Imagens em Reumatologia

012 - WHEN BONE VANISHES: A RARE SYNDROME

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Introduction: Gorham-Stout syndrome, also named vanishing bone disease, is a rare disorder characterized by progressive osteolysis, with unclear aetiology1-2. We report a case of bilateral hip reabsorption, suggestive of Gorham-Stout Syndrome.

Clinical case: A 54 years old woman with morbid obesity and type 2 diabetes, with no smoking, alcohol or toxophiliac habits was referred to our consult from Orthopedy service due to radiographic abnormalities.

The patient reported chronic arthralgia, predominantly in knees and hip joints, and had already been diagnosed with coxarthrosis and gonarthrosis, for which she was medicated with non-steroidal anti-inflammatory drugs and opioids for pain management. When questioned, she described worsening of the bilateral hip pain since 2022, with progressive gait limitation, requiring crutches for gait support. Thus, hip plain radiographies were performed, which showed bilateral reabsorption of the acetabular head (picture 1A), not present in radiograms from 2018.

A computer tomography (CT) scan was performed (picture 1B), indicating severe acetabular dysplasia with deformity and resorption of the femoral heads, particularly on the left side, and significant sclerosis on the right, with intra-articular thickening and potential inflammatory pannus on the right side. The magnetic resonance imaging (MRI) (picture 1C-D) demonstrated bone loss in both femoral heads, bone marrow infarction in the proximal right femur, small bilateral joint effusions, and synovitis, more pronounced on the right; these findings suggested rapidly destructive osteoarthritis with underlying degenerative changes and marginal acetabular osteophyte remodelling.

There were no signs or symptoms suggestive of inflammatory arthropathy and except for vitamin D deficiency, there were no changes in phospho-calcium metabolism (normal calcium, phosphate and parathyroid hormone).

The radiological and clinical findings are consistent with Gorham-Stout Syndrome (Gorham's disease), a rare condition characterized by progressive osteolysis of unknown aetiology.

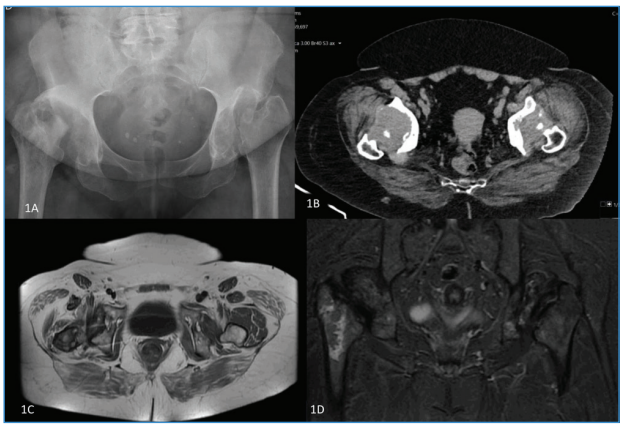

Figure 1. lA, hip plain radiographies showing bilateral reabsorption of the acetabular head; 18, CT-scan presents severe acetabular dysplasia with deformity and resorption of the femoral heads; 1C-D, MRI scan showing bone loss in both femoral heads.

Discussion: Gorham-Stout syndrome is a rare disorder, characterized by destruction of osseous matrix and proliferation of vascular structures, resulting in osteolysis1, which can involve one or multiple bone sites2. Its aetiology remains unclear and can have multiple clinical presentations, depending on affected bone, even though many symptoms are rather unspecific, which can delay the diagnosis1-2. The prognosis is variable, depending on the existence of other manifestations, such as pleural effusion or development of complications, such as infection or spinal cord involvement1. Reporting these cases and imaging findings is central for enhancing understanding and recognition of this condition.

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020 - HYDROCEPHALUS-RELATED DEMENTIA IN PAGET'S DISEASE OF THE SKULL

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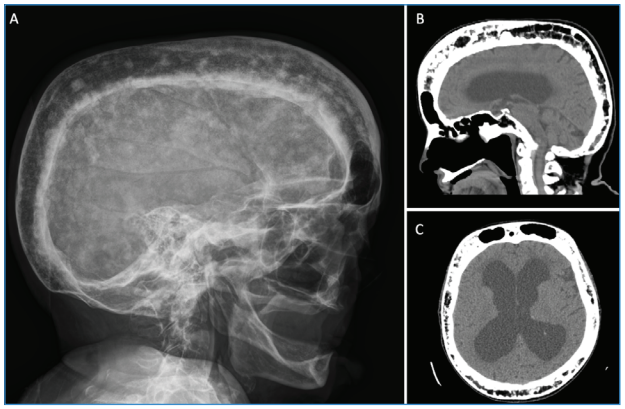

Figure. Radiography and computed tomography (CT) images

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An 83-year-old woman, fully dependent for daily activities, had a history of progressive dementia over several years, with marked loss of short-term memory, progressive decrease in spontaneous activity and speech, and apathy. The neck was short, and the head was markedly enlarged with a frontal bulge. Radiography showed an overall enlargement of the cranium with diploic widening, cotton wool appearance, and the Tam o' Shanter sign (panel A). Computed tomography disclosed platybasia with midbrain pons deformity, including ventral rectification, and a significant supratentorial ventricular enlargement due to obstruction of the posterior fossa cisterns and distortion of the Sylvius aqueduct (panels B and C). Biochemical tests revealed normal levels of calcium (10 mg/dL), phosphate (3.3 mg/dL), alkaline phosphatase (76 IU), P1NP (48.9 µg/L), and CTx (0.38 µg/L).

This case describes a rare presentation of advanced Paget's disease of the skull, biochemically quiescent, with platybasia and hydrocephalus-related dementia. Platybasia, characterized by flattening of the skull base, can result in cranial nerve compression and obstructive hydrocephalus. Clinicians should consider Paget's disease in elderly patients presenting with cognitive decline and skull abnormalities, as early diagnosis and management are crucial for preventing further neurological complications.

024 - TETRAPARESIS RELATED TO ATLANTOAXIAL SUBLUXATION: INDEX HOSPITAL PRESENTATION OF AN UNDIAGNOSED CASE OF RHEUMATOID ARTHRITIS

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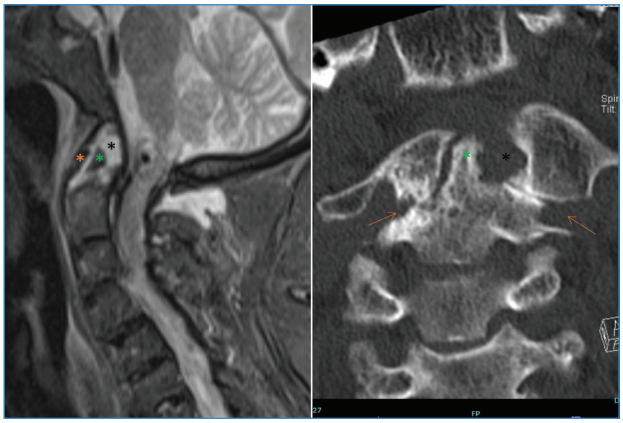

Figure. Sagital T2 STIR MRI sequence (left image) showing severe inflammatory pannus (black asterisk) with erosive destruction of the odontoid process (green asterisk); coronal CT scan (right image) showing dislocation of the C1-C2 lateral joint masses (arrows).

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A 77-year-old woman with a long-standing history of pain and progressive deformity of the hands, wrists, and knees, was admitted to the internal medicine inpatient clinic following tetraparesis that had developed gradually over the past few weeks. She complained of inflammatory neck pain during the last year. Physical examination showed subluxation and severe deformity of the wrists and proximal interphalangeal joints of the hands. Laboratory workup showed anemia of chronic disease, elevation of inflammatory markers and high titers of rheumatoid factor (3260 UI/mL) and anti-citrullinated peptide C antibodies (>300 UA/mL). CT-scan and MRI of the neck revealed posterior atlas subluxation with erosive destruction of the odontoid process of the axis and prominent inflammatory pannus. The diagnosis of seropositive rheumatoid arthritis with cervical spine involvement was made.

Anterior atlantoaxial subluxation is the most common form of cervical spine involvement in rheumatoid arthritis and has been decreasing in the past decades.1 We here present a case of advanced posterior atlantoaxial subluxation with odontoid erosion and inflammatory pannus, reinforcing the importance of maintaining awareness for the serious complications of the disease.

025 - PAGET'S DISEASE OF BONE: A CASE OF BILATERAL INVOLVEMENT OF THE RADIUS

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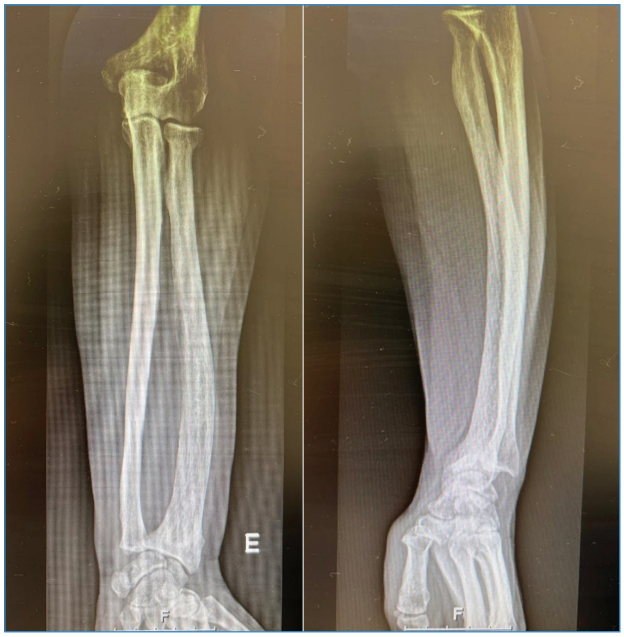

Figure. Left forearm x-ray (frontal and lateral views)

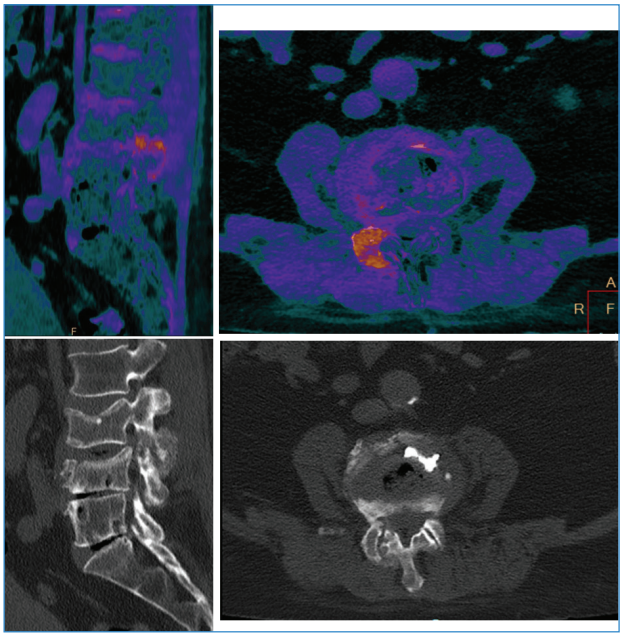

Figure. Tophaceous gout on lumbar spectral CT scan

A 55 year-old woman presented to the rheumatology outpatient clinic with a history of painless progressive deformity of the left forearm over the last year and of the right forearm over the last month. Physical examination showed a hard swelling of the distal third of the left forearm and right wrist in their radial aspects. The blood tests showed alkaline phosphatase elevation with normal calcium levels. Radiogram of the left forearm performed previously to the right-side complaints showed bowing and sclerosis of the left radius. The diagnosis of polyostotic Paget's disease of bone (PDB) was made.

PDB is the second most common bone remodeling disease following osteoporosis. The most involved sites are the axial skeleton, femur and tibia. Although the upper limbs are infrequently affected, here we present a bilateral involvement of the radius.

028 - TOPHACEOUS GOUT: AN ATYPICAL MANIFESTATION AND VERSATILITY OF SPECTRAL CT

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A 56-year-old man with a history of gout and alcohol consumption presented to the emergency department complaining of long-standing low back pain that worsened in the past few weeks. He was treated with colchicine and oral corticosteroids for the last 3 years. He had deformities of the hands, elbows and feet with the presence of significant tophi. Neurological exam detected no motor or sensitive deficits and palpation of the lumbar spine was painful. MRI of the lumbar spine depicted fractures of the L3 and L4 vertebral bodies and a bony lesion at the L3-L4 level. Spectral lumbar CT-scan was performed to better characterize this lesion that turned out to be uric acid deposition, in relation to tophaceous gout.

Gout is a common inflammatory arthropathy characterized by an inflammatory response to the deposition of monosodium urate crystals (MUC) in the synovium. Tophaceous gout is a type of manifestation of chronic gout, characterized by collections of solid MUC in soft tissue accompanied by chronic inflammatory and, often, destructive changes in extra-articular connective tissue. Tophi are most common in peripheral joints, tendons and bursae. The spine is an uncommon place for urate deposition, with a few dozen cases reported in the literature. In recent studies, spectral CT, through its ability in isolating specific molecules in the body, showed a high sensitivity and specificity for diagnosing MUC deposition (above 90%) with promising results regarding its usage for determining response to therapy and prognosis. According to recent literature only two cases of spine tophaceous gout identified on spectral CT were reported.

