

Case Report

Primary Synovial Chondrosarcoma of the Right Knee: Case Report and Literature Review.

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Abstract: Synovial chondrosarcoma, either primary or secondary, is very rare. These two entities share a very similar profile, not only epidemiologically and clinically, but also on the radiological and pathological level. Thus the distinction between synovial chondromatosis and synovial chondrosarcoma, is a real dilemma. However, this distinction has a major interest in view of the evolutionary and therapeutic modalities are very different. In this work, we report a case of synovial chondrosarcoma, a woman of 62 years old, whose diagnosis was confirmed by histological examination. We will then discuss the contribution of individual authors, and all scientific testing, to establish specific criteria and objectives, that suggest the aggressiveness of chondrosarcoma, hidden behind the benignity of synovial chondromatosis.

Keywords: synovial chondrosarcoma; synovial chondromatosis; Malignant transformation; MRI; histology.

INTRODUCTION

Primary synovial chondromatosis is a benign synovial dystrophy, characterized by the formation of cartilaginous nodules that can later ossify (osteochondroma), within a joint or, more rarely, a synovial sheath.

It corresponds to a metaplasia of the synovial membrane of unknown origin, producing chondromes which can be pediculated and detached forming free foreign bodies in the articular cavity.

It affects classically the young adult of 20 to 50 years, more willingly masculine [1]. The knee joint is the most common location, about 50%, [2] followed by the hip, shoulder, elbow, ankle and finally the wrist. However, this synovial chondromatosis, long considered benign, may be complicated by malignant degeneration of synovial chondrosarcoma, rare cases of which have been reported [3- 6].

CASE REPORT

60-year-old patient, with a history of pain in the right knee, associated with episodes of joint swelling and shrinkage, and whose clinical examination

demonstrated a morphotype in genu varum, moderate articular effusion, flexum A few degrees and a positive plane.

A radiographic assessment made of standard x-ray of the left knee face and profile (Figure 1 & 2), which showed a significant diffuse bone demineralisation of the femoral condyles as well as the tibial plates and the patella, a pinching of the femoro-Patella with presence of suprapatellar calcifications. Presence of osteophytosis of the femoral condyles.

A IRM of the right knee plus the thigh (Figure 3) which showed a voluminous tumor process heterogeneously heightened after contrast, tissue and cystic component, the largest of the tumor developing at subquarripital recess and overflowing. On the sides of the lower femoral metaphysis, especially on the internal side. No femoral invasion or intra-articular extension. This lesion is relatively well defined and also respects the patella, the quadricipital tendon as well as the adjacent muscular planes, in particular the Large external quadriceps.

An extension of the thoraco-abdomino-pelvic CT does not find secondary sites (Fig. 4) and a bone scintigraphy showing no attachment of the remainder of the skeleton.

It was decided to perform a biopsy. However, during the surgical cure, the macroscopic appearance was that of chondromatosis and it was decided to perform a synovectomy (Figure 5)

The surgical specimen (FIG. 6) was addressed to the anatomopathology which speaks of a tumor proliferation posing the problem between a chondrome and a chondrosarcoma of low grade but given the high cellularity, the binucleation and the atypies, the diagnosis is in Favor of a low-grade chondrosarcoma.

