Case report

Medical Ultrasonography
2012, Vol. 14, no. 1, 67-70

Milwaukee shoulder syndrome associated with pigmented villonodular synovitis. Case report.

Horaţiu I. Popov[1], Claudia Gherman[2], Liliana Rogojan[3], Carolina Botar-Jid[4], Cristina Barna[1], Daniela Fodor[1]

[1] 2nd Internal Medicine Department, [2] 2nd Surgical Department, [3]Histopathology Department, [4]Radiology Department "Iuliu Haţieganu" University of Medicine and Pharmacy, Cluj-Napoca, Romania

Abstract

Milwaukee shoulder syndrome (MSS) describes a destructive shoulder arthropathy associated with of calcium hydroxyapatite and calcium pyrophosphate dihydrate crystals deposition found mainly in elderly women. Pigmented villonodular synovitis (PVNS) is a benign proliferative disorder of the synovium usually seen in young adults, found frequently in the knee joint. We present the case of a 63 year old Caucasian woman admitted for severe swelling of the left shoulder and mild pain, with 10 years history. Clinical signs, laboratory tests, imaging studies, and histopathological examinations established the diagnosis of MSS associated with PVNS. Surgical partial synoviectomy followed by radiotherapy (total dose 36 Gy) was considered with local improvement. This is the first report in literature about this association.

Keywords: Milwaukee shoulder, pigmented villonodular synovitis, shoulder

Introduction

The Milwaukee shoulder syndrome (MSS) consists of the association of rotator cuff complete tear with osteoarthritic changes, noninflammatory joint effusion containing calcium hydroxyapatite and calcium pyrophosphate dihydrate crystals, hyperplasia of the synovium, destruction of cartilage and subchondral bone, and multiple osteochondral loose bodies [1,2]. It classically affects patients aged over 70 years, with predominance in females, and it clinically manifests as a rapidly progressive and destructive arthritis of the shoulder. Pigmented villonodular synovitis (PVNS) is a rare benign hyper-proliferative disorder characterized by an exuberant overgrowth of synovial cells, causing recurring joint non-serosanguineous effusions and erosive lesions of the adjacent bony structures in advanced stages of the disorder. Most frequently, it affects the knee joint, whereas only 2 % of cases have involvement of shoulder joint [3,4]. Patients present with insidious onset of progressive joint swelling and discomfort [5]. Radical synovectomy is the treatment of choice, which is possibly limited by the complex anatomic structure of the involved joint, and/or by the histological type of the disorder (diffuse type of PVNS). This is responsible for the elevated recurrence rates and has led to the use of postsurgical external radiotherapy [6].

In this case report we described a patient with MSS associated with PVNS.

Received 24.01.2012 Accepted 31.01.2012
Med Ultrason
2012, Vol. 14, No 1, 67-70
Corresponding author: Daniela Fodor, MD, PhD
2nd Internal Medicine Department
"Iuliu Haţieganu" University of Medicine
and Pharmacy, Cluj-Napoca, Romania
2-4 Clinicilor str, 400006 Cluj Napoca, Romania
Phone: 004 0264591942/442
Email: dfodor@umfcluj.ro

Case report

A 63 year old woman was admitted for severe swelling of the left shoulder started 10 years before. During this time she did not refer to any doctor or follow any treatment, due to the lack of pain. The disability of the shoulder was moderate and had not interfered with her daily activity until one year prior presentation. In the last year she noted impairment in the shoulder movements,

especially in abduction, mild pain described as local pressure, a supplementary increase in dimensions of the shoulder and, intermittent, ecchymosis around the shoulder. She had no history of trauma, diabetes, syphilis or other significant systemic diseases.

Clinical examination revealed important swelling of the left shoulder extending up to the arm and chest wall, and limitation of joint movements, especially the abduction. The swollen area presented multiple small dilated blood vessels and was bordered by an extended ecchymosis appeared 2 days before admision, after a moderate physical activity (fig 1). Her cardiac, pulmonary and abdominal examination were normal.

Routine laboratory tests showed inflammatory syndrome (erythrocyte sedimentation rate 71 mm/h, fibrinogen 564 mg/dl). No other laboratory tests were abnormal.



Fig 1. Clinical aspect of the patient. Note the multiple small dilated blood vessels and the extended ecchymosis.

