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Malignant Degeneration of an Osteogenic Exostosis in Multiple Exostatic Disease (About a Case)

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Abstract **Case Report**

Introduction: Osteochondroma, otherwise known as osteogenic exostosis, is a benign surface tumor; it develops from an enchondral ossification bone, which and in contact with the epiphysis. Its most dreadful complication is chondrosarcoma. ***The aim:*** of our observation is to report a rare and dreadful complication of osteochondroma. ***Observation:*** This is an 08 year old female patient, without any particular pathological history, who consulted for a painful swelling of the lower extremity of the left thigh with a chronic evolution. ***Conclusion:*** Once the diagnosis of exostosis is made, an annual radiological control is mandatory. Carcinological resection is mandatory as soon as malignant transformation is suspected.

Keywords: Exostosis, Complications, Imaging.

INTRODUCTION

Osteogenic exostosis or osteochondroma; which falls within the framework of multiple exostant disease, is considered as a benign surface tumor; which develops from a bone with enchondral ossification, in contact with the epiphysis [1]. Its serious complication is chondrosarcoma. The transformation to a malignant tumor develops from the cartilage [2, 3]. The accentuation of the thickness of the cartilaginous plaque is the early and alarming sign that should be sought by imaging.

MEDICAL OBSERVATION

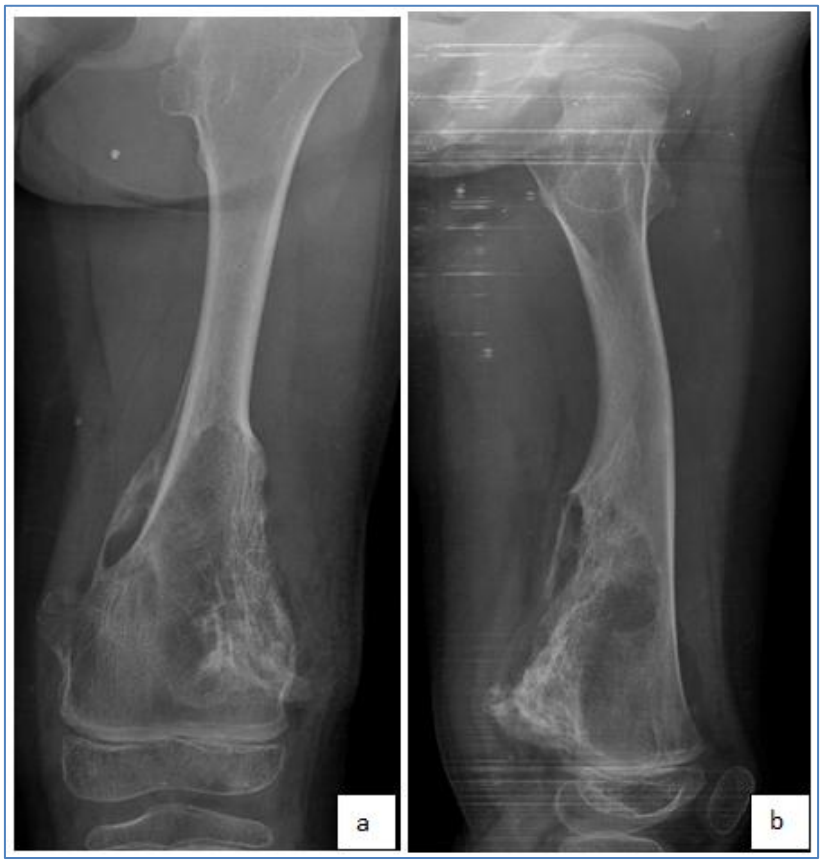
We propose to explore the case of an 08 year old female patient, without any particular pathological history, resident in Casablanca, who presents a painful swelling of the lower extremity of the left thigh. On clinical examination, a hard swelling of the lower extremity of the left thigh was found, immobile in relation to the superficial and deep planes, with no inflammatory signs opposite.

MRI of the left thigh shows a metaphyseal lesion process of the lower third of the posterior cortico-cancellous femur with sessile exophytic development, a broad base of implantation and a diaphyseal direction of the long axis.

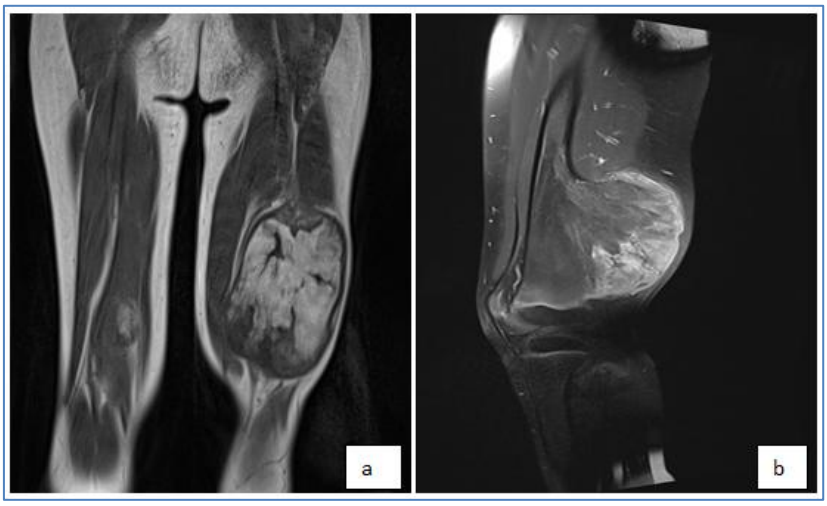
The surgical treatment was for aesthetic purposes. The resection was partial due to the proximity of the tumour to the popliteal vascular pedicle. 80% of the tumour volume was removed with 20% remaining posteriorly

The diagnosis of chondrosarcoma was confirmed by macroscopic and histological examination of the surgical specimen.

