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Case Report

Simultaneous pigmented villonodular synovitis and synovial chondromatosis of the hip: case report

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ABSTRACT

This report presents a case of a 37-year-old female with a history of hip pain. Magnetic resonance arthrography revealed loose bodies within the joint and synovial hypertrophy indicative of synovial chondromatosis (SC). Hip arthroscopy revealed free chondral bodies and focal villonodular synovial proliferation. The focal synovial proliferation was excised, a total synovectomy performed, and all cartilaginous free bodies removed. A post-operative histological examination of the removed nodular mass and synovium yielded evidence of both SC and pigmented villonodular synovitis (PVNS). A 1-year post-operative clinical examination showed marked clinical improvement and no signs of recurrence on MR images. Despite the clinical similarities, PVNS and SC are two distinct conditions that, to our knowledge, have never been reported as simultaneously occurring in a hip joint. The simultaneous presence of both pathologies may suggest a common origin of synovial metaplasia.

INTRODUCTION

Pigmented villonodular synovitis (PVNS) is a rare, benign synovial disease characterized by unique characteristics including polyhedral cells surrounded by collagen, giant cells, and hemosiderin. Despite being defined as a benign process, PVNS causes direct degradation of the joint through bone and cartilage penetration. The disease presents most commonly in 20- to 50-year-old patients but is otherwise demographically non-specific. The knee is the most routinely affected joint, followed by the hip, ankle, shoulder and elbow. Clinically, PVNS presents with joint pain, swelling, and a limited range of motion [1].

PVNS diagnosis generally relies on magnetic resonance imaging (MRI). The focal nodules appear as low signal heterogenous masses. Radiographically, late stage PVNS may present as bony erosions in the joint of the diseased synovium which can be seen on plain radiographs as well as MRI [1].

Synovial chondromatosis (SC), is a benign joint disease characterized by the formation of intra-articular cartilaginous nodules. The resulting nodules can detach from the synovial lining, becoming free bodies which may undergo secondary calcification and ossification. Once ossified, the free bodies cause degradation of the articular cartilage. This disease commonly presents between the ages of 20 and 70 and is twice as likely to affect men. Clinically, SC also presents with joint pain, swelling and a limited range of motion [1].

Radiographically, the clearest indication of SC is the presence of free bodies within the joint, however, if the bodies have not ossified, the presence of such lesions may not be readily apparent on plain radiographs. In the hip, medial joint space widening may also be suggestive of SC as a result of loose bodies within the acetabular cotyloid fossa or a lateral change in femoral head positioning due to long-term structural changes [2]. SC is associated with the

development of cam type dysplasia of the femoral head which leads to femoroacetabular impingement (FAI) and is commonly accompanied by a labral tear which can be seen on MRI.

Though SC and PVNS often present indistinguishably in clinical examinations, each disease maintains a unique mechanism of joint degradation. The authors believe that this may be the first report in literature of a simultaneous occurrence of both diseases within the same hip joint.

CASE REPORT

A 37-year-old female presented to the senior author with atraumatic left hip pain and limited range of motion for 13 years. Despite the pain and stiffness, the patient retained an active lifestyle until 6 months prior to the surgery, at which point the patient's symptoms had progressed and she sought evaluation. The patient noted ambulation with a noticeable limp, complained of pain about the left groin, and noted a positive C-sign. Upon physical examination, the patient demonstrated limited range of motion with hip flexion of 110°, external rotation of 60° and internal rotation of 10°. The patient had painful limitation of motion during the flexion adduction internal rotation tests (FADDIR) and in the flexion abduction external rotation tests (FABER). Radiographs showed mild medial joint space widening, mild arthritic changes and an alpha angle of 70° (Fig. 2).

The MRI revealed numerous loose bodies within the joint and acetabular cotyloid fossa, a distinct longitudinal oval mass of soft tissue at the anterior aspect of the femoral neck, bone edema in the anterior aspect of the femoral head–neck junction, and a partial antero-superior labral tear (Fig. 1a and b). The radiological findings were consistent with medial joint space widening and cam type FAI (Fig. 2).

Arthroscopic hip surgery was performed through an anterolateral viewing portal and a modified anterior working portal according to the basic principles of hip arthroscopy for SC [3].

