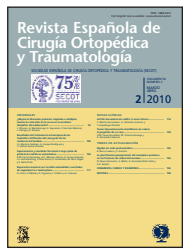
Rev esp. cir. ortop. traumatol. 2010;**54(2)**:120-125

Revista Española de Cirugía Ortopédica y Traumatología

www.elsevier.es/rot



CASE REPORT

Liposclerosing myxofibrous hip tumour. A case

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Received April 3, 2008; accepted October 8, 2008
Available on the internet from May 9, 2009

KEYWORDS
Tumor;
Liposclerosing;
Myxofibrous;
Hip;
Fibrous dysplasia;
Lytic lesion

Abstract
Introduction: To look into the liposclerosing myxofibrous tumour, a rare benign tumour formation with a predilection for the proximal femur.
Clinical case: We present a case treated in our hospital. We performed curettage and filling of the defect with an iliac crest graft; the lesion had initially been diagnosed as a one cyst/enchondroma in the hip. The pathological study revealed the presence of a liposclerosing myxofibrous tumour. Although this diagnosis was not suspected, the treatment administered was correct and the patient evolved satisfactorily.
Conclusions: The liposclerosing myxofibrous tumour is a rare condition. Its presence may be suspected when the X-ray reveals a lytic lesion, with variable sclerotic margins, lodged in the femoral epiphysis or proximal shaft. There is no unanimous acceptance of this entity in the literature, with some authors claiming that it could be a variant of fibrous dysplasia.

PALABRAS CLAVE
Tumor;
Lipoesclerosante;
Mixofibroso;
Cadera;
Displasia fibrosa;
Lesión lítica

Tumor lipoesclerosante mixofibroso de cadera. A propósito de un caso

Resumen
Introducción: El objetivo de esta investigación es estudiar el tumor lipoesclerosante mixofibroso (TLEMF), una tumoración benigna y rara con predilección de asiento en el extremo proximal femoral.
Caso clínico: Se presenta un caso clínico atendido en este centro con realización de raspado y relleno de la cavidad, con injerto de cresta ilíaca de una lesión que inicialmente fue etiquetada de quiste óseo o encondroma en cadera. La anatomía patológica informó de TLEMF; aunque no se sospechaba el diagnóstico, se realizó un tratamiento correcto y el sujeto evolucionó de manera adecuada.

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Conclusiones: El TLEMF es infrecuente y se sospecha ante una lesión radiológica lítica, geográfica, con márgenes de esclerosis variable y asiento en la epífisis y metáfisis proximal femoral. No hay una aceptación unánime de esta entidad en la literatura médica, pero hay autores que argumentan que es una variante de la displasia fibrosa.

Introduction

Liposclerosing myxofibrous tumour (LSMFT)[2-8] is a rare entity with a predilection for the proximal femur in young adults in the fourth decade of life.[1-3,8]

Other names for it include polymorphic fibro-osseous tumour, polymorphic fibrocystic disease of the bone, polymorphic fibro-osseous lesion of the bone,[2,3] polymorphic fibro-osseous lesion of the proximal femoral bone[9] or atypical fibrous dysplasia.[4,5]

On plain radiography it appears as a lytic lesion with a geographic appearance, well-defined borders, and margins of variable sclerosis that contain areas of calcification.[1-3,8]

A differential diagnosis that includes other entities is obligatory, such as bone dysplasia, bone cyst, lipoma, non-ossifying fibroma, enchondroma, and bone infarct.[2,8]

Its treatment is the same as that for many benign osseous tumoural lesions: scraping and curettage of the cavity, filling with bone graft, and fixation if necessary.[8]

In addition, periodic follow-up should be performed since sarcomatous degeneration has been described in between 10 and 16% of cases;[2-4] although the exact percentage is unknown and a lower number is suspected (as will be seen later on).

Clinical case

A 43-year-old male who was sent from the Internal Medicine offices after presenting with arthralgias and a ferritin elevation.

