Case Report

Liposclerosing myxofibrous tumor: A series of 9 cases and review of the literature

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ABSTRACT

Background: Liposclerosing myxofibrous primary bone tumor is a rare benign bone lesion that was characterized by complex mixture of various histological elements.

Methods: We have studied the radiological, clinical and pathological features of nine patients with this disorder. Pain and limping were the main symptoms.

Results: Radiographic images typically showed a geographic lytic lesion with thick sclerotic margin, reflecting a pattern of slow growth. Histological sections revealed a polymorphic neoplasia characterized by predominant proliferation of stellate and fusiform cells aimed the myxoid matrix.

Conclusions: These features suggest that the lesion may represent a variant of fibrous dysplasia with a high risk of malignant transformation.

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1. Introduction

Liposclerosing myxofibrous primary tumor (LSMFT) is a benign lesion of the bone. First described by Sweet and Ragsdale in 1986, it is characterized by combination of several histological elements including lipoma, fibroxanthoma, myxoma, myxofibroma, fibrous tissue, cyst formation, fat necrosis, ischemic bone tissue, and rarely cartilaginous tissue.^{1,2} Despite its typical characteristics, it is considered an extremely rare tumor.³ It has been shown that the LSMFT should not be considered as an isolated entity, but rather a variation of other tumors, such as fibrous dysplasia, intraosseous lipoma, healed solitary bone cysts and fibromyxoma osseous.⁴ Genetic analysis suggested that the lesion may represent a variant of fibrous dysplasia.⁵ The risk of transformation to malignant fibrous histiocytoma or osteosarcoma was estimated between 10% and 16%. The exact term for malignant transformation is not known but occurs over a long time with slow progression of the lesion.^{4,6} The purpose of this study is to show series of LSMFT treated in a referential oncologic hospital in Rio de Janeiro/Brazil and compare our observations with other studies.

2. Methods

A total of 9 reported cases of LSMFT were treated. The data collected consisted of clinical records, operative notes, radiographic images, microscopic imaging, as well as pathological reports.

2.1. Consent

Written informed consent was obtained from each patient in order to allow the publication of these series of case and accompanying images. A copy of the written consent document is available for review by the Editor-in-Chief of this journal. The Brazilian ethics committee permission for research – CAAE: 39771214.0.0000.5273

3. Results

Nine patients with LSMFT were evaluated and treated. The objects of study were composed by two men (22.2%) and seven women (77.8%); mean age was 39 years (19–53), with five black patients (55.5%) and four white patients (44.4%). The time between the onset of symptoms and the histopathological diagnosis varied between six and sixty months (mean 27 months) (Table 1). Pain and limping were the first symptoms. Other clinical and radiological data reported were history of trauma (two patients – 22.2%), pathological fracture (three patients – 33.3%) and local edema (two patients – 22.2%). The

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Table 1

Location, size and treatment of liposclerosing myxofibrous tumor per patient.

Case	Sex	Age (Year)	Ethnicity	Location	Size (Method)	Image (RX)	Symptoms/ diagnosis (Months)	Follow-up (Months)	Treatment
1	F	50	White	Right neck and femoral head	3 cm × 3 cm (MRI)	Lytic lesion, well-defined, eccentric and sclerosis edge	6	14	Curettage and grafting
2	M	53	Brown	Right distal tibia	6 cm × 6 cm (CT)	Lytic lesion with bone formation, eccentric, blowing, sclerosis edge and disruption of cortical	36	97	Transtibial amputation
3	M	19	Brown	Right distal femur	8.5 cm × 8 cm (CT)	Lytic lesion, eccentric, blowing, and sclerosis edge	36	76	Curettage and grafting
4	F	45	Brown	Right intertrochanteric region and femoral neck	4 cm × 3 cm (CT)	Central lesion, sclerotic and well-defined	12	66	Curettage and grafting
5	F	42	Brown	Left intertrochanteric region	3 cm × 3 cm (CT)	Lytic lesion, well-defined, eccentric and sclerosis edge	60	66	Curettage and grafting
6	F	50	White	Left iliac and acetabulum	6 cm × 7 cm (CT)	Lytic lesion and multiple injuries and sclerosis edge	12	40	No treatment
7	F	39	White	Right intertrochanteric region and femoral neck	7 cm × 4 cm (CT)	Eccentric mixed lesion and sclerosis edge	48	29	Curettage and grafting
8	F	30	Brown	Right intertrochanteric region	5 cm × 6 cm (CT)	Lytic lesion, well-defined, eccentric and sclerosis edge	24	28	Curettage and cementation
9	F	27	Brown	Left neck, femoral head and intertrochanteric region	10 cm × 8 cm (CT)	Eccentric mixed lesion and sclerosis edge	12	14	Curettage and grafting