037 - A NEW ANGLE - A CASE OF SPINAL CURVATURE CHANGE IN ACHONDROPLASIA

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Achondroplasia is the most common form of bone dysplasia, with a prevalence of 1:20000. (1) This disorder is caused by a gain-of-function pathogenic variant in the FGFR3 gene and the diagnosis is confirmed by molecular testing. Its most typical features are a disproportionately short stature with long-bone shortening mainly affecting the proximal aspects of the extremities, midface hypoplasia, macrocephaly and exaggerated lumbar lordosis. Thoracolumbar kyphosis is typical in infancy. It usually resolves with age, as there is an increase in lordosis. The overall prevalence of thoracolumbar kyphosis is 35%, but there is no estimation of its prevalence in adults. The prevalence of lumbar spinal stenosis increases with age, being around 80% in adults. (2) Moreover, multiple other complications can arise from this condition. Therefore, following of these patients should be ensured by a multidisciplinary team, preferably with expertise in the area, with focus on education of the patient, adjustment of daily activities, physical therapy for functional gain and pain control and surveillance of common complications and comorbidities such as obesity, hypertension, recurrent otitis media and sleep apnea. (3)

A 55-year-old female presented to a Rheumatology clinic due to chronic aggravating back pain and neurogenic claudication. She had a known diagnosis of achondroplasia, but no specialized medical care. On examination she presented disproportionate short stature (height of 112cm), with shortened limbs, midface retrusion. She had a flattened dorsum but with a kyphotic, inverted angulation of the thoracolumbar transition. Imaging of the spine with computed tomography and magnetic resonance showed a L1 hemivertebra with an anteroposterior wedge and a lumbar stenotic spine due to degenerative changes.

The patient started follow-up in the genetics, otorhinolaryngology, physical medicine and rehabilitation and orthopedics clinics. Together with the patient, the decision for a initial non-surgical approach was made, with significant improvement of symptoms with physical therapy.

Achondroplasia is a form of bone dysplasia that has a high impact on the quality of life of patients. Musculoskeletal deformities and complaints contribute highly to the morbidity of this disorder and can lead to chronic pain and declined physical function. A multidisciplinary approach is best suited for the care of these patients.

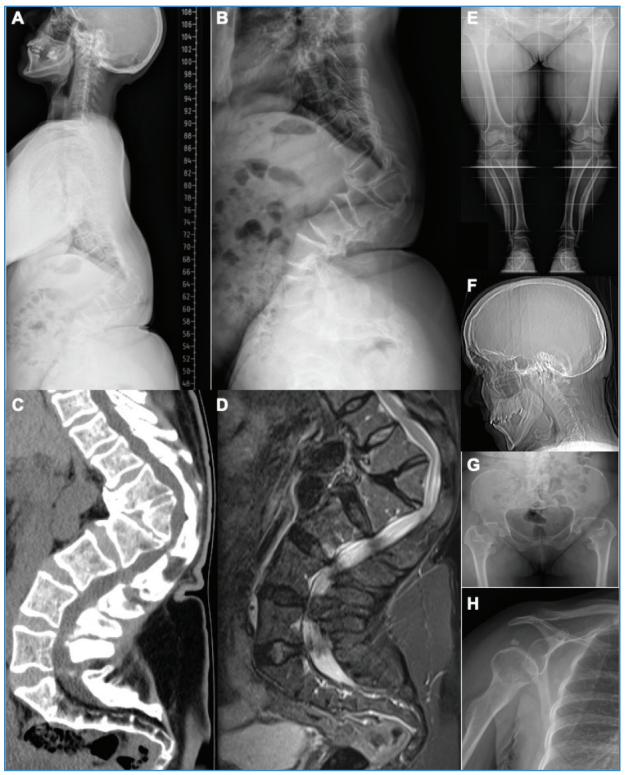

Figure. Radiographic, tomographic and magnetic resonance findings in achondroplasia; subtitle with greater detail in abstract

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046 - SCHWANNOMA OF THE POSTERIOR TIBIAL NERVE: A (NOT SO) COMMON CAUSE OF TARSAL TUNNEL SYNDROME

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Presentation: A 68-year-old woman was followed in a rheumatology appointment with long-standing radio-

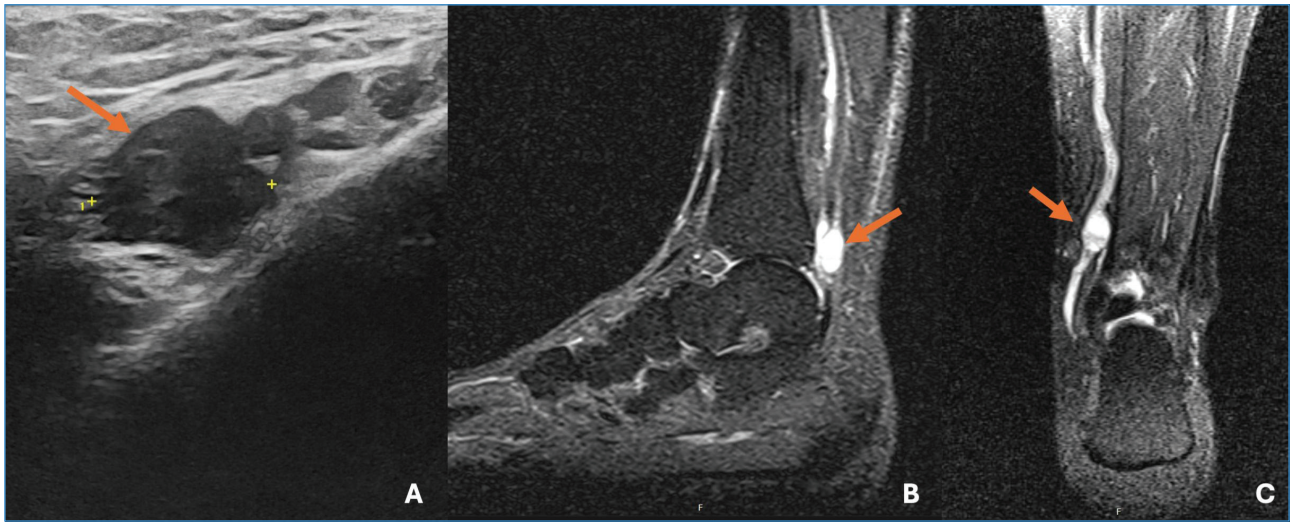

Figure. (A) Ultrasound of the posterior tibial nerve showing the nerve tumor as a hypoechoic mass within the nerve sheath (arrow); (B) Sagittal, turbo inversion recovery magnitude sequence magnetic resonance imaging of the ankle showing the nerve tumor as a hypoechoic structure posterior to medial malleolus (arrow); (C) Coronal, T2-weighted resonance imaging of the ankle with the nerve tumor (arrow).

graphic axial spondyloarthritis currently treated with adalimumab 40mg subcutaneously every two weeks. Her personal history was relevant for asthma, depression, hepatic steatosis, and dyslipidemia.

She presented in the office with pain and dysesthesias in the medial region of the left ankle with evolution of some months and had experienced little relief with nonsteroidal anti-inflammatory drugs (NSAIDs). She reported no history of trauma. On physical examination of the left ankle, she had tenderness above the medial malleolus and a positive Tinel test over the posterior tibial nerve (PTN). She had no arthritis, cutaneous lesions, or other relevant findings.

A musculoskeletal ultrasound was performed (figure 1, A), revealing a hypoechoic ovoid image with 18x9x10mm inside the tarsal tunnel and in intimal relationship with PTN, suggestive of a nerve sheath tumor. The magnetic resonance imaging (MRI) showed a nodular formation with 8x10mm in the axial plane and 15mm in the longitudinal plane in close relationship with the anterior surface of PTN, hypointense in T1-weighted sequences and hyperintense in T2-weighted and turbo inversion recovery magnitude sequences (figure 1, B and C).

A diagnosis of probable schwannoma of the posterior tibial nerve was made and the patient was proposed for surgical excision of the tumor, and is currently waiting for the procedure.

Discussion: The tarsal tunnel is a fibro-osseous structure that contains the posterior tibial, the flexor digitorum longus or the flexor hallucis longus tendons and the neurovascular bundle. Any distension within this structure can cause compression of the PTN, resulting in pain, dysesthesias, paraesthesias or hyperesthesias along the trajectory of the nerve. Some common causes of tarsal tunnel syndrome (TTS) include tendinopathy or tenosynovitis, osteophytes, and trauma.

A schwannoma is a slow growing benign peripheral nerve tumor that arises from the schwann cells of the nerve sheath and most commonly occurs in the head and neck region. Schwannomas of the lower extremities are rare and account for less than 10% of this nerve sheath tumors.

Given their slow growing and rarity, diagnosis can be delayed, but they must be considered in the differential diagnosis of patients presenting with TTS. Musculoskeletal ultrasound can help in this diagnosis, contributing to an effective identification and treatment decision.

060 - DISPLASIA TCHECA MIMETIZANDO ARTRITE REUMATOIDE

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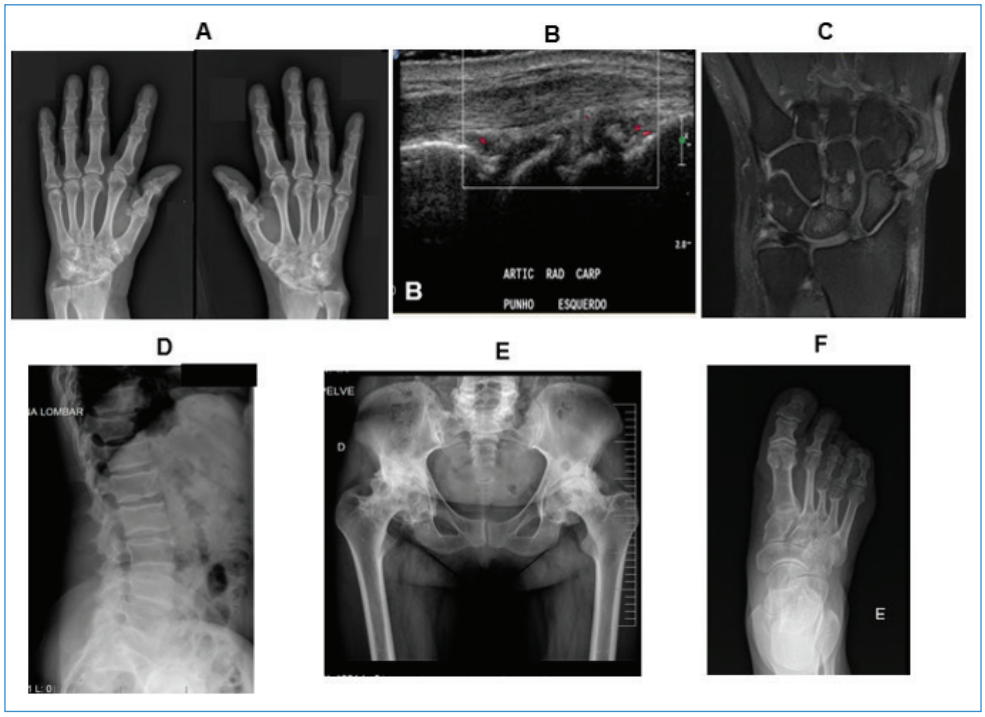

Figure. Exames de imagens dos pacientes portadores de Displasia Tcheca

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Introdução: A Displasia Tcheca (OMIM #609162) é uma rara colagenopatia tipo 2, autossômica dominante, causada por uma única mutação missense (R275C, c.823C>T) no gene COL2A1. Caracteriza-se por estatura normal, osteoartrite precoce, platispondilia, perda auditiva e metatarsos curtos. Doze pacientes, membros de uma família de brasileiros afetados pela Displasia Tcheca que apresentam características semelhantes a artrite reumatoide, foram descritos e publicados na revista Modern Rheumatology em 2023 (Czech dysplasia mimicking rheumatoid arthritis: Case series and literature review).
Objetivo e métodos: Demonstrar algumas imagens dos pacientes da família de brasileiros portadores da Displasia Tcheca.
Resultados: A) Radiografia da mão de uma paciente de 54 anos evidenciando osteoartrite do punho, de região intracárpica, de 1ª metacarpofalangeana e 1ª interfalangena. É possível visualizar as epífises das metacarpofalangeanas alargadas; B, C, D e E) Radiografias de uma paciente de 58 anos; B) Ultrassonografia de punho de paciente com sinovite; C) Ressonância do punho do paciente com derrame articular e edema ósseo; D) Radiografias da coluna lombar do paciente com platispondilia; E) Radiografia da pelve com osteoartrite do quadril bilateral; F) Radiografia dos pés evidenciando 3º e 4º metatarsos curtos de uma paciente 37 anos.
Conclusões: É importante considerar a displasia Tcheca como um potencial diagnóstico diferencial para artrite reumatoide. Esta displasia esquelética autossômica dominante está associada à altura normal, metatarsos curtos, platispondilia, perda auditiva, epífises aumentadas e osteoartrite precoce. Achados inflamatórios como artrite e sinovite também podem estar presentes em indivíduos com Displasia Tcheca.

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098 - (MÁS) LÍNGUAS REUMÁTICAS

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Apresentam-se, nesta imagem, dois doentes com alterações na cavidade oral secundárias a terapêuticas reumáticas (ambos sob hidroxicloroquina (HCQ). Estes casos ilustram a necessidade da realização de um

Figura. Alterações na cavidade oral secundárias à terapêutica com hidroxicloroquina (caso 1 à esquerda e caso 2 à direita)

exame objetivo geral em todas as consultas de Reumatologia, bem como a importância de informar os pacientes que vão iniciar terapêuticas reumáticas sobre os possíveis efeitos colaterais inerentes. A HCQ pode, raramente, causar hiperpigmentação da mucosa oral, sendo as regiões anatómicas mais afetadas o palato (mais frequente), a mucosa jugal e labial, e a língua.

Caso 1 - Doente de 66 anos, do sexo feminino, seguida em consulta de Reumatologia desde há 4 anos, por síndrome de Sjögren. Após 2 anos de terapêutica com HCQ 400mg/diária, referiu o aparecimento de estrias violáceas na língua (ver figura 1, à esquerda). A terapêutica com plaquinol foi suspensa, e a doente foi referenciada à Otorrinolaringologia. Com a suspensão da terapêutica, a coloração da língua voltou a normalizar após 6 meses de interrupção do fármaco.

