Scholars Journal of Medical Case Reports

Abbreviated Key Title: Sch J Med Case Rep ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online) Journal homepage: <u>https://saspublishers.com</u>

Imaging of Primary Synovial Osteochondromatosis: A Case Report F. Abakka^{1*}, M. OualiIdrissi¹, Y.Zouine¹, B.Boutakioute¹, N. CherifIdrissi El Ganouni¹, A. Benhima², I. Abkari², H.

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DOI: <u>10.36347/sjmcr.2021.v09i06.005</u>

| Received: 23.03.2021 | Accepted: 02.05.2021 | Published: 08.06.2021

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Abstract

Synovial chondromatosis (primary or idiopathic) is characterised by the formation of multiple nodules of hyaline cartilage under the synovial membrane. These nodules may be released into the joint cavity and are typically numerous and of similar shape and size. They may agglomerate with each other and form masses that are sometimes large. Radiographically, isolated non-specific synovial swelling, multiple intra-articular calcifications or osteochondromes, extrinsic erosions of the bone opposite or reactive hyperostosis may be seen. The joint space is normal or enlarged, which is suggestive of the diagnosis (chondromal interposition/articular cartilage hypertrophy). The arthroscanner or, better still, the 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 mineralisation and ossification. The treatment consists essentially of a synovectomy as complete as possible.

Keywords: Synovial chondromatosis; Arthroscanner; Magnetic resonance imaging.

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INTRODUCTION

Synovial osteochondromatosis is a rare and benign condition, characterised by metaplasia of the synovial tissue which produces small, roughly rounded cartilaginous masses (chondromas) protruding from the medial aspect of the synovium which pediculate and finally detach from it becoming free intra-articular chondromas.

The term "chondromatosis" is used for simple chondromas, but the cartilaginous nodules may then ossify and justify the term osteochondromatosis.

This metaplasia can occur at any site where there is synovial tissue, but can also occasionally affect the bursa and tendon sheaths [1, 2].

A distinction is made between primary forms (about 10% of cases), whose etiology and pathogenesis are unknown, occurring in healthy joints, and secondary forms occurring in joints affected by a degenerative process (osteoarthritis, synovitis, osteochondritis, trauma, etc.).

MATERIALS AND METHODS

Retrospective study of 5 cases of primary synovial osteochondromatosis collected at the Arrazi

radiology department in collaboration with the orthopaedic traumatology department of the Mohamed VI University Hospital of Marrakech and spread over a period of 3 years (2017-2020).

The positive diagnosis of synovial osteochondromatosis is retained on clinical, radiological and histological arguments.

All our patients underwent radiological exploration by standard radiography (2 cases), ultrasound (3 cases), Arthroscan (3 cases) and MRI (5 cases).

The purpose of this article is to review the radiological aspects of synovial osteochondromatosis

RESULTS

Synovial osteochondromatosis was selected in 5 patients in whom the clinical features were dominated by chronic pain and joint swelling.

The locations found were the knee (2 cases), elbow (1 case), hip (1 case) and hand (1 case). Osteochondromas were found in 4 patients, they corresponded in standard radiography to calcified oval foreign bodies, in CT to hypodense formations of

Citation: F. Abakka *et al.* Imaging of Primary Synovial Osteochondromatosis: A Case Report. Sch J Med Case Rep, 2021 Jun 9(6): 629-636.

variable shape moulded by the PDC while in MRI their signal was variable according to their degree of ossification.

Synovial hypertrophy was described in 1 patient associated with a medium-sized joint effusion. As for the widening of the joint space, it was noted in one patient and was associated with a synovial mass with calcifications.



Fig-1: Primary synovial osteochondromatosis of the lateral gastrocnemius tendon bursa.
a- Radiographie standard: Formations ovalaires calcifiées en arrière du genou
b- Ultrasound: Multiple oval hyperechoic formations with clear center (→
c- MRI: Oval formations within the bursa of the lateral gastrocnemius tendon in hyposignal(↓



 Fig-2: Primary osteochondromatosis of the right hip a-Standard X-Ray: widening of the right joint space(↓ b-CT: tissue mass with calcification.
 b-TDM: masse tissulaire siège d'une calcification.
 c-MRI: synovial mass in heterogeneous hypersignal after injection of PDC, with areas of signal void d- surgical specimen: osteochondromatosis cartilaginous nodules.





Fig-3: Synovial osteochondromatosis of the knee

A-Ultrasound: multiple oval hyperechoic formations with intra-articular effusion (🔿

B-CT: hypodense foreign bodies in the subquadricipital pouch moulded by the pdc.

C-MRI: synovial hypertrophy with medium-sized joint effusion. Multiple foreign bodies in the subquadricipital cul-de-sac with intermediate t1 and t2 signals and in the popliteal fossa, round and oval bodies of variable size with low t1 and t2 signals ()



Fig-4: Osteochondromatosis of the elbow MRI: oval tissue lesion in the proximal tendon sheath of the left brachialis muscle with intermediate T1 signal (A), STIR and DP hypersignal (B), Intensely and heterogeneously enhanced after PDC injection (C).



