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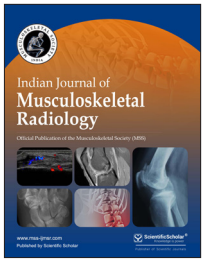

Case Report

Primary synovial osteochondromatosis of hip joint in a teenager

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ABSTRACT

Primary synovial osteochondromatosis (PSOC), also known as idiopathic synovial chondromatosis, is a rare benign articular disease with synovial proliferation and metaplasia without any predisposing factor. Clinical and imaging features play a major role in the diagnosis of this condition. We report a case of a young girl presenting with chronic hip pain, and suspected to have PSOC on imaging and confirmed on histopathology. Removal of loose bodies without any creation of surgical femoral dislocation was done as part of her treatment. There was no requirement of synovectomy due to complete synovial destruction secondary to the chronic disease process and such a case has not been reported till date. Patient has completely recovered from her symptoms after surgery.

Keywords: primary synovial osteochondromatosis, synovial metaplasia, hip pain

INTRODUCTION

Primary synovial osteochondromatosis (PSOC) is commonly seen in middle aged individuals and has been rarely reported in patients <20 year of age.[1] PSOC of hip in young needs optimal diagnosis and treatment to avoid recurrence and secondary osteoarthritis also to maintain the hip joint stability, integrity, and function. MRI remains the diagnostic imaging modality of choice and to look for any complications. Treatment of hip PSOC is standard which include removal of loose bodies and synovectomy.

CASE REPORT

A teenage girl aged 19 year came to the outpatient department with history of chronic onset of pain in the left hip for 3 years and difficulty walking for 6 months. There was no history of any major trauma in the past or pre-existing arthritis/predisposing inflammatory conditions. Medical history was unremarkable other than pain killers for the present condition which were not effective. Pain was mechanical rather than inflammatory in nature. Examination of the patient showed that there was difficulty to ambulate and severe pain while standing. Hip range of motion was asymmetric with guarding on extremes of motion. Significant pain was produced in the left hip during gentle internal and external rotation of hip while in 90° flexion. Knee range of motion was satisfactory. Her complete blood picture was perfectly normal.

She was evaluated earlier at a primary health-care center for her symptoms and preliminary x-ray done at that center showed erosion of the subchondral aspect of acetabulum and erosion

of the left femoral neck on radiography with few suspicious loose bodies seen on oblique view [Figure 1a and b]. Femoral head did not show any sclerosis, subchondral cysts, or contour irregularity. Joint space was normal. Due to lack of advanced imaging facilities at the primary health-care center, patient was referred to a tertiary hospital for further management. In view of her history, examination, and prior radiographic findings, MRI was advised. CT was also performed to look for the erosions and loose bodies [Figure 2a]. Only few calcified small loose bodies were seen on CT within the acetabular notch. On MRI, there were multiple T1 iso to hypointense and T2 hyperintense intra-articular loose bodies involving the left hip joint without any signal void on gradient images [Figure 2b-e]. Synovial lining was not appreciated. There was erosion of left acetabular notch and femoral neck without any bone marrow edema. There was no femoral head deformity/contour irregularity/impingement or secondary osteoarthritic changes. Initial diagnosis of PSOC was done on imaging. Differential diagnosis of pigmented villonodular synovitis (PVNS) was considered as there were bone erosions involving the left femur and acetabulum with associated multiple loose bodies. However, lack of susceptibility artifact from hemosiderin was evident against PVNS.

Patient was offered surgical treatment. Femoral head was exposed and reflected by modified Hueter anterior approach, separated from the joint capsule with further retraction. "I" shaped capsular arthrotomy was performed centered over the femoral neck. Synovial membrane was already destroyed by the disease process and synovectomy was not necessary. Loose bodies were removed from superior and inferior aspects of the femoral neck. Loose bodies in the posterior aspect were also removed through this incision at the level of the neck. However, those at the level of acetabulum surrounding the ligamentum teres could not be removed as dislocation was necessary for the removal. Labrum was preserved. Surgical femoral dislocation was not performed. Finally, closure of the arthrotomy was done. Gross specimen shows multiple hard to gritty loose bodies of similar shape and size [Figure 3a].

On histopathology, there were multiple circumscribed lobules of hyaline cartilage, surrounded by fibrotic synovial membrane. Attenuated synovial cells were seen lining the synovial membrane. Individual lobules consisted of mildly atypical chondrocytes arranged in clusters [Figure 3b and 3c]. There was no evidence of malignancy.

She is completely asymptomatic following 15 months post-surgery and has gained complete range of movements. MRI was advised after 6 months of follow-up. However, the patient and her family refused to undergo any further imaging as she was free of symptoms and due to financial constraints.

