecífica, com sintomas constitucionais, evoluindo com quadro sugestivo de doença arterial oclusiva ou isquémica. O diagnóstico precoce é dificultado pela falta de marcadores específicos ou sinais patognomónicos e pela variabilidade de apresentação dependente da gravidade e localização das lesões vasculares. O tratamento varia de acordo com a gravidade de doença, iniciando-se com glucocorticóides (GC) e agentes modificadores da doença (DMARD) convencionais. Nos casos refratários deve ser equacionada terapêutica biotecnológica o mais precocemente possível.

Caso clínico: Mulher de 21 anos previamente saudável, com um quadro indolente de 3 anos de astenia, dores tóraco-abdominais difusas e temperaturas sub-febris. Internamento num serviço de Medicina Interna, por agravamento, em jan2021. Analiticamente salientavam-se anemia (Hb 7.7g/dL), VS 126mm/h e PCR 15,3mg/dL e na avaliação por angioRM redução focal de fluxo do segmento cervical alto da artéria carótida interna esquerda e espessamento da parede vascular, levando ao diagnóstico de ATK. Nesta altura iniciou prednisolona (PDN) 50mg que manteve durante 3 meses com controlo dos sintomas constitucionais, mas com recidiva das queixas aquando do seu desmame.

Em maio 2022, recorreu a Consulta de Reumatologia para segunda opinião. Mantinha sintomas constitucionais, apesar de 50mg/dia de PDN. Observaram-se mãos frias, sem pulsos radial e cubital palpáveis, estrias cutâneas abdominais arroxeadas e IMC 18,2Kg/m2. Analiticamente sem anemia com leucocitose, VS 11mm/h e PCR 3.7mg/dL. Na ecografia carotídea com d-ppler, havia espessamento parietal difuso isoecogénico e concêntrico nas paredes arteriais ao nível das artérias carótidas primitivas, discretamente na bifurcação carotidea direita e tronco braquicefalico.

Iniciou metotrexato (MTX) 20mg semanal e indicação para reduzir PND paulatinamente até 25mg. Em julho com PDN 25mg e MTX 20mg, sintomas mais controlados, mas exame objetivo sobreponível e VS e PCR elevados. Foi proposto início de tocilizumab (TCZ) 162mg semanal e na avaliação dos 3 meses foi objetivado pulso radial palpável pela primeira vez. Em dezembro, pulso radial e cubital palpáveis bilateralmente. Nos meses seguintes foi possível o desmame de PDN progressivo estando nesta fase sob TCZ 162mg, MTX 20mg e PDN 17,5mg, sem sinais ou sintomas de doença ativa.

Discussão: Neste caso clínico, o diagnóstico de ATK, com recurso a angioRM, ocorreu antes de lesões isquémicas graves e não se verificou atraso no início de terapêutica de indução com PDN 1mg/Kg/dia, que controlou a progressão da doença e melhorou os sintomas constitucionais. No entanto, a diminuição de dose de PDN acompanhou-se de novos surtos inflamatórios. Assim, a doente manteve PDN em monoterapia durante 15 meses, sem atualização de terapêutica ou controlo da doença.

Nessa altura a doente procurou uma segunda opinião e foi decidido iniciar um DMARD biológico, o tocili-

zumab. A doente iniciou TCZ 20 meses após início dos sintomas, com alteração marcada do curso da doença na avaliação ao 3º mês de terapêutica biológica.

Conclusão: AKT é uma doença rara que pressupõe um alto grau de suspeição e articulação multidisciplinar para o diagnóstico. O tratamento é de fundamental importância para impedir lesões isquémicas graves que podem modificar de forma significativa a qualidade de vida e o prognóstico vital destes doentes, sendo fundamental uma estratégia de "T2T".

242 - INIBIDORES DE CHECKPOINT IMUNITÁRIO COMO FORMA DE MIMETISMO DE SÍNDROME DE SJÖGREN, A PROPÓSITO DE UM CASO CLÍNICO

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

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

Introdução: O Síndrome de Sjögren (SS) é uma doença crónica imunomediada, caracterizada por envolvimento de glândulas exócrinas, com xerostomia e xeroftalmia, podendo ainda afetar outras mucosas. Pode surgir isoladamente (primário) ou associada a outra doença imunomediada, como a Artrite Reumatoide. As queixas extraglandulares, apesar de mais raras, são muito relevantes, quer pelo impacto na qualidade de vida, quer no prognóstico do doente, sendo também um potencial confundidor, uma vez que podem preceder as manifestações secas, que são a base desta patologia[1].