Laboratory testing was requested (complete blood count, biochemistry, erythrocyte sedimentation rate, C reactive protein), which showed hyperuricaemia.

A bone series was performed which showed the existence of an osteolytic lesion consistent with enchondroma and chondrosarcoma at the level of the left femoral metaphysis.

The diagnosis was made of bone cyst of the left femur and hyperuricaemia, and rheumatologic disease and haemochromatosis were ruled out. The patient was discharged to the Internal Medicine offices and referred for consultation with Trauma and Orthopaedic Surgery (fig. 1).

Magnetic resonance (MR) was ordered based on the consultation at this centre. The complete report was as follows:

"A well-defined tumour is appreciated in the proximal metaphysis of the left femur, with hypointense border on all sequences, with a matrix that is hypointense on T1 and hyperintense on T2, and with hypointense areas in all sequences probably relating to calcification.

The cortical bone is not affected.

No other abnormalities are appreciated in the rest of the osteomuscular structures in the area studied.

Tumour formation in the proximal metaphysis of the left femur with benign appearance consistent with enchondroma" (figs. 2, 3, 4 and 5).

Surgical intervention was decided on based on this diagnosis.

An external Bauer approach was utilised under spinal anaesthesia and fluoroscopic control, followed by curettage and cleaning of the cavity in the proximal area of the left femur. The resulting bony cavity was filled with cancellous bone graft from the ipsilateral iliac crest and a bone cerclage was placed to fix the bone window. The specimen was sent for pathological study.

The patient received analgesic treatment, antibiotic prophylaxis, antithrombotics, and gastric protective treatment, and was discharged 72 hours after the intervention.

During the first post-operative month, the patient walked with crutches and kept weight off the affected extremity. Partial load-bearing was allowed after this first month and full load-bearing was started at the third month.

Informed consent was obtained from the patient for all the diagnostic tests, their later use or dissemination, as well as for the surgical intervention.

The pathologist from this centre made the pathology diagnosis prior to consulting the Anatomic Pathology Service of the reference hospital (Yagüe de Burgos General Hospital), where several cuts were sent and the diagnosis confirmed.

The description from the report was the following: "Microscopic examination of several fragments of an osseous lesion showing course, newly-formed bony trabeculae with geographic areas of pseudo-Paget's that are lacking in osteoblastic and osteoclastic linings. In addition, it shows areas of curvilinear and circular ossicular trabeculae that mimic fibrous dysplasia. The background of the lesion is fibromyxoid, with scant cellularity, without devitalised cells or plasma cells. Adipose areas and focal areas of xanthomatous cells can be seen. The fibromyxoid tissue enters, occasionally, into the Haversian spaces of the pre-existing peripheral compact bone".

Diagnosis: "Liposclerosing myxofibrous tumour (polymorphic fibro-osseous tumour)" (figs. 6, 7, 8 and 9).

Follow-up consultations were performed every 3 months and, one year after the surgery, the patient was asymptomatic and had completely resumed social and work-related activities.

The radiographic evolution was successful with cortical restoration at the level of the femur and osseous integration of the graft and, on computerised tomography at one year, a residual cavity was seen without any evidence of disease recurrence (an MRI was not ordered due to the possibility of artifact from the cerclage).

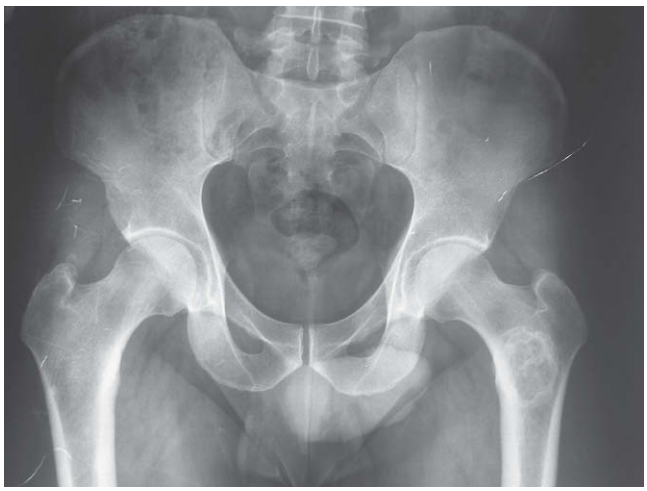

Figure 1 Plain anteroposterior pelvic radiograph in which a well-defined geographic lytic lesion is seen, with a sclerotic border at the level of the left femur, which respects the cortex and contains areas of calcification.