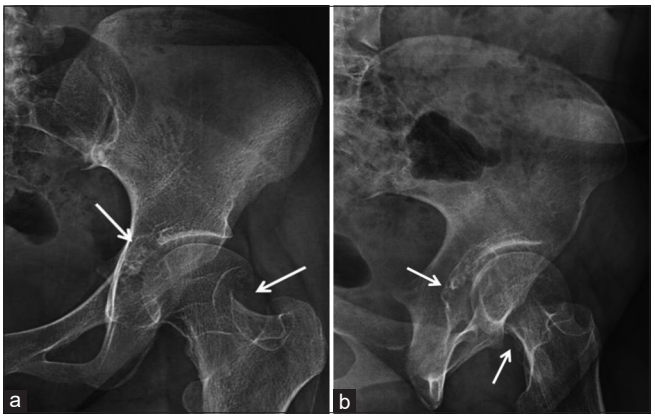

Figure 1: A 19-year-old female with primary synovial osteochondromatosis (PSOC) presented with hip pain. X-ray of pelvis Antero-posterior (a) and oblique views (b) show erosion of the left acetabular notch and neck of the left femur (arrows).

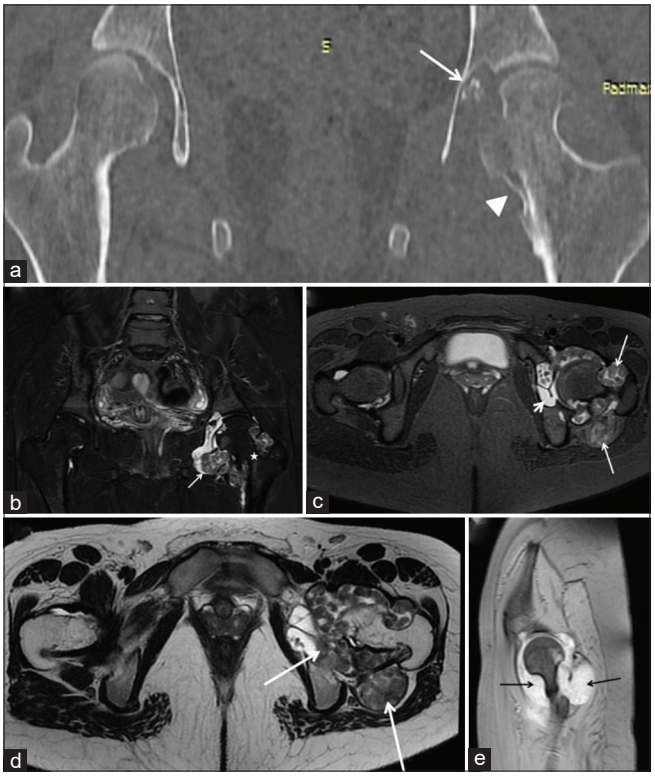

Figure 2: A 19-year-old female with PSOC presented with hip pain. (a) CT coronal section bone window image shows erosion of left acetabular notch and few adjacent small loose bodies (arrow) and erosion of femoral neck of the left femur (arrowhead). (b and c) Coronal and axial STIR images of pelvis show left hip joint effusion, multiple small hypointense intra-articular loose bodies (arrows) and erosion of the acetabulum (short arrow) and femoral neck (asterisk). (d) Axial T2-weighted image of pelvis shows left hip joint effusion with multiple small hypointense intra-articular loose bodies (arrows). (e) Gradient echo sagittal sequence of pelvis shows hip joint effusion without signal void (arrows).

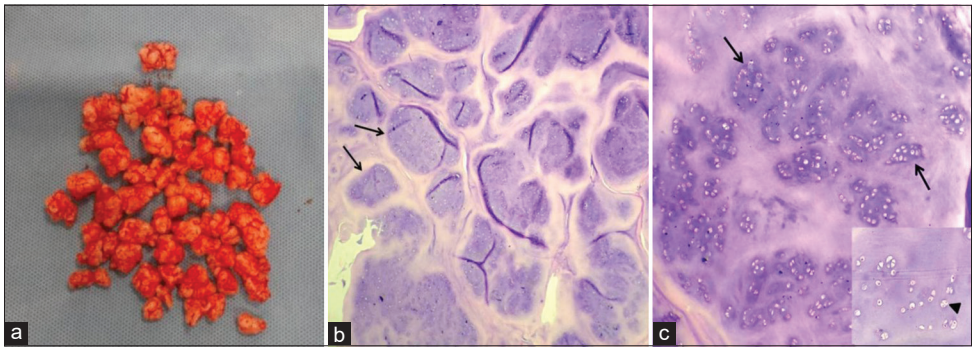

Figure 3: A 19-year-old female with PSOC presented with hip pain. (a) Photomicrograph of gross specimen showing multiple chondral bodies of similar size and shape. (b) Photomicrograph shows multiple lobules of hyaline cartilage embedded within the synovial connective tissue (arrows). (c) Photomicrograph showing high power view of one of the nodule displaying typical clustered arrangement of chondrocytes (arrows) and few of them showing binucleation (arrowhead in the inset).