Anteroposterior radiograph of the left shoulder and CT scan was performed (details in fig 2). Magnetic resonance imaging (MRI) evaluation was not posible due to the patient's claustrofobia.

Shoulder ultrasound depicted a massive fluid collection in the subacromial-subdeltoidian (SASD) bursa, large synovial proliferation (fig 3) with multiple calcification and multiple irregularities of the humeral head, clavicle and acromion. Evaluation of the biceps, infraspinous, supraspinous, and subscapularis tendons was difficult to be performed. A small amount of fluid was identified into the glenohumeral joint. Given the large effusion, US guided arthrocentesis of the shoulder was performed and 200 ml hemorrhagic fluid from the SASD bursa was aspirated and local corticosteroid was administrated. This led to obvious improvement in the affected area. Cytologic examination of synovial fluid showed blood elements and the culture was negative.

Ultrasound guided needle biopsy of the proliferated synovial was performed in the same session in order to exclude a soft-tissue sarcoma, particularly synovial sarcoma, haemangioma, or a fibroma. The examination of the biopsy specimen revealed a small number of lymphocytes and polymorphonuclear cells, focal hyperplasia of the synovial cells and villous proliferation, many plasma cells around the vessels, multiple hemosiderin deposits, multinucleated giant cells and hematic infiltration suggesting PVNS. Calcium pyrophosphate dihydrate crystals were also found on polarized light microscopy (fig 4). The conclusion was that our patients had the findings for the both diseases: MSS and PVNS.

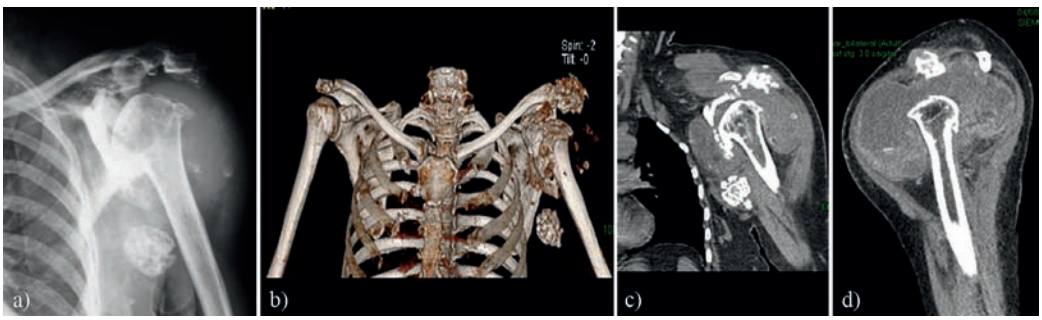


Fig 2. X-Ray and CT scan of the left shoulder. a) anteroposterior radiograph: soft-tissue swelling, irregular calcifications, large calcificated mass in left axilla, subchondral sclerosis, and bone cysts; b) CT scan, coronal plane reconstruction, bone substraction: joint space narrowing, bone sclerosis and destruction, capsular and soft-tissue calcifications, large calcified mass in the left axilla; c) CT scan, coronal plane reconstruction: large soft-tissue inhomogenous hypodense mass around the left humeral head, with calcifications and some hypodense spots (hemosiderin accumulation?), subchondral sclerosis with cysts in the left humeral head, bone structural changes of the left acromion, multiple irregular intra and periarticular calcifications, and a large multiple, well defined calcifications in left axilla; d) CT scan of the left shoulder, sagital plane: large hypodense, inhomogenous soft-tissue mass around the humeral head with calcifications, and bone structural changes.

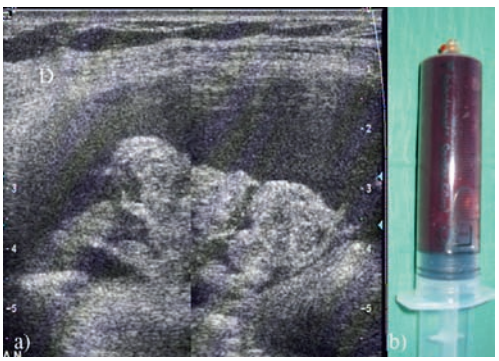


Fig 3. Ultrasonography of the left shoulder: a) longitudinal scan from the anterior part of the shoulder-fluid collection into subacromial-subdeltoidian bursa, marked synovial proliferation, D- deltoid muscle; b) the macroscopic aspect of the hemorrhagic fluid aspirated from bursa.

