



Case Report

Giant synovial Osteochondromatosis – A rare case report

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Abstract

Synovial osteochondromatosis (SOC) is one of the benign conditions which gives rise to intra or extra articular osteochondral loose bodies. Though the exact aetiology is not known, SOC may be an outcome of trauma, avascular necrosis, osteoarthritis, rheumatoid arthritis, osteochondritis dessicans. Usually the disease is monoarticular affecting the knee joint. Treatment of choice of SOC is surgical excision with open or arthroscopic approach. Overall prognosis is good but recurrence is inevitable if removal is incomplete.

Keywords: Synovial osteochondromatosis, Giant osteochondromatosis, Intra articular, Loose bodies, Osteochondromatosis.

Introduction

Synovial osteochondromatosis (SOC) is a benign synovial lesion which is characterized by multiple pearl like intra-articular osteochondral loose bodies. It was first described by Leannec in 1813; however, its current description was put forth by Jaffe in 1958. Though the exact aetiology is not known, SOC may be an outcome of trauma, avascular necrosis, osteoarthritis, rheumatoid arthritis, osteochondritis dessicans.¹ The incidence is double in male population with peak incidence in 5th decade of life.² It can be classified as generalized when the disease is diffused in multiple compartments of the joint and localized when it affects a specific spot. Usually the disease is monoarticular affecting the knee joint.³ Edeiken et al coined the term Giant SOC in 1904 to define synovial chondromas of more than one cm and occasionally reaching up to 20 cm of diameter. The pathological process proceeds with neoplasia of subsynovial

cartilage, hyperplasia in the synovium and the production of round cartilaginous nodules known as chondromas. The nodules tend to grow, nourished by synovial fluid and those that calcify are then termed as osteochondromatosis.⁴ Most commonly Knee joint is involved followed by shoulder, elbow and hip joint.

Case Report

A 75 years old male patient presented with chief complaints of pain over left knee while walking since one year which was gradual in onset and progressive in nature. The pain aggravated since last two months while walking for even a short distance. Patient noticed a mobile mass over medial aspect of left knee since 15 days, he had no history of trauma, fever, weight loss etc. No associated medical conditions like diabetes mellitus, hypertension, tuberculosis, bronchial asthma, epilepsy were noted. Also was not taking any long term medication.

Family history was not significant. Patient had no habits of alcohol consumption, tobacco smoking or chewing.

On clinical examination of left knee, a palpable mobile swelling present over the medial aspect of left knee joint measuring about 6x4 cm, not fixed to the skin, soft tissue or underlying bone (Figure : 1&2). The consistency of the swelling was hard and lobulated, non-tender. Range of motion of knee joint was restricted & painful. Distal sensations and pulsations were normal, there was no compression of adjacent neurovascular

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Conflict of Interest: None

Financial Aid: Nil

structures. Plain radiograph of left knee AP/Lateral view confirmed the presence of giant osteochondromatosis diagnosis (Figure: 2).

He underwent arthroscopic assisted removal of the mass. An 8 cm incision was made over lateral aspect of left knee joint (Figure: 3&4). Six small and one large loose bodies were recovered of various sizes, the largest being the size of 6x4cms(Figure: 4,5&6). A joint lavage was given and postoperatively the patient did well and recovered knee range of movements. The intra operative sample sent for histopathology which confirmed the diagnosis with the microscopic features of osteochondromatosis (Figure:8&9).The presence of Osteocartilagenous nodule suggested that its from synovial in origin (Figure: 10&11).



Figure 1 & 2: clinical image of the swelling in patient from front and side view.



Figure 3 & 4: Antero-posterior and lateral radiographs of left knee joint.



Figure 5, 6 & 7: Gross appearance of the osteochondroma measuring 6x4cm.

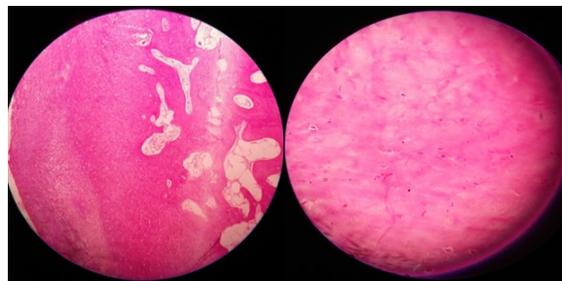


Figure 8 & 9: Microscopic appearance of synovial osteochondromatosis.

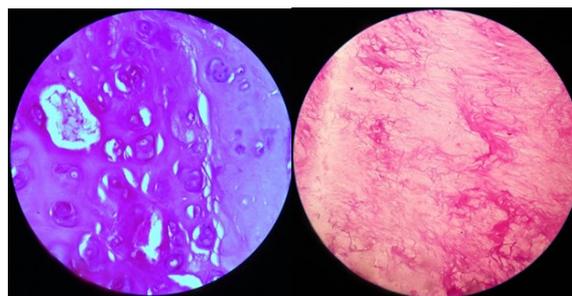


Figure 10 & 11: Microscopic picture showing osteocartilagenous nodule suggestive of synovial origin.