femur was the most affected bone (77.7%), with six cases (66.6%) in the proximal metaphysis and one (11.1%) case in the distal metaphysis. One case (11.1%) of LSMFT was observed in distal tibia and the other (11.1%) in the iliac (Table 1).

In general, radiographs typically showed a well-defined geographic lytic lesion with thick sclerotic margin, reflecting a pattern of slow growth. The bone contours were normal or showed expansion of cortical, but did not break it. Accelerated growth and cortical break were indicative of malignancy. The computed tomography (CT) images showed lytic lesions with sclerotic edge. Moreover, sclerotic and circinate patterning into tumor was observed. However, intralesional calcifications and break cortical were not found. The signal identified in magnetic resonance imaging (MRI) was high in T2-weighted and low in T1-weighted, with well-defined margins. The contrast injection usually shows a low or moderate intensity, as occurs in patients with fibrous dysplasia (Fig. 1). These characteristics can be explained probably due to the small amount of fat tissue and the abundance of myxofibrous and osteofibrous tissue.²

The surgical treatment was curettage with graft or cementation (intralesional margin). Six patients underwent allograft bone from tissue bank and one case was used bone cement in order to fill the tumor cavity. The case of malignancy was treated with transtibial amputation. After the diagnosis, one patient refused surgical treatment. This patient is being followed up in outpatient routine undergoing exams after 30 months of follow-up showed no evidence of malignancy, growth or metastasis.

The mean follow-up from histopathological diagnosis was 47 months (6–88 months). There were no complications related to surgery, recurrence, metastasis or death. No patient required chemotherapy or radiotherapy. One patient presented pain and deformity in the distal tibia. The biopsy was performed after 36 months of symptoms. Histopathological analysis showed several cellular aspects of malignancy. Thus, the patient underwent transtibial amputation, and after 88 months of follow-up, the recurrence or metastases were not observed (Fig. 2).

Histological sections revealed a polymorphic neoplasm characterized by predominant proliferation of stellate, fusiform, bipolar and oval cells arranged singly or in strings, which resulted in the myxoid matrix where there were also fat cells and histiocytes. It is also characterized by the formation of cystic cavities and septa. Bone foci of necrosis with fibrosis and amorphous calcifications were observed (Fig. 3).

The tissue from the patient with malignancy showed adipose tissue fragments with myxoid areas and proliferation of fusiform cells arranged in fibrous matrix and metaplastic bone formation and thick trabecular of poorly structured bone, with increased cellularity and atypical nuclei (Fig. 4).

Fig. 1. Liposclerosing myxofibrous primary bone tumor in proximal metaphysis in the right femur. Anteroposterior view showing circumscribed lytic lesion with well-defined sclerotic halo without cortical involvement (A). Computed tomography showing lytic lesion with sclerotic halo and small dense points into the lesion without cortical involvement (B). T1-weighted magnetic resonance showing hypointense lesion (C). T2-weighted magnetic resonance showing hyperintense signal (D).

Fig. 2. Radiological aspects of the malignancy of liposclerosing myxofibrous tumor in the distal metaphysis of the right tibia. Anteroposterior and lateral view showing destructive lytic lesion with intense board sclerosis and disruption of the lateral cortical (A and B). Computed tomography showing lytic lesion with break cortical and points of calcification inside (C).