DISCUSSION

The benign synovial metaplasia may be primary or secondary depending on the presence of the cause. PSOC is idiopathic and commonly intra-articular in nature. However, extra-articular involvement is also noted in the subsynovial tissue of the tendon sheath and bursae and are named tenosynovial and bursal chondromatosis accordingly.[2] Involvement of the major joints like knee and hip are common, though it can involve smaller joints including temporomandibular, acromioclavicular, and interphalangeal joints. Secondary osteochondromatosis occurs due to trauma or osteoarthritis.[2]

The initial phase of the condition includes nodular proliferation of synovial membrane resulting in synovitis with no obvious loose bodies, followed by dissociation of the metaplastic nodules with formation of loose bodies ranging from few millimeters to few centimeters and finally synovial proliferation settles down with resolution of synovitis.[2] PSOC presents with late onset of symptoms between third to fifth decade of life and is predominantly seen in men. It is rarely seen below the age of 20 year as in our case.[1,3] It presents with pain, swelling, and restricted movements. Abnormalities in fibroblast growth factors 2 and 3 and also chromosome 6 are described in patients with PSOC.[4]

Complications include secondary osteoarthritis which may result in deformity, femoroacetabular impingement, recurrence, and malignant transformation to chondrosarcoma. Recurrent PSOC predisposes to chondrosarcoma. However, malignant transformation has relatively lesser incidence rate and the median period of such transformation being 20 year following the initial diagnosis.[5] MRI plays a crucial role in distinguishing synovial chondromatosis from chondrosarcoma.

Initial diagnosis of PSOC is made either radiographically or by cross-sectional imaging. Radiographs show multiple or more than five, similarly shaped, intra-articular loose bodies with typical ring, and arc pattern of mineralization.[6] These loose bodies may also show peripheral rim of calcification with the central lucent focus. However, visualization of loose bodies on radiograph depends on the degree of calcification of the chondrocytes. There may be widening of the hip joint space due to joint effusion. Secondary osteoarthritic changes are seen in the chronic or recurrent cases.[7] Ultrasound has not been given much importance for diagnosing the condition, though it may show loose bodies as non-vascular hyperechoic foci with or without posterior acoustic shadowing depending on the amount of mineralization. Thickened and nodular synovium is another important finding on sonography. Extensive mineralization of the loose bodies is a potential pitfall on sonography.[8] However, in diseases such as calcium pyrophosphate deposition and gout, ultrasound is sensitive in detecting the crystals.[9,10] Computed tomography (CT) reveals hypodense non-mineralized synovial thickening and loose bodies with ring and arc or punctate mineralization. It may show enhancement of the vascularized synovium with non-enhancing loose bodies. CT is very useful in depicting the bony anatomy including the articular surface in all three planes aiding in pre-operative planning.[11] Characteristic features on MRI include synovial thickening, intra-articular bodies, and bone erosions. It may appear as homogeneous lobulated T2 hyperintense intra-articular signal with low signal intensity foci. These foci may depict hypointense peripheral rim with central hyperintensity depending on the degree of mineralization. Bone erosions can be deeper as in our case where there was involvement of the acetabulum.[6] If the osteoarthritic changes have already set in, it is difficult to distinguish PSC from secondary synovial chondromatosis.

The two most common imaging differential diagnoses are PVNS and lipoma arborescens. Signal void along the synovial lining and the loose bodies are characteristic

of PVNS. Lipoma arborescens shows fat containing T1 hyperintense frond like synovial projections which suppress on fat saturation sequences.[12]

Loose bodies on MRI may be confused with rice bodies seen in chronic inflammation or infection. Rice bodies are the fibrin bodies of dead synovial tissue and are seen as hypointense on T2-weighted images without any mineralization.[12]

The synovial loose bodies are 2–10 mm in diameter on gross pathology. They contain chalky yellow material due to calcification and/or ossification. On histopathology, the chondrocytes show typical clustered arrangement with mild atypia in PSOC whereas in secondary type, there will be laminar arrangement of the chrondrocytes.[6] Concentric calcification of the hyaline bodies is seen in secondary osteochondromatosis as opposed to typical ring and arc calcification of PSOC.

Treatment of this condition involves removal of loose bodies, synovectomy, and debridement.[1] Partial synovectomy or removal of loose bodies alone may result in recurrence and hence total synovectomy along with removal of the loose bodies is considered as the standard treatment for PSOC, though synovectomy may not be of much use in late phase of the disease. However, PSOC of hip necessitates open surgical approach. This includes surgical dislocation through anterior or posterior approach.[13] In our case, as the synovium was not visualized due to complete destruction by the underlying disease process. The modified Hueter direct anterior approach was applied to fix the femoral head without any surgical dislocation. This approach causes less trauma and short operation time with an advantage of visualization of clear anatomical structures.[14] Secondary osteoarthritic changes in hip osteochondromatosis are less likely post-debridement if the changes are not seen at or before the surgery in the absence of impingement.

CONCLUSION

PSOC is a benign neoplastic process resulting from synovial metaplasia and proliferation. On imaging, intra-articular loose bodies with popcorn or ring and arc calcification are pathognomonic and may not always be present. Clustering of the chondrocytes on microscopy confirms the disease. PSOC is commonly treated by performing synovectomy and removal of loose bodies. However, surgical approach requires modification in few of the cases where there is no synovium left in the joint.

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Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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