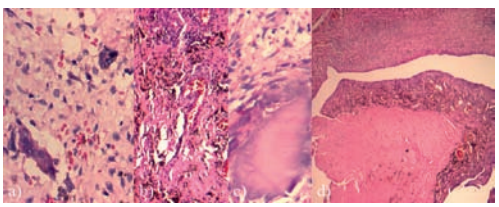


Fig 4. Histopatologic examination: a) multinucleated giant cells; b) hemosiderin deposits and hematic infiltration; c) calcific deposits; d) villous proliferation of the synovia.

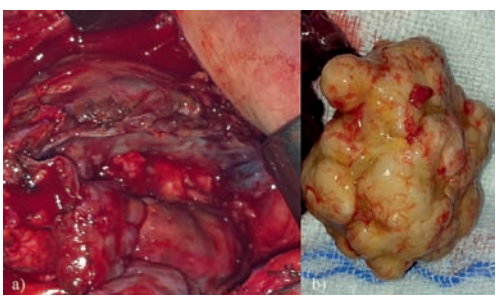


Fig 5. Intra-operative photographs showing a) the macroscopic aspect of proliferated synovia; b) the macroscopic appearance of the calcification described on imagistic evaluation in the axilar region

At first, due to objective reasons, the patient was treated conservatively with oral colchicine and nonsteroidal anti-inflammatory agents but two weeks later the shoulder effusion was restored on physical and ultrasound examination. Surgical treatment was considered – bursectomy and partial synovectomy of the left shoulder was performed. The largest calcareous conglomerate extracted from the SASD bursa had 6 cm in diameter (fig 5). The histopathologic examination of the proliferated synovia confirmed the previous bioptic findings. The patient received also postsurgical radiation therapy in total dose of 36 Gy (6x6Gy/every 3 days). The evolution was favorable with improvement of the local changes and range of movements of the left shoulder. In one year follow-up period the patient remained stable.

Discussion

MSS is a well defined clinical entity characterized by destructive arthropathy associated with calcium hydroxyapatite and calcium pyrophosphate dihydrate crystals depositions. The etiology of the disease is unclear. Trauma on the affected side (fall on the outstretched hand), calcium pyrophosphate dihydrate crystals deposition disease, cervical neuropathy due to syringomyelia or severe spondylosis [7], long-term dialysis [8] were involved. In many cases no causative factor can be identified [9]. Histopathological and ultrastructural studies of synovium in MSS demonstrated villous and synovial lining cell hyperplasia, giant cell formation, fibrin, and basic calcium phosphate crystals deposition. No inflammatory cell infiltration is observed, at most only a few lymphocytes [10]. The histopathologic findings make the differentiation with other crystal deposition arthropathies possible. The synovial fluid have a xanthochromatous or frank hemorrhagic aspect usually in large quantities, with noninflammatory cell count, basic calcium phosphate crystals and sometimes calcium pyrophosphate dihydrate crystals [7,10].

PVNS is a rare condition, not a true neoplasm, but a slowly progressive inflammatory monoarthropathy characterized by exuberant synovial overgrowth and joint erosion. The etiology of PVNS is unknown and may be attributed to a repetitive inflammatory process [11]. There is a significant association between PVNS and repetitive trauma [12] but it is consider that intraarticular hemorrhage is not the trigger factor [13]. Synovial tissue in PVNS is characterized by extensive haemosiderin deposition and the presence of numerous macrophages and giant cells which expressed an osteoclast-phenotype responsible for the periarticular bone destruction [11].

Both diseases are rare conditions. The estimated yearly incidence of PVNS is 1.8 per one million inhabitants [14], but the prevalence of MSS is not noted [9]. Their coincidence has a low probability. We found in literature no report about association of MSS with PVNS. We assume that PVNS was the first disease of the shoulder due to the slow progression and preservation for a long period of the shoulder range of motion [15] with supra-

added of MSS due to release into the synovial fluid of basic calcium phosphate crystals from the bone erosions, crystals secondary phagocytosed by synovial lining cells [11]. MSS can be unilateral or bilateral, but in unilateral disease the shoulder on the dominant side is uniformly involved [7]. The order of the disease's sequences can explain why the non-dominant shoulder was affected in our case. The absence of pain for a long period (in the absence of syringomyelia or other nervous system pathology) followed by mild pain describe as local pressure, is another particularity of the case. This was the main reason way the patient had not seek medical advice in the past 10 years.