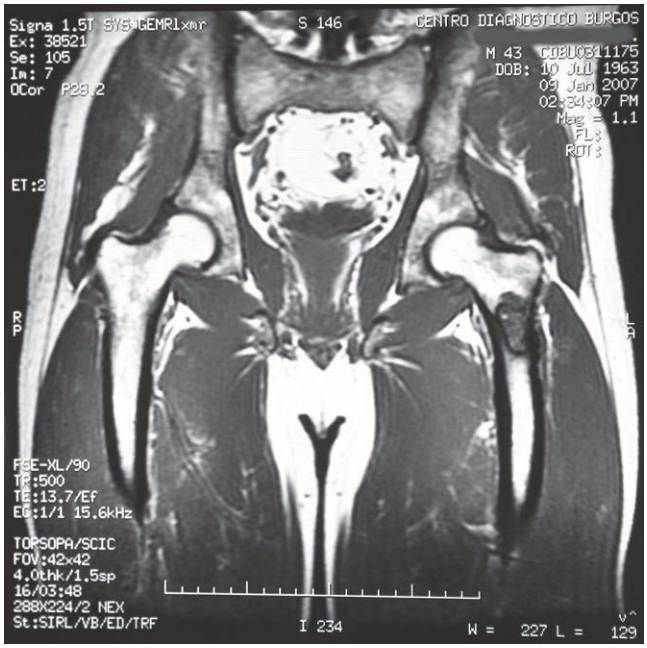

Figure 2 Coronal cut from the magnetic resonance that shows a well-defined hypointense signal in the left hip in the T1 sequence.

Discussion

LSMFT of the hip is an atypical, benign fibro-osseous lesion with a complex variety of histological patterns that may include lipoma, fibroxanthoma, myxoma, characteristics of fibrous dysplasia, cyst formation, fat necrosis, ischaemic ossification, and, rarely, cartilage.[1-4,8] Moreover, stromal hyalinisation and niduses of Paget's-like bone may be seen.[2-5]

The osseous trabeculae may show sclerosis, mosaicism, ischaemic ossification, and well-formed laminar bone.[2,3,9] Its

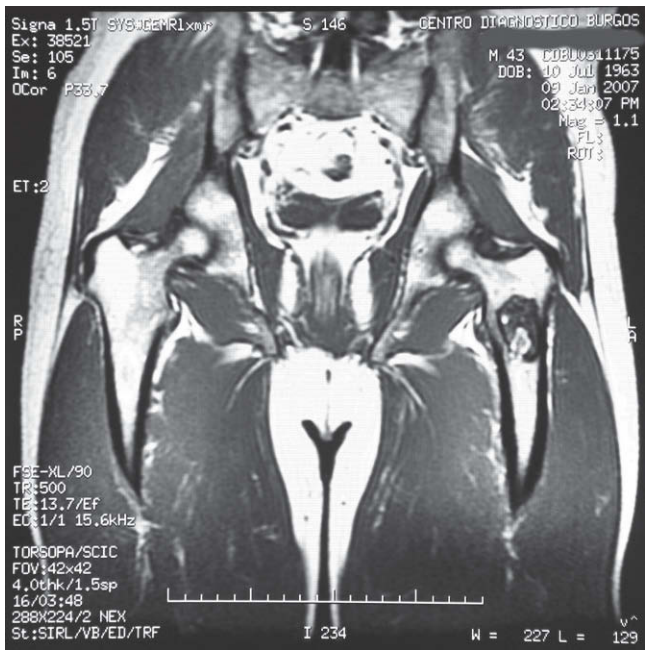

Figure 3 Another magnetic resonance image from the T1 sequence of the same case.