The hip was distracted for joint access. Loose bodies were removed from the central compartment with a 5.5-mm arthroscopic cannula and then the outer sleeve of a 5.5-mm-long abrader burr (Smith & Nephew, Andover, MA, USA) (Fig. 3a and b). After debriding the underlying acetabular margin, the labrum was repaired with two anchors. Once central compartment work was completed, traction was released and the hip was flexed. The peripheral joint was addressed, revealing brownish-yellow mass of soft tissue resembling a giant cell tumor, which was removed and sent to pathology for examination (Fig. 3a, c and d). All remaining loose bodies were removed from the joint and a synovectomy of the diseased tissue in the peripheral compartment was performed. An osteoplasty of the head neck junction was executed to correct the morphology of the femoral head. A dynamic assessment under direct arthroscopic viewing was subsequently performed to ensure adequacy of head neck resection. The capsule was closed with two number 2 Vicryl sutures (Johnson & Johnson, New Brunswick, NJ, USA). Post-operatively, the patient was allowed immediate full weight bearing as tolerated with the support of crutches for the first 2 weeks. The patient was initially prescribed daily range of motion exercises and advanced to strengthening

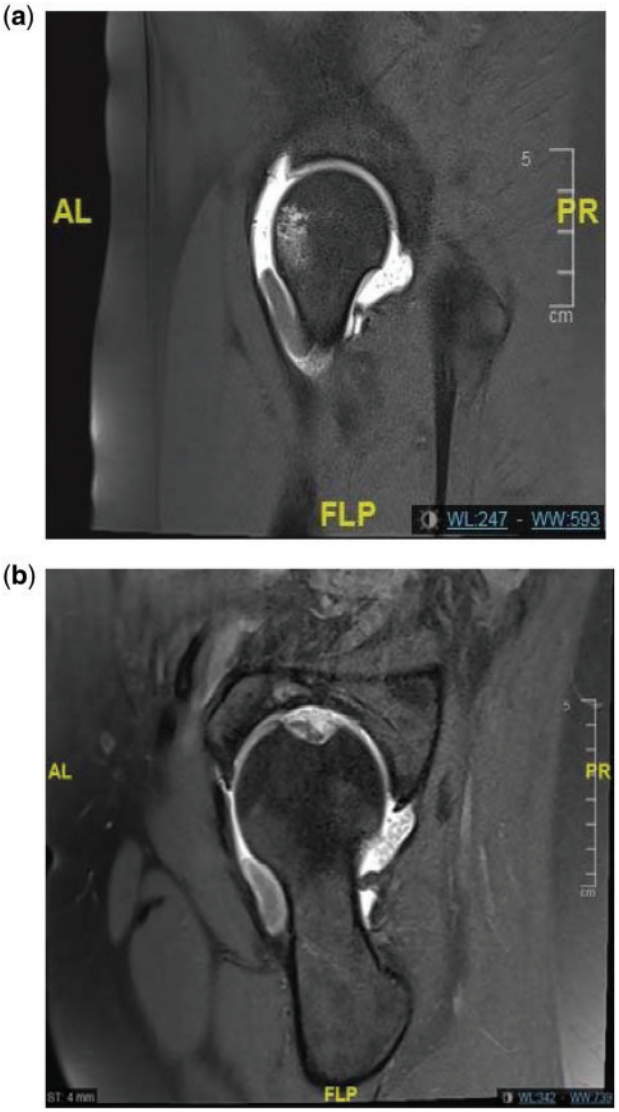

Fig. 1. (**a**) Preoperative MRI left hip. Free bodies present in the posterior joint space. An oval soft tissue mass occupies the space anterior to the femoral neck, edema seen at anterior femoral head as well as a labral tear. (**b**) Preoperative MRI. Free bodies present in the posterior joint space and in acetabular fossa. An oval soft tissue mass occupies the space anterior to the femoral neck.

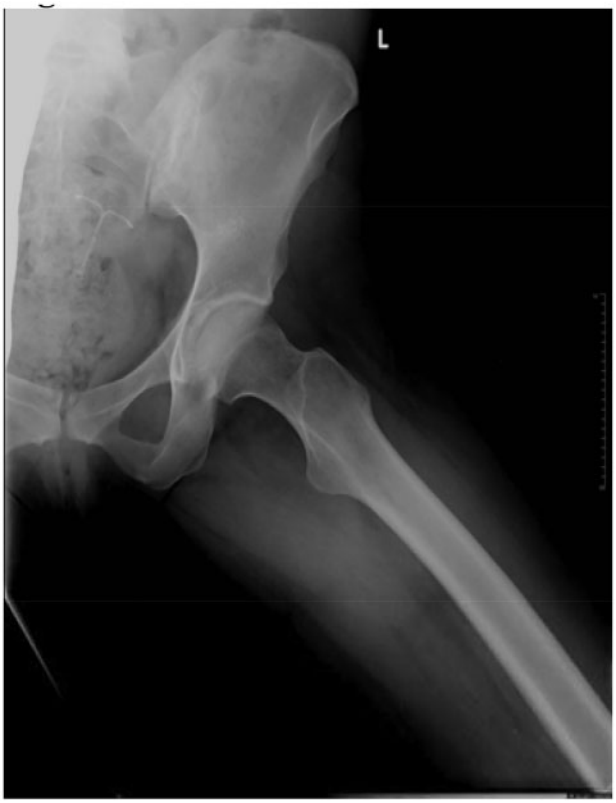

Fig. 2. An axial (frog) view of the left hip. The radiograph shows widening of the medial joint space and cam type FAI.

exercises in accordance with the senior author's routine protocol.

