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Localized Pigmented Villonodular Synovitis of the Shoulder: a Rare Presentation of an Uncommon Pathology

Sinovite Vilonodular Pigmentada Circunscrita do Ombro: uma Apresentação Rara de uma Patologia Incomum

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ABSTRACT

Pigmented Vilonodular Synovitis is a rare clinical entity characterized as a synovial membrane benign tumour, despite possible aggressive presentation with articular destruction. The localized variant is four times less frequent and the shoulder involvement is uncommon. We present the case of a Caucasian 59 year-old patient, who presented with left shoulder pain, of uncharacteristic quality, with local swelling and marked functional limitation of 1 month duration. Shoulder ultrasonography showed subacromial bursitis. An ultrasound-guided aspiration was performed: synovial fluid was citrine-colored and translucid. One month later, the patient maintained swelling, pain and functional impairment of the left shoulder. New shoulder ultrasound revealed exuberant subacromial bursitis, which was again aspirated using ultrasound guidance. The synovial fluid was haematic, without changes in the cell count or biochemical analysis and cultural exams. We performed an injection with 60 mg of hexacetonide triamcinolone. Two months later there was a relapse, with shoulder ultrasonography once more showing subacromial bursitis with extensive synovial membrane proliferation. Shoulder MRI revealed subacromial bursitis involving the anterior, posterior and medial recesses, with deltoid distension, but without tendinous or intra-articular involvement. In the interior of the bursa hypointense images in T2 were observed, suggesting the diagnosis of Pigmented Vilonodular Synovitis. The patient had surgical bursectomy with success and without complications. The histological exam of the operatory piece confirmed the imaging diagnosis. Pigmented Vilonodular Synovitis is uncommon, rarely affecting the shoulder in a localized variant. It is a diagnosis to be considered in shoulder pain, especially if associated with recurrent subacromial bursitis.
Keywords: Synovitis, Pigmented Villonodular; Shoulder Joint.

RESUMO

A Sinovite Vilonodular Pigmentada é uma entidade clínica rara caracterizada como um tumor benigno da membrana sinovial, apesar de possível apresentação agressiva com destruição articular. A variante circunscrita é quatro vezes menos frequente e o envolvimento do ombro é incomum. Apresentamos o caso de uma doente de 59 anos de idade, que apresentava omalgia à esquerda, incaracterística, com tumefacção local e limitação funcional de um mês de duração. A ecografia do ombro mostrou bursite subacromial. Foi efectuada uma aspiração ecoguiada: o líquido sinovial era translúcido e de cor citrina. Um mês mais tarde, a doente mantinha tumefacção, dor e incapacidade funcional do ombro esquerdo. Foi efectuada nova ecografia do ombro, que revelou bursite subacromial exuberante, que foi novamente aspirada com apoio ecográfico. O líquido sinovial era hemático, sem alterações nos exames culturais e citoquímico. Efectuou-se injecção ecoguiada na bursa com 80 mg de hexacetonido de triamcinolona. Dois meses mais tarde houve recorrência de sintomas e a ecografia mostrou uma vez mais bursite subacromial com proliferação sinovial extensa. A RMN do ombro revelou bursite subacromial envolvendo os recessos anteriores, posterior e interno, com distensão do deltóide, mas sem envolvimento tendinoso ou intra-articular. No interior da bursa observaram-se imagens hipointensas em T2, sugerindo o diagnóstico de Sinovite Vilonodular Pigmentada. A doente foi sujeita a bursectomia cirúrgica, com sucesso e sem complicações. O exame histológico da peça operatória confirmou o diagnóstico imagiológico. A Sinovite Vilonodular Pigmentada é incomum, raramente afectando o ombro na variante circunscrita. É um diagnóstico a ser considerado na omalgia, especialmente se associado a bursite subacromial recorrente.
Palavras-chave: Articulação do Ombro; Sinovite Vilonodular Pigmentada.

