

Etiology of a painful knee: Primary synovial chondromatosis

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ABSTRACT

Primary synovial chondromatosis is characterized by a benign synovial proliferation associated with a metaplastic process. It remains a rare benign pathology of unknown cause. It evolves in three phases and the diagnosis is most often late. Imaging plays an essential role, particularly conventional radiology, computed tomography (CT)/arthroscan and magnetic resonance imaging (MRI). The imaging findings vary according to the stage. Synovial nodules and intra-articular foreign bodies are found. The knee is the most frequent localization. The treatment is surgical with the contribution of arthroscopy. We report the case of a 36-year-old woman who presented with gonalgia without any notion of trauma and who was diagnosed with primary chondromatosis by MRI with histologic confirmation. The recurrence rate remains low postoperatively and is often due to incomplete removal of synovial nodules and intra-articular foreign bodies.

Keywords: Chondromatosis, Knee, MRI, Primary

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INTRODUCTION

Primary (or idiopathic) synovial chondromatosis or Reichel–Jones–Henderson syndrome is characterized by the formation of multiple hyaline cartilage nodules under the synovial membrane [1–3]. These nodules may be released into the joint cavity and are then typically numerous, of approximately the same shape and size. Its etiology is poorly understood. It may be secondary or the result of a pre-existing pathology. This pathological condition is generally mono-articular and the knee is the most frequently affected joint. The evolution is most often progressive with a delay in diagnosis. It may occur less frequently in other joints, notably the hip, elbow, and shoulder [4]. Magnetic resonance imaging (MRI) allows a precise assessment of the local extension. The semiology is influenced by the subsynovial or free nature of the chondromas, and by their degree of mineralization and ossification. However, the diagnosis confirmation is made on histology. The treatment consists essentially in a synovectomy as complete as possible.

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CASE REPORT

A 35-year-old female patient consulted for knee pain with a medial meniscal syndrome without any notion of trauma. The evolution was marked by a recurrent pain for two years with a moderate swelling of the knee. The patient didn't present a restriction of range motion. An X-ray had been performed one year earlier without any abnormality. In view of the worsening of the symptomatology, a knee MRI was released in search of an etiology and revealed signal anomalies: Intra-articular nodular formations, Hoffa's fat, and the sub-quadricepsal recessus, in T1 hyposignal, in T2 hypersignal with peripheral enhancement after injection, the most voluminous located behind the posterior cruciate ligament (PCL). Joint effusion and synovial enhancement are associated (Figure 1).

The diagnosis of primary synovial chondromatosis of the knee was evoked. Arthroscopy was performed and a histologic analysis found chondromas, which confirmed the diagnosis of chondromatosis (Figure 2).

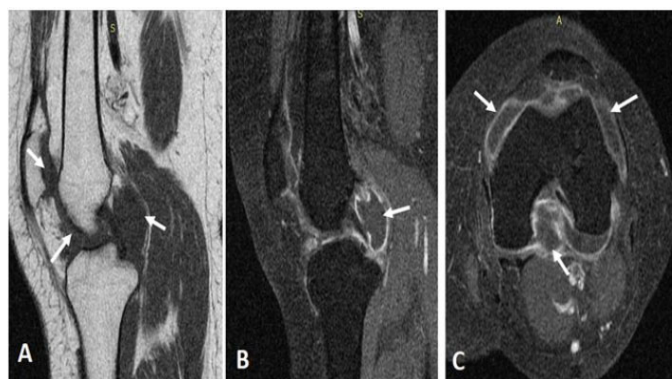


Figure 1: Knee MRI in T1 sagittal plane (A), T1 Fat Sat sagittal plane (B), and axial plane (C) showing well-limited synovial and intra-articular nodular formations in hyposignal (arrows).

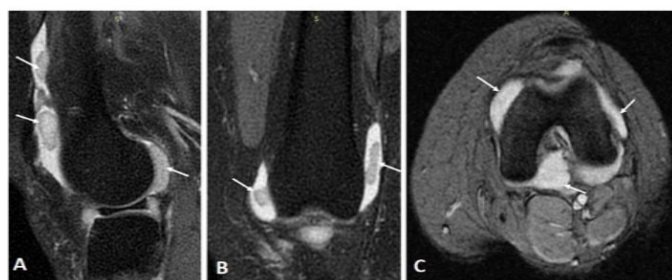


Figure 2: Knee MRI in proton density (PD) sagittal plane (A), coronal plane DP sequence (B), and T2* axial plane (C) showing well-limited synovial and intra-articular nodular formations in hypersignal with a rice grain appearance (arrows).