Caso 2 - Doente de 34 anos, do sexo feminino, seguida em consulta de Reumatologia desde os 19 anos por lúpus eritematoso sistémico. Ao exame objetivo geral, apresentava estrias violáceas na língua (ver figura 1 , à direita). A doente referiu já ter esta alteração há cerca de 12 anos, coincidindo com o início da terapêutica com HCQ 400mg/diária. Neste último caso, a terapêutica com HCQ manteve-se.

101 - CRAB-LIKE HANDS AS AN UNUSUAL PRESENTATION OF A RHEUMATIC DISEASE

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A 68-year-old male presented with intense pain in the

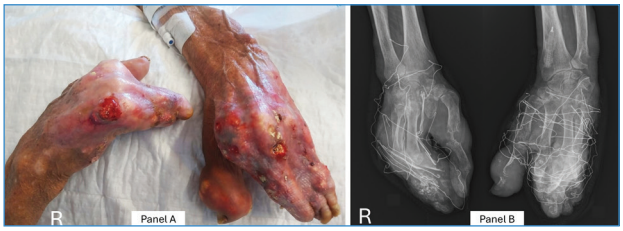

Figure 1. Panel A: A giant tophus is observed in the thumb of the left hand. Panel B: X-ray of the hands

upper and lower limbs for several weeks. He reported no limb weakness or history of trauma. His past medical history included tophaceous gout (non-compliant to allopurinol), chronic renal insufficiency, heart failure and a poor social condition. On physical examination, the normal anatomy of the hand was lost due to joint destruction and fusion of the interdigital spaces and had active drainage of gout tophi. Also, a giant tophus can be observed in the first finger of the left hand (figure 1- panel A). The patient displayed limited range of motion in the wrists, fingers, and elbows, with hindered mobilization of knees, ankles, and feet due to pain. X-rays of the hands, elbows, and feet revealed multiple “punched-out” erosions (figure 1 – panel B). The patient was initiated on antibiotics, corticosteroids, and urate-lowering therapy. This case illustrates the severe consequences of chronic untreated gout, which led to extensive joint destruction and tophi formation. This underscores the critical need for timely medical intervention and comprehensive care in managing aggressive gout.

130 - CALCIUM PYROPHOSPHATE DEPOSITION IN THE CERVICAL SPINE: AN UNUSUAL IMAGE AT AN UNUSUAL AGE

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Axial cervical spine CT image of a 44-year-old female with previously known knee chondrocalcinosis showing curvilinear calcifications of the transverse ligament of the atlas (Fig. A, arrow).

The patient presented chronic neck pain without morning stiffness or fever. Inflammatory markers were negative, not fulfilling criteria for crowned dens syndrome. Iron and phosphocalcium metabolism studies were unremarkable. Her past medical history also included obesity, diabetes mellitus, arterial hypertension and hyperuricemia. There was no relevant family history.

Calcium pyrophosphate deposition disease (CPPD) is a crystal deposition arthropathy involving the ar-

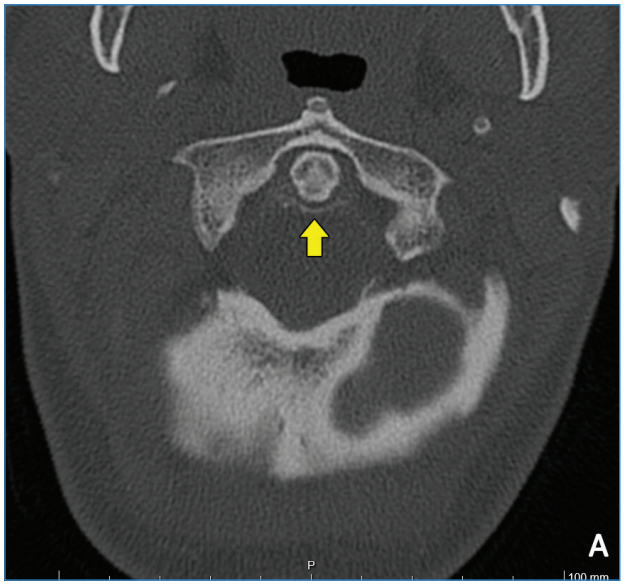

Figure 1. Axial cervical spine CT image of a 44-year-old female

ticular and periarticular tissues (1). The prevalence of radiographic chondrocalcinosis ranges from 4% to over 10% among older adults, though the prevalence of symptomatic CPPD remains unclear (2). The knees, wrists and hips are the most commonly affected joints, while spinal CPPD is uncommon. It is typically a disease of the elderly, being rare in patients younger than 60 years of age. When present in young individuals, it should motivate investigation of familial association or metabolic disease (3), which were excluded in our patient.

We highlight this image due to the rarity of this finding, particularly at a young age.

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138 - NAIL-PATELLA SYNDROME

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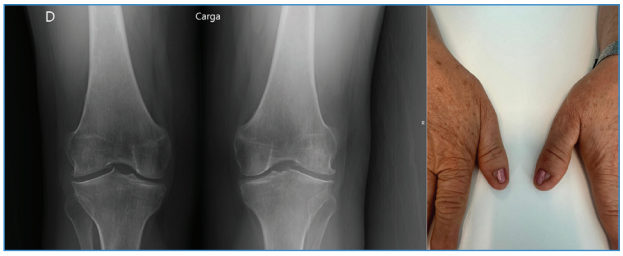

Figure 1. Left: Radiography of the knees; Right: Picture of the thumb fingernails

Patellar hypoplasia or absence and nail dystrophy are pathognomonic signs of Nail-patella syndrome (NPS), a rare autosomal dominant condition caused by a heterozygous loss-of-function mutations in the LMX1B gene. This condition, first described in 1897 by Little, affects both ectodermal and mesodermal structures: elbow dysplasia and iliac horns are the most prevalent manifestation, followed by renal impairment – raging from mild proteinuria to end-stage renal failure – and lastly, ocular hypertension and/or glaucoma.

A 59-year-old woman presented to a Rheumatology consultation with bilateral mechanical gonalgia. On examination, preserved range-of-motion in both knees was noted, but the physician was not able to identify the patella on palpation. Additionally, the patient presented bilateral 5th finger camptodactyly, nail hypoplasia of both thumbs and limited extension and pronosupination of both elbows. Radiographs of the knees (figure 1) depict severe bilateral hypoplasia of the patella, and mild marginal osteophytes in the lateral compartments. Moreover, radiographs of the elbows showed bilateral radio-head hypoplasia and subluxation. All blood tests including acute phase reactants, urea and creatinine, and urinary sediment analysis were normal. The distinct clinical features allowed a prompt diagnosis, which was confirmed via DNA analysis – heterozygous pathogenic mutation c.178>T (pQ60X) in the exon 2 of the LMX1B gene - in the interest of an accurate genetic counselling.

141 - SÍNDROME DOLOROSO REGIONAL COMPLEXO TIPO I: ACHADOS RADIOGRÁFICOS

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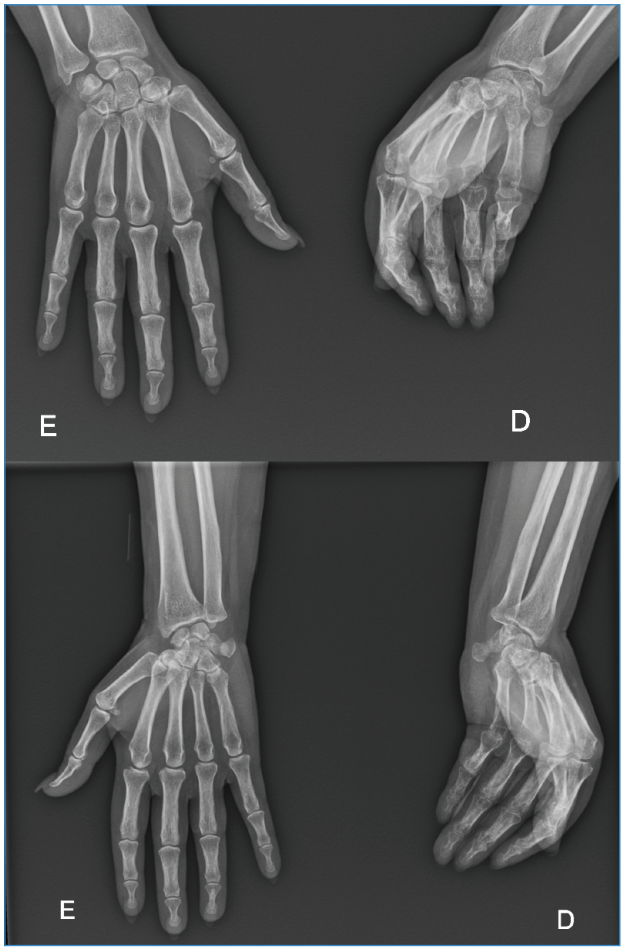

Figura. Radiografia das mãos com achados sugestivos do SDRC da mão direita.

Doente do sexo feminino, de 51 anos, com quadro de dor e tumefacção da mão direita, acompanhado de hipersudorese, alterações da coloração da pele e parestesias/disestesias, com início 3 meses após cirurgia de libertação do tendão flexor do 5º dedo à direita.

Ao exame objetivo, apresentava tumefação da mão e palidez cutânea à inspeção, com diminuição da temperatura e alodínia/hiperalgesia ao toque. Destacava-se incapacidade de realizar mobilização ativa dos dedos, com dor intensa e limitação franca à extensão e flexão passivas, principalmente do 3º ao 5º dedo. O estudo radiográfico (figura) evidenciou osteopenia em padrão mosqueado sobretudo das falanges da mão direita. Face à apresentação clínica típica, e achados radiográficos, foi possível assumir o diagnóstico de Síndrome Doloroso Regional Complexo tipo I.

143 - SÍNDROME DE FAHR SINTOMÁTICO EM DOENTE COM LÚPUS ERITEMATOSO SISTÉMICO (LES): UM DESAFIO DIAGNÓSTICO

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Doente do sexo feminino, de 59 anos, com quadro de instabilidade da marcha, com quedas, tremores simétricos das mãos, cefaleias, tonturas sem vertigens e disfonia progressiva. Antecedentes de Overlap Lúpus eritematoso sistémico / Doença de Sjögren, sob terapêutica com hidroxicloroquina (200mg id), azatioprina (100mg id) e Rituximab endovenoso semestral (2 tomas de 500mg, em 2 semanas).

Ao exame objetivo, apresentava apraxia imitativa ligeira em tarefas complexas, tremor postural simétrico, dificuldade na sequenciação, adiadocinesia bilateral, "starring eyes" e hipofonia com esforço produtivo e dificuldade na persistência vocal. Destacava-se ainda hiperreflexia miotática generalizada, com grande extensão de área reflexogénica, reflexo cutâneo plantar em extensão bilateralmente e lentificação da marcha sem desvio do eixo.

Sem quaisquer sinais clínicos ou analíticos sugestivos de atividade da doença reumatológica.

O estudo imagiológico por tomografia computorizada crânio-encefálica (A) evidenciou calcificações bilaterais dos gânglios da base, núcleo dentado e ponte, complementada com ressonância magnética do neuro-eixo (B) que descreveu os mesmos achados, e uma hiperintensidade FLAIR ligeira da substância branca subcortical e periventricular, sem alterações da restrição de difusão e sem anormalidades medulares.

Face à apresentação clínica e achados do estudo complementar, foi possível assumir o diagnóstico de Síndrome de Fahr sintomático, um distúrbio neurológico raro caracterizado pela presença de calcificações bilaterais dos gânglios da base e do córtex cerebral, que se associa a uma variedade de anormalidades endócrinas,

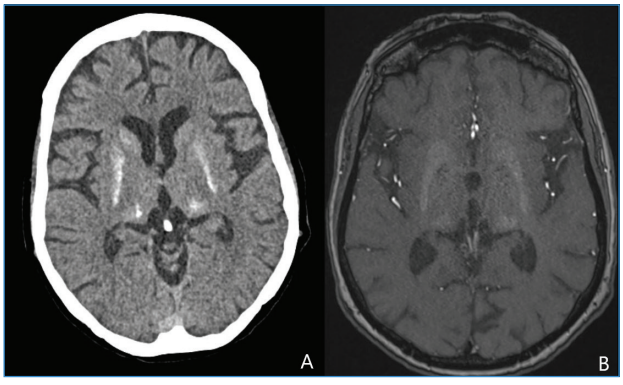

Figura. TC CE (A) e RM CE (B) com calcificações bilaterais nos gânglios da base.

condições genéticas, infeções e toxinas, mas estando também descrita na bibliografia em associação ao LES.

147 - MOREL-LAVALLÉE LESION: A RARE CAUSE OF LEG PAIN

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Case Presentation: A 68-year-old female patient, diagnosed with spondyloarthritis with peripheral involvement (treated with methotrexate and acemetacin), presented to the Rheumatology appointment with a 3-weeks history of pain in the posterior and inner region of the right leg associated with a sensation of swelling. She denied recent trauma. On physical examination, she had asymmetric edema of the right lower limb, with slight pain upon palpation of the posterior-inner region. There were no evident signs of redness, warmth, fluctuations, or palpable masses, nor other skin changes, and no peripheral arthritis was observed, particularly in the right knee.

The patient had previously been observed in the emergency department where she underwent a CT angiogram of the right lower limb to exclude vascular injury, which revealed a hypodense collection (65x29 mm in diameter) on the inner side of the right leg, adjacent superiorly to a Baker's cyst, in relation to the medial gastrocnemius muscle, raising the hypothesis of muscle rupture. She was seen by Orthopedics, discharged with analgesia and advised to rest.

