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IP Archives of Cytology and Histopathology Research

Journal homepage: <https://www.achr.co.in/>

Case Report

Synovial chondromatosis of the foot: A case report with brief review

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ARTICLE INFO

Article history:

Received 07-07-2021

Accepted 11-08-2021

Available online 02-12-2021

Keywords:

Synovial chondromatosis

Cartilaginous nodules

Small joints

Synovectomy

ABSTRACT

Synovial chondromatosis is a rare, benign disorder which commonly involves the large joints and is characterised by the proliferation of cartilaginous nodules beneath the synovial membrane. We report a case of an elderly male who presented with pain and swelling of the right foot since five years. Proper clinico-radiological and histological evaluation of this case led to the diagnosis of synovial chondromatosis which was managed surgically and patient's symptoms were relieved. The rare occurrence of this entity in the small joints of hands and feet and the presence of several close differentials, warrants a thorough clinico-radiological and histological work-up to prevent unnecessary surgical exploration.

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1. Introduction

Synovial chondromatosis is a rare, benign disorder of the joints, tendon sheath or bursae, characterised by proliferation of multiple, small, variably sized metaplastic cartilaginous nodules within the synovial membrane. These nodules may detach from the synovium and form loose bodies in the joint space or they may undergo ossification (synovial osteochondromatosis).^{1,2} Common sites of involvement are large joints like knee, hip, elbow etc. Rarely, small joints of hand and feet may be involved. The index case is a rare presentation of synovial chondromatosis of the foot. This case is discussed to highlight the clinical, radiological and pathological features of this rare entity.

2. Case Report

A sixty-year old male presented to the orthopaedic out-patient department with a dull aching pain and diffuse

swelling of the right foot since five years. The symptoms were insidious in onset and patient had recently developed itching and watery discharge from the skin of foot. Patient had developed gradual restriction of movements of the right foot. No history of trauma was elicited, nor were there any similar complaints in other joints. Patient also complained of low grade fever on and off, which prompted the clinician to the clinical diagnosis of chronic osteomyelitis and conservative management with NSAIDs and antibiotics was started. Plain radiographs were performed which showed multiple calcific foci diffusely scattered around the first metatarsal [Figure 1], suggestive of synovial chondromatosis. Thereafter, surgical resection was planned. Intra-operatively, multiple loose bodies were identified, which were subsequently removed and a synovectomy was performed. The specimen was sent for histo-pathological examination. Grossly, the tissue was received in two pieces which were lobular and yellowish-white in colour, measuring 2.5 x 2 x 1 cm & 1.5 x 0.5 x 0.5 cm. On cut section, glistening, yellowish, gritty areas were noted [Figure 2]. H&E stained microscopic sections showed lobules of hyaline cartilage surrounded by fibro-

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vascular stroma lying beneath the synovial lining. Benign appearing chondrocytes were clustered together along with focal calcification. No chondrocyte atypia, atypical mitosis or areas of necrosis were seen in the section examined [Figure 3]. Hence, the microscopic findings were found to be consistent with the radiological diagnosis of synovial chondromatosis. Patient is doing well on follow-up.

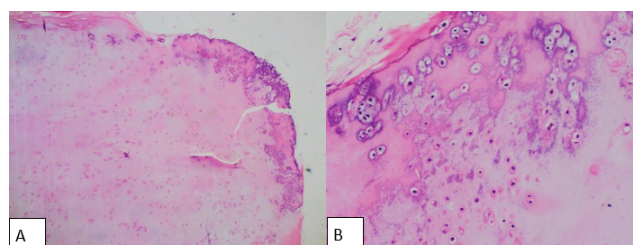


Fig. 3: H&E sections showing A) lobule of hyaline cartilage beneath synovial lining B) chondrocytes clustered together.



Fig. 1: Plain radiograph showing diffuse calcifications around the first metatarsal.

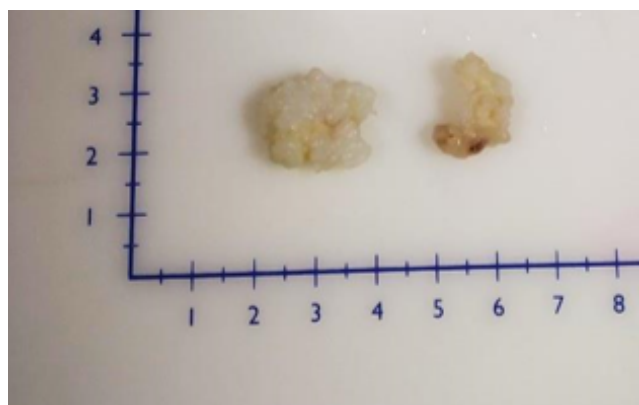


Fig. 2: Synovectomy specimen, grossly showing two yellowish-white, lobular pieces of tissue.

