

# Failure of arthroscopic removal of loose bodies in knee mandating synovectomy, later diagnosed histopathologically as synovial chondromatosis (Reichel-Jones-Henderson syndrome)

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## Abstract

Synovial chondromatosis is a rare, idiopathic, highly destructive and aggressive, yet generally benign condition, mostly monoarticular, commonly involving the synovial membranes of large joints, most commonly the knee joint, with preponderance in males of 4th to 6th decade. Presence of multiple cartilaginous nodules and loose bodies in the synovium or cavity are characteristic. We present a case of 38 year old male with swollen stiff right knee joint for 2 years, presenting with sudden locking sensation. After clinical and radiological assessment, diagnostic arthroscopy was planned which was converted into arthrotomy and complete synovectomy, in lieu of large loose bodies, histopathological examination of which confirmed the diagnosis.

**Keywords:** synovial chondromatosis, loose bodies, knee joint, synovectomy

## Introduction:

Primary synovial chondromatosis[1] is an uncommon benign condition of unknown etiology. As a rule, the disease is monoarticular in origin characterized by cartilaginous nodules involving the synovial lining of joints, synovial sheaths and bursae. This metaplastic process of synovium converts it into cartilage and gets detached to become a loose body[2]. Though smaller joints and bursae[3,4] may be affected, large joints like knee, hip, shoulder, wrist, ankle are the common sites[5,6,7]. Typical presentation is 4th to 6th decade (M:F ratio of 2:1 to 4:1). It is rare in childhood with only a few reports in literature. Presentation is mainly painful and swollen joint with

variable degree of stiffness. Surgical management is the chief plan of treatment which may be arthroscopic or open. Large loose bodies may mandate arthrotomy and synovectomy in case of failure by arthroscopic means.

## Case report:

Here we present a case of 38 years old male with history of insidious onset swelling and pain with stiffness in right knee joint. Pain was aggravated by physical activities and relieved by rest. In lieu of these complaints patient presented to our institute, and underwent joint aspiration for suspected collection. The aspirate was documented to be approximately 50 ml of yellowish clear viscous synovial fluid without any evidence of suspended particles or bloody tinge. Routine and microscopic examination of the aspirate was found out to be unremarkable. Microbiological profile of the fluid was unyielding. Then,

patient became symptom free for approximately 2 years except an occasional grating sensation while walking and a persistent mild swelling. Patient had an episode of sudden locking of the knee joint on standing from sitting position, with severely restricted joint movement. The patient presented to our institute again with generalized swelling of the knee, with fullness in popliteal fossa, with no antecedent history of trauma, or history suggestive inflammatory or infectious etiology. There was a diffuse swelling and tenderness all around the knee without any redness or local rise of temperature, medial joint line being the tendermost site. Instability tests were negative, and the distal neurovascular status was normal on examination. RA and other seronegative arthritis including acute phase reactants were assessed and were found to be negative. X Rays (figure 1) show stippled periarticular calcification and multiple intra-articular chondral bodies with

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© Authors | Journal of Clinical Orthopaedics | Available on [www.jcorth.com](http://www.jcorth.com) | doi:10.13107/jcorth.2456-6993.2017.443

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**Fig.1:**stippled periarticular calcification and multiple intra-articular chondral bodies with “ring-and-arc” chondroid mineralization suggestive of loose bodies

“ring-and-arc” chondroid mineralization suggestive of loose bodies. MRI (figure 2,3,4) findings are suggestive of osseous loose bodies posterior to the posterior horn of medial meniscus and in the anterior intercondylar notch with osteoarthritic changes in the adjacent articular surfaces, and suprapatellar bursitis[ 11]. Patient was planned for a diagnostic arthroscopy. Diffuse irregular nodular outgrowths were found all around the synovial margin with many large loose bodies (Figure 5) , the largest one measuring approximately 4 × 2.5 cm in the intercondylar notch. Initially the nodular bodies were removed piecemeal but due to large loose bodies, the joint was opened by medial parapatellar approach through an anterior midline incision. The synovium with its adherent nodular bodies was stripped in its all extent and sent for histopathological examination (figure 6) .Wash was given and closure

was done in layers. Postoperatively, patient was instructed about protected weight bearing, knee mobilization and muscle strengthening exercises. Patient’s disease was classified as phase 2 according to milgram’s staging. The patient was followed up regularly. Patient’s range of movement was 120. without pain at 2 months postoperatively and there were no newer complaints or any evidence of recurrence.

