



## Synovial Chondromatosis of Knee

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### ABSTRACT

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Synovial chondromatosis (also called synovial osteochondromatosis) is a type of non-cancerous tumor that arises in the lining of a joint. It has been divided into primary and secondary forms. Primary synovial chondromatosis was originally considered to represent chondroid metaplasia in the synovium of a joint with resultant formation of multiple intraarticular chondral bodies. Secondary synovial chondromatosis is associated with joint abnormalities, such as mechanical or arthritic conditions, that cause intraarticular chondral bodies. Primary synovial chondromatosis typically affects adults, predominantly men, in the third to fifth decades of life. Synovial osteochondromatosis manifests clinically with joint pain, swelling, and limitation of motion. Although the condition is not cancerous, it can severely damage the affected joint and, eventually, lead to osteoarthritis. As conclusion, this case is typical of secondary synovial chondromatosis that is the result of a degenerative change in the joint.

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### Introduction

The exact prevalence of synovial chondromatosis is unknown, but the disorder is rare worldwide. Most reported series indicate a male-to-female ratio of 2:1. In addition, most cases are reported in patients aged 20-50 years; only a few case reports have described the condition occurring in children. Synovial chondromatosis usually affects the larger diarthroidal joints of the axial skeleton, typically the knee 35%, elbow 22%, wrist 11% and hip 4% (Jeyaraj and Vineet, 2017).

Synovial chondromatosis is a benign monoarticular disorder condition that involves the synovium, which is the thin layer of tissue that lines the joints. Synovial osteochondromatosis is an uncertain cause characterized by proliferation and metaplastic transformation of the synovium with formation of multiple cartilaginous nodules. Synovial chondromatosis is a rare condition in which foci of cartilage develop in the synovial membrane of joints, bursae, or tendon sheaths as a result of metaplasia of the subsynovial connective tissue. These ectopic foci of cartilage can result

in painful joint effusions and, on the generation of loose bodies, mechanical symptoms. The synovial lining of a joint, bursa, or tendon sheath undergoes nodular proliferation, and fragments may break off from the synovial surface into the joint. There, nourished by synovial fluid, the fragments may grow, calcify, or ossify. The intra-articular fragment may vary in size from a few millimeters to a few centimeters.

Plain radiographs frequently show characteristic feature including multiple (usually >5) calcified or osseous bodies within the joint or bursa. When fragments are not calcified, intrasynovial fragments may not be seen on plain images, and arthrographic studies are required to demonstrate the bodies. The differential diagnosis includes degenerative joint disease in which osteophytes have broken off into the joint. However, synovial chondromatosis tends to have a larger number of bodies in the joint. Other differential diagnoses include soft tissue and intra-articular chondromas. In advanced stages of synovial chondromatosis, secondary degenerative changes are often observed. For early diagnosis, plain radiographs help the doctor to make next examination and treatment plan.

### **Case Presentation**

We present a 52-years-old woman with main complain of pain dan swelling at right knee 1 day before. No traumatic history. Blood pressure was 160/100 and pulse was 80 x / mnt, temperature 40 C. Plain x-ray examination revealed irregular amorph cartilage lesion in bursal tissue and/or in tenosynovial tissue in proximity to an involved join. It was also revealed sclerotic and irregular facies articular, also narrow joint space.

Physical examination will be looking for swelling, tenderness, limited range of motion, and creaking or grinding noises during movement, an indication of bone-on-bone friction. Imaging studies will help differentiate synovial chondromatosis from osteoarthritis. X-rays provide images of dense structures, such as bone. Large loose bodies are usually calcified or ossified and can be seen on x-ray. Smaller loose bodies and those that are not calcified or ossified may not show up. If the loose bodies are not visible on x-ray, the doctor may suggest a magnetic resonance imaging (MRI) scan or computerized tomography (CT) scan to better evaluate the joint. Loose bodies can typically be seen on both MRI and CT scans.

Depending on the symptoms, simple observation can sometimes be a treatment option. The doctor will carefully consider a number of factors in determining whether observation is appropriate in your case. Treatment for synovial chondromatosis typically involves surgery to remove the lose bodies of cartilage. In some cases, the synovium is also partially or fully removed (synovectomy) during surgery.