Discussion

Synovial osteochondromatosis (SOC) is a rare benign condition which gives rise to intra or extra articular osteochondral loose bodies. Synovial osteochondromatosis is also called as Reichel's syndrome. Theory behind this pathology is reactivation of residual embryonic cells, traumatic initiation and benign neoplastic transformation have been advocated in causation. Primarily the pathology proceeds as subsynovial cartilage neoplasia, hyperplasia of subsynovium and the formation of round cartilaginous nodules known as chondromas. These chondromas tend to further grow and calcify to form osteochondroma.⁵

Edeiken et al coined the term Giant SOC in 1904 for the synovial chondromas of diameter 1 cm up to 20 cm. The incidence is twice common in males than females with peak incidence in 5th decade of life.⁶⁻⁸ SOC is usually monoarthritic. Knee joint is involved in 60- 70% of cases which in view of incidence is followed by shoulder, elbow and hip. Two forms of Synovial osteochondromatosis have typical histological differences. In primary lesions, cartilage nodules and loose bodies manifest as irregular nests, usually with binucleated cells formed by sporadic diffuse calcification and metaplastic cartilage. By

contrast, in secondary lesions, calcifications are band like with uniform, evenly distributed chondrocytes; furthermore, fragments of subchondral bone or articular cartilage may be present at the centre of loose bodies.⁹ The primary Synovial osteochondromatosis is associated with the benign reactive metaplasia of the synovial membrane whereas secondary SOC is associated with the setting of pre-existing joint pathology such as trauma, osteonecrosis, tuberculosis, rheumatoid arthritis, osteoarthritis, synovitis, osteochondritis desiccans and neuropathic arthropathy.¹⁰

Milgram et al established staging system by suggesting three phased evolution of SOC. Phase I-active intra-synovial lesion without loose bodies. Phase II - Active intra-synovial pathological tissue mixed with loose bodies. Phase III- Is with multiple free osteochondral bodies with no synovial disease process.¹¹⁻¹³ Extra-articular (bursa or tendon sheath) involvement is extremely rare.

A recent study showed role of TGF (Transforming growth factor) and TN (Tenascin) in synovial chondromatosis. TGF increases the differentiation of mesenchymal cells, the production of proteoglycans, the replication of chondroblasts and the stimulation of extra-cellular matrix production. Tenascin is important for chondrogenesis and transformation of cartilage to bone in extra cellular matrix. Another study revealed FGFR-3, FGF-9, a specific ligand of FGFR-3 have been found in synovial fluid of synovial osteochondroma. These are absent from normal synovium and cartilage and may explain the pathogenesis of Synovial osteochondromatosis.¹⁴

Clinical picture of SOC is subtle - tenderness with reduced range of motion and palpable mass are often found. The SOC diagnosis is mainly depends on concurrent clinical, radio graphical and histo-pathological findings and exclusion of other conditions. Imaging plays an important role in diagnosis of Synovial osteochondromatosis, with calcification present in standard radiographic examination in 70-95% of cases. Immunohistochemistry shows markers to differentiate primary Synovial osteochondromatosis and low grade chondrosarcoma but, certain studies have shown no definitive immunostaining as the gold standard technique.¹⁵

Multiple smooth, round, calcified bodies of variable size are found within the joint capsule are findings with diagnostic significance. Nevertheless radiographs can only reveal increased soft tissue density affected joint. Thus calcification or ossification, MRI is particularly useful.

Differential diagnoses of SOC includes chronic joint infection, degenerative condition like osteoarthritis, inflammatory conditions like-

monoarthritis, pigmented villonodular synovitis, juxta-articular neoplasms like lipoma, meniscal cyst, ganglion, malignant fibrous histiocytoma, synovial haemangioma and synovial sarcoma.

Complications of Synovial osteochondromatosis may be osteoarthritis, recurrence and malignancy. With a potential risk of transformation to malignancy is 5%, Synovial osteochondromatosis is an important differential diagnosis of prognostic importance. Shearer et al, 2007 found that difference between the two conditions may be difficult because of the similar clinical and radiological appearances.¹⁶ A cell proliferative activity study concluded that, primary Synovial osteochondromatosis appeared to occupy a position which is intermediate between benign chondroma and malignant chondrosarcoma which may explain the occasionally aggressive clinical behaviour. Synovial osteochondromatosis typically presents with irregular contours, clumping calcifications and bony destructions. Permeative and destructive margins than erosive margins with adjacent marrow invasion suggest malignancy. Sudden clinical deterioration in long standing cases, bony destruction by imaging study or cases with recurrent synovial chondromatosis should alert the clinician to the possibility of malignant transformation. According to Maurice et al the recurrence rate is 11.5% but can be as high as 23%.¹⁷

Treatment of choice of SC is surgical excision with open or arthroscopic approach. Two techniques have been suggested, the first being excision of nodule only. The second being excision associated with extensive synovectomy. Though Church et al reported that synovectomy does not guarantee success. Beside surgical intervention, a study from Chong et al. demonstrated the effectiveness of post-operative radiotherapy for the treatment of recurrent Synovial osteochondromatosis of the knee implies that radiotherapy might be a solution for knee Synovial osteochondromatosis. But still studies with higher level of evidence are needed to support these hypothesis. Overall prognosis is good but recurrence is inevitable if removal is incomplete.