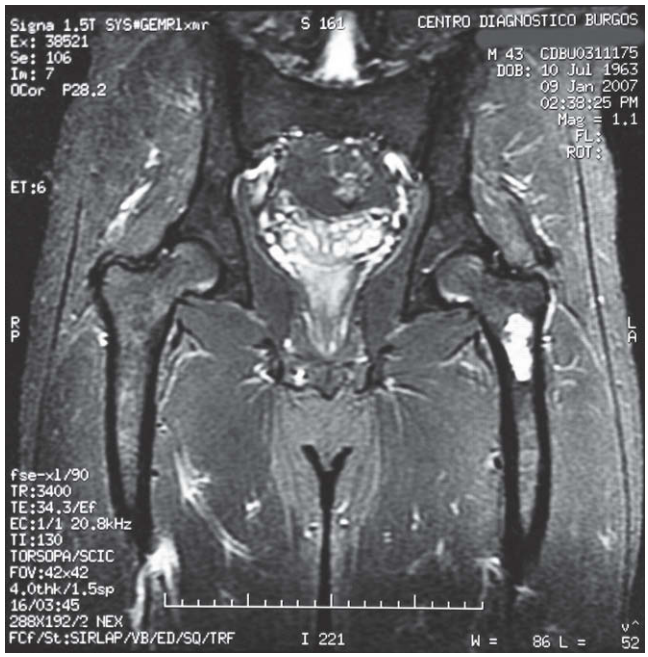

Figure 4 Magnetic resonance image in which a lesion is appreciated in the proximal metaphysis of the left femur with hyperintense signal in the T2 sequence, with well-defined border and that respects the cortex.

favoured location is the proximal area of the femur in 80 to 90%of cases,[1-3,8,9] but it has been described in other locations less frequently, such as the femoral shaft or other bones such as the iliac, humerus, tibia, and the second rib.[2-4]

Some authors state that the most common occurrence of this entity is that of an incidental finding on radiography.[2]

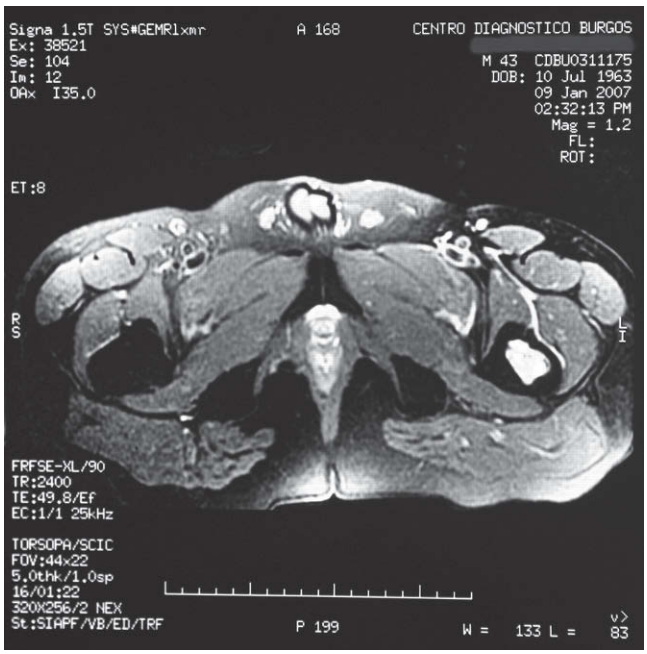

Figure 5 Sagittal cut of T2 magnetic resonance sequence that shows a hyperintense image that appears well-defined at the level of the proximal end of the left femur.