Figure 1. Multiple opacities appear on the soft tissue on the cranial aspect of the patella dextra, size 1-2 m, rounded shape, irregular edges. Multiple osteophytes are seen in the osseous genu dextra system. Joint space narrows. Irregular articular facies. Oseous genu dextra with multiple opacities of calcification in the soft tissue region of the cranial patella dextra leading to synovial chondromatosis type 2 genu dextra.

Surgery can be done using either an open procedure or an arthroscopic procedure. In a traditional open procedure, the doctor will usually make one or two large incisions. In an arthroscopic procedure, the doctor will make smaller incision and use miniature surgical tools to remove the loose bodies. The end results of both open and arthroscopic procedures are the same. Synovial chondromatosis may return in up to 20 percent of patients. For a period of time after surgery, the doctor will schedule regular follow-up to visits to check for any recurrence.

## Discussion

This study reports a case of synovial chondromatosis is unknown. Some research suggests that trauma may play a role in its development because the condition primarily occurs in weight-bearing joints. Infection has also been considered as a contributing factor. The condition is not inherited.<sup>1</sup> Synovial chondromatosis is a benign condition that can result in severe disability and dysfunction of an involved synovial joint. Observation of involved cases indicates that this benign condition rarely undergoes malignant degeneration (Olufemi, 2020; Health Jade Team, 2019).

Case reports have described the coexistence of chondrosarcoma and synovial chondromatosis, sparking debate as to whether the chondromatosis is a cause or the result of chondrosarcoma. Only a few case reports have documented malignant degeneration of synovial chondromatosis. Synovial joints are differentiated from cartilaginous and fibrous joints by the existence of capsules surrounding the articulating surfaces of the joint and the presence of lubricating synovial fluid within the capsule. The synovium is a thin, specialized tissue that lines noncartilaginous spaces within the joints (e.g., hips, knees, elbows). This tissue is a few cell layers thick and plays a major role in regulating the fluid and cellular environment within the joint. A highly vascular capillary system within the synovium provides joint lubricating fluid containing hyaluronan and lubricin.

The synovium mediates molecular and cellular changes within the joint and protects the joint from physiologic and biomechanical stresses. Additionally, the synovium provides macrophage access to the joint. Activated macrophages perform several immunologic functions within the joint space and are important mediators of both joint homeostasis and certain disease states.

The most common disorders include synovial chondromatosis and pigmented villonodular synovitis (PVNS). Both of these conditions are benign tumors of synovial origin. Failed treatment and recurrent disease can be associated with articular cartilage degeneration and secondary osteoarthritis. These joints usually present as having a large effusion and sometimes appearing deformed due to swelling or synovial hypertrophy. They are painful at rest and are more painful with motion. The range of motion is usually decreased, which often progresses slowly for several years (Olufemi, 2020; Health Jade Team 2019) Synovial chondromatosis begin as small nodules of cartilage. These nodules can separate and become loose within the joint. Some tumors may be no larger than a grain of rice.

The disease process begins with metaplastic differentiation of mesenchymal cells in the synovial membrane of the joint. These cells mature into chondroblasts and form small nodules of cartilage in intimal layers of the synovial membrane. These nodules subsequently enlarge and detach to lie within the joint space, resulting in the formation of multiple intracapsular and extracapsular loose bodies. These loose bodies continue to grow in diameter via multiplying chondrocytes and may calcify in their central zones, leading to the term osteochondromatosis. Milgram further described synovial chondromatosis as a self-limited intrasynovial process that occurs in three phases: early, transitional, and late. (Bacher, NB, 2020). In the early phase, only active intrasynovial disease is present, with no loose bodies. The transitional phase includes both active intrasynovial proliferation and free loose bodies. The late phase shows multiple free osteochondral bodies extruded in the joint space but no demonstrable intrasynovial disease. These phases become important in clinical decision making in determining the necessity of performing synovectomy to treat symptoms and prevent disease recurrence. (Baecher NB, 2020)

Synovial chondromatosis occurs as either a primary or secondary form. Although the molecular basis is still unclear, high levels of BMP-2 and BMP-4 have been isolated from diseased synovium and free bodies. These growth factors may be involved in the pathologic metaplasia observed in synovial chondromatosis. Primary synovial chondromatosis also referred to as Reichel syndrome or Reichel-Jones-Henderson syndrome. The primary and secondary forms have different presentations, and as such are treated different (Health Jade Team,2019). Primary synovial chondromatosis, which is rarer, occurs spontaneously and does not appear to relate to any pre-existing conditions.