INTRODUCTION

Pigmented Villonodular Synovitis (PVNS) is a benign tissue proliferation that presents as a frontier case between a reactive and a neoplasic process and emanates from the tenosynovial layers, joint capsule or the synovial bursa. It was first defined in 1941[1] in a series of patients with proliferative lesions arising from the synovium of various joints.

Pigmented Villonodular Synovitis is an uncommon clinical entity, with an estimated prevalence of 1,8:1 000 000 people.[2] It usually occurs in young adults, between the 3rd and 4th decades of life, with men and women equally affected.[3] Some authors found a female predominance.[4] It can present as a diffuse or a localized/circumscribed form, and it is usually monoarticular. Poliarticular involvement is exceptionally rare.[3,5] The extra-articular form of pigmented

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villonodular synovitis is very rare and only a few cases were reported in which the lesion was found exclusively outside the joint without intra-articular segment.[6]

The most common affected locations by PVNS are the knees (80% of all cases), but it is occasionally found in the hips, ankles and finger joints.[4,5] In the indexed medical literature we found less than 40 described cases of Pigmented Villonodular Synovitis of the shoulder. Of those, only 2 were circumscribed,[6,7] exclusively extra-articular. The first description of such a case was published in 1997.

CASE REPORT

A 59 years old female patient, Caucasian race, born and living in Portugal, domestic worker, came to the Rheumatology appointment due to persistent moderate left shoulder pain, 1 month duration, with mixed characteristics. The pain was more intense in the morning, with partial relief with rest, aggravation with effort and limited response to non-steroid anti-inflammatory drugs. This condition significantly limited her daily routines. She denied any other symptoms or history of trauma.

At observation she presented swelling of the shoulder, with manifest functional limitation. A shoulder radiograph did not reveal any anomalies beside marked subacromial bursitis (Fig. 1). Shoulder ultrasonographic (US) exam revealed subacromial bursitis, which was punctured using ultrasound guidance: approximately 70 ml of citrine-colored translucid synovial liquid was aspirated.

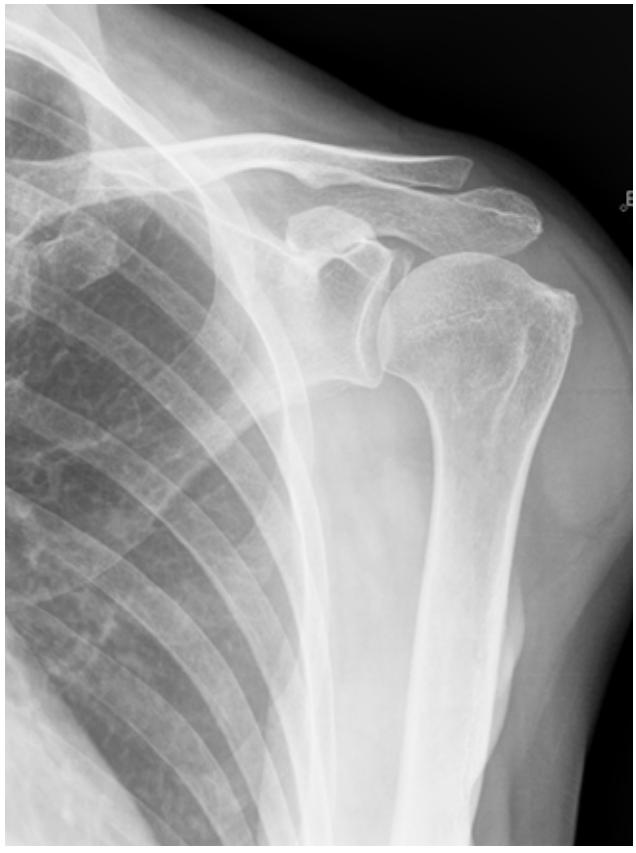


Figure 1 – Shoulder radiograph shows soft tissue swelling with a hypertransparent line next to the lateral contour of the left shoulder, without noticeable bone changes.