References

1. Fornaciari P, Schai P, Niehaus R, Exner U. Intra Articular Giant Synovial Osteochondroma: Case Reports of the Ankle and Knee Joint. *Case Rep Orthop* 2015;2015:320139. doi: 10.1155/2015/320139.
2. Sedeek S, Choudry Q, Garg S. Synovial Chondromatosis of the Ankle Joint: Clinical, Radiological and Intraoperative Findings. *Case Rep Orthop* 2015; 2015:359024. doi: 10.1155/2015/359024.

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3. Chung JW, Lee SH, Han SB, Hwang HJ, Lee DH. A synovial osteochondroma replacing the anterior cruciate ligament at the intercondylar notch. *Orthopedics* 2011; 34(2):136. doi: 10.3928/01477447-20101221-26.
4. Tahmasebi MN, Bashti K, Sobhan M, Ghorbani G. Bilateral Synovial Knee Chondromatosis in a Patient with Rheumatoid Arthritis: Case-report and Literature Review. *Arch Bone Jt Surg* 2014; 2(4):260-4.
5. Giannetti S, Santucci A, Patricola A, Stancati A, Di Sanzo V. Neglected synovial osteochondromatosis of the elbow: a rare case. *World J Surg Oncol* 2013;11:233. doi: 10.1186/1477-7819-11-233.
6. Alaqeel MA, Al-Ahaideb A. Synovial osteochondroma originating from the synovium of the anterior cruciate ligament. *BMJ Case Rep* 2013;2013:bcr2012008218. doi: 10.1136/bcr-2012-008218.
7. Jiménez-Martín A, Zurera-Carmona M, Santos-Yubero F, Pérez-Hidalgo S. Arthroscopic Treatment of Synovial Chondromatosis, an Unusual Cause of Shoulder Pain. *Reumatol Clin* 2014;10(6):416-7. doi: 10.1016/j.reuma.2013.12.008.
8. Kukreja S. A Case Report of Synovial Chondromatosis of the Knee Joint arising from the Marginal Synovium. *J Orthop Case Rep* 2013 ;3(1):7-10.
9. Yu X, Li W, Dai M, Zhang B, Zou F, Liu H. Giant extra-articular synovial osteochondromatosis of the left proximal thigh: A case report. *Oncol Lett* 2015;10(6):3577-80. doi: 10.3892/ol.2015.3746
10. Lui T. Giant Solitary Synovial Osteochondroma of the Subtalar Joint. *The J Foot Ankle Surg* 2016 ;55(1):183-7. doi: 10.1053/j.jfas.2015.02.003.
11. Jeffreys T. Synovial Chondromatosis. *J Bone Joint Surg Br* 1967;49(3):530-4..
12. Maurice H, Crone M, Watt I. Synovial chondromatosis. *J Bone Joint Surg Br* 1988;70(5):807-11. doi: 10.1302/0301-620X.70B5.3192585.
13. Mackenzie H, Gulati V, Tross S. A rare case of a swollen knee due to disseminated synovial chondromatosis: a case report. *J Med Case Reports* 2010;4(1):113. doi:10.1186/1752-1947-4-113
14. Chen CY, Chen AC, Chang YH, Fu TS, Lee MS. Synovial chondromatosis of the hip: management with arthroscopy-assisted synovectomy and removal of loose bodies: report of two cases. *Chang Gung Med J* 2003;26(3):208-14.
15. Wei J, Wu XT. Disseminated Knee Synovial Chondromatosis treated by Arthroscopy and combined Anterior and Posterior Approaches. *J Clin Exp Orthop* 2015;1(1):1:5. doi: 10.4172/2471-8416.100005
16. Evans S, Boffano M, Chaudhry S, Jeys L, Grimer R. Synovial chondrosarcoma arising in synovial chondromatosis. *Sarcoma* 2014;2014:647939. doi: 10.1155/2014/647939.
17. Jamshidi K, Barbuto R, Shirazi MR, Abolghasemian M. Giant Solitary Synovial Chondromatosis Mimicking Chondrosarcoma : Report of a Rare Histologic Presentation and Literature Review. *Am J Orthop* 2015 ;44(8):E286-90.