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A Clinical Dilemma in the Diagnosis of Primary Synovial Osteochondromatosis of Knee

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Abstract: Primary synovial osteochondromatosis is an uncommon benign condition involving mostly the large joints such as the knee, hip, and elbow characterized by proliferation of the synovium with cartilaginous metaplasia within the bursa, tendon and joints. The disease usually presents in three stages, the first phase is nonspecific with only intraarticular cartilage deposition without loose bodies, which presents with very non specific symptoms and easily missed based on clinical and radiological features. It's relatively easier to diagnose the second and third phase which shows the presence of loose bodies in the joints. We present a case in a 40 year old male, who presented with a history of swelling and pain in the knee joint on and off, the disease diagnosed as early osteoarthritis based on clinical and radiological findings. The purpose of this case report is to document that histopathology is helpful in giving the correct diagnosis and managing the symptoms as the reported evidence of malignancy to synovial chondrosarcoma in the setting of synovial osteochondromatosis is around 6.4%.
Keywords: Primary synovial osteochondromatosis, knee joint, phase one.

INTRODUCTION

Synovial osteochondromatosis was first described by Leannac in 1813[1]. However, its current description was not applied until 1958 by Jaffe [2].

Primary synovial chondromatosis is a very rare monoarticular synovial disease characterized by nodular proliferation of cartilage in the synovial membrane of joints, bursae or tendon sheaths; due to metaplasia of intimal layer of synovium. [3] The majority of intraarticular disease involves the knee (50%), with the hip, elbow & shoulder less commonly affected [4]. Involvement of smaller joints has also been reported, which includes distal radioulnar, tibio fibular, metacarpophalangeal and metatarsophalangeal joint [5]. The majority of patients affected by synovial osteochondromatosis are primarily in the third or fifth decade of life [6]. Primary synovial osteochondromatosis has an incidence of 1:100,000. The primary clinical manifestations of synovial osteochondromatosis are non-specific, including swelling, pain, a palpable mass, tenderness and restricted movement of the joint, which may develop slowly over several years [7]. Due to low incidence & nonspecific symptoms of synovial osteochondromatosis in first phase, diagnosis of this disease may be extremely difficult and it should be considered as one of the differential diagnosis of knee swelling in middle aged individuals.

CASE REPORT

A 40 year old male patient presented at the Orthopaedics OPD at Oxford Medical College, Hospital & Research Centre, Bangalore, with symptoms of swelling and pain of left knee joint on & off since 1year, which aggravated over the period of 2 months. There was no history of antecedent trauma, fever, weight loss and night sweats with no past medical history and he was only taking analgesics for the pain Physical examination revealed local tenderness & swelling over left knee joint, fullness in popliteal fossa with mild restriction of joint movement. Plain X-ray of left knee joint lateral view showed a ill-defined soft tissue haziness (opacity) in the posterior aspect of knee with an eccentric irregular calcification anterosuperiorly

with suprapatellar effusion. (Fig 1) Ultrasonography of left knee joint revealed a well-defined, oval, hypoechoic lesion (2.3 x 1.7 cm) at the posterior aspect of knee (posterior lower thigh level) & the lesion showed patchy foci of vascularity on colour Doppler imaging. (Fig 2) Patient was not responding well with analgesics. Open synovectomy & debridement was planned. Hence, preoperative lab investigations were within normal limits except for mildly elevated ESR & CRP levels. Anterior mid-line incision was given & medial parapatellar approach was done to expose the knee joint. Intraoperatively synovial hypertrophy was noted & subtotal synovectomy along with debridement was performed. The excised tissue was sent for histopathological examination. Postoperative period was uneventful.

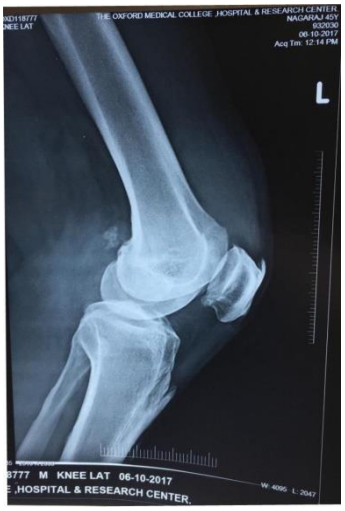

Fig-1: X Ray shows ill-defined soft tissue opacity in the posterior aspect of knee with an irregular calcification