Fig-5: Tenosynovialosteochondromatosis of the left hand

A-Ultrasound: Multiple oval hyperechoic formations generating a posterior shadow cone (b-CT: calcified tissue formations on the dorsal surface of the left hand opposite the metacarpophalangeal joint of the 2nd and 3rd radii c-MRI: tissue formations opposite the metacarpophalangeal joint of the 2nd and 3rd radii of the dorsal surface of the left hand in T1 hypersignal and T2 hypersignal enhanced after injection of Gadolinium().

DISCUSSION

Epidemiology

Primary synovial chondromatosis is rare (7-10% of synovial chondromatoses) [3]. It affects young adults between 20 and 40 years of age, and men more often than women [4-7].

The condition is almost always monoarticular, rarely multiarticular. According to Lequesne [8], it affects, in order of frequency, the knee (40%), elbow (35%), hip (15%), shoulder (5%) and, more rarely, the ankle, wrist and hand joints and the temporomaxillary joint [9, 10]. Rare cases of multi-joint involvement have been described [11].

Secondary synovial chondromatosis is much more common (90%) and affects older subjects. It can be multi-jointed and is frequently seen in the hip, knee and elbow.

DIAGNOSTICS

Clinical

Symptomatology varies depending on the size and number of foreign bodies, the joint involved and whether the chondromatosis is primary or secondary [12].

Patients with CS may be asymptomatic or present with stiffness, swelling, recurrent effusions,

pain or locking. They may also experience mechanical pain or joint tenderness.

Palpation of one or more chondromas may involve superficial joints such as the knee, elbow or wrist Instability, joint effusions or signs of damage to surrounding neurovascular structures are less common. If left untreated, the primary disease progresses to osteoarthritis. Chondrosarcomatous degeneration is exceptional [13]. Biological tests are normal.

Imaging o Standard radiography

Osteochondromas may be single or multiple and of variable size. They project into the joint in 60-80% of cases [3].

They are round or oval in shape, with a calcified peripheral shell and clear centre. More rarely, they are ossified with real bone trabeculae or present more or less agglomerated punctiform calcifications [12]. They may be attached to the cortical bone or cause erosion of the cortical bone.

Standard X-rays can easily establish the diagnosis of osteochondromatosis (fig.1a) when the chondromas are calcified or ossified in the form of multiple intra-articular radio-opaque bodies, often of comparable size (osteochondromas), or dotted articular or periarticular calcifications.

However, X-rays are normal in the case of pure chondromatoses where the chondromas are not ossified, which makes the diagnosis more difficult (19 to 51% of cases depending on the series). In this case, indirect signs should be carefully looked for: [12-14].

Enlargement of the joint space compared to the healthy side secondary to foreign body interposition (Lequesne's mega-spacer form) (Fig. 2a).

Bone erosions in the synovial reflection zones indicating mechanical aggressiveness of the cartilage islands and erosion by the hypertrophic synovium; may raise suspicion of the diagnosis in "tight" joints such as the hip.

An increase in periarticular density linked to the numerous chondromas, joint effusion and synovial thickening can sometimes be detected.

Late in the course of the disease, secondary osteoarthritis, usually macrogeodic, may be observed. The diagnosis should be discussed if it occurs in the hip in a young person without dysplasia. If this osteoarthritis is advanced, it is often difficult to know whether it is the cause or the consequence of chondromatosis.

Arthroscan

The arthroscan remains the best examination for the diagnosis of chondromatosis. This examination should be requested in the case of moderate but chronic mechanical pain, with normal radiography and doubtful MRI, in a young subject.

It allows free chondromas to be visualised in negative, moulded by the contrast medium, to be counted and located in order to guide the therapeutic attitude (fig.3b). Radiolucent foreign bodies (purely cartilaginous) appear as rounded, oval or polyhedral gaps in the contrast medium. Partially ossified bodies take on a cocoon-like appearance, with a radiopaque ossified centre and a radiolucent cartilaginous peripheral shell floating within the contrast medium.

The synovial membrane, when active, appears thickened and irregular. In the initial stage of chondromatosis, the diagnosis is more difficult as only the non-specific appearance of hypertrophic synovitis is observed (chondromas are embedded in the metaplastic synovium and not yet released into the joint cavity) [15].

The interest of arthroscanner is crucial because it allows to [12]

- Make the diagnosis by showing the existence of radiolucent cartilaginous foreign bodies
- Show the presence of associated retractile capsulitis;

- Clarify the exact location of the foreign bodies, which is essential before surgical or arthroscopic removal.
- Look for early chondropathy.

Unprepared CT scan

The CT appearance of synovial chondromatosis depends on the stage of the disease [15]. When plain films are normal, the CT scan may show a periarticular soft tissue mass of muscle-like density, consistent with chondromas and synovial hypertrophy. Incipient calcifications and bone erosions may be detected earlier than on plain films [12].