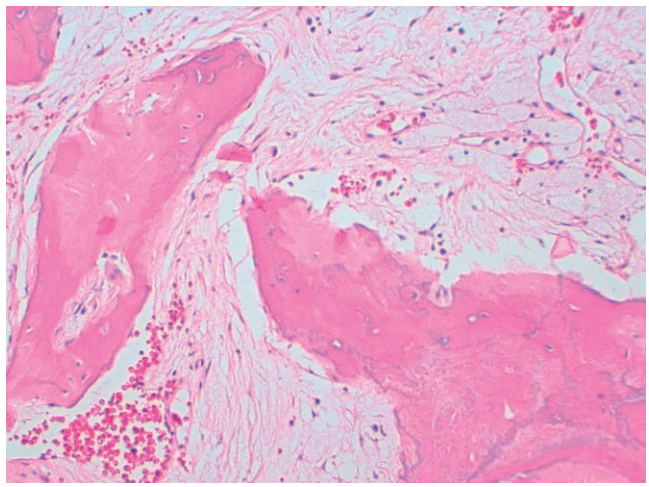

Figure 6 Histological image with haematoxylin and eosin stain at 40× in which areas of pseudo-Paget's can be seen (matrix and osseous trabeculae without osteoblastic or osteoclastic lining) as well as the fibromyxoid stroma. Areas of pseudo-Paget's are indicated.

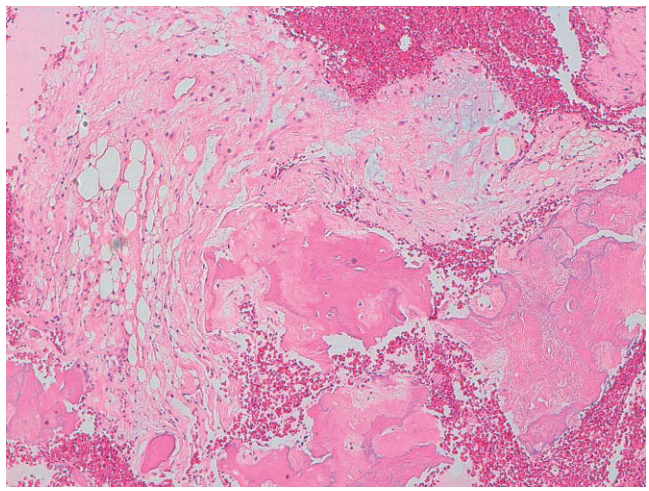

Figure 7 Histological image with haematoxylin and eosin stain at 20× in which pseudo-Paget's formations can be seen that are lacking in osteoblastic and osteoclastic lining. In addition, a fibromyxoid matrix is seen with scant cellularity.

According to other authors, the first symptom is pain, with variable intensity and duration.[3,8] It may also start as a pathologic fracture.[2,3]

The age range is from the second to the seventh decade of life, with an average of approximately 40 years.[1-3,8] It may develop during infancy and its clinical manifestation or diagnosis may take place in adulthood[2] without a preference for either sex.[1-3,8]

Plain radiograph of this entity shows a geographic lytic lesion with a well-defined border that is frequently sclerotic, with internal mineralisation and, in some cases, mild bone expansion.

Thus, the differential diagnosis from plain radiography is made up of those entities that have well-defined lytic lesions at the proximal end of the femur: osseous dysplasia, bone cyst, lipoma, non-ossifying fibroma, enchondroma, and bone infarct, among others.[2,8]

In a paper from 1993, Ragsdale based the differential diagnosis on other entities from plain radiography and on pathology findings. It is now appropriate to delve into and discuss the appearance of LSMFT on CT and MRI, according to the reviewed medical literature.