Due to the persistence of symptoms, a magnetic resonance imaging (MRI) was requested and showed an elongated (20cm x 3cm) and fusiform collection on the medial and posterior side of the right leg, between the medial gastrocnemius and the superficial fascia, with hyperintense signals on T2-weighted images (not suppressed by fat suppression), suggesting hemorrhagic content, compatible with a Morel-Lavallée lesion (ML), likely originating from a post-traumatic fascial rupture/detachment as there were no evident signs of muscle rupture. Additionally, the collection extended, without continuity, to the posterior aspect of a Baker's cyst (35mm x 24mm x 13mm) without indirect signs of rupture (which would cause primary exudation in the surrounding tissues of the popliteal region involving the cyst). Moderate joint effusion in the ipsilateral knee was also evident (fig. 1).

Given the diagnosis, the patient was evaluated by the Plastic Surgery team, which excluded the need for drainage or surgical treatment. The patient continued under conservative treatment, including analgesia, cryotherapy, and elastic compression, and has not experienced any complications to date.

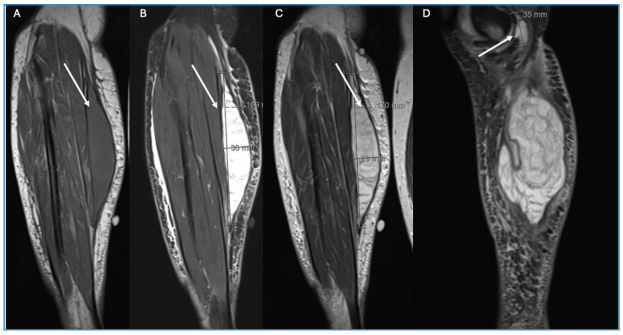

Figure. Coronal T1-weighted (A), T2- weighted (B and C) of right leg revealing a large well defined elongated lesion (white arrow) on the medial and posterior side of the right leg, between the medial gastrocnemius and the superficial fascia, with hyperintense signals not suppressed by fat suppression, compatible with a Morel-Lavallee lesion. It is additionally shown a Baker's cyst (D, white arrow) without indirect signs of rupture.

Discussion: The Morel-Lavallée lesion is a rare condition and results from the abrupt separation of the skin and subcutaneous tissue from the underlying fascia, leading to the detachment of soft tissues. MRI is the modality of choice for diagnosis and assessment of ML lesions, which can present different imaging patterns (hyperintensity in T2 and hypointensity in T1 or hyperintensity in both sequences). The ML lesion is most often found at the level of the greater trochanter or the anterolateral aspect of the hip (forming part of the differential diagnoses of greater trochanteric pain syndrome), but it can rarely affect the knee or lower leg. Although it mostly occurs associated with blunt trauma, there are few cases described where no prior or recent trauma was identified, similar to our patient. Identification of the lesion is essential to prevent complications such as compression of surrounding structures which may lead to soft tissue infection or skin necrosis. Conservative treatment with analgesia and compression bandaging is indicated in mild cases (ML lesion without a capsule), but percutaneous drainage or surgery may be necessary for complete evacuation of the lesion.

151 - WHEN FEMURS LOSE THEIR HEADS

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Introduction: Avascular necrosis (AVN) of the femoral head occurs due to the interruption of the vascular supply to the proximal femur, leading to bone ischemia. It can result from traumatic or non-traumatic

systemic conditions and, if left untreated, ultimately leads to femoral head collapse and secondary hip osteoarthritis. Glucocorticoids (GC) and chronic alcohol abuse are the most frequently reported non-traumatic causes. Smoking is considered to be correlated with AVN. A non-operative management is recommended for patients in early pre-collapse stages, while advanced stages require surgical treatment.

Case report: A 55-year-old man, with undifferentiated spondylarthritis for 10 years, taking prednisolone 5mg/day, calcium 1500mg/day, vitamin D 880U/day and acemetacin 90mg on demand, had been complaining of hip pain for 3 months. The pain radiated to the groin and was briefly relieved with analgesics and non-steroidal anti-inflammatory drugs. The patient also had a history of alcohol abuse and a smoking history of 40 pack-years. The pelvic X-ray showed severe irregularities in the femoral heads with sclerosis and collapse of the articular surface, corresponding to avascular necrosis at stage IV of the ARCO staging system (figure 1). The dose of GC was reduced and the patient underwent total hip arthroplasty.

Discussion: AVN resulting from GC therapy manifests in up to 38% of patients and exhibits a dose-dependent relationship. There are few case reports related to low dose of GC (≤5mg prednisone equivalente per day). In this case, the long-term therapy, the history of alcohol abuse and smoking may have contributed in a synergistic way to the occurrence of AVN.

Conclusion: In a patient receiving GC therapy, even at a low dose, the emergence of hip pain should raise the suspicion of AVN. If feasible, GC administration should be halted; alternatively, the dosage should be reduced. Evaluation by an orthopedist is essential to assess the need for surgical intervention.

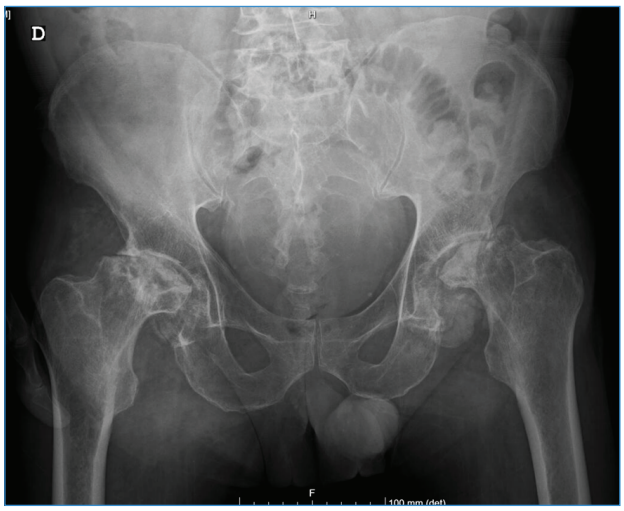

Figure 1.

166 - DOENÇA DE MADELUNG, UMA ENTIDADE RARA

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Introdução: A Doença de Madelung caracteriza-se pela presença de múltiplas massas lipomatosas simétricas, constituídas por tecido adiposo subcutâneo não capsulado, geralmente a envolver a região do pescoço, ombros, braços e tronco. É mais prevalente no sexo masculino (15:1) e em aproximadamente 90% dos casos associa-se a hábitos alcoólicos. Embora descrita como benigna, pode gerar síndromes compressivos a condicionar disfagia e dispneia se houver envolvimento laríngeo ou mediastínico, existindo casos descritos de evolução para lipossarcoma. O tratamento habitual passa por lipoaspiração, com aumento considerável da funcionalidade e qualidade de vida.

Descrição: Doente ex-fumador (38 UMA), com hábitos alcoólicos mantidos, consumindo cerca de 0,75 a 1 L de vinho tinto/dia e consumo esporádico de cerveja.

Medicado habitualmente com pantoprazol, ácido fólico e vitamina D, azilsartan + clorotalidona e analgesia em sos.

Do exame objectivo geral em consulta, doente com volumosas massas de consistência lipomatosa, não aderentes aos planos profundos, não dolorosas nem pulsáteis, sem alterações cutâneas associadas, com localização bilateralmente simétrica ao nível do dorso e cintura escapular, clinicamente compatíveis com Doença de Madelung, como se pode observar na imagem anexa. Associadamente, o doente apresentava estudo ecográfico de partes moles compatível com

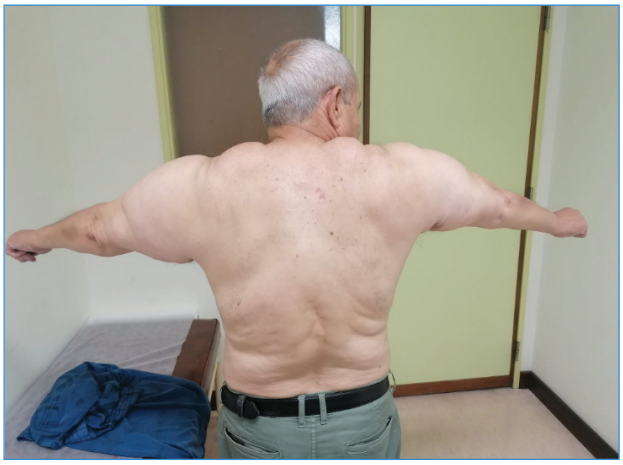

Figura 1. Lipomatose simétrica difusa

infiltração lipomatosa difusa e estudo analítico com anemia macrocítica com défice de ácido fólico e aumento significativo de GGT (497 U/L) e FA (156 U/L), interpretados no contexto do seu etilismo crónico. O doente não apresentava qualquer limitação funcional secundária, constituindo as alterações lipomatosas apenas um problema estético a causar diminuição da sua qualidade de vida, tendo sido por isso referenciado a cirurgia estética para lipoaspiração das lesões.

168 - SKIN MANIFESTATIONS IN SYSTEMIC LUPUS ERYTHEMATOSUS: AN IMPRESSIVE AND SAFE RESPONSE TO ANIFROLUMAB

Bárbara Fernandes Esteves[1], Miguel Correia Natal[1], Maria Seabra Rato[1], João Carlos Almeida[2], Miguel Bernardes[1], Lúcia Costa[1], Raquel Miriam Ferreira[1]
[1]Rheumatology Department, Unidade Local de Saúde de São João, Porto, Portugal, [2]Pathology Department, Unidade Local de Saúde de São João, Porto, Portugal

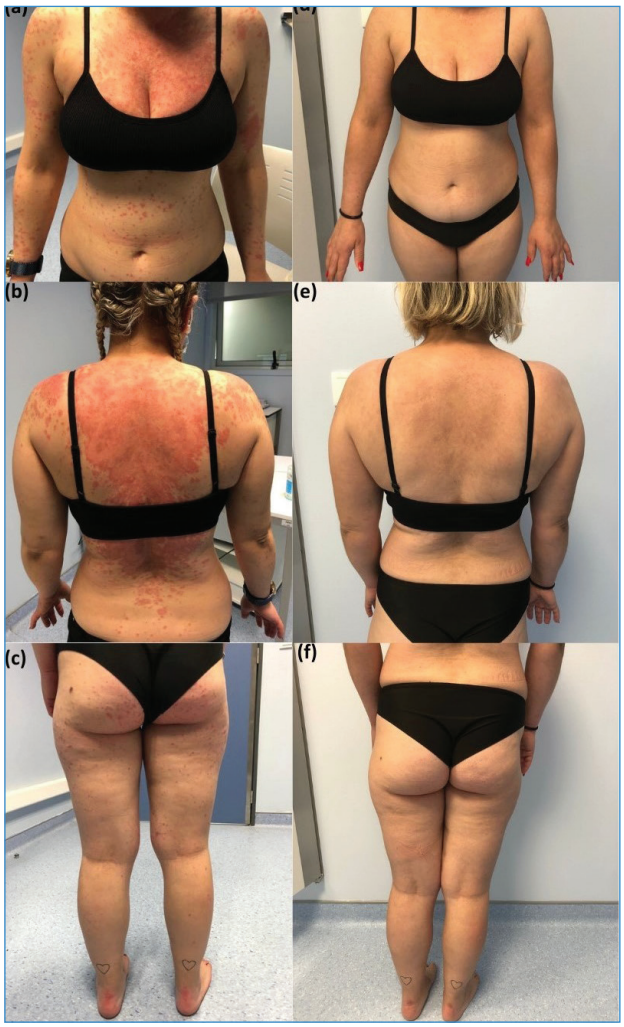

Figure 1. (a, b, c) Subacute cutaneous lupus before treatment and (d, e, f) after two infusions of anifrolumab with total resolution

We present the case of a 37-year-old woman with a one year history of extensive erythematous annular lesions on the upper thorax, back, extensor surfaces of arms and forearms, gluteal region and posterior upper thigh (fig 1 a, b, c) and non-scaring alopecia. A diagnosis of SLE was established and hydroxychloroquine initiated. Azathioprine was tried, however it had to be discontinued due to severe lymphopenia. Lymphopenia was once again observed with mycophenolate mofetil. Given the patient's significant disease activity despite multimodal therapy, anifrolumab 300mg infusion was started every 4 weeks. Six weeks later, the patient showed a dramatic improvement of the skin lesions (Fig 1 d, e, f). The dose of prednisolone was progressive tapered to 5mg/day over three months. No adverse effects have been noted during biologic therapy. This case highlights the value of anifrolumab in the treatment of cutaneous lupus after the failure of multiple therapies, due to ineffectiveness or adverse effect.

170 - DOUBLE VERTEBRAE KUMMELL'S DISEASE

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Sagittal and frontal view of computed tomography (CT) of the thoracolumbar spine in a 56-year-old woman with vertebral pseudarthrosis (VP). The patient had rheumatoid arthritis, treated with methotrexate 17.5mg/week and prednisolone 5mg/day, and osteoporosis medicated with alendronate. However, the therapeutic adherence to bisphosphonate was poor and she refused other treatment options. Due to recurrent falls and new back pain, a thoracolumbar radiography was performed, uncovering multiple vertebral fractures. A subsequent CT scan displayed dorsal hyperkyphosis, with vertebral body flattening from D5 to

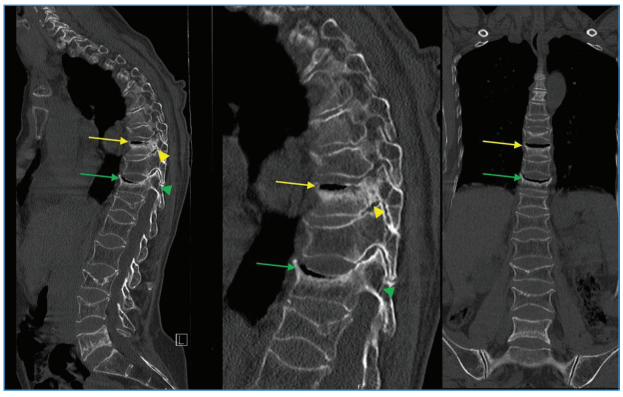

Figure 1. Double Vertebrae Kummell Disease

L5, and surprisingly, VP was evident in a double-level: D9 and D11. At D9, an Intervertebral Cleft (IVC) was visible (arrow) with ossification of the pedicle (yellow arrowhead). At D11, an IVC (arrow) was found with a stress fracture of the spinous process (green arrowhead). Both present marginal sclerosis of the IVC. These are some of the most characteristic findings in VP, others include double-line sign and paravertebral callus formation. VP, historically referred as Kummell's Disease, is rare and considered a complication of osteoporotic vertebral compression fractures. It usually affects only one vertebrae, being the involvement of a double-level vertebrae even more uncommon, as shown in this case.