ICONOGRAPHY



Fig-1: FACE X-ray of right knee



Fig-2: Profile X-ray of right knee

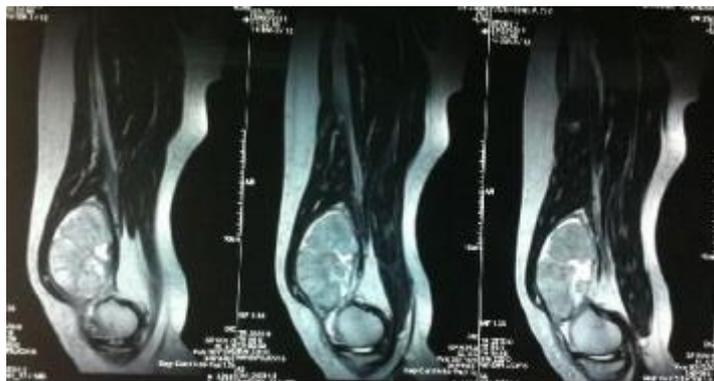


Fig-3: MRI of right knee

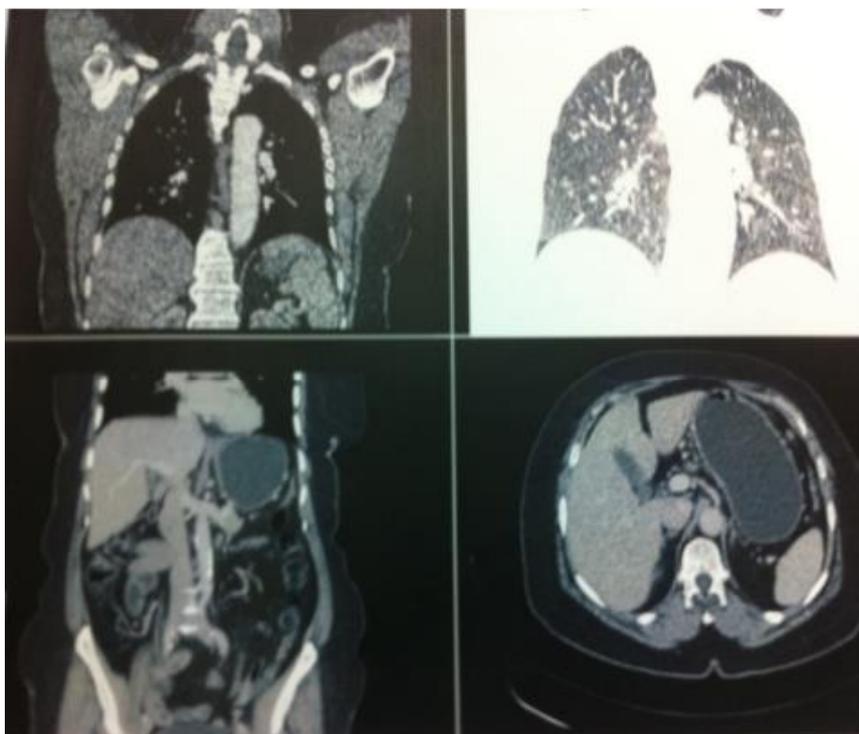


Fig-4 : Thoraco-abdomino-pelvic scanner



Fig-5: intraoperative picture



Fig-6: Resection part

DISCUSSION:

Primary synovial chondromatosis is a benign synovial dystrophy, characterized by the formation of cartilaginous nodules (chondromas) that can osseochromatically (osteochondromas), within a joint or, more rarely, a synovial sheath.

It corresponds to a metaplasia of the synovial membrane of unknown origin, producing chondromes which can be pediculated and detached forming free foreign bodies in the articular cavity. It affects classically the young adult of 20 to 50 years, more willingly masculine [1]. The knee joint is the most frequent location, about 50%, [2] followed by the hip, shoulder, elbow, ankle and finally the wrist. However, this synovial chondromatosis, long considered benign, may be complicated by malignant degeneration of synovial chondrosarcoma, rare cases of which have been reported [3- 6].

Of the 42 cases reported in the literature: Patients' age ranged from 25 years [7] to 82 years [6]. With an average age of 50 years. A male predominance with a sex ratio at 2.23 with 29 men and 13 women.

The knee is the most affected with 50%, followed by the hip 30.9%, then, in descending order, one finds again the ankle, the shoulder, the elbow and finally the wrist.

We were able to identify the initial clinical symptoms of 21 patients among the 42 reported: Pain

50% Swelling 21.87% Mass 21.87% and Stiffness 6.26%

Duration of clinical symptoms before the first consultation in 15 cases. This duration varies between 2 months [3] and 15 years [8], with an average duration of 5 years.

Radiologically, the Bertoni series [3], covering 10 cases: only nine had X-ray images. Of the nine pictures, the image of a mass of the soft tissue was found at the level of the articulation in question. Four of them had calcifications in this mass. And in three patients there was marginal erosion of the bone at the articulation in question.

In IRM, synovial chondrosarcoma would describe a lobulated and intraarticular mass of soft tissue, with a hypointense T1 signal, becoming hyperintense in T2 [9, 10].

Histologically, Bertoni's histological criteria [3] for the diagnosis of synovial chondrosarcoma: 1. The arrangement of cartilaginous tumor cells in sheets and not in clusters. 2. A myxoid change at the level of the matrix. 3. Hypercellularity with crowding of the nuclei at the periphery 4. Necrosis unrelated to calcifications and in the absence of pathological fractures. 5. Synovial chondrosarcoma shows permeation of the trabecular bone with filling of the medullary space, which must be differentiated from bone erosion caused by synovial chondromatosis.

From a therapeutic point of view, the ten cases in the Bertoni series [3]: A tumor recurrence was noted for nine patients, these patients would have benefited only from a local excision of the tumor.

For further radical treatment 8 patients underwent amputation 1 patient underwent extensive excision (without lesion margins) with arthroplasty. This, despite these aggressive surgical measures: 5 patients developed pulmonary metastases: 4 of them They died 1 survived after pulmonary metastasis removal The remaining 4 patients remained alive without developing metastases 11 years after surgery.

Indeed most authors have retained that an inadequate surgical excision involves a high risk of local recurrence and subsequent metastasis [3- 6].

Our clinical case: In the light of these data The epidemiological age of our patient is 62 years. Our patient belongs to the third age group, with an estimated distribution of 21.40%. Sex: Our clinical case concerns a woman. However, the literature shows a clear masculine predominance. Localization - Our clinical case concerns a chondrosarcoma of the knee. - And the knee is proven the most frequent localization, it is interested in 50% of the cases. Clinically: Our patient complained of knee pain with swelling. In the literature pain is the main symptom at 50%, swelling second at 21.87% .the duration of development before the first consultation - For our case the duration is 5 years - The average duration in literature is Also 5 years. On the radiographic plane The imaging means made it possible to identify, in our patient, the presence of a mass of the soft tissue with diffuse calcifications, which agrees with the authors' findings. - However, there was no evidence of marginal bone erosion. - Ni, other criteria suggestive of malignancy described by the authors [4]. Histologically, pathologists were more in favor of low-grade synovial chondrosarcoma. From the therapeutic point of view, in view of the absence of bone invasion, and in the absence of histological evidence of malignancy, a simple excision of the mass with synovectomy was chosen. The authors, on the other hand, recommend a wide resection, for a better local control of the tumor [3, 5, 6, 8]. On an evolutionary level -Two months after the operation, the patient reports a clear clinical improvement, in particular a decrease in the painful condition of her knee. It should also be noted that no metastasis was detected on the extension.

CONCLUSION:

Our clinical case presents the profile of a primary synovial chondrosarcoma, a very rare tumor, which, according to the review of the literature, is the sixth reported case. However, the differential diagnosis between synovial chondromatosis and synovial chondrosarcoma marks several limits, leaving the doors wide open for further investigations.

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