DISCUSSION

Primary chondromatosis is a rare benign monoarticular pathology of the articular synovium

characterized by synovial metaplasia of unknown origin. It is also known as Reichel–Jones–Henderson syndrome [4]. This synovial metaplasia and proliferation resulting in the formation of multiple cartilaginous foreign bodies of relatively similar size, not all of which are ossified. The term synovial chondromatosis is therefore preferred to primary synovial osteochondromatosis. It is distinguished from secondary synovial chondromatosis which is the result of a degenerative change in the joint. Secondary osteochondromatosis results from a pre-existing pathology [5]. The combination of osteoarthritis and osteocartilaginous nodules in the joint cavity corresponds exactly to this definition of secondary osteochondromatosis.

It affects men two to four times more than women, with a peak between the ages of 20 and 40 [4, 6]. Rare cases of familial association (2% cases) have been described in relation to collagen type 2 abnormalities, as described in Wagner–Stickler syndrome (hereditary arthro-ophthalmopathy) [7].

It classically evolves in three phases [8]:

- Initial phase: metaplastic formation of cartilaginous nodules in the synovium.
- Transitional phase: detachment of these nodules and formation of free intra-articular foreign bodies.
- Inactive phase: resolution of the synovial proliferation, but the foreign bodies remain in the joint and can increase in size by feeding on the joint fluid by diffusion.

The clinical signs frequently identified are pain (85–100%), joint swelling (42–58%), restriction of joint mobility (35–55%). There is a delay in diagnosis, estimated on an average at five years after the onset of symptoms [5, 6].

The majority of cases are located in the joints. The knee is the most frequent joint affected, with 50–65% of cases. Other locations are the hip, elbow, shoulder, and ankle. Occasionally the bursa and tendons may be affected [1–3].

Imaging reveals characteristic signs of the pathology, notably the presence of multiple intra-articular chondral bodies with “chondroid mineralization in the form of rings and arches, and extrinsic bone erosion on both sides of the bone.”

On conventional radiography, isolated non-specific synovial swelling, multiple intra-articular ring, and circle calcifications or osteochondromas, extrinsic erosions of the bone opposite or reactive hyperostosis may be seen [6, 8]. The joint space is normal or enlarged, which is suggestive of the diagnosis (interpositions of chondromas/articular cartilage hypertrophy may show abnormalities only in the calcification stage [6, 8].

On ultrasonography, they appear as round or oval nodules that are well defined and echogenic, with an effusion layer [7].

Computed tomography (CT) is the examination of choice for the demonstration of calcified intra-articular nodules and bone notches. Arthro-CT is described as the best examination for visualizing chondromas [5, 6].

The MRI appearance varies according to the stage of mineralization and ossification of the chondral bodies. There are three subtypes with a variable signal on MRI [4–6]:

- Non-mineralized chondromas (16%): “grains of rice” synovitis in homogeneous T1 hyposignal, T2 hypersignal, T2 Fat Sat, and in T1FS hyposignal after gadolinium injection.
- Calcified chondromas (75%): homogeneous T1 hypersignal, T2 hypersignal, T2 Fat Sat heterogeneous, and T1FS hyposignal after gadolinium injection.
- Ossified chondromas (9%): osteochondromas, presence of areas of heterogeneous T1 hyposignal, T2 hypersignal, T2 Fat Sat heterogeneous, and T1FS hyposignal after gadolinium injection.

Some differential diagnoses can be evoked in front of this picture namely [3, 5, 8]: secondary osteochondromatosis, arborescent lipoma (subquadriceps fatty swelling, without bone erosion with hypertrophy of synovial bangs with fatty signal in ± T2 hypersignal and enhancement of non-fatty areas, without effusion), synovial chondrosarcoma, pigmented villo-nodular synovitis (diffuse or localized with synovial thickening showing T2 hypointense areas with blooming on gradient echo images due to hemosiderin), synovial hemangioma (poorly systematized serpiginous lesion with extra-articular extension, no mass effect in hypo T1 hyper T2 iso + hypo T2 septa, intra-lesional fatty patches with heterogeneous intense enhancement).

The treatment of choice remains surgery. This treatment consists of removal of intra-articular foreign bodies. In case of active synovitis, a synovectomy can be performed [6].

The recurrence rate varies between 3% and 23% and is often associated with incomplete excision in most cases [4].

Arthroscopy with resection is becoming increasingly important in the management of arthroscopic injuries. Among other things, it reduces the mortality rate associated with the various postoperative rehabilitation procedures [4].

CONCLUSION

Primary chondromatosis of the knee remains a rare pathology. It presents as intra-articular foreign bodies originating from a metaplasia of the synovium. The cause remains unknown. Imaging plays an essential role in the diagnosis through the various means of exploration, in particular CT/arthroscan and MRI, which allow the diagnosis to be made with pathognomonic radiological signs. However, histological confirmation is sometimes

necessary. The diagnosis is most often made late. The treatment of choice remains surgery by removal of the nodules. Arthroscopy is playing a major role in the management of the disease through the development of surgical techniques.

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Romeo Thierry Yehouenou Tessi – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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