O seu caráter multissistémico leva a uma grande diversidade de manifestações, que podem constituir um desafio diagnóstico, acrescido pela possibilidade de existência de fatores confundidores como determinados fármacos. Neste contexto, destacam-se os inibidores de checkpoint imune (ICI), terapêutica em expansão no ramo da Oncologia, estando descrita síndrome sicca associada a estes fármacos[2].

Caso Clínico: Mulher, 71 anos, com antecedentes de hipertensão arterial, hipotiroidismo e diagnóstico de neoplasia vesical em maio/2022, submetida a esquema de Pembrolizumab em setembro/2022 (fez toma única), após falha da terapêutica de 1ª linha. Em novembro/2022, recorreu ao serviço de urgência (SU) por clínica de parotidite com xerostomia e xeroftalmia. Assumido quadro provavelmente iatrogénico, tendo iniciado prednisolona 60 mg/dia e pilocarpina.

Por ter sido equacionada hipótese de SS, foi referenciada à consulta de Reumatologia, em janeiro/2023, onde foi avaliada. Nessa altura, não apresentava sintomatologia sugestiva de SS, além das queixas secas, tendo-se iniciado desmame da corticoterapia (CCT).

Cerca de 2 semanas após avaliação inicial, foi admitida no internamento de Nefrologia para estudo etiológico de Lesão Renal Aguda anúrica (creatinina: 5.2 mg/dl à admissão para basal: 0.59 mg/dl) e início de técnica dialítica. Nas análises realizadas, apresentava positividade para Anticorpos Antinucleares (ANA) (1/160 – AC 2,4,5 mosqueado), Velocidade de Sedimentação de 96 mm/h, sem outras alterações relevantes, incluindo subclasses de IgG normais.

Foi realizada biópsia renal que demonstrou Nefrite Tubulointersticial com sinais de atividade, coincidindo esta agudização com a redução da CCT. Apesar do baixo título de ANA, ausência de outras queixas e quadro inicialmente sugestivo de toxicidade medicamentosa, a hipótese de SS mantinha-se, quer pelo flare da doença após redução de CCT, quer pela forma de atingimento renal, uma das mais comuns nesta doença.

Por persistir dúvida etiológica, realizou-se biópsia das glândulas salivares minor, dada a especificidade do padrão histológico no SS, onde se constatou a presença de infiltrado inflamatório inespecífico, pouco sugestivo deste diagnóstico.

Com base nesta evidência, considerou-se que a hipótese de síndrome seca secundária a fármacos seria a mais provável, tendo mantido CCT sistémica, com normalização da função renal e resolução das queixas secas.

Conclusão: O caso descrito evidencia a dificuldade em estabelecer critérios de diagnóstico precisos e fiáveis em doenças multissistémicas e complexas como o SS, que podem representar um verdadeiro desafio diagnóstico. Com este caso, pretende-se alertar para o diagnóstico diferencial, muitas vezes amplo, em doentes com manifestações que, apesar de sugestivas, podem ser associadas a outras entidades, nomeadamente fármacos. É expectável que, com o uso crescente dos ICI, casos como este se tornem cada vez mais prevalentes, pelo que é essencial conhecer os seus efeitos adversos.

250 - PAGET DISEASE OF BONE - A DIAGNOSTIC CHALLENGE

Miguel Correia Natal[1, 2], Chenyi Zhu[1], Ana Isabel Morais[3], Joana Tender Vieira[1], Jorge Almeida[1]

[1]Internal Medicine, Centro Hospitalar de S. João, E.P.E, Porto, Portugal, [2]Serviço de Reumatologia, Centro Hospitalar de São João, Porto, Portugal, [3]Serviço de Ortopedia e Traumatologia, Centro Hospitalar São João, Porto, Portugal

Introduction: Paget disease of bone (PDB) is a focal disorder of bone metabolism in which an accelerated bone turnover (believed to be mainly associated with abnormalities in pathways related to osteoclast activa-

tion and differentiation) leads to abnormal deposition and overgrowth of bone in affected sites.