172 - SYNCHRONOUS BROWN TUMORS: AN EXTREME FORM OF HIGH-TURNOVER BONE DISEASE

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A 46 year-old female patient with a stage 5 chronic kidney disease, in peritoneal dialysis, developed in march 2022 a non painful swelling of the right jaw. The jaw X-ray (Figure 1a) showed an expansive lytic bone lesion (blue arrow) on the posterior aspect of the right mandibular body. Dental computed tomography scan (CT) (Figures 1b and c) revealed a multilocular expansive lytic lesion (blue arrow) at the mandibular angle, with several areas of cortical thinning and internal septa. Shortly afterwards, she also presented a palpable tender mass in the right rib cage. A chest X-ray (Figure 1d) revealed an expansive lytic lesion (green arrow) on the posterior aspect of the 6th right rib, that was confirmed by thoracic CT (Figure 1e), raising the possibility of osteitis fibrosa cystica. A biopsy of the mandibular lesion showed a high cellular density, fibroblastic proliferation with abundant multinucleated giant cells of the osteoclast type, and slight focal hemorrhage, consistent with a brown tumor. Laboratory test confirmed a tertiary hyperparathyroidism (HPT). Through the early diagnosis and treatment of HPT, Brown Tumors of Hyperparathyroidism (BTHs) have become progressively less frequent. When present, they are associated with more severe and prolonged forms of HPT. BTHs can pose diagnostic challenges, often being mistaken for bone metastases. With this case, the authors aim to demonstrate that although increasingly rare, BTHs can still occur and may present synchronously in various distinct locations.

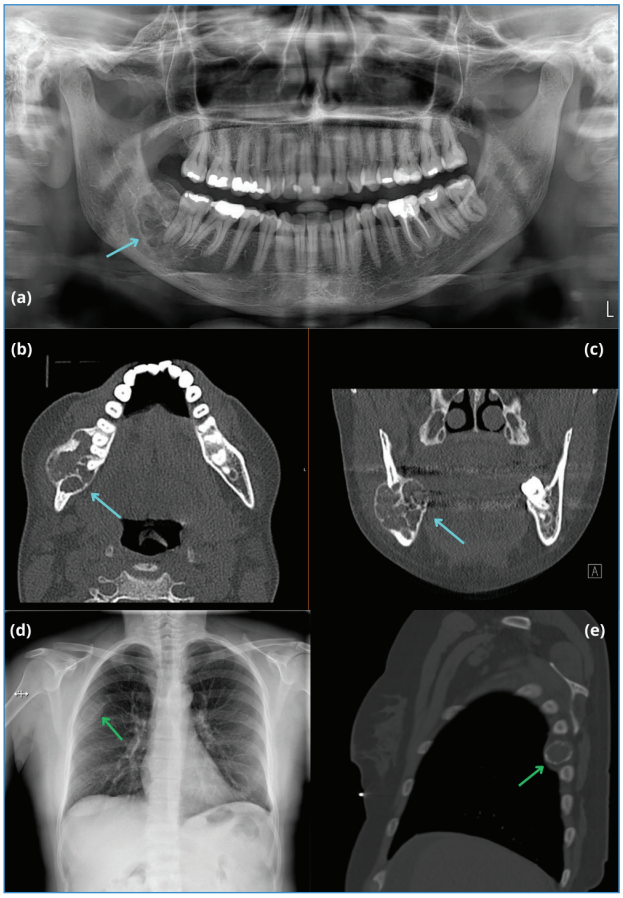

Figure 1. Synchronous Brown Tumors

188 - FRATURAS DE FRAGILIDADE EM DOENTE COM SÍNDROME DE RETT

Maria de Sá Pacheco[1], Ana Filipa Rocha Águeda[1, 2], Miguel Guerra[1, 2], Rita Pinheiro Torres [1], Joana Ramos Rodrigues[1], Margarida Oliveira[1, 2]
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Introdução: O Síndrome de Rett é um distúrbio do neurodesenvolvimento raro e multissistémico, mais comum em indivíduos do sexo feminino, associado a mutações do gene MECP2. Caracteriza-se por um período de desenvolvimento normal durante aproximadamente 6 meses, com posterior desaceleração e regressão das capacidades adquiridas, perda do uso voluntário das mãos, com comportamento compulsivo e repetitivo de torcer ou lavar as mãos, desaceleração do crescimento da cabeça e encéfalo, convulsões, dificuldade de deambulação e défice intelectual. É uma patologia sem cura, com evolução dividida em 4 estadios até à deterioração

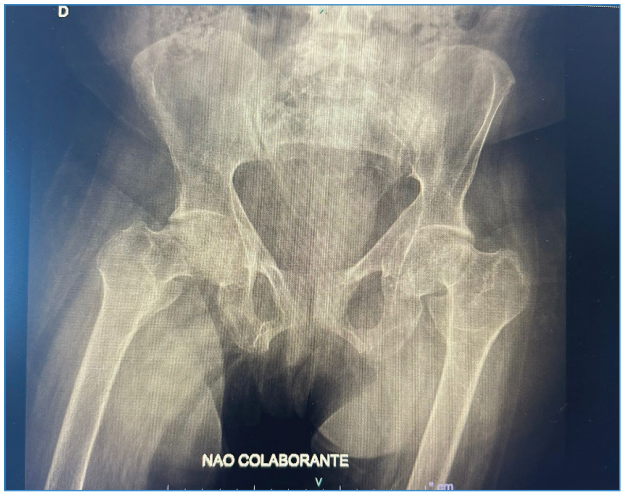

Figura 1. Fratura bilateral do colo do fémur e ramos isquiopúbicos

motora quase completa. O seu tratamento envolve uma abordagem multidisciplinar focalizada primariamente no tratamento sintomático e melhoria de qualidade de vida.

Descrição: Doente de 39 anos, sexo feminino, com Síndrome de Rett e epilepsia. Sintomas iniciais com 1 ano de idade, com declínio progressivo, marcha comprometida desde os 4 anos de idade e perda total de capacidade de marcha aos 35 anos.

Doente com múltiplos fatores de risco para osteoporose, entre os quais a própria patologia, mas também o comprometimento do atingimento de um bom pico de massa óssea durante o crescimento e uso de antiepiléticos desde a infância.

Para estudo de queixas álgicas da bacia que surgiram à mobilização durante a higiene, realizou radiografia da bacia, que documentou fratura do colo do fémur e ramos isquiopúbicos bilateralmente. Sem qualquer noção de traumatismo prévio.

189 - DESTRUIÇÃO DE SEIOS PERINASAIS EM DOENTE JOVEM COM GRANULOMATOSE COM POLIANGEÍTE

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Introdução: A Granulomatose com Poliangeíte é uma vasculite sistémica de pequenos vasos, com atingimento preferencial do trato respiratório superior/inferior e renal, com formação de granulomas e necrose.

Descrição: Doente de 26 anos, sexo feminino, com antecedente pessoal de Granulomatose com Poliangeíte PR3+ com quadro de hemorragia alveolar e lesões pulmonares cavitadas na apresentação inicial. Para além do envolvimento pulmonar, a doente apresentava também atingimento da via aérea superior, com alterações imagiológicas marcadas em TAC dos seios perinasais. Na figura 1 evidencia-se erosão das paredes mediais dos seios maxilares, com destruição de células etmoidais, particularmente à esquerda. Na figura 2, adicionalmente, observa-se erosão da lâmina papirácea da órbita esquerda, possivelmente relacionada com o processo etmoidal inflamatório crónico, evidenciada por ocupação das células etmoidais anteriores esquerdas.

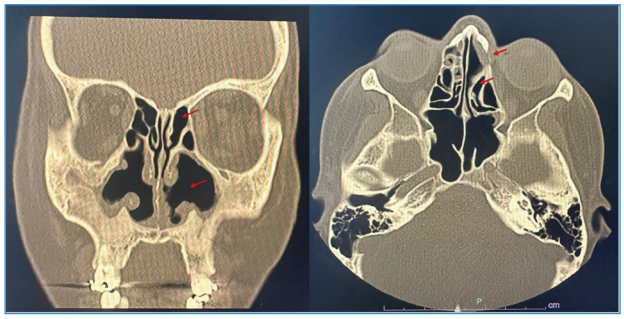

Figura 1 e 2. TAC dos seios perinasais

200 - DERMATOSE NEUTROFÍLICA EM DOENTE COM LÚPUS ERITEMATOSO SISTÉMICO

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As dermatoses neutrofílicas são um grupo de doenças caracterizadas na histologia da biópsia de pele pela presença de um infiltrado rico em neutrófilos na derme e epiderme, leucocitoclasia variável e ausência de vasculite primária. Existem vários tipos de dermatoses neutrofílicas que podem estar associadas ao Lúpus Eritematoso Sistémico (LES). A dermatose neutrofílica do tipo-Sweet é uma condição rara associada ao LES.

Doente do sexo feminino, 52 anos, raça negra, seguida na consulta de reumatologia por LES com mais de 10 anos de evolução, em remissão de longa data, medicada com hidroxicloroquina 400 mg por dia, anti-hipertensor diário e vitamina D mensal. Recorreu ao SU por quadro de febre não diária, com 2 meses de evolução (máximo 39,2º). Ao exame objetivo era evidente a presença de rash malar e a presença de lesões

eritematosas papulares nas palmas das mãos bilateralmente (figura 1). Analiticamente destacava-se anemia normocítica, normocrómica (hemoglobina 10,9 g/dl), leucopenia com linfopenia (2900/ul e 800/ul, respetivamente), elevação da velocidade de sedimentação (82 mm/h), sem aumento significativo da proteína C reativa (9 mg/L), creatinina 1,1 mg/dl e proteinúria de 24h de 340 mg. Realizou estudo exaustivo que excluiu presença de intercorrência infeciosa (nomeadamente endocardite, espondilodiscite e tuberculose) e de neoplasia (realizou biópsia de adenopatia axilar que revelou linfadenite lúpica). Para melhor esclarecimento das lesões cutâneas, foi realizada biópsia da palma da mão direita que revelou tratar-se de uma dermatite de padrão perivascular superficial, com características "do tipo" Síndrome de Sweet histiocitóide. Além disso, foi ainda realizada biópsia renal que revelou nefrite lúpica classe V. Assim, assumiu-se que o quadro seria enquadrado no contexto de um flare lúpico, com envolvimento hematológico, cutâneo, linfadenopático e renal. Foi medicada com pulsos de metilprednisolona endovenosa 500 mg durante 3 dias, seguidos de prednisolona oral 0.5mg/Kg/dia, com melhoria progressiva do estado geral e das lesões cutâneas.

As lesões cutâneas com infiltrado neutrofílico mais comum no LES são o LE bolhoso e a vasculite leucocitoclásica. No entanto, recentemente têm sido descritas lesões neutrofílicas da pele mais variadas, onde se inclui a dermatose neutrofílica do tipo Sweet. Nos casos descritos até à data, esta dermatose caracteriza-se pela presença de placas ou nódulos eritemato-edematosos em áreas foto-expostas, podendo ser pruriginosas ou não. O curso é habitualmente subagudo ou crónico e pode haver recorrência das lesões, ao contrário de outras dermatoses. O aparecimento destas lesões está intimamente associado à presença de flare do LES e pode acorrer como manifestação inicial, sendo que o tratamento passa pelo uso de imunossupressores.

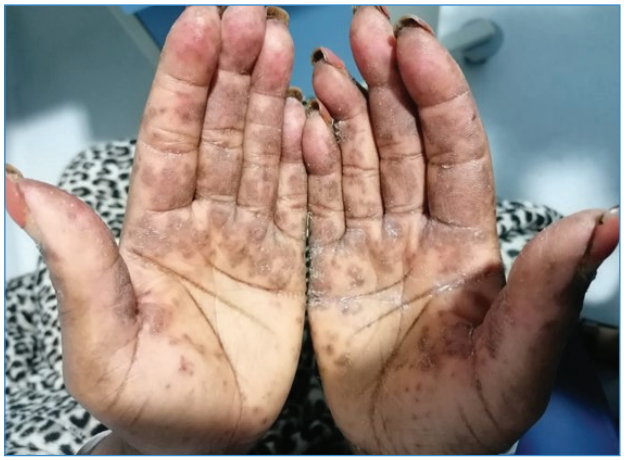

Figura 1. Lesões palmares, bilaterais, eritemato-papulares em doente com flare lúpico.

204 - NUVENS NA RADIOGRAFIA - ENFARTE ÓSSEO COMO DIAGNÓSTICO DIFERENCIAL

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Introdução: A osteonecrose consiste num processo de isquemia óssea resultante de suprimento vascular insuficiente.1 Quando este processo ocorre na metáfise e/ou diáfise de um osso longo designa-se por "enfarte ósseo". Os principais fatores de risco são os glicocorticóides, o consumo excessivo de álcool, a dislipidemia e infeção por VIH.