The imagistic techniques (radiology, CT, US) allowed a proper pre-surgical evaluation. MRI would have been more useful in describing the synovial proliferation but it was not possible due to the severe claustrophobia of the patient. US was especially useful for US guided synovial biopsy and synovial fluid extraction. In this way the sarcoma or other types of synovial proliferation were excluded before surgery.

Interesting is the apparition of multiple small dilated blood vessels (venous) around the shoulder, causing repeated subcutaneous hemorrhages with extensive ecchymosis. We do not have a satisfactory explanation for this modification. Of noted that, after surgical intervention, the number and volume of the vessels significantly decreased and no other hemorrhages appeared.

The patient is still under observation due to the possibility of recurrence or of the worsening of the shoulder function. In this situation shoulder arthroplasty will be take in consideration, as being the radical treatment for both diseases [9,15] .

References

1. Llauger J, Palmer J, Rosón N, Bagué S, Camins A, Cremades R. Nonseptic monoarthritis: imaging features with clinical and histopathologic correlation. RadioGraphics 2000; 20: S263-S278.
2. Ea HK, Liote F. Calcium pyrophosphate dihydrate and basic calcium phosphate crystal-induced arthropathies: update on pathogenesis, clinical features, and therapy. Curr Rheumatol Rep 2004; 6: 221-227.
3. Mahieu X, Chaouat G, Blin JL, Frank A, Hardy P. Arthroscopic treatment of pigmented villonodular synovitis of the shoulder. Arthroscopy 2001; 17: 81–87.
4. Dorwart RH, Genant HK, Johnston WH, Morris JM. Pigmented villonodular synovitis of synovial joints: clinical, pathologic, and radiologic features. AJR Am J Roentgenol 1984; 143: 877-885.
5. Dowart RH, Genant HK, Johnston WH, Morris JM. Pigmented villonodular synovitis of the shoulder: radiologic-pathologic assessment. AJR Am J Roentgeno. 1984; 143: 886–888.
6. Heyd R, Micke O, Berger B, Eich HT, Ackermann H, Seegenschmiedt MH. Radiation therapy for treatment of pigmented villonodular synovitis: results of a national patterns of care study. Int J Radiat Oncol Biol Phys 2010; 78: 199-204.
7. McCarty DJ. Milwaukee shoulder syndrome. Trans Am Clin Climatol Assoc 1991; 102: 271-283.
8. Yano E, Takeuchi A, Yoshioka M. Hydroxyapatite associated arthritis in a patient undergoing hemodialysis. Int j Tissue React 1985; 7: 527-534.
9. Epis O, Viola E, Bruschi E, Benazzo F, Montecucco C. Milwaukee shoulder syndrome (apatite associated destructive arthritis): therapeutic aspects. Reumatismo 2005; 57: 69-77.
10. Halverson PB, Garancis JC, McCarty DJ. Histopathological and ultrastructural studies of synovium in Milwaukee shoulder syndrome--a basic calcium phosphate crystal arthropathy. Ann Rheum Dis 1984; 43: 734-741.
11. Taylor R, Kashima TG, Knowles H, Gibbons CL, Whitwell D, Athanasou NA. Osteoclast formation and function in pigmented villonodular synovitis. J Pathol 225: 151- 6.
12. Myers BW, Masi AT. Pigmented villonodular synovitis and tenosynovitis: a clinical epidemiologic study of 166 cases and literature review. Medicine (Baltimore) 1980; 59: 223–238.
13. Docken WP. Pigmented villonodular synovitis: a review with illustrative case reports. Semin Arthritis Rheum 1979; 9: 1–22.
14. Rao AS, Vigorita VJ. Pigmented villonodular synovitis (giant-cell tumor of the tendon sheath and synovial membrane). A review of eighty-one cases. J Bone Joint Surg Am 1984; 66: 76–94.
15. Petsatodis G, Karataglis D, Kapoutsis DB, Papadopoulos P, Christodoulou AG. Hemiarthroplasty for pigmented villonodular synovitis of the shoulder: a report of two cases. J Orthop Surg (Hong Kong) 2011; 19: 116-119.