Histopathology of Primary Synovial Osteochondromatosis: A Rare Case Report

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Abstract: Synovial Osteochondromatosis of the knee is an uncommon disease affecting knee joint in middle aged males. We report a case of synovial Osteochondromatosis in the postero medial and lateral corner of the knee; which was diagnosed clinically and confirmed histopathologically. Patient underwent open synovectomy and loose bodies removal. He has been in regular follow up for two years and there is no evidence of recurrence or malignant transformation.

Keywords: Synovial Osteochondromatosis, Knee joint, Loose bodies.

INTRODUCTION
Synovial Osteochondromatosis is a rare metaplastic proliferative disorder of the synovium, characterised by the formation of multiple cartilaginous nodules in the synovium which detach and become loose bodies [1]. It is a benign self-limiting disease of adults, typically affecting the larger joints like knees, shoulders, hips, and elbows in decreasing order of frequency [1,2]. It is more common in males between third to fifth decade [2].

CASE REPORT
Case History: A 40yr old male patient presented with progressive pain and swelling in the left knee joint of two years duration. There was no history of pre-existing joint disease in the past. On physical examination swelling, tenderness and effusion was present in the left knee joint. Range of movements was restricted. Plain X-ray revealed multiple radio opaque round bodies in the left knee joint. Rest of the joints did not show any other changes. CT scan showed multiple conglomerated intraarticular calcified loose bodies in the posteromedial and lateral aspect of the left knee joint. Patient underwent open synovectomy and the tissue was sent for histopathological examination.

Grossly thickened membranous polypoidal grey white tissue fragments m/s 15x9x3cms was present, with an embedded focal bony mass m/s 3x3cms adherent to the synovial membrane (Fig. 2). Multiple spherical, hard, chalky-white loose bodies with irregular surface was also seen. Cut section was gritty grey white in centre and translucent grey white at the periphery.

Microscopy showed hyperplastic synovial tissue lining composed of nodules of cartilage seen embedded within the synovial tissue (Fig. 3). The nodules comprised of chondrocytes of varying cellularity having pyknotic dark - staining nuclei (Fig. 4). Areas of increased cellularity and focal areas of calcification and ossification were also seen (Fig. 5). There was absence of nuclear atypia. A histopathological diagnosis of Synovial Osteochondromatosis was made.

Fig. 1: CT scan showing multiple conglomerated intraarticular calcified loose bodies in the medial and lateral aspect of the left knee joint
DISCUSSION

Primary synovial osteochondromatosis (SOC), also known as synovial chondrometaplasia, is an uncommon entity, characterized by the formation of osteocartilaginous bodies in the synovial membrane [3].

SOC has been classified as primary and secondary. Primary SOC involves otherwise normal joints and secondary SOC includes those cases in which there is underlying degenerative joint disease, osteochondritis dissecans or neuropathic arthropathy [2].

Though aetiology is unknown, irritation due to minor trauma, infection or some chromosomal aberrations have been suggested as possible causative factors [4]. Through histopathologic analysis, Milgram hypothesized that primary synovial osteochondromatosis begins with undifferentiated stem cells in the stratum synovial and fibroblasts forming a primitive chondral matrix [5].

IL-6 and VEGF-A in the synovium is known to produce a vicious circle with synovitis and does not seem to disappear even when the process of cartilaginous metaplasia is completed [3]. The active growth of loose bodies can occur due to nourishment by synovial fluid [6].

Primary synovial osteochondromatosis most commonly occurs in people aged 30 to 40 years and is more prevalent in men. Because of their abundance of synovial tissue, larger joints are more likely to be affected than smaller joints. Knees are the most commonly affected joints, followed by, in no specific order, shoulders, hips, and elbows. The ankle and the joints of the hand are seldomly involved [7] Bursae, tendon sheath and soft tissues may rarely be involved.
Presentation is usually unilateral, but bilateral involvement has also been seen [8]. Symptoms include pain, swelling, loss of motion and locking. The most common physical signs consist of soft tissue swelling, crepitation, palpable loose bodies and limitation of movement [1]. Radiographic findings include multiple radio-opaque loose bodies with osteoarthritis changes, although sometimes normal [6].

Milgram described early, transitional, and late phases in the development of primary synovial osteochondromatosis. In the early phase, synovitis is active, and nodular loose bodies are absent; the transitional phase involves both synovitis and loose bodies; in the late phase, loose bodies are present, and active synovitis is absent. Understanding this progression helps in correlating radiographic imaging and clinical symptoms [5].

Synovial osteochondromatosis appears as numerous glistening blue-gray nodules in the synovial membrane. Nodules range from 2.0 mm to more than 1.0 cm and are firm on cut section. They often demonstrate chalky yellow regions that represent calcification, foci of enchondral ossification, or both [9].

Microscopically, nodules of hypercellular hyaline cartilage are embedded in the synovial connective tissue. Individual nodules are circumscribed and round and demonstrate greater cellularity than articular cartilage. Chondrocytes typically are clustered rather than evenly distributed throughout the matrix. The individual cells exhibit a considerable size range and nuclear chromaticity. Most have pyknotic dark-staining nuclei, but many cells have atypical features, including large nuclei, dispersed chromatin, and nucleoli. Occasional mitoses may be found. The degree of cellularity and nuclear atypia found in synovial chondromatosis in most cases equals or exceeds that seen in low- and intermediate-grade intraosseous chondrosarcoma. Hence, care must be taken to avoid an erroneous diagnosis of malignant neoplasm in this setting [9, 13].

SOC is a non-aggressive condition with a fairly good prognosis. Rare cases of malignant transformation of primary SOC have been documented, the relative risk being 5% according to a study by Davis [11]. Treatment includes synovectomy (open or arthroscopic) with loose body removal. Synovectomy alone without loose body removal leads to local recurrence [12].

Complications of synovial osteochondromatosis can be secondary osteoarthritis, malignant transformation and recurrence. Pigmented villonodular synovitis, synovial hemangioma and lipoma arborescens are few conditions which can mimic synovial chondromatosis. Radiology and histopathology may help to differentiate amongst them [8].

CONCLUSION

Primary Synovial Osteochondromatosis is a rare benign condition involving the synovial lining of joints, synovial sheaths and bursae. One of the most important complications of synovial osteochondromatosis is its malignant transformation to Chondrosarcoma. Hence it is important to diagnose this condition by histopathological study with certainty.

REFERENCES