Caso clínico: Homem de 82 anos, seguido em consulta de Reumatologia por artropatia gotosa, com antecedentes de adenocarcinoma da próstata, fibrilhação auricular, doença pulmonar obstrutiva crónica e hábitos etílicos marcados. O doente referia gonalgia mecânica bilateral de longa data, sem noção de tumefação. Ao exame físico apresentava amplitudes articulares preservadas e sem evidência de tumefação articular. Realizou radiografias simples dos joelhos onde era evidente diminuição da interlinha articular, esclerose subcondral, envolvendo sobretudo o compartimento medial, em relação com gonartrose ligeira a moderada, mas também lesões radiopacas serpiginosas centrais na metáfise e parte da diáfise, compatíveis com enfartes ósseos bilaterais.

Discussão e conclusão: Na maioria dos casos, os enfartes ósseos são assintomáticos, sendo maioritariamente diagnosticados de forma incidental em estudos de imagem realizados por outro motivo. Podem, no entanto, ser causa de dor óssea, sobretudo nas formas agudas que ocorrem em síndromes de hiperviscosidade, como é o caso das hemoglobinopatias ou doenças de depósito. Os locais mais frequentemente acometidos são as metáfises ou a região metáfise-diáfise do joelho (fémur distal, tíbia proximal e perónio proximal), sendo o membro superior raramente envolvido. O envolvimento isolado da diáfise é extremamente raro. Um sinal radiológico clássico é a presença de imagens escleróticas heterogéneas na metáfise ou região metadiafisária, no centro do canal medular, poupando o córtex e circundadas por esclerose em concha ou semelhante a uma nuvem de fumo. Numa fase inicial, as radiografias podem ser normais ou apresentar al-

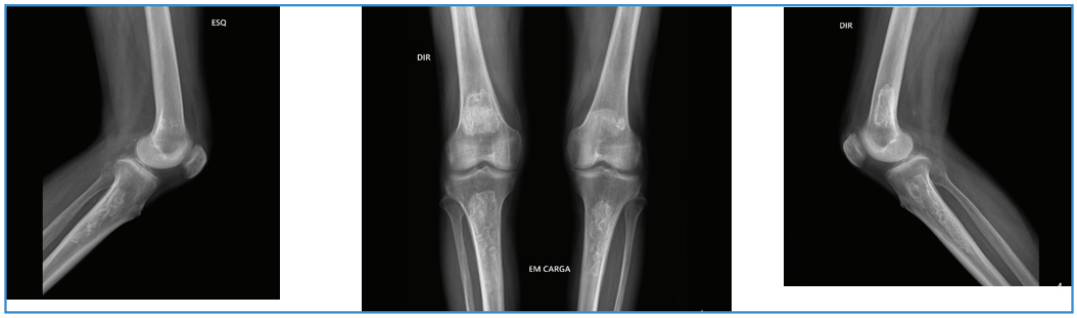

Figura 1. Radiografias dos joelhos com enfartes ósseos na tíbia e fémur bilateralmente

terações inespecíficas, observando-se geralmente uma reação periosteal paralela à diáfise femoral ou tibial. Semelhante à radiografia, a tomografia computadorizada pode ser normal numa fase inicial ou mostrar lesão medular circundada por uma orla esclerótica poupando o córtex. O exame histológico da lesão geralmente não é necessário. Apesar do enfarte ósseo ser uma condição benigna, pode ser necessário o diagnóstico diferencial com lesão tumoral ou infeção, de acordo com o quadro clínico. Raramente podem complicar com transformação maligna, ocorrendo mudança das características imagiológicas prévias.

No caso clínico descrito, o diagnóstico foi estabelecido por radiografia simples, sem necessidade de estudo adicional por apresentar imagem radiológica típica e não apresentar sintomas de alerta. Neste caso, considerou-se que os antecedentes de hábito etílicos marcados representam o principal fator de risco para enfartes ósseos.

209 - PANICULITE LÚPICA – UM CASO DE MANIFESTAÇÃO RECORRENTE E SEVERA

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Apresenta-se o caso de uma doente de 58 anos com diagnóstico de Artrite Reumatoide (FR/Anti-CCP positivo) desde o ano 2000, seguida em consulta de Reumatologia, em remissão durante vários anos sob terapêutica com Metotrexato e Etanercept, sem outros antecedentes de relevo até 2008. Nessa altura, por quadro de lesões cutâneas sugestivas de Lúpus discoide e AVC isquémico em idade jovem, internada para estudo etiológico, do qual resultou diagnóstico de overlap com Lúpus Eritematoso Sistémico (LES), dado atingimento cutâneo, evento trombótico e presença de marcadores serológicos (ANA e DsDNA positivos). Nesse sentido, iniciou também Hidroxicloroquina para controlo da doença subjacente (que manteve até hoje), bem como vários ciclos de Rituximab, por controlo insuficiente da doença, nomeadamente do ponto de vista articular. Encontrava-se em remissão desde 2012, após otimização do seu esquema terapêutico com cDMARD's.

Durante o seguimento habitual, por quadro respiratório incipiente, foi requisitado estudo imagiológico, no qual se detetou densificação em vidro despolido no lobo pulmonar superior direito e várias formações ganglionares (com PET normal e 2 biópsias negativas para transformação maligna), pelo que manteve vigilância imagiológica regular, dada a possibilidade de doença linfoproliferativa (quer pelos achados em exame de imagem, quer pelo risco inerente associado a doenças imunomediadas).

Desde meados de 2020, descreve lesões cutâneas de novo, em placa, endurecidas, dispersas, com agravamento progressivo em extensão e intensidade, com locais com atrofia e depressão tecidual (Figura 1). Apesar da hipótese de paniculite lúpica, era imperativo excluir hipótese de linfoma cutâneo de células T (um dos diagnósticos diferenciais, associado a grande morbimortalidade), motivo pelo qual se optou por realizar biópsia cutânea, que demonstrou "paniculite lúpica em fase tardia", encontrando-se a doente, atualmente, a aguardar reavaliação pela Dermatologia e Reumatologia, para definição de estratégia conjunta.

Note-se que o LES, enquanto doença multisistémica, pode afetar virtualmente qualquer órgão sendo, por isso mesmo, uma doença de grande complexidade e multidisciplinaridade. Entre as manifestações possíveis, as cutâneas acabam por ser das mais comuns e, nalguns casos, um grande desafio diagnóstico.

Exemplo disso, é a paniculite lúpica/lúpus profundo, que apesar de menos comum, representa uma entidade com um diagnóstico diferencial complicado, vasto e cujo tratamento compreende o controlo da doença de base. Nesta extensa lista, inserem-se entidades como o Linfoma subcutâneo de células T, cujo diagnóstico tardio/falhado, pode implicar morbimortalidade signif-

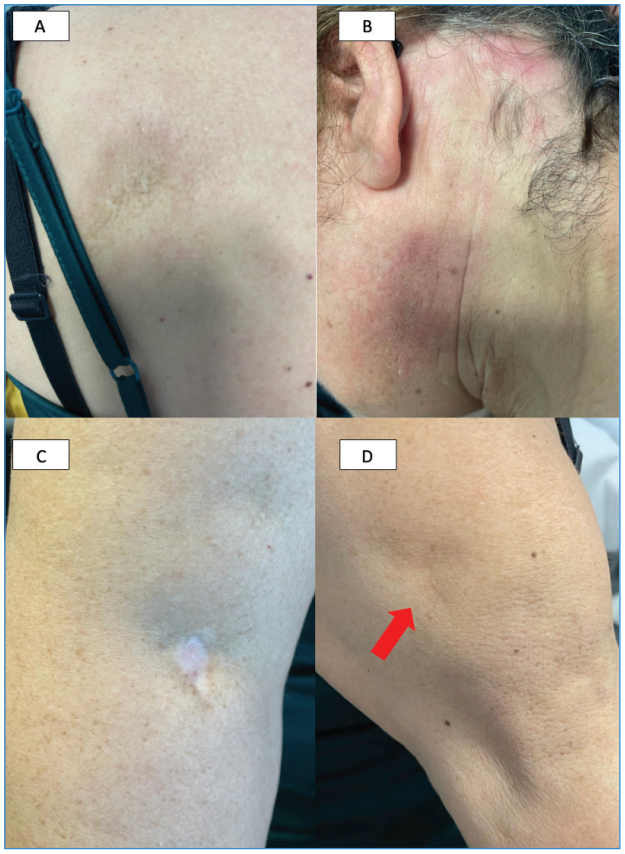

Figura 1. Lesões descritas na região dorsal (A), cervical (B) e nos membros superiores (C e D), já com depressão central.

icativa para o doente.

Nesse sentido, este caso reveste-se de especial importância, pela elevada extensão das lesões, mas também pelo enquadramento clínico da doente, que poderia apontar sobretudo para um quadro neoplásico/linfoproliferativo – atendendo à doença de base com vários anos de evolução e presença de adenopatias.

Do ponto de vista terapêutico, representa também um desafio, uma vez que não existe uma orientação preconizada, se trata de uma manifestação rara e, geralmente, as opções terapêuticas não são particularmente eficazes, o que se torna ainda mais relevante numa doente que já esgotou várias opções no campo dos cDMARD e bDMARD (como o Rituximab).

Por fim, ilustra perfeitamente a complexidade e multidisciplinaridade de algumas doenças Reumáticas, como as doenças do tecido conjuntivo, no qual o diagnóstico e abordagem terapêutica não são, muito frequentemente, diretos.

224 - SÍNDROME DE SOBREPOSIÇÃO POLICONDRITE RECIDIVANTE COM ARTRITE PSORIÁTICA

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Descrição do caso: Doente do sexo feminino, 44 anos, referenciada à consulta de Reumatologia por quadro de artrite das pequenas articulações das mãos e esternocostais, pericondrite auricular (figura 1A) e nasal e episódio de esclerite, com resposta parcial ao metotrexato. Do estudo laboratorial, sem alterações no hemograma e na bioquímica, sem elevação da velocidade de sedimentação ou da proteína-C reativa, sem elevação da gamaglobulina e com serologias para vírus e sífilis negativas. O estudo imunológico foi negativo, incluindo para ANA, FR e anti-CCP. Em consulta de seguimento, descreve segundo episódio de esclerite e apresenta disfonia de novo, tendo sido aumentada dose de metotrexato para 25mg por semana e proposta para adalimumab 40mg a cada 2 semanas, com melhoria significativa da sintomatologia após o seu início. Colocada como hipótese mais provável um síndrome de sobreposição policondrite recidivante com artrite seronegativa. Evolução com aparecimento de lesões cutâneas eritematosas não descamativas nos cotovelos e couro cabeludo (figura 1B) sugestivas de psoríase e episódios de artralgias sem artrite evidente.

Discussão: A policondrite recidivante é uma doença imuno-mediada associada a inflamação sistémica com envolvimento predominante de estruturas cartilagíneas. O atingimento auricular é considerado o mais característico, com inflamação unilateral ou bilateral da orelha externa, que caracteristicamente poupa o lóbulo da orelha. Apesar de até um terço dos doentes com policondrite recidivante poder apresentar outra patologia de forma concomitante, como vasculite, doença do tecido conjuntivo, doença maligna ou imunodeficiência, a associação com a artrite seronegativa é pouco conhecida.

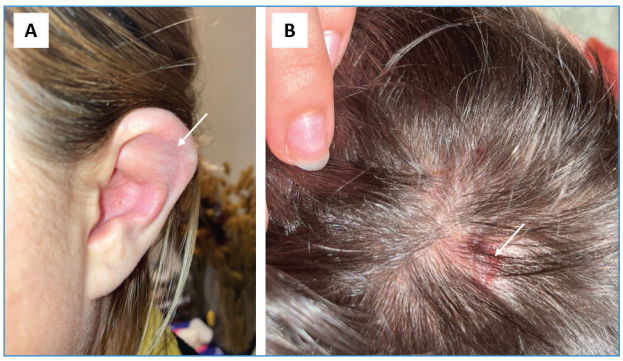

Figura 1. Pericondrite auricular (A). Lesão cutânea eritematosa não descamativa no couro cabeludo (B).

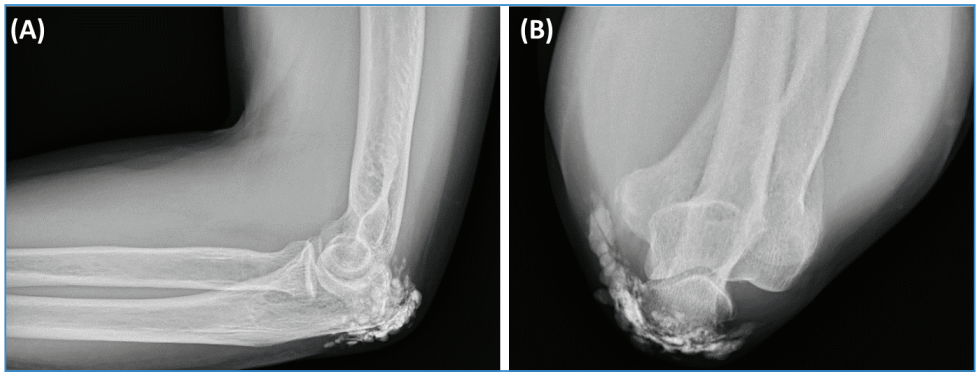

Figure 1. (A) X-ray of the left elbow, side view. (B) X-ray of the left elbow, axial view.