When plain films show calcific intra-articular foreign bodies, plain CT scans can be useful in pinpointing the location of chondromas (Fig. 2b). In the hip, it is important to determine the location of intra-articular foreign bodies in the acetabulum [15].

Magnetic resonance imaging

MRI has the advantage of avoiding intraarticular puncture. The MRI appearance depends on the stage of the disease [16].

Although it can sometimes provide suggestive images, it is usually difficult to interpret in chondromatosis, as the cartilage nodules have a signal close to that of the synovial membrane and synovial fluid in both T1 and T2 [17].

- Non-calcified chondromas have an intermediate signal in T1 and appear as T2 isosignal within the hydarthrosis which has a slightly higher signal. Overall, a discretely heterogeneous appearance of the joint effusion can sometimes be suggestive of the diagnosis.
- Osteochondromas have a variable signal (Fig. 4).
- Partially calcified, they give the image of punctate or cocarde nodules with central hyposignal in T1 and T2.
- Massively ossified, the nodules take on a suggestive appearance in T1 and T2 hypo signal, contrasting with the higher synovial signal, especially in T2. However, at this stage, the osteochondromas are already clearly visible on standard radiographs.
- IV gadolinium injection can sometimes show enhancement of the synovium (if it is active and well vascularised), which moulds and detects the chondromas whose signal is not altered [14].
- In case of free chondromas, late sections show negative chondromas, moulded within the joint fluid. These aspects can now be observed directly by arthro-MRI.

MRI also assesses bone erosions and the extraarticular extension of chondromatosis and can detect malignant transformation in the case of bone invasion [18]. Oedema of the bone may occur as a reaction to the pressure exerted by foreign bodies. There is no true invasion of the bone marrow in this condition. A classification of MRI semiology has been proposed in the literature [19]:

- Type A: cartilaginous tissue without detectable calcifications;
- Type B: type A with calcifications;
- Type C: type A or B with osteochondromas showing a central fatty signal. Type B is by far the most frequent [19].

Ultrasound

Ultrasound may be useful for screening in forms with extracapsular extension, particularly in the bursa. Foreign bodies appear as echogenic beads surrounded by a thin layer of effusion, mobile on compression. Calcified chondromas are accompanied by a posterior shadow cone [15] (Fig. 5a).

Ultrasound can also detect a joint effusion, which is frequently present and which, if punctured, yields a clear mechanical fluid with a count of less than 2000 elements [20].

DIFFERENTIAL DIAGNOSIS Secondary synovial chondromatosis

Much more common than the primary form, secondary synovial chondromatosis is a reaction to joint disease that has released one or more (osteo) cartilage bodies into the synovial cavity [21]. These include destructive degenerative or arthropathy, or osteochondral fracture. There is no detectable cytogenetic abnormality within the synovium. On imaging, osteochondromatous nodules are fewer in number (typically less than 10), more variable in size (suggesting a different origin over time), but often larger than in synovial chondromatosis [22]. In addition, they may have multiple calcified rings (in contrast to the single ring, osteochondromatous nodules of primary synovial chondromatosis), with more marked degenerative and dystrophic changes [23]. Finally, arthropathy is typically associated.

• Rice grain'' nodules

This is a classic trap in MRI or arthroscanner. It is the accumulation, within a synovial cavity, of multiple whitish nodules composed of an amorphous acetowhite material coated with fibrin and collagen. They are seen in chronic inflammatory conditions such as rheumatoid arthritis and tuberculosis [22]. They may mimic multiple osteochondromas on MRI, as their signal is hypointense in T2, but they are not calcified. The subsynovial changes characteristic of synovial chondromatosis are also not present.

Other pseudo-tumourous conditions of the synovium

These conditions have in common the sometimes considerable thickening of the synovium or the subsynovial tissue, with the possibility of erosions of the adjacent bone. However, certain semiological elements allow them to be differentiated [24]:

- Pigmented villonodular synovitis: T2 hyposignal of the synovium typically more diffuse than the few spots sometimes detectable in synovial chondromatosis, absence of calcifications, absence of intra-articular foreign bodies
- Amyloidosis: T2 hyposignal of the synovium, absence of calcifications, polyarticular involvement, absence of intra-articular foreign bodies, suggestive terrain (renal insufficiency in particular) - gout: T2 hyposignal of the synovium, sometimes high density of tophus, but no cartilaginous-type calcifications, polyarticular involvement, absence of intra-articular foreign bodies
- Arborescent lipoma: fatty signal, absence of calcifications, exceptional erosions apart from those of associated arthropathy;
- Synovial haemangioma: serpiginous T2 hypersignal, without lobulation, of the synovium, absence of intra-articular foreign bodies, phlebolith-like calcifications;
- Tuberculosis: frequent osteopenia, associated soft tissue abscesses.

CONCLUSION

The radiological semiology of synovial chondromatosis is influenced by the subsynovial or free character of the chondromas, and by their degree of mineralization and ossification. This semiology is the source of radiographic signs suggestive of the diagnosis. Arthroscanner or, better still, MRI, allow a precise assessment of the local extension.

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