**Discussion**

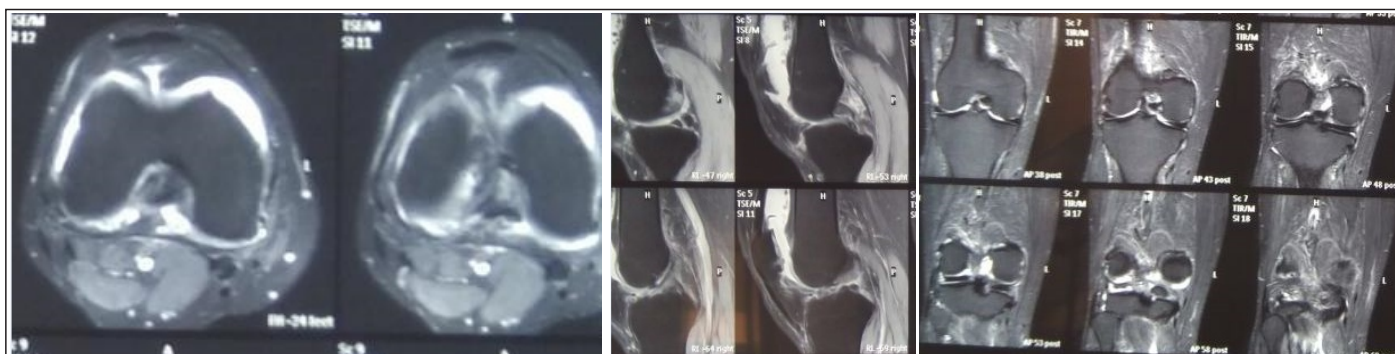
Normal synovial membrane contains no cartilage cells, but metaplasia of the synovial lining cells into chondrocytes, generates foci of cartilage in synovium resulting in this situation, called Primary chondromatosis, also called articular chondrosis, synovial osteochondromatosis, synoviochondrometaplasia, synovial chondrosis, which is a rare entity characterised by cartilaginous bodies forming within the synovium with subsynovial connective tissues. In addition to the explained theory of metaplastic transformation, another explanation for the appearance of masses of cartilage and bone, in the synovial lining lies in the well known phagocytic ability of the lining membrane, which absorbs detritus of cartilage and bone from degenerated articular surface of an osteoarthritic joint. These fragments however lack the round or oval appearance of chondromatous foci, and lack the

staining characteristics of normal cartilage and bone and lack sharp demarcation from the surrounding synovium. Milgram described 3 phases of the disease[8].

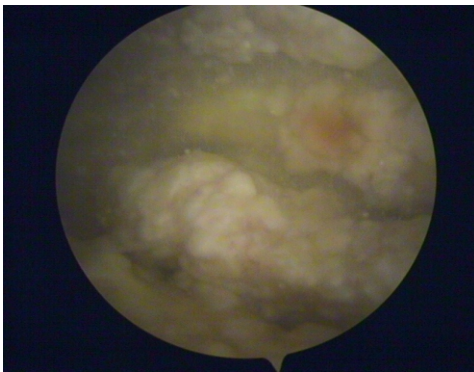
**Phase I :** (active intrasynovial phase) - cartilaginous metaplasia of the synovial intimal cells occurs with trauma which is considered to be an inciting stimulus. Active synovitis and nodule formation is present without any evidence of calcification.

**Phase II :** (Transitional lesion phase.) Nodular synovitis and loose bodies which are still cartilaginous.

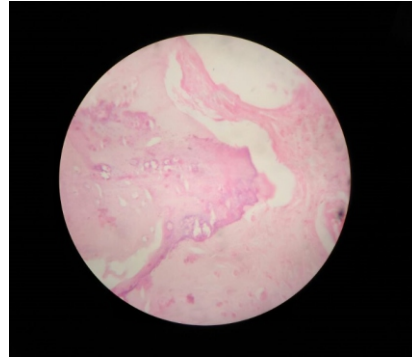
**Phase III:** (quiescent/inactive intrasynovial phase) Loose bodies remain but synovitis has resolved. Metaplastic growth of the synovium is present only in first two phases while loose bodies are present in the later two phases. However, on the basis of the currently known molecular abnormalities, Chromosome 6 abnormalities which are identified by the cytogenetic and molecular cytogenetic analyses, have been a recurrent finding in primary synovial chondromatosis. Multiple cartilaginous nodules, are commonly formed. The formation of loose bodies can be explained by the expelled out chondrocytes which have been pedunculated and encrusted inside the synovium, growth, and calcification of which may continue by the synovial fluid in nearly 60% of the cases. This disease is commonly seen in the 4rd to 6th decades of life, with a male to female