3. Discussion

Known by many names like synovial osteochondromatosis, synovial chondrosis, synoviochondrometaplasia and articular chondrosis, synovial chondromatosis is a disease of large joints like knee (most commonly affected), hip, shoulder and ankle. Less frequently affected joints include temporomandibular, metacarpophalangeal, interphalangeal, proximal tibiofibular and distal radioulnar joints. It is twice more common in males, seen mostly in their fourth to sixth decade of life although female predominance is seen in older age. Insidious in onset, it usually presents with pain, swelling and restricted movements in the affected joint along-with crepitus, diffuse joint swelling, joint tenderness and often palpable nodules on joint examination.^{3,4}

Synovial chondromatosis may arise as a primary disease or may develop secondary to a pre-existing pathology like degenerative disease, infection or trauma. Primary cases are rare and often reported to be mono-articular, although they may involve other joints and extra-articular sites, may recur and are known to be aggressive. Conversely, secondary cases are commoner and non-aggressive. Extra-articular involvement may be tenosynovial or may arise in the bursae, but is quite rare.^{5,6}

Millgram has classified the disease into three phases. Phase one was described as an active intra-synovial disease with absence of loose bodies and no demonstrable calcification on plain radiographs. Phase two was reported to be a temporary, transitional phase where loose bodies were also seen along-with active intra-synovial disease. Lastly, phase three had multiple demonstrable loose bodies which tend to calcify and calcifications could be visualized on plain radiographs but no active intra-synovial disease was present.⁷ The index case presented in its third phase with discernible loose bodies on plain radiographs.

Synovial chondromatosis had been previously thought to arise from over activity of embryonic cell rests. It has largely been considered a metaplastic or a neoplastic process with recent evidence supporting the latter. Hedgehog signalling pathway and some growth factors like fibroblast growth factor 2 and 3 as well as chromosome 6 abnormalities have been demonstrated to have a role in the pathogenesis of primary synovial chondromatosis.⁶

Diagnostic evaluation of synovial chondromatosis requires radiological investigations and histo-pathological confirmation. Radiological detection of calcifications depends on disease stage. The early lesions are unmineralized and appear normal or equivocal on plain radiographs while advanced lesions develop mineralization and in majority of the cases show multiple, similarly shaped calcifications throughout the joint. However, computed tomographic scan and magnetic resonance imaging are considered to be better imaging modalities that delineate the location and extent of lesion in the joint as well as offer better detection of loose bodies, joint effusion and bone erosion. Confirmation of diagnosis is through histo-pathological examination. Macroscopically, the lesion appears as a conglomerate of multiple nodular bodies which are greyish-white to blue in colour and of variable shape and size, embedded within the synovial membrane. On cut section, they are firm in consistency and often show yellowish chalky areas of calcification or ossification. Microscopically, they are seen as multiple, circumscribed, rounded nodules of metaplastic hyaline cartilage underneath an attenuated (and sometimes hyperplastic) synovial lining. These nodules appear hypercellular with chondrocytes clustered together that often show atypical features like nuclear enlargement, hyperchromasia and prominent nucleoli along with occasional mitosis. Some cases may demonstrate areas of calcification and enchondral ossification that proceeds from periphery to centre. Similar findings were also seen in the index case. Malignant transformation to chondrosarcoma is a rare (5%) complication that shows overlapping features with synovial chondromatosis because of cytologic atypia. However, chondrosarcoma is suspected when there is cortical destruction and permeation of the marrow while the circumscribed nature of the lesion and occurrence of organized ossification favour benign disease. Clinical and radiological data also help to make the distinction. Other important differentials to consider are pigmented villonodular synovitis, extra-skeletal chondroma, osteochondritis dissecans and synovial vascular malformation.^{1–3,6–9}

Shearer et al reported bilateral synovial chondromatosis of the ankle with calcific nodules in tendon sheaths in a young male, which was treated conservatively without surgical exploration.¹⁰ Galat and colleagues reviewed eight patients with synovial chondromatosis of the foot and/or ankle with similar histological findings and chief complaints of pain, locking and stiffness. They reported that four patients underwent synovectomy and loose body removal and were rendered pain free on follow up, while three patients underwent below knee amputation, two for transformation to chondrosarcoma and one for multiple recurrences.¹¹ Ryan et al reported a 36 year old male patient with synovial chondromatosis of the first metatarsophalangeal joint who underwent surgical

excision.¹² The index case was a sixty year old male who had a history of pain and swelling of foot since five years. Radiological and histological evaluation rendered the diagnosis of synovial chondromatosis and he underwent surgical removal of loose bodies and synovectomy.

The early phase of disease may be managed conservatively by NSAIDs but the treatment of choice for established disease remains surgical resection with removal of loose bodies and anterior and posterior synovectomy. There is risk of recurrence with primary synovial chondromatosis which depends on completeness of resection and the phase of disease. Newer modalities, like arthroscopic removal of loose bodies, may also be performed, with close follow up for recurrence. A long term complication of surgical management is development of degenerative disease like osteoarthritis. Long standing disease also has a rare chance of malignant transformation. Therefore, long term follow up is warranted.^{4,6}

4. Conclusion

Synovial chondromatosis of the foot is extremely rare. The rarity of its occurrence in small joints of hands and feet and presence of several close differentials, warrant a thorough clinico-radiological and histological work-up to reach to the correct diagnosis and prevent unnecessary surgical exploration. The role of MRI is important to detect first phase of disease. Surgical resection and removal of loose bodies is the treatment of choice. Close follow up is required in view of recurrence and malignant transformation.

5. Conflicts of Interest

The authors declare that there are no conflicts of interest regarding the publication of this paper.

6. Source of Funding

None.

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Cite this article: Shams A, Das A, Sinha M, Shams A. Synovial chondromatosis of the foot: A case report with brief review. *IP Arch Cytol Histopathology Res* 2021;6(4):275-278.