The typical history of a patient with primary synovial chondromatosis of the knee is that of a

middle-aged man with monoarticular pain, swelling, and stiffness with or without mechanical symptoms in the knee. No history of acute trauma is usually reported, but the patient may have a distant history of knee injury. No systemic signs of infection or illness are apparent (Baecher NB,2020). Primary synovial chondromatosis represents as a benign neoplastic process with hyaline cartilage nodules in the subsynovial tissue of a joint, tendon sheath, or bursa and as loose bodies in the joint cavity with or without calcification and without an identifiable joint pathology (Murphey 2007, Baecher NB, 2020) The nodules may enlarge and detach from the synovium. The knee, followed by the hip, in male adults are the most commonly involved sites and patient population. The pathologic appearance may simulate chondrosarcoma because of significant histologic atypia, and radiologic correlation to localize the process as synovially based is vital for correct diagnosis (Murphey, 2007). Secondary synovial osteochondromatosis is seen with coexistent osteoarthrosis. A general distinction in the secondary form is multiple bodies of differing sizes with concentric rings of growth.

The primary form of synovial chondromatosis is characterized by numerous small, round loose bodies that are uniform in size. It is not precipitated by any identifiable joint pathology and likely occurs secondary to metaplasia. Lesions are often aggressive and are associated with a high incidence of recurrence (Olufemi, 2020; Dutt, 2020). Secondary form (occurring in the setting of joint degeneration) is made in the literature. The secondary form occurs more common and usually, in older individuals (Health Jade Team,2019)

In rare cases, an extra-articular form of the condition can be identified. In these cases, lesions are found to be in bursal tissue and/or in tenosynovial tissue in proximity to an involved joint. This number varies anywhere from two or three loose bodies to several dozens. They present in varied sizes and several loose bodies may combine to form larger bodies. The number of intraarticular lesions is greater in the primary type than the secondary type The loose bodies may create pathological mechanical wear on joint surfaces and result in various types of erosion of articular surfaces (Health Jade Team, 2019)

The disease progresses from an active initial phase, with synovial proliferation and formation of intrasynovial cartilaginous nodules, to a final phase, which is characterized by inactive synovial disease and persistent nodules, which may break off into the joint space (Baecher, NB, 2020). The nodules may contain unmineralized cartilage alone (synovial chondromatosis), cartilage and bone, or mature bone with fatty marrow. Calcification is absent in approximately 25%–30% of patients. Treatment is surgical synovectomy; however, the recurrence rate is over 25% (Health Jade Team, 2019).

When calcified bodies are present, the radiographic findings of synovial osteochondromatosis are pathognomonic, consisting of multiple intraarticular calcified nodules, characteristically

uniform in size. Synovial osteochondromatosis may be associated with a small joint effusion, marginal erosions, and late secondary degenerative joint disease. Conversely, the secondary form is characterized by fewer, larger, more variably sized cartilaginous masses. This form is more likely to occur in the setting of preexistent osteoarthritis, rheumatoid arthritis, osteonecrosis, osteochondritis dissecans, neuropathic osteoarthopathy, tuberculosis, or osteochondral fracture. The underlying disease process generates chondral fragments that implant into synovium, inducing metaplastic cartilage formation.

Primary synovial chondromatosis is a self-limiting benign neoplastic. Three phases of articular disease have been identified:

1. Initial phase: metaplastic formation of cartilaginous nodules in the synovium
2. Transitional phase: detachment of those nodules and formation of free intra-articular bodies
3. Inactive phase: resolution of synovial proliferation, but loose bodies remain in the joint, and may increase in size obtaining nourishment from the joint fluid by diffusion (Olufemi, 2020; Dutt, 2020)

## Conclusion

Radiographic findings enable differentiation of primary from secondary types. If radiographs indicate no underlying joint pathology, the primary type can be diagnosed. Secondary synovial chondromatosis is felt to occur as a result of mechanical changes in a joint due to arthropathy. The formation of loose chondral bodies is thought to be part of the degenerative process in these joints. MRI and MRI arthrography are also helpful in making the diagnosis. MRI can help differentiate and diagnose bursal extension of the disease process. An evaluation of patients with findings that suggest secondary synovial chondromatosis should include an attempt to identify underlying arthritic processes.

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