**Fig. 2,3,4:**MRI Shows osseous loose bodies posterior to the posterior horn of middle meniscus and in the anterior intercondylar notch with osteoarthritic changes in the adjacent articular surfaces, and suprapatellar bursitis



**Fig.5:** Diffuse irregular nodular outgrowths were found all around the synovial margin



**Fig.6:** Histopathological Examination

ratio of 2:1 [9]. The onset is described as insidious and it occurs over months to years, mostly monoarticular, has a preponderance specifically for the knee joint, yet other sites like tendon and bursa are not uncommon. The primary synovial chondromatosis occurs in an otherwise normal joint, revealing undifferentiated stem cell proliferation in the stratum synoviale [19], which is a progressive and recurrent condition resulting in degenerative arthritis at the end. Secondary synovial chondromatosis is, mostly non-recurrent, thought to be caused by the irritation of the synovial tissue, by either of degenerative joint disease, trauma, inflammatory and noninflammatory arthropathies, avascular necrosis, and osteochondritis dissecans. Before the diagnostic confirmation is made the patients are generally planned for diagnostic arthroscopy in which loose bodies are found with multiple subsynovial nodules. Arthroscopic removal of these lesions may fail, necessitating arthrotomy for complete synovectomy as well as removal of some of the large loose bodies. Thus, in majority of the cases, arthrotomy and complete synovectomy is mandatory.

The tissue derived is sent for histopathological examination, which confirms the diagnosis. In minimally involved cases, the mere removal of suspected lesions arthroscopically, in which the histopathological diagnosis comes out to be synovial chondromatosis requires a revisional surgery for complete synovectomy. Though macroscopically normal, some areas of synovial membrane may show typical microscopic changes, failure to remove these areas may lead to future recurrences. The X Rays are only helpful in the third phase of the disease, once the calcification has occurred revealing radiopaque, round, or oval, loose bodies within the joint [10]. There are only a few cases of malignant transformation. Surgical excision is the treatment of choice [17], as radiotherapy and chemotherapy have no effect on synovial chondromatosis. In asymptomatic patients, the nodules may resorb over time and invasive procedures should be avoided in localized intra articular disease, the complete excision of the abnormal synovium is curative. In the phase III disease, the removal of the loose bodies alone is sufficient, with recurrence rate

approximately 10 to 25%. Complications like secondary degenerative osteoarthritis due to chronic mechanical irritation and bone destruction by the loose bodies is the rule. Surgery predisposes the patients to tissue scarring, subsequently compromising the joint function. Recurrence [13,14,15,16] is also a known complication. Chondrosarcoma arising from pre-existing synovial chondromatosis is an extremely rare event with prevalence rate of only 5%. The differential diagnoses of primary synovial osteochondromatosis include pigmented villonodular synovitis, lipoma arborescens, secondary synovial osteochondromatosis, rheumatoid, or other seronegative arthritis, septic arthritis which includes granulomatous infections, synovial hemangioma, synovial chondrosarcoma and osteochondromas with adjacent secondary bursal osteochondromatosis. Blood tests and the arthritis profiles can also help in ruling out the specific differential diagnoses.

### Conclusion:

Primary intra-articular synovial chondromatosis is an uncommon, highly destructive and aggressive disease. Misdiagnosis and unwarranted surgery may result due to lack of awareness of this condition. As radiographic or MRI findings are not of much help, histopathology is must for the confirmation. Removal of loose bodies with stripping of and complete excision of the synovium remain the mainstay of treatment, but secondary osteoarthritis and recurrences are not uncommon

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**Conflict of Interest: NIL**  
**Source of Support: NIL**

#### How to Cite this Article

Bhavsar NM, Desai B, Trivedi A, Amin P, Patel P R. Failure of arthroscopic removal of loose bodies in knee mandating synovectomy, later diagnosed histopathologically as synovial chondromatosis (Reichel-Jones-Henderson syndrome). *Journal of Clinical Orthopaedics* Jan-June 2018; 3(1):25-28