It most often affects patients over the age of 55 and has a predilection for the skull, thoracolumbar spine, pelvis, and long bones of the lower extremities. The most frequent clinical manifestations of PDB are pain (either from the pagetic lesion or from secondary causes, including osteoarthritis, nerve compression, and tumor) and bone deformities, with an elevated risk of fracture. However, the majority of patients are asymptomatic and the disease is incidentally detected.

A combination of clinical and/or laboratory (such as an isolated elevation of alkaline phosphatase) findings and suggestive radiographic changes is usually diagnostic of PDB.

Clinical case: Woman, 66 years old, with no relevant personal medical history, was admitted for etiological investigation of a pathological fracture of the left humerus (Fig.1), diagnosed after a fall from her own height. She had no complaints prior to the episode, particularly related to pain. The analytical study highlighted only an elevated level of alkaline phosphatase (267 U/L, reference range: 30-120 U/L). On the other hand, the analytical study of serum protein electrophoresis, liver transaminases, thyroid-stimulating hormone (TSH), parathyroid hormone (PTH), vitamin D, calcium, and phosphorus were within the reference values. The possibility of metastatic injury was ruled out with a whole-body CT scan and appropriate cancer screenings for the patient's age group. The skeletal X-ray showed a single lesion with heterogeneous trabecular bone and sclerotic areas in the proximal end of the left humerus. A directed CT scan was performed, which identified heterogeneous bone texture with lytic and sclerotic areas, regions with ground-glass densification, and compensatory hyperostosis in the proximal humerus, raising the suspicion of PDB. Due to the potential need for surgical intervention and the absence of unequivocal signs of the disease, bone biopsy and MRI were performed. The bone biopsy showed reactive changes to the fracture, while the MRI confirmed the presence of findings suggestive of PDB, with bone expansion and coarse heterogeneous trabeculation involving the proximal half of the humerus up to the fracture site. After multidisciplinary discussion, considering the duration of the condition and evidence of good consolidation in the follow-up X-ray, conservative treatment was decided upon, with a referral to a Rheumatology consultation.

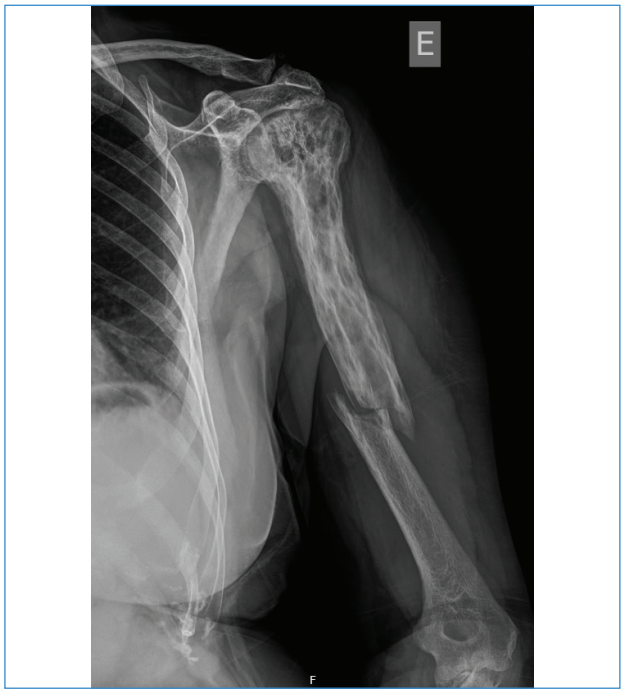

Figure 1.

Discussion: PDB is still not fully understood, and its pathogenesis remains unclear. Although the diagnosis is relatively straightforward in many cases, the variability in clinical and laboratory presentations, heterogeneity in bone involvement, and possible overlap with other conditions such as osteoporosis, osteoarthritis, or bone tumors can pose a diagnostic challenge. The described case represents an example of an atypical presentation of this disease, as the humerus is an infrequently affected site, monostotic involvement is present in less than 20% of cases and it was accompanied by less pronounced elevations in markers of bone metabolism. Therefore, it is important to maintain a high suspicion for PDB in patients with unexplained bone pain or pathological fractures.