227 - CALCINOSIS CUTIS AS A FIRST STEP INTO DIAGNOSIS OF SYSTEMIC SCLEROSIS

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Clinical case report. A 64-year-old woman with a personal history of chronic ischemia of the lower limbs was referred to the Rheumatology department due to free drainage of whitish liquid content from the left elbow. She reported inflammatory arthralgias of both hands and shoulders and denied cough, chest pain, dyspnea, dysphagia or other complaints. On the physical exam, she presented with puffy hands, digital ulcers on the third and fourth fingers of the right hand, cold extremities and hard, subcutaneous and painful nodules of the left olecranon. An x-ray of the left elbow showed several radiopaque foci over the olecranon region compatible with calcinosis (Figure 1. (A), Figure 1. (B)). After analytic workup a diagnosis of anti-centromere antibody-positive limited systemic sclerosis was confirmed. Cutaneous calcinosis is a symptom of systemic sclerosis that affects particularly older female patients with longer disease duration. However, it frequently goes unnoticed and is therefore underdiagnosed and undertreated. In fact, a recent study carried out in Portugal reports that around 40% of the cases are subclinical. Drawing attention to this manifestation of systemic sclerosis can contribute to greater diagnostic accuracy of the disease and its treatment.

237 - BAND ACRO-OSTEOLYSIS: A RARE MANIFESTATION OF SYSTEMIC SCLEROSIS DISEASE

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A 71-year-old caucasian woman with a diagnosis of diffuse cutaneous systemic sclerosis manifesting as triphasic Raynaud phenomenon, proximal skin thicken-

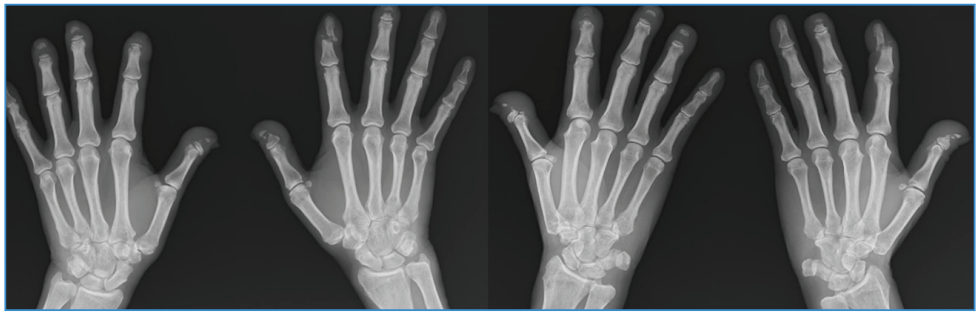

Figure 1. X-ray of both hands demonstrating band acro-osteolysis

ing, microstomia, arthritis, and oesophageal dysmotility presented to a routine Rheumatology appointment. Her immune profile consisted of positive antinuclear antibodies (in a titer of 1/1280) and positive anti-RP11 antibodies. Upon physical examination, shortening of multiple fingers was noted and a hand x-ray (Figure 1) was ordered, which revealed resorption in the midshaft of the 1st, 2nd, 3rd, and 4th right distal phalanges and the 1st and 3rd left distal phalanges. Destruction of the left 2nd distal interphalangeal joint was noted. These findings were consistent with band acro-osteolysis. History of exposure to polyvinyl chloride, trauma, or a family history of Psoriasis or Hajdu-Cheney syndrome were excluded. Parathyroid hormone, calcium, and vitamin D serum levels were within the normal range. Acro-osteolysis is a well-known feature of systemic sclerosis, with terminal tuft resorption being more frequently reported than band acro-osteolysis. The pathophysiological mechanisms behind acro-osteolysis are not fully understood, but it is believed that vascular changes and decreased capillary density leading to tissue hypoxia may be involved.

245 - DIFFUSE IDIOPATHIC SKELETAL HYPEROSTOSIS AND DYSPHAGIA: WHAT'S THE LINK?

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Diffuse Idiopathic Skeletal Hyperostosis (DISH) is a systemic, non-inflammatory clinical entity characterised by calcification of ligaments and/or entheses. The diagnosis is established if there is calcification of the anterior longitudinal ligament of at least 4 contiguous vertebral bodies and in the absence of: pronounced discopathy; ankylosis of facet joints; sacroiliitis. It most often occurs in men between the ages of 50 and 60. The aetiology is unknown, but it is associated with type 2 diabetes (DM2), hyperuricemia and metabolic syndrome. It predominantly affects the thoracic spine but can also affect the lumbar and cervical spines. Clinically it can be asymptomatic, or it can be presented with low back pain and morning stiffness. When it affects the cervical spine, in addition to cervicalgia, it can present with dysphagia and/or dysphonia. We present a case of a patient with bilateral cervicobrachialgia and progressive dysphagia for solids.

A 76-year-old man with a history of DM2, metabolic syndrome, hyperuricemia, cervical discopathy and surgery for a herniated lumbar disc. Medicated with oral anti-diabetics, acetylsalicylic acid, antihypertensives, esomeprazole 20mg/day and allopurinol 100mg/day. No drug allergies. He went to the neurosurgery consultation due to progressive dysphagia for solids, with six months of evolution and continued to complain of bilateral cervicobrachialgia, which had been going on for six years. There were no other complaints of pain or systemic signs and symptoms. A cervical CT scan showed exuberant anterior osteophytosis from C4 to C6, compatible with DISH, and the cervical MRI showed disc herniations with myelomalacia at C5-C6. The oesophageal barium study (EBS) revealed the same anterior cervical ossifications moulding the posterior wall of the oesophagus, with no obstacle to the passage of contrast. Given the diagnosis of DISH leading to dysphagia, the patient underwent surgery, with resolution of the dysphagia.

According to the literature, there are 400 described cases of dysphagia associated with DISH, with an incidence of 10.6 per cent if older than 60 years old. Solid dysphagia is the most common. It is associated with anterior osteophytosis at C5-C7 due to mechanical compression or a local inflammatory reaction. The decision to opt for surgery depends on the extent of the disease, which is why CT, cervical MRI and EBS should be carried out. CT is essential to determine the repercussions on the digestive tract and airway, while MRI is useful if spinal cord compression is suspected. EBS makes it possible to assess the degree of dysphagia. Most authors argue that if there are small osteophytes and if dysphagia does not cause weight loss, patients should start conservative treatment. In this clinical case, the patient had all the risk factors associated with DISH. DISH was responsible for the dysphagia, and other differential diagnoses such as spondylosis, cervical neo-

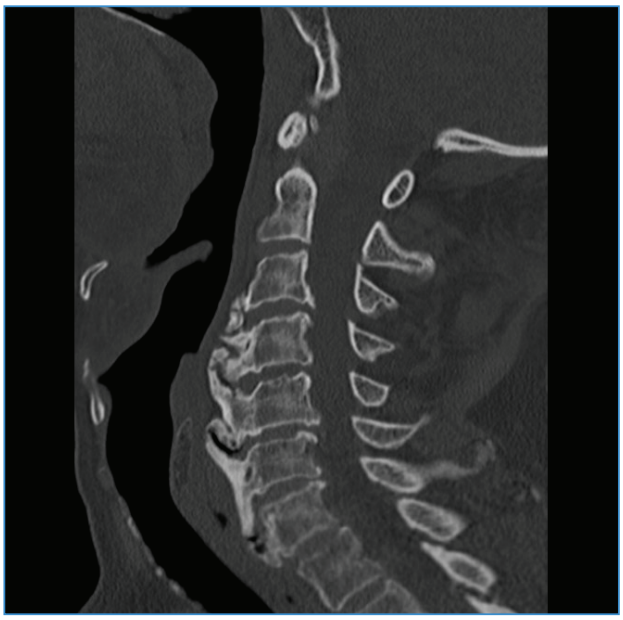

Figure 1. DISH at C3-C7 leading with dysphagia

plasia and oesophageal pathology were ruled out. The involvement of C4-C6 led to dysphagia for solids, as described in the literature. As for treatment, the patient underwent surgery straight away due to spinal cord involvement and concomitant dysphagia.

In short, DISH can be present with dysphagia for solids, which, if not diagnosed in timely manner, can lead to life-threatening situations such as aspiration pneumonia. Surgical treatment consists of osteophytectomy and postoperative regression of dysphagia is observed in 96 per cent of cases.

250 - HEMOLACRIA- A RARE SIGN OF A RARE CLINICAL SYNDROME

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Introduction: Gardner-Diamond syndrome was first described in 1955 as a psychological and dermatologic syndrome involving painful, ecchymotic, purpuric lesions that usually occur after a period of stress or minor trauma. It is a rare condition with unclear pathophysiology that affects mainly young women. This is exclusion diagnosis, so hematological evaluation is needed. An early approach could reduce pain and emotional impact of the disease. The aim of the treatment is to control anxious and depressive disease leading to control of lesions. We present a case with impressive ocular and cutaneous manifestations.

Clinical case: A 43 years old female patient was send to the outpatient rheumatology clinic due to myalgia, arthralgia and positive antinuclear antibodies 1/320.

In the last months, after a miscarriage, worsening of the widespread pain, with morning stiffness of less than 30 minutes, lead to an interruption in the daily life activities and word.

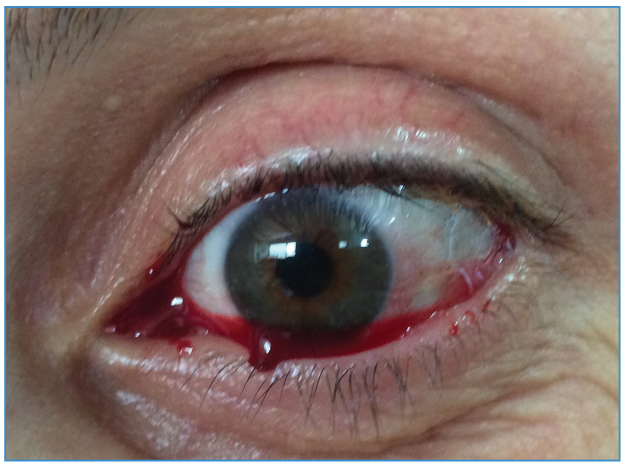

Figure. Hemolacria

She did not have any other symptoms suggestive of connective tissue disorder but had a remarkable story of bloody tears, spontaneous hair hemorrhage and easy bruising.

Also had a G10 P0 history, pregnancy losses of the first trimester, without a cause and a previous major depression episode 10 years ago.

Had been followed in several imunohemotherapy clinics (Lisbon, Oporto and Coimbra) with extensive laboratory investigations but without a definite diagnosis.

In the CHUC Imunohemotherapy clinic, a probable diagnosis of Gardner Diamond syndrome was suggested.

The clinical evaluation of this patient presented a decrease C3 in the first evaluation but dsDNA, ENAs, LAC test and other anticardiolipin antibodies were negative.

This patient was first started on duloxetine 30mg but the increased to 60mg was not tolerated. Duloxetine controlled the pain but increased the bruising and was stopped, C3 was normal in the second evaluation.

She is now starting agomelatine which was the drug she tolerated best in the depression episode.

Conclusion: Gardner-Diamond syndrome condition has been rarely described and some of these patients do received Systemic Lupus Erythematosus diagnosis first due to low complement levels and positive anticardiolipine antibodies.

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253 - CALCINOSIS UNIVERSALIS – UM ACHADO RARO

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Os autores apresentam um caso de calcinosis universalis, numa doente do sexo feminino, com 56 anos de idade, seguida em consulta de Reumatologia com diagnóstico de Síndrome de sobreposição entre Esclerose Sistémica e Artrite Reumatóide.

Este diagnóstico foi baseado na presença de

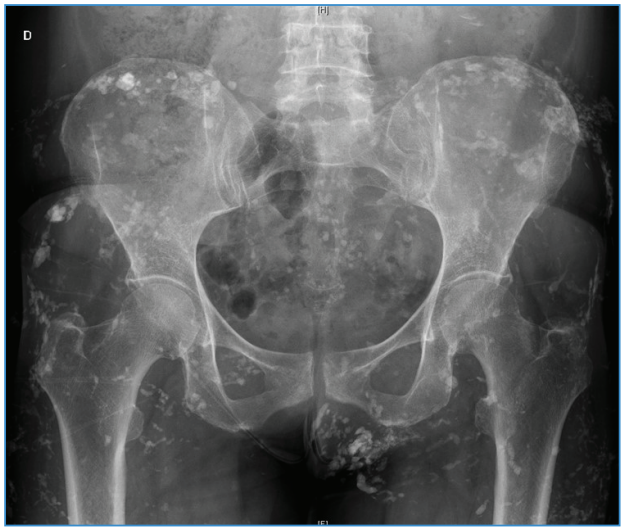

Figure. Calcinose cutis universalis visível em radiografia de anca

poliartrite simétrica e erosiva com positividade em elevado título para fator reumatóide e anticorpos anti-citrulinados e a presença de Raynaud trifásico das mãos e pés de instalação tardia, telangiectasias múltiplas, esclerodactilia, lesões cutâneas de tipo sal e pimenta, calcinose cutânea, doença intersticial pulmonar (padrão NSIP) e positividade de ANAs com doseamento de anti-PM-Scl100.

As radiografias solicitadas no sentido de avaliar o dano estrutural revelaram a existência de extensas lesões de calcinose envolvendo os planos fasciais da pele e tecido celular subcutâneo, compatíveis com calcinosis universalis. Esta forma extensa de calcinose cutânea constitui uma apresentação rara, estando associada a significativa morbilidade e impacto funcional.

A doente está atualmente sob tratamento com abatacept ev 500mg mensal, prednisolona 5mg id, hidroxicloroquina 200/400mg id, calcifediol 0.266mg mensal e iniciou colquicina 1mg id, no sentido de prevenir surtos inflamatórios pela calcinose, com bom resultado clínico.