One month later, the patient presented swelling, pain and functional impairment of the left shoulder. Another ultrasound exam was performed, which showed exuberant subacromial bursitis. An ultrasound-guided aspiration of the subacromial bursa was performed, collecting slightly haematic synovial fluid. An infiltration with 60 mg of hexacetonide triamcinolone was successfully executed. The synovial fluid cell count and biochemical analysis were normal and cultures were negative.

Two months after this procedure, there is a relapse of the previous described symptoms and clinical findings. Another ultrasound reveals exuberant subacromial bursitis, with marked synovial membrane proliferation. This finding, along with the recurrent bursitis despite treatment leads us to think in Pigmented Vilonodular Synovitis as a probable diagnosis. A MRI of the shoulder (Fig.s 2a, 2b and 3a, 3b) showed increased volume of the subacromial bursa, involving the anterior, posterior and medial recesses, with deltoid muscle distension, but without tendinous or articular involvement. In the interior of the bursa, the existence of hypointense images in T1 and T2 modes confirmed the diagnosis of Pigmented Villonodular Synovitis.

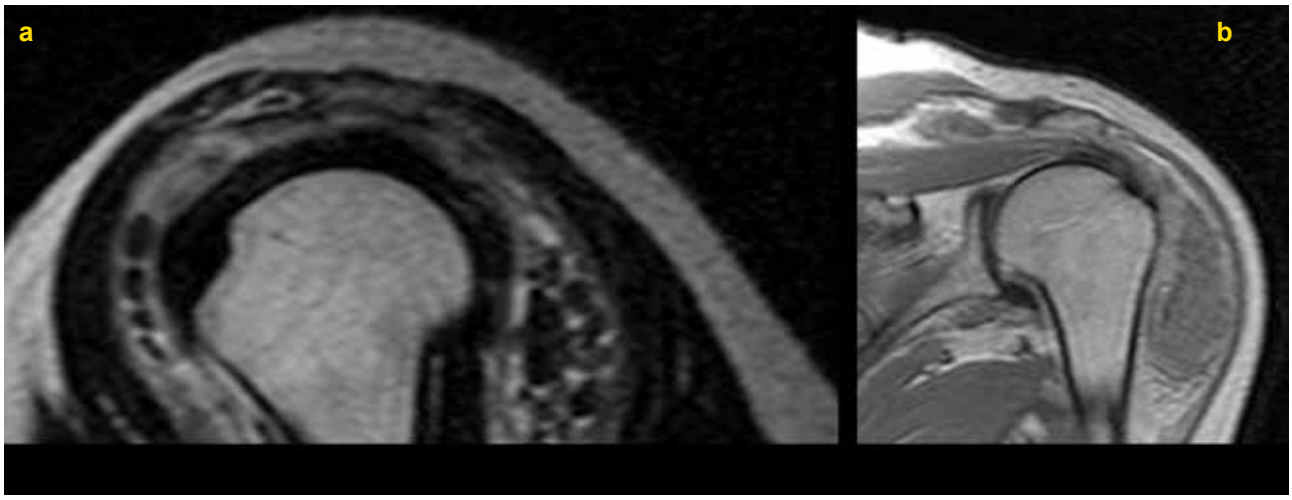
An open subacromial bursectomy was performed on the patient six months afterwards, with success and without surgical or post-surgical complications. The histological examination of the operatory piece confirmed the diagnosis. So far the patient has no symptoms or functional limitation of the affected shoulder. Two years later the patient remains asymptomatic and no recurrence has been shown in radiographs, ultrasound and MRI of the shoulder.