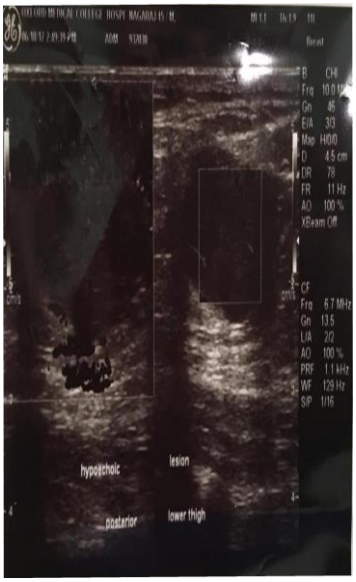

Fig-2: Ultasound showed a well-defined, oval, hypoechoic lesion (2.3 x 1.7 cm) at the posterior aspect of knee

Macroscopic examination of specimen revealed multiple pearly white to membranous soft tissue masses, largest measuring 4 x 3 x 1cm. On sectioning, it was gritty with yellow to white translucent nodular areas. Microscopic examination of haematoxylin & Eosin stained sections showed exuberant synovial hyperplasia along with subepithelial granulation tissue. The nodules were composed of peripheral synovial lining & fibrous tissue surrounding multiple lobules of cartilage along with irregular bony trabeculae. High power view of the same showed lobules of cartilage composed of clusters of mature binucleate to multinucleated benign appearing chondrocytes. Osteoid tissue consists of immature osteoid matrix with osteoblastic rimming with no loose bodies in the synovial tissue. Thus a final diagnosis of primary synovial osteochondromatosis was arrived depending upon histopathology, clinical and radiological findings.

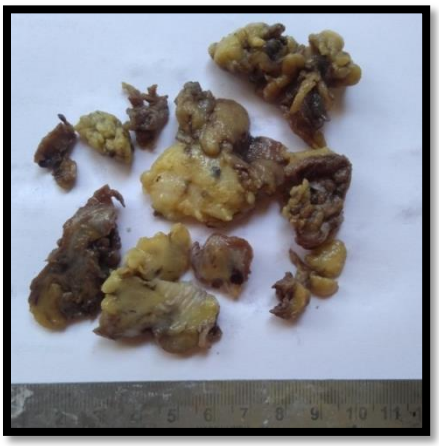

Fig-3: Gross specimen cconsist of multiple grey-white to membrane soft tissue masses

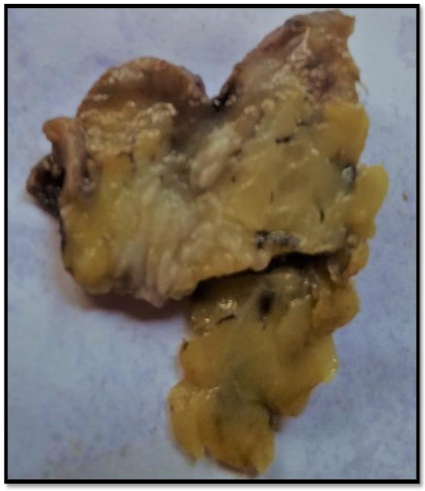

Fig-4: Gross on cut surface Gritty with Membranous & yellowish areas

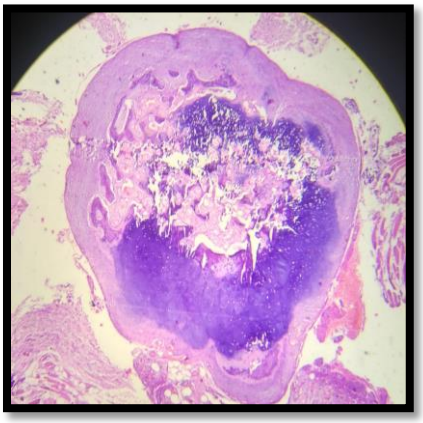

Fig-5: HP: H & E sections showed lobules of cartilage surrounded by osteoblastic rimming

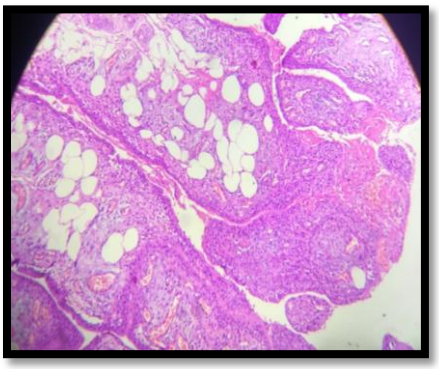

Fig-6: LP: H & E sections showed exuberant synovial hyperplasia along with subepithelial granulation tissue