A histological report of loose body specimens removed from the joint during the surgery confirmed SC as the diagnosis. An examination of the unidentified soft-tissue mass/nodule excised during the surgery showed a proliferation of mononuclear cells, giant cells and foamy histiocytes set in collagenous stroma with focal hemosiderin pigment consistent with PVNS. The patient returned for a routine 1-year follow-up, pain free despite a return to full physical activity (including sports), and a resulting MRI showed no indication of recurrence of synovial disease (Fig. 4a and b).

DISCUSSION

Studies involving the mono-articular synovial diseases SC and PVNS tend to only have a few subjects and report mixed or inconclusive results [4].

SC is a benign joint disease characterized by the formation of intra-articular cartilaginous nodules. When the nodules detach from the synovium and undergo ossification,

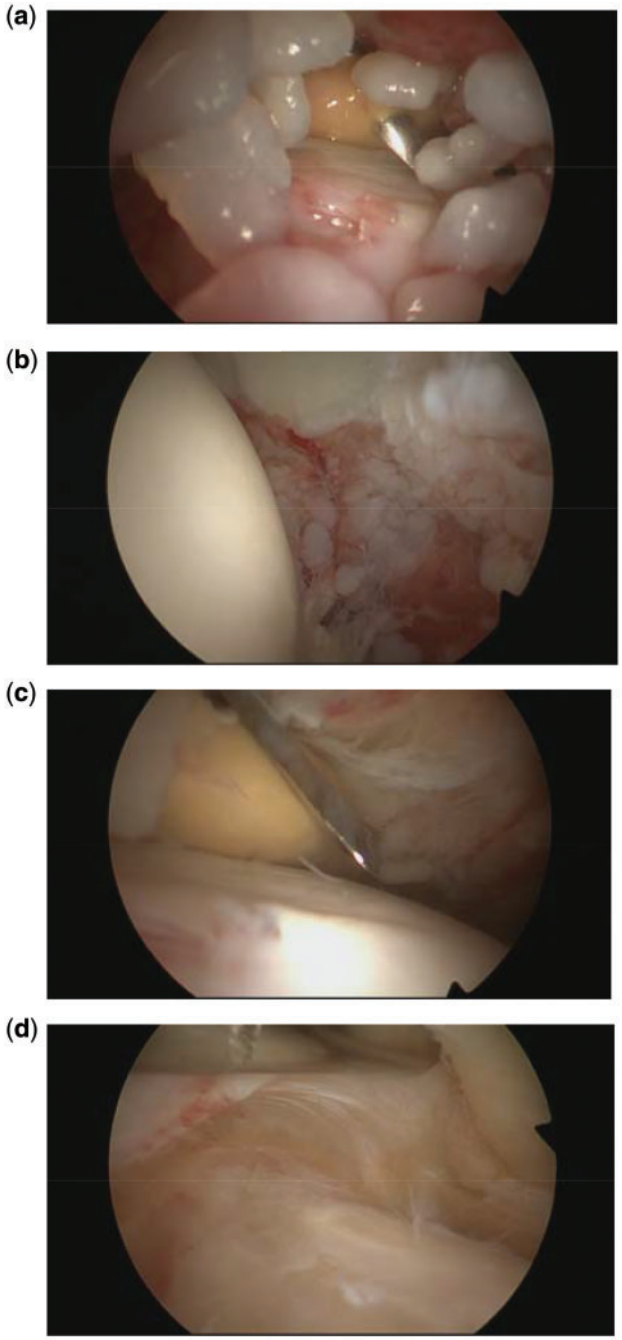

Fig. 3. (**a**) Arthroscopic image. Numerous loose bodies can be seen in the peripheral joint. A needle is piercing the brownish-yellow soft tissue mass. (**b**) Arthroscopic image. Loose bodies are obstructing the acetabular fossa. (**c**) Arthroscopic image. A view of the oblong mass. A histological report revealed the mass as a PVNS nodule. (**d**) Arthroscopic image. Peripheral hip space after complete removal of free bodies, subtotal synovectomy and the removal of the PVNS lesion.