DISCUSSION

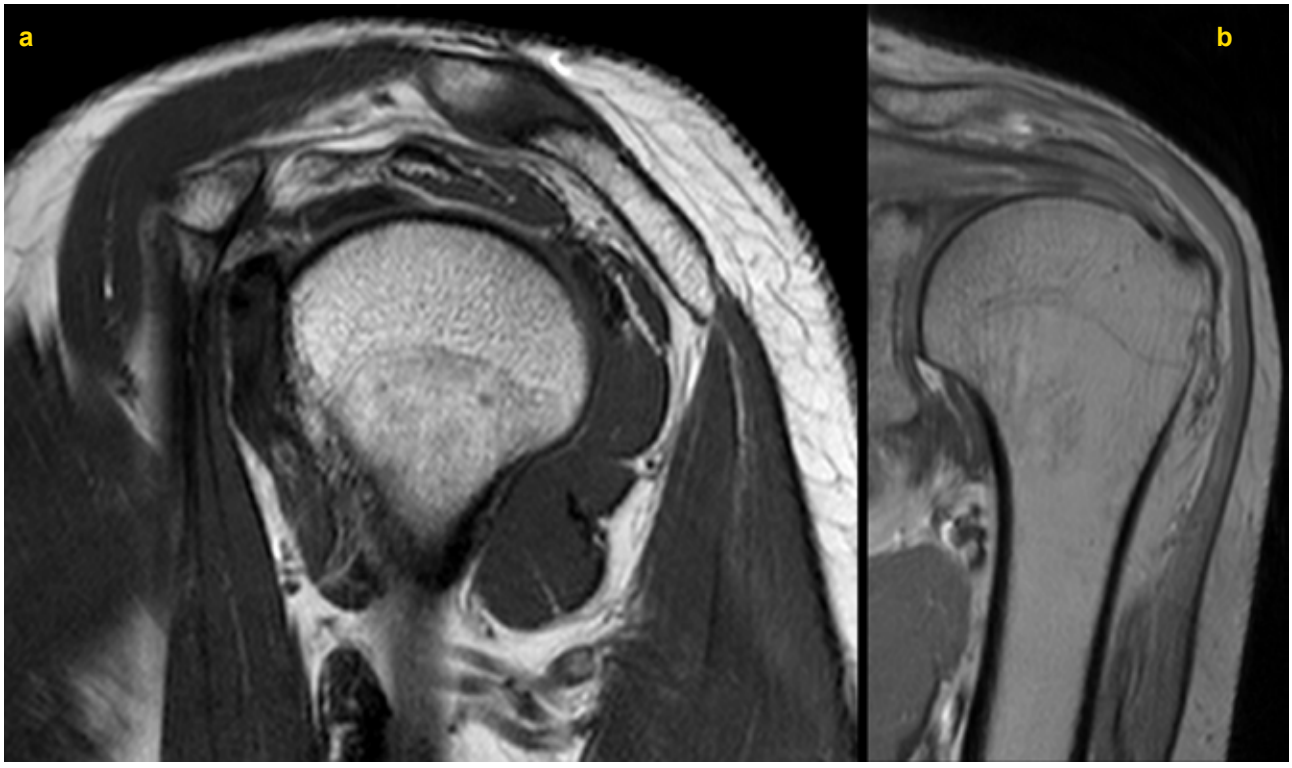
Pigmented Villonodular Synovitis is an uncommon clinical entity, with an estimated prevalence of 1.8: 1 000 000 people.[2] It usually occurs in young adults, between the 3rd and 4th decades of life, with men and women equally affected.[3] The most common locations are the knees (80% of all cases), and it is occasionally also found in the hip, ankle and finger joints.[4,5] In the indexed medical literature we found less than 40 described cases of PVNS of the shoulder. The extra-articular form of PVNS is very rare and often represents an extension of a primary intra-articular process. Only a few cases have been reported in which the lesion was found solely outside the joint, without intra-articular involvement.[6]

Of the PVNS cases affecting the shoulder only 2 were circumscribed, with all remaining cases presenting articular commitment.[6,7] Our case, as far as we know, is the first depiction of circumscribed Pigmented Villonodular Synovitis of the shoulder in the Portuguese population and the third worldwide.

Although Pigmented Villonodular Synovitis usually presents as a benign entity, in some cases it can destroy the joints leading to significative limitation or functional impairment.[8] Most series show only a small number of recurrences, with no metastatic disease or death.[9] However there are



Figures 2a and 2b – MRI (T1) of the shoulder reveals subacromial bursitis with exuberant circumscribed (not intra-articular) synovial membrane proliferation and hypointense nodular images, in grapefruit disposition, which correspond to haemosiderin deposits in the synovial.