251 - ENVOLVIMENTO EXTRAGLANDULAR NO SÍNDROME DE SJÖGREN – UMA SÉRIE DE CASOS

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

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

Introdução: A síndrome de Sjögren (SS) é uma doença crónica imunomediada, pautada por queixas secas, decorrentes da afetação de glândulas exócrinas. Pode afetar praticamente qualquer órgão e, assim, cursar com clínica muito variada que pode representar um sério desafio diagnóstico. Dada a grande variabilidade de manifestações possíveis, é necessário um elevado grau de suspeição clínica para o diagnóstico.

Caso 1: Pneumonite Intersticial Não Específica (NSIP) Mulher, 77 anos, seguida em Pneumologia para estu-

do de dispneia crónica e Tomografia Computorizada de Tórax com padrão de NSIP. A doente referia ainda artralgias inflamatórias nos punhos e tibiotársicas, xerostomia e xeroftalmia de longa data. Analiticamente com Anticorpos Antinucleares (ANA) e Anti-SSA positivos. Realizou-se biópsia das glândulas salivares minor (GSM), com presença de infiltrado inflamatório sugestivo de SS, tendo-se assumido esse diagnóstico.

A doença intersticial pulmonar foi enquadrada neste contexto e a doente iniciou hidroxicloroquina (HCQ) encontrando-se, à data, com doença estável.

Caso 2: Glomerulonefrite Membranoproliferativa (GNMP) e Nefrite Tubulointersticial (NTI)

Mulher, 83 anos, com antecedentes de Doença Renal Crónica (DRC) por nefropatia diabética, foi admitida no serviço de urgência (SU) em março/2023 com síndrome urémico por Lesão Renal Aguda anúrica (creatinina: 5.7 mg/dl e basal de 1.2 mg/dl), tendo sido internada para estudo etiológico e início de diálise.

Nas análises, identificado consumo de complemento e positividade para ANA, Anti-SSA e -SSB. A biópsia renal evidenciou GNMP e NTI crónica, tendo-se colocado a hipótese de SS com atingimento renal, sendo requisitada avaliação por Reumatologia.

A doente negava queixas secas ou outra clínica sugestiva de SS, pelo que se realizou biópsia das GSM, compatível com SS. Iniciou então corticoide (CCT) e Micofenolato (MMF), com recuperação parcial da função renal e normalização do débito urinário.

Caso 3: Acidose Tubular Renal (ATR) e Nefrocalcinose

Mulher, 79 anos, com história de DRC, vários episódios de nefrolitíase com necessidade de intervenção e Lúpus Eritematoso Sistémico (cutâneo e articular) de longa data, em remissão com HCQ. A doente referia xeroftalmia e xerostomia crónicas, agravadas nos últimos anos. Analiticamente, positividade para ANA, Anti-SSA, -SSB, Fator Reumatoide (FR) e hipergamaglobulinemia.

Realizou-se biópsia das GSM, dada a suspeita de SS, que revelou ser compatível com esta hipótese (vários focos), assumindo-se SS secundário.

Durante seguimento, apresentou proteinúria e função renal em agravamento, com acidose metabólica, hipercalemia e hipocitraturia, compatível com ATR. Iniciou-se MMF, com melhoria significativa da função renal e proteinúria.

Caso 4: Tetraparésia secundária a hipocalemia grave por ATR

Mulher, 33 anos, admitida no SU por tetraparésia espástica enquadrada em hipocalemia grave, com necessidade de suplementação de potássio e bicarbonato; foi diagnosticada ATR tipo 1 por Nefrologia e, na marcha diagnóstica, detetada positividade de anti-SSa e -SSb, sendo solicitada avaliação por Reumatologia. Apesar da ausência de clínica sugestiva de SS, apresentava positividade de FR, hipergamaglobulinemia e linfopenia. Realizou biópsia de GSM compatível com SS, tendo-se assumido diagnóstico.

Iniciou CCT em alta dose, com redução sucessiva da necessidade de suplementação iónica, encontrando-se atualmente com MMF, em esquema de redução de CCT, com monitorização da função renal, fator de resposta principal, que tem vindo a melhorar.