DISCUSSION

Synovial osteochondromatosis is a reactive metaplastic process of unknown pathogenesis that is characterized by the formation of multiple cartilaginous nodules in synovium, many gets detached & float within joint space [3]. It primarily occurs in the third or fifth decade of life. [6] It is usually a monoarticular disease, although polyarticular involvement has also been reported in up to 5% of cases [8]. The primary clinical manifestations of synovial osteochondromatosis are non-specific, including swelling, pain, a palpable mass, tenderness and restricted movement of the joint, which may develop slowly over several years [7]. In present case, the patient was middle aged with monoarticular involvement and an early and non specific presentation; and insignificant ossification process radiologically resulting in misdiagnosing as synovitis with effusion, secondary to osteoarthritis.

Synovial osteochondromatosis can be primary or secondary. Primary synovial chondromatosis, also known as Reichel syndrome is of unknown origin with no underlying joint disease. But secondary chondromatosis developed following underlying degenerative changes of joint as in osteoarthritis or neuropathic joint [9]. The molecular basis of the disease is unclear though high levels of BMP-2 and BMP-4 have been isolated from diseased synovium and loose bodies. Fibroblastic growth factors 2 and 3 and chromosome 6 abnormalities have been found in cases of primary synovial chondromatosis and may also play a factor in metaplasia. These growth factors and cytogenetic abnormalities have not been found in secondary synovial chondromatosis [10]. Recent studies also proved clonal chromosomal alterations in synovial osteochondromatosis, suggesting that this is a neoplastic lesion rather than a metaplastic &/ or reactive process. Cell proliferation studies of synovial osteochondromatosis have shown proliferative activity that is intermediate between enchondroms and chondrosarcomas [11].

Most of the cases of synovial chondromatosis can be diagnosed clinicoradiologically, depending on extend of ossification & stage of disease. Milligram [12] classified the disease into three phases based on the presence or absence of intrasynovial disease: Phase 1: Early phase, Metaplasia of synovium with active synovitis but no loose bodies. Phase 2: Transitional phase, Active synovitis with formation of loose bodies, which are still cartilaginous Phase 3: Late phase, multiple loose bodies tend to calcify but no intrasynovial disease. The present case was in the initial phase of disease; where radiological and clinical features suggested a diagnosis of osteoarthritis. But the excision biopsy of the lesion proved the lesion to be osteochondromatosis and not the calcium deposition secondary to the degenerative changes of osteoarthritis.

Since osteoarthitis can be, a predisposing factor in secondary chondromatosis as well as a complication in primary synovial chondromatosis; challenging management dilemma arises when confronted with both SC & osteoarthritis [13].

Microscopically, loose bodies in osteoarthritis have characteristic microscopic picture composed of central ossification surrounded by peripheral concentric ring of calcification & cartilage. The other complications of this condition are extension to underlying structures, soft tissue & tendon, bone erosion, local recurrence and also secondary degenerative joint diseases like osteoarthritis with loose bodies in synovial fluid. [7] However, synovial osteochondromatosis may be locally aggressive with a tendency to recur, but has no metastatic potential. Malignant transformation of preexisting primary synovial osteochondromatosis to synovial chondrosarcoma is recognized to be a rare event with the reported incidence of up to 6.4% [14].

The neoplastic conditions to be considered in the differential diagnosis are pigmented villonodular synovitis, synovial hemangioma, and lipoma arborescens are few conditions which can mimic synovial chondromatosis. The other important condition to be considered if synovial hyperplasia is associated with metaplastic proliferation of cartilage is osteochondritis dissecans. In osteochondritis dissecans,

loose bodies are made up of avascular bone with overlying articular cartilage [15].

The most effective treatment is total synovectomy with removal of loose cartilaginous bodies. Following synovectomy, the incidence of recurrence in primary synovial chondromatosis is 23%. [16]. In present case, subtotal synovectomy with debridement was done; follow up after 8 months showed no residual or recurrent lesions.

CONCLUSION

Primary synovial chondromatosis is a very rare monoarticular synovial disease. The initial phase of the disease is difficult to diagnose clinically as well as radiologically; since there are no loose bodies and metaplastic change has already commenced along with intrasynovial disease. Hence, histopathology is mandatory in guiding the orthopaedic surgeons regarding the entity to prevent the misdiagnosis as osteoarthritis and to manage the symptoms as the chances of reoccurrence is very lean. Overall, synovial osteochondromatosis is a benign disease, and prognosis following removal of the nodules is reported as excellent.

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