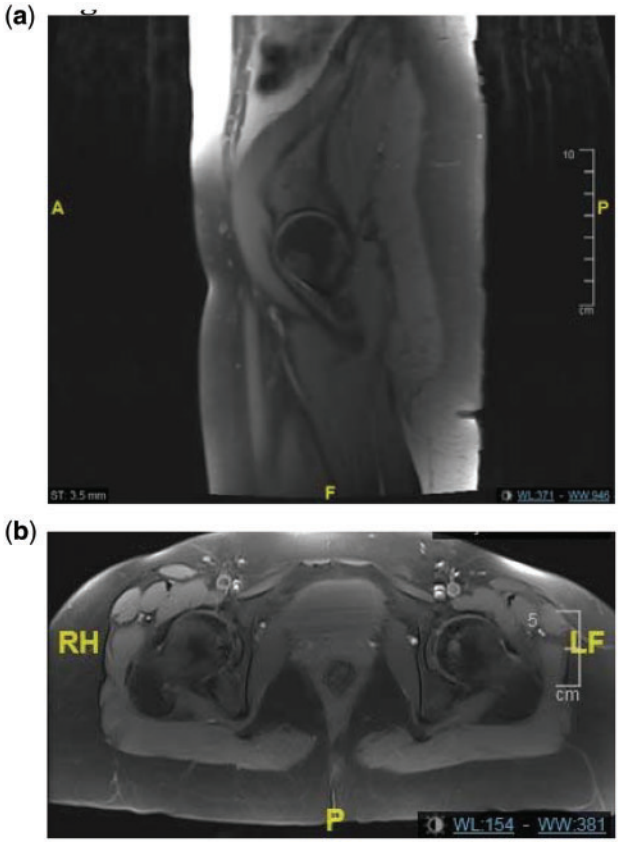

Fig. 4. Post-operative MRI 1 year from surgery. There is no recurrence of free bodies or synovial proliferation is seen.

the loose bodies cause degeneration of the articular cartilage. Despite unproven origins, SC is thought to originate from the metaplastic change of cells in the synovium to cartilaginous tissue [5]. A cytogenic study of SC revealed consistent abnormalities within chromosome 6 that suggests a possible neoplastic origin [6]. Clinically, SC presents with joint pain, swelling and a limited range of motion. Radiographic evidence of SC includes free bodies (sometimes calcified or ossified), medial joint space widening, cam type FAI and labral tears [1, 3, 4, 7].

Arthroscopic intervention and subtotal synovectomy to remove all free bodies and diseased synovium is the recommended management and has been shown to be beneficial for the patient [1, 3, 4, 8]. The aforementioned procedure to arthroscopically remove chondral lesions, damaged tissue, resolve FAI, and repair the labrum, was followed to treat the SC [3]. The recurrence rate of SC is 7.1% after arthroscopic intervention and does not depend on the stage of the disease. However, in their comprehensive systematic review, de Sa *et al.* suggest that the rate may increase relative to the age of the patient [8].

PVNS is a rare, benign synovial disease characterized by yellow-brown nodular lesions containing hemosiderin-laden macrophages, multinucleated giant cells, and inflammatory cells [9]. PVNS causes direct degradation of the joint which can be seen on plain radiographs as bone erosion. Although the origin of this disease is unknown, recent finding suggests a possible neoplastic etiology [10]. Furthermore, an immunohistochemical examination of PVNS confirmed that the pathology originates from the synovial surface cells [11]. PVNS can be diffuse hypertrophy of the synovium or a focal (nodular) lesion. In either case, the diseased synovium must be removed. If the patient presents with the diffuse form of PVNS, there is a significantly higher risk of recurrence of the disease regardless of surgical intervention and may require adjuvant radioactive injections. Focal PVNS does not generally require adjuvant radiotherapy [2]. There is no accepted recurrence rate for PVNS as studies have reported values anywhere between 8 and 46% [12].

PVNS and SC are separate diseases and must be treated individually. The SC was treated with arthroscopic loose body removal and synovectomy [3]. The PVNS was a focal lesion indicating that excision of the nodule and diseased tissue should be sufficient, but warrants follow up monitoring with MRI. SC leads to cam type dysplasia of the femoral head, FAI and labral tears which must be addressed to preserve the native hip.

The data regarding the origins of both diseases are suboptimal and there is no undeniable proof of any particular theory. However, both diseases stem from cells within the synovium and there is evidence that both may derive from neoplastic origins [5, 6, 11].

In conclusion, this case presents a rare simultaneous occurrence of both SC and PVNS in a hip, which was treated arthroscopically by a subtotal synovectomy, removal of free bodies and excision of the focal PVNS lesion. The simultaneous occurrence brings about the possibility that PVNS and SC may have a common origin. Despite the symptomatic similarities, treatment must account for the idiosyncrasies of both pathologies.

CONFLICT OF INTEREST STATEMENT

None declared.

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