According to Resnick, LSMFT may manifest on CT and on MRI as a mixture of internal elements without signs of fat.[1] O'Dwyer describes it on CT as a lesion with a sclerotic margin and non-specific matrix.[8] According to Kransdorf, the globular nature of the mineralised matrix is seen on axial cuts of the mid-section of the lesion.[3]

MRI images showing moderate heterogeneity on the T2 sequence are cited in later works, as well as showing high intensity or partially low intensity for fat, as in a well-defined lesional border (at least in part).[7]

On the T1 sequence, the lesion has been described as a hypointense signal and on T2 sequence as a hyperintense signal (consistent with the present case). The non-specific lesional image and the hyperintensity on the T2 sequence (due to the high myxoid content) present internal irregularities due to possible calcifications.[4,8]

Kransdorf notes an image on the T1 sequence with a signal similar to the muscular skeleton that is moderately heterogenous on the T2 sequence, with a well-defined margin and areas of high or intermediate intensity in this signal.[3] Kransdorf further states that a technetium-99m bone scan shows accumulation of the tracer at the site of the lesion.[3] No CT or MRI image was found in the medical literature that could assist with the definitive or differential diagnosis of this entity as it relates to other processes.

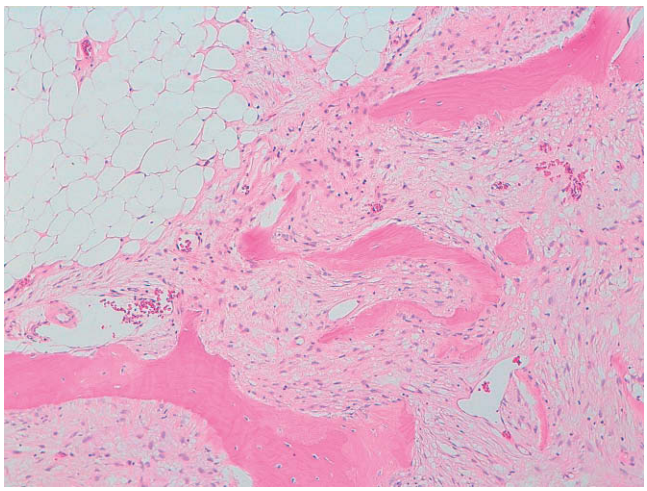

Figure 8 Histological image with haematoxylin and eosin stain at 40× that shows adipose cells.

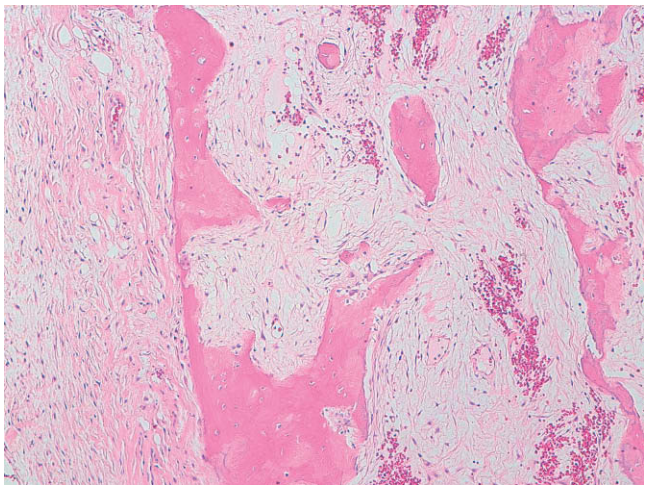

Figure 9 Histological image with haematoxylin and eosin stain at 20× in which the fibromyxoid matrix and areas of osseous necrosis are seen.

According to O'Dwyer,[8] the diagnosis of LSMFT should be based on plain radiographic findings, image-guided biopsy, and pathology.

It is important to remember that plain radiography always shows some constant characteristics of this entity: a well-defined lytic lesion with sclerotic border, probable internal mineralisation, and a predilection for the intertrochanteric region.[1-3,8]

Ragsdale's work in 1993 puts the malignant percentage at 16%, even while noting that the exact incidence of sarcomatous degeneration is unknown and probably overestimated, which could show that the ischaemic desmoplasia present in the lesion favours tumoural genesis.[2,3,8]

In his work from 1999, Kransdorf cites 10% malignancy, but advises that the value must be taken with caution since it may be overestimated (cases of sarcomatous degeneration with a previous diagnosis of LSMFT were included in his study), or the possibility of malignancy may be considerably lower due to it often being an incidental finding.[3] However, for some authors such as Heim-Hall, the risk may be lower since it is considered a variant of fibrous dysplasia (0.5% based on the malignancy rates of that entity).[5]