254 - ULTRASOUND-GUIDED DIAGNOSIS AND MANAGEMENT OF RECURRENT ACROMIOCLAVICULAR JOINT CYSTS: A CLINICAL IMAGE

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Introduction: Acromioclavicular joint (ACJ) cysts are uncommon lesions that arise in the region of the ACJ. These cysts are typically associated with chronic degenerative conditions of the shoulder, especially rotator cuff tears that allow joint fluid to extend into the subacromial-subdeltoid bursa. This process can result in the formation of a cystic structure. In addition, the increased local pressure may lead to the development of cutaneous fistulas, posing a risk of infection. Symptoms typically include swelling and discomfort in the shoulder region, often exacerbated by physical activity. Diagnosis is commonly achieved through imaging techniques like ultrasound, which help in assessing the condition of the shoulder joints, the rotator cuff, and the presence of the cyst. Treatment options vary based on the underlying condition, the severity of symptoms, and patient characteristics, ranging from conservative approaches to more invasive procedures.

Case Report: We report a case of 82-years-old man with medical history of elderly-onset Rheumatoid Arthritis controlled by prednisolone 2.5 mg/day, DM type II, essential hypertension, prostate cancer treated by radiotherapy, dementia, Severe symmetric axonal sensorimotor polyneuropathy. Presented to Rheumatology appointment with an impressive mass in the left shoulder area, without cutaneous signs suggestive of inflammation/infection. Ultrasound was performed, diagnosing an ACJ cyst. We aspirated around 40cc of fluid and 6cc of a more gelatinous synovial material and performed intra-articular steroid injection. We applied a pressure dressing after the procedure. There was no microbe growth. This was the third recurrence of the same cyst, so we referred him to Orthopedic care and potential surgical treatment.

Discussion: Usually, in degenerative shoulder joint conditions, the cyst occurs due to communication between the glenohumeral joint and ACJ. Synovial fluid flows out of the glenohumeral joint and into the ACJ. It is believed that the cyst enlarges due to the presence of a "one-way valve". This was a typical case, associated to a chronic rotator cuff tear and elderly-onset Rheumatoid Arthritis, confirmed by elevated CRP, ESR, RF and ACPA. However, in all cases we should consider other differential diagnosis, as cystic neoplasm, epidermoid cyst or subcutaneous lipoma. That's why bed-side ultrasound presents as an immediate and convenient tool to ensure accurate diagnosis and effective treatment planning, significantly enhancing patient care and outcomes in cases involving shoulder pathologies.

Conclusion: Ultrasound imaging is important for diagnosing and managing ACJ cysts due to its non-invasive nature, cost-effectiveness, and real-time imaging capabilities. It excels in identifying associated shoulder disorders, such as rotator cuff tears, and is instrumental in guiding minimally invasive procedures like aspi-

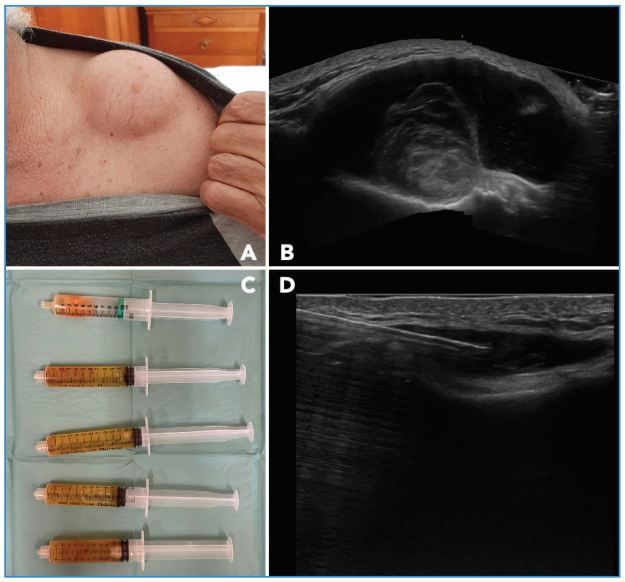

Figure. Image of Recurrent Acromioclavicular Joint Cyst: physical examination, ultrasound-diagnosis and ultrasound-guided management

rations or injections. This case image is paradigmatic of the exposed, depicting a massive cyst.

Image Description - Recurrent Acromioclavicular Joint (ACJ) Cyst: A – macroscopic aspect of the mass on physical examination; B – A well-defined, anechoic fluid-filled sac with a lower-central hyperechoic heterogenous mass located in the ACJ at the top of the shoulder on ultrasound; C – Five barrels with the content extracted from the ACJ cyst (macroscopic appearance – viscous translucent yellowish liquid); D – ultrasound-guided puncture of the ACJ cyst.

257 - ATYPICAL MECHANIC'S HANDS IN A CASE OF ANTISYNTHETASE SYNDROME-SYSTEMIC LUPUS ERYTHEMATOSUS OVERLAP

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Introduction: Antisynthetase syndrome (ASyS) is an inflammatory myopathy associated with anti-aminoacyl transfer RNA synthetase antibodies and typically

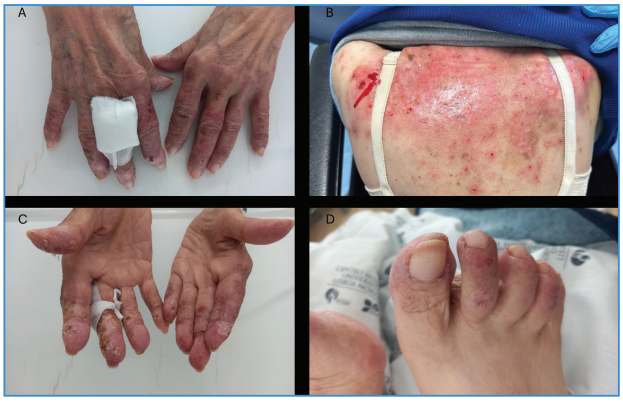

Figure 1. (A, C and D) Diffuse purpuric and scaly macules and patches on the fingers of the hands and feet. (B) Shawl sign.

characterized by arthritis, myositis and lung involvement. Skin (mechanic's hands, hiker's feet, heliotrope rash, Gottron's sign, shawl sign, V-neck sign) and vascular (Raynaud's phenomenon) manifestations are also frequent. ASyS can occur as an overlap with other connective tissue diseases and, in those cases, further skin changes might be noted, such as purpuric lesions, digital ulcers and subacute cutaneous lupus.

Clinical vignette: An 83-year-old-woman presented to the emergency department with a three-month history of a nonpruritic, hyperkeratotic, erythematous and scaly skin rash of the hands, feet, neckline and back, associated with vasculitis lesions (Figure 1). The exuberance of these lesions on the dorsum and sides of the fingers resembled the previously mentioned mechanic hands, with an additional finding of concomitant purpuric lesions. She also had proximal muscle weakness (Manual Muscle Testing - MMT8 142/150), Raynaud's phenomenon and a productive cough for 2 months. Laboratory testing results were notable for anaemia (Hb 9.1 g/dL), leukopenia (3.6 x 109/L), lymphopenia (0.38 x 109/L), elevated C-reactive protein (9.36 mg/dL), an alpha-1 and alpha-2 peak in the serum protein electrophoresis and complement consumption (C3 = 35 mg/dL, C4 = 8 mg/dL). Additionally, antinuclear antibodies (titre 1:1280, AC-1 pattern), anti-double stranded-DNA (230.6 UI/mL), anti-P ribosomal (>86.9), anti-Ro52, anti-Ro60 and anti-PL-7 (strongly positive by immunoblot) antibodies were positive.

Chest x-ray and computed tomography (CT) scan revealed acute pneumonia.

The patient was diagnosed with ASyS and Systemic Lupus Erythematosus (SLE) overlap, complicated with an infectious pneumonia, and was treated with prednisolone 30mg/day, hydroxychloroquine 400mg/day, intravenous immunoglobulin (IVIg) 2g/Kg (divided in 5 consecutive days) and antibiotics, with progressive

and significant improvement.
Conclusion: With this case, we want to feature the skin involvement in a rare case of ASyS and SLE overlap, highlighting the similarity between the typical ASyS cutaneous manifestations such as mechanic's hands and the shawl sign, but also the less common exuberant vasculitis lesions, a reminder to always consider alternative or additional diagnosis.

271 - POLICONDRITE RECIDIVANTE – DO NARIZ À TRAQUEIA

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Descrição do caso: Doente do sexo feminino, 45 anos, com antecedentes pessoais de asma persistente de difícil controlo e rinossinusite crónica com polipose nasal, tratada com dupilumab desde junho de 2022. Inicia quadro de dor torácica episódica e dispneia para esforços em julho de 2023, a motivar internamento por suspeita de asma descompensada. Realiza tomografia computorizada do tórax neste contexto, que evidencia estenose e espessamento difuso das paredes da traqueia e dos brônquios (figura 1A, 1B e 1C) e pequenas adenopatias infracarinais. O quadro evoluiu com persistência de queixas respiratórias associado a tumefação e destruição rápida da cartilagem nasal (figura 1D), uveíte anterior e artrite. Inicia tratamento com metotrexato em dose crescente, mas sem melhoria, e manutenção de dose elevada de prednisolona diária. O estudo imunológico foi negativo e a biópsia do septo nasal, sem representação de tecido cartilagíneo, não evidenciou lesões de vasculite. Iniciou ciclofosfamida pela gravidade do envolvimento cartilagíneo do sistema respiratório.

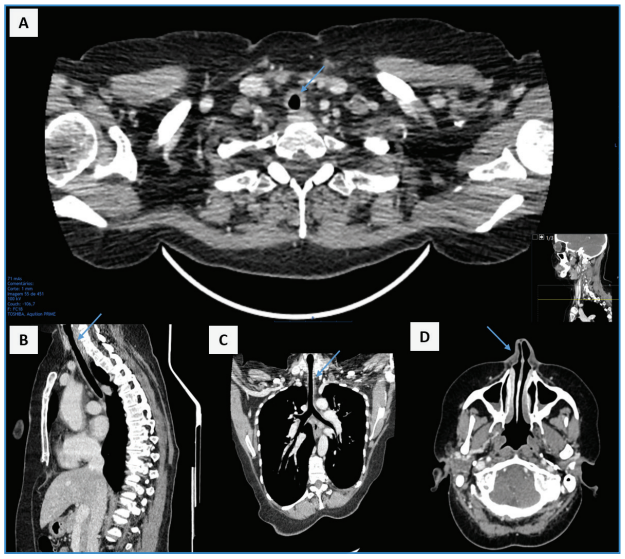

Figure. Estenose e espessamento das paredes da traqueia e dos brônquios principais (A, B e C) e colapso da asa do direita do nariz (D)

Discussão: A policondrite recidivante é uma doença imuno-mediada associada a inflamação sistémica com envolvimento predominante de estruturas cartilagíneas. Embora o atingimento auricular seja o mais característico, pode envolver estruturas como nariz, olhos, articulações, cartilagem costal, trato respiratório, coração, vasos, rim e sistema nervoso. Em doentes com envolvimento grave das vias aéreas, a abordagem terapêutica inicial envolve geralmente uma combinação de corticoide e ciclofosfamida (em alternativa, metotrexato ou azatioprina), com redução gradual da dose de corticoide quando controlada a atividade da doença. Em casos de resistência à terapêutica inicial, podem ser usados agentes biológicos ou ciclosporina.

278 - THE SUCCESSFUL TREATMENT OF REFRACTORY CHRONIC CUTANEOUS LUPUS WITH ANIFROLUMAB

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Systemic lupus erythematosus (SLE) is a heterogeneous autoimmune disease with different organ manifestations. Chronic cutaneous lupus (CCL) can present in 70% to 80% of patients with SLE. Given its potential for permanent scarring and dyspigmentation, CCL has a significant negative impact on quality of life. Its treatment is often challenging. Anifrolumab was recently approved for the treatment of SLE. Prior evidence has shown a significant benefit in patients with SLE, but data regarding the efficacy of CCL is lacking.
A woman, 39 years old, presented to the outpatient rheumatology clinic with diffuse erythematous, crusty, pruritic plaques on her upper and lower extremities and face (figure 1A), photosensitivity, and alopecia. She also described prolonged morning stiffness and arthritis in her fingers, wrists, and right knee. She was positive for antinuclear and anti-SSA/Ro60 antibodies and had low C3 and C4 proteins. The skin biopsy was

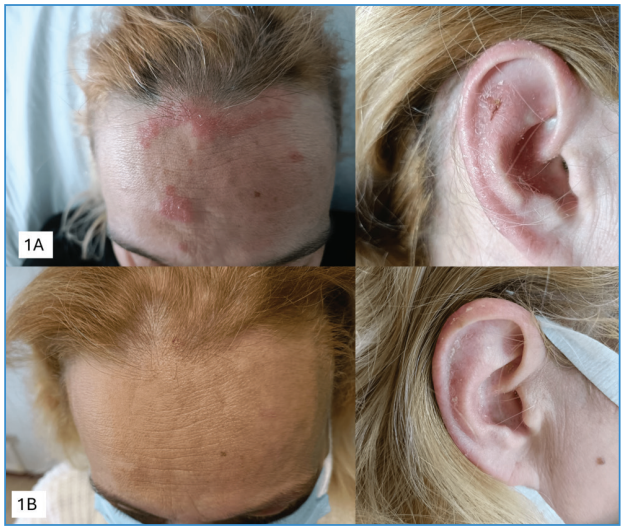

Figure 1. Erythematous, crusty, pruritic plaques corresponding to refractory chronic cutaneous lupus in the forehead and ear of a patient before (1A) and after (18) Anifrolumaft.

consistent with discoid lupus erythematosus, and she was diagnosed with SLE. Skin lesions were refractory to treatment with topical corticosteroid, hydroxychloroquine, methotrexate, and cyclosporine. After 7 years, Anifrolumab infusions were initiated with a complete resolution of cutaneous symptoms within 3 months (figure 1B).

Anifrolumab has been shown to be a therapeutic option that should be considered in patients with refractory or severe CCL.