FIGURES



- **Fig-1: Radiograph of the left thigh: front view (a) side view(b) : metaphyseal formation of the lower third of the femur with sessile exophytic development, a broad base of implantation and a large axis with a diaphyseal direction**



- **Fig-2: T1 coronal (A) and T1 FAT SAT sagittal (B) sequences of the left thigh showing a metaphyseal lesion process in the lower third of the posterior cortico-spongiosa femur with sessile exophytic development, a broad base of implantation and a diaphyseal direction of long axis. It has a heterogeneous signal mostly similar to that of the cancellous bone with which it continues from the femur and is covered by a thick irregular crown in T1 hyposignal.**

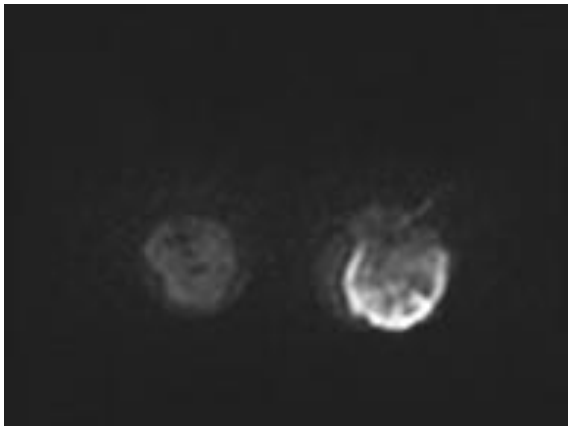


Fig-3: Diffusion sequence showing hyposignal within the lesion process

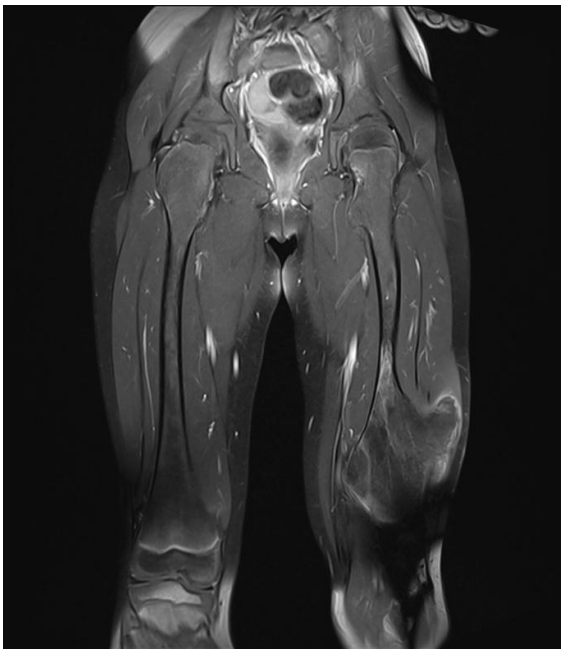


Fig-4: T1 coronal injected sequence showing contrast enhancement of the process after gadolinium injection.

DISCUSSION

Malignant transformation is the most dreaded complication of osteochondroma. The prevalence of this transformation is 1%, most often it is chondrosarcoma, however, other rare cases of sarcoma have been observed [4,2]. The malignant degeneration develops from the cartilage cap [2, 3]. The increase in thickness of the cartilage cap is the earliest sign, which must be investigated by imaging, in particular by MRI or ultrasound. Clinically, the symptoms that suggest degeneration are an increase in size, increased local pain and inflammation [5].

Radiologically, a cartilaginous cuff with a thickness of less than 1 cm is considered normal, whereas a cuff with a thickness of more than 2 cm suggests a chondrosarcomatous transformation.

A margin between 1 and 2 cm is considered doubtful. The presence of other radiological signs should make us think of degeneration, such as the existence of calcifications that are irregular and heterogeneous and that extend beyond the edges of the exostosis, and that may appear larger and more numerous from one incidence to another, the presence of a mass that develops from the soft tissue surrounding the tumor.

Due to the intense fixation of the radioactive marker, bone scans help to identify active osteochondromas. However, differentiation between benign osteochondromas, which are the site of active enchondral bone formation, and degenerative exostoses is not possible. However, a normal scan does not rule out a malignant transformation [6]. Whenever there is a symptomatic or radiologically suspicious exostosis, it should be respected.

The pelvis and the shoulder are the locations most at risk of degeneration [7]. The average age of malignant transformation is 30 years [8].

Anatomopathological exploration of the surgical specimen is the key examination for diagnostic confirmation. Frequently, chondrosarcomas are of low grade with a slow evolution. In about 10% of cases, sarcomatous transformation into dedifferentiated chondrosarcomas with a poor prognosis and possible metastatic evolution [9]. This transformation remains rare in children, in whom the indications for excision are essentially functional.

CONCLUSION

With the progress of current imaging, the diagnosis of exostosis is easily made. As soon as we are sure of the pathology, an annual radiological control is mandatory. Carcinological resection is mandatory as soon as malignant transformation is suspected.

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