Figures 3a and 3b – MRI (T1) of the shoulder after surgery shows slight thickening of the lateral recess of the subacromial bursa (without distension) and partial rupture of the supraespinatus tendon; no intra-articular lesions are found.

case reports of malignant transformation.[10] One study confirmed chromosome changes in the surgical specimens of PVNS, suggesting that this disease is most likely neoplasic in nature.[11] The same authors postulate that localized and diffuse forms of tenosynovial giant cell tumor might represent two morphologic manifestations of the same entity.[11]

Patients typically present with swelling, stiffness, and progressive pain in the involved joint,[3] as did our patient. Aspirated fluid is typically xanthochromic or serosanguineous. Involvement of the shoulder typically occurs in older patients and is characterized by the absence of serosanguineous effusion.[3,5] In our case the first aspiration of the subacromial bursa revealed normal synovial fluid, which supports this claim. Later aspirations showed the typical serosanguineous fluid found in PVNS.

The radiographic appearance of the diffuse form of PVNS is usually normal. The localized form varies from a normal osseous anatomy with soft tissue mass, subacromial effusion or small cystic erosions of the subchondral bone in the early stages of disease, to juxta-articular cystic lucency of the glenoid and humeral head in the late stages.[7,12]

The clinical findings in PVNS are non-specific and the diagnosis is made either by MRI, due to its characteristic images, or by anatomophatological examination of the operatory piece. In MRI the diffuse form of PVNS is usually presented with joint effusion fluid in the shoulder bursa,

dark haemosiderin pigmented synovium accumulation in T1-T2W sequences (particularly in the glenohumeral joints) and rotator cuff tear can also be seen in some cases.[2,12] The localized nodular form is generally presented with a soft tissue mass involving the shoulder capsule and rotator cuff muscle tendons.[12] Sawmiller et al[7] presented a case of extra-articular PVNS where the MRI of a 57 year old female patient showed complete rotator cuff tear, large glenohumeral joint effusion with fluid in the subacromial and subdeltoid bursae, and a large acromial spur in the right shoulder. In this case, the rotator cuff tear was most likely secondary to a classic impingement syndrome due to the large spur and not by a direct invasion of PVNS. In our case we found partial rupture of the supraespinatus tendon only after surgery (without any acromial spur), which we consider as a surgical outcome setback. Similar to the case reported by Chul-Hyun Cho et al,[6] the disease in our patient occurred in the subacromial space, had no communication between the subacromial space and glenohumeral joint and no full-thickness tear of the rotator cuff was found.

Considering MRI, differentiating calcifications from haemosiderin-laden foci in the setting of PVNS may prove to be difficult. Radiographs can be used in this context to confirm or deny the presence of calcifications.[13] Calcifications are not a usual feature of PVNS but rarely, foci of dystrophic calcifications can be found. Nevertheless MRI has a crucial role and allows differentiating several pathologies which compromise the joint and adjacent structures like synovial hemangioma, synovial osteochondromatosis, synovial arborescence lipoma and synovial sarcoma.[14] MRI is the imaging modality of choice in PVNS and is useful for diagnosis, surgical planning, and follow-up.

Regarding treatment options and considering our experience in this patient, hexacetonide triamcinolone seems ineffective and adds little value to bursa ultrasound-guided aspiration. The latest is effective in relieving symptoms, but only on a temporary basis until the possible definitive solution: synovectomy/bursectomy. Still, in many reported cases, surgery is not the definitive treatment, as relapses can occur in as much as 50% of cases of diffuse PVNS.[2,8] Recurrence has not been reported in extra-articular PVNS of the shoulder.[6]

Untreated PVNS can invade the surrounding soft tissue and joint, leading to articular destruction, in which case arthroplasty may be the solution.[14] Therefore, early diagnosis and prompt treatment are important for optimal disease management.

CONFLICT OF INTERESTS

None stated.

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None state.

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