There are authors who clearly recognise LSMFT as its own entity despite its varied histogenesis, based on its typical characteristics on plain radiograph and its typical location in the intertrochanteric region of the proximal femur.[1-3,8,9]

Other authors believe that we may be dealing with a posttraumatic variant of fibrous dysplasia and maintain that it is the evolution of this entity that is located in the proximal end of the femur due to the rigorous mechanical demands placed on that anatomic region, which then causes fatigue factures on fibrous dysplasia, and that there is no radiologic difference between one lesion and the other.[5]

There are also authors who believe that the difference or relationship between one entity and the other is still not clear, and that more research is needed in order to avoid diagnostic errors.[4,6,7]

In addition, it is argued whether these entities are related to intra-osseous lipomas[1,3] or if they are involutional changes caused by a prior benign osseous lesion of a fibromyxoid or fibro-osseous nature.[3,4] On the other hand, three cases of LSMFT have been recognised (two in Matsuba's work and one in Corsi's) that have a mutation in the *GNAS* gene, an alpha subunit of the G protein at the Arg^{201} codon that increases the creation of cyclic adenosine monophosphate.

This mutation is the cause of McCune-Albright syndrome and is also present in polyostotic fibrous dysplasia and in some cases of monostotic fibrous dysplasia.[6-8] Based on this, more uncertainty is added to the question of whether or not LSMFT is an entity that is truly separate from fibrous dysplasia.

Treatment of LSMFT consists of curettage of the lesion and filling of the cavity with bone grafts; osteosynthesis may be associated, if necessary.

Cases that present with pathologic fracture may require hip arthroplasty.[8] Those cases that are found incidentally or those that are not clinically painful may simply be watched.[8] Furthermore, one must balance the benefit and risk of performing a prophylactic excision on a benign lesion, with the risks of a larger surgery,[3] especially in an area like the proximal end of the femur and the significant loads that it bears.

Periodic follow-up of the lesion must be performed, since malignant transformation has been described in this entity (sarcomatous degeneration in 10 to 16% of cases),[2-4,8] although this issue has not been completely clarified.

The orthopaedic surgeon must be able to create a differential diagnosis of skeletal tumoural lesions, first differentiating them from other lesions that are dysplastic, infectious, etc., and then assessing if they show benign or malignant characteristics if a tumour is suspected.

It is clear that, if there is any doubt, the patient should be sent to a specialised centre where definitive diagnosis can be offered though biopsy and where therapeutic strategies may be presented.[10] Thus, diagnostic surprises such as that of the present case may be avoided, along with the serious consequences that incorrect therapeutic planning and implementation may bring to these patients.

In conclusion, one must think about the possible diagnosis of LSMFT when faced with a lytic lesion with well-defined

geographical margins and variable sclerosis located in the proximal region of the femur.

The differential diagnosis is based on those entities that show well-defined lytic images in the proximal end of the femur; those include osseous dysplasia, bone cyst, lipoma, non-ossifying fibroma, enchondroma, bone infarct, etc.[2,8]

Given the rarity of this process, which may not be found in the conventional medical literature, there is no unanimity regarding what an entity like this might be. Nor, based on the work and experience of our group, can we say that LSMFT is really its own nosological entity or a variant of fibrous dysplasia.

When concerns arise about the nature of the lesion or the differentiation between the benign or malignant nature of the lesion, or if it is thought at the outset that a lesion is malignant, a biopsy study is needed and the patient should be sent to a reference centre that has experience with osseous tumoural disease.

Conflict of interest

The authors have not received any financial support for this work. Nor have they signed any agreement through which they would receive benefits or fees from any commercial entity. Furthermore, no commercial entity has paid nor will pay any foundations, educational institutions, or other non-profit organisations with which the authors are affiliated.

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