4. Discussion

The LSMFT is a benign lesion of the bone which is characterized by complex combination of several histological elements including lipoma, fibroxanthoma, myxoma, myxofibroma, fibrous tissue, cyst formation, fat necrosis, ischemic bone tissue, and rarely cartilaginous tissue. Despite its histological complexity, the radiological features and skeletal distribution are typical.¹ LSMFT affects men and women equally in the fourth decade of life, with no ethnical predilection. Although this series had a predominance of

females (77.7%), the age range was consistent with other studies.¹ The lesions are usually discovered for reasons unrelated to injury (41% of cases). Pain and pathologic fractures are the most common symptoms seen in 48% and 10% of cases, respectively.^{1–3,7} As noted in this study, three patients (33.3%) had pathological fracture.

LSMFT has important predilection for the femur (85% of lesions), with 90% in the intertrochanteric region. Our results showed that seven patients (77.7%) had tumor in the femur, where six of them (85.7%) were in the proximal femur. The other sites are ilium, humerus and tibia. Radiographs show a well-defined lytic

Fig. 3. Histological sections of liposclerosing myxofibrous tumor stained with hematoxylin and eosin. Poorly structured bone trabeculae and numerous fusiform or oval cells setting pagetoid appearance supported by myxoid stroma (A). Osteoblasts forming osteoid matrix with irregular calcification (B). (Magnification of 4× in A and 40× in B.)

Fig. 4. Histological sections of liposclerosing myxofibrous tumor suggest low-grade osteosarcoma stained with hematoxylin and eosin. Trabeculae of metaplastic bone immersed in fibrous stroma, in which are observed groups of adipocytes and histiocytes (A). Bone trabeculae aspect in pagetoid and myxofibrous stroma with proliferation of fusiform cells (B). (Magnification of 10× in A and 20× in B.)

lesion, often with thick sclerotic margins, and sometimes with calcification into the inner part, reflecting slow growth pattern. The bone contour is normal or shows the expandable design in 30% of cases.^{1,2,4}

The matrix mineralization is common, and present in 72% of radiographs. A bone scan shows increased activity, but the uptake is usually mild to moderate, not as intense as typically seen in fibrous dysplasia. The presence of myxoid tissue explains the low attenuation in CT and hyperintense signal in MRI T2-weighted images. In addition, CT and MRI showed a well-defined lesion with marginal sclerosis and globular and irregular mineralized matrix.^{1,2}

Intraosseous lipoma is easily differentiated because the MR and CT offer the possibility of documenting intralesional adipose tissue. CT demonstrates low attenuation of fat. In MRI, the adipose tissue of intraosseous lipoma shows image similar to subcutaneous fat in T1 (hyperintense) and T2 (hyposignal).¹

Fibrous dysplasia reveals less sclerosis and increased activity in bone scintigraphy. The intensity signal of fibrous dysplasia in MRI varies. The standard of signal is low or moderate in fibrous dysplasia using T2-weighted images; however, the same signal is not often seen in LSMFT.^{1,2}

Pathological fracture of the proximal femur may require arthroplasty. These tumors have a high risk of malignant transformation, compared with fibrous dysplasia.^{1,2,4} Malignant transformation occurs because of malignant fibrous histiocytoma or osteosarcoma, and it was estimated to occur between 10% and 16% of cases. In this study, one patient (11.1%) had malignant tumor for low-grade osteosarcoma. The exact term for malignant transformation is not known but occurs over a prolonged period of time due to the slow progression of the lesion. Cortical destruction and soft-tissue extension are aggressive characteristics that may indicate malignancy.⁵ The tumors with these characteristics should be always diagnosed by percutaneous biopsy and histopathology analyses and treated by curettage and bone grafting. After histological diagnosis, the patient can choose the surgical or non-surgical treatment. In non-surgical treatment, the patient should be evaluated every three months in the first two years and the third year in each six months by magnetic resonance and computed tomography alternately.

Average follow-up for the patients with LSMFT was 39 months (range of 6–88 months). No patient in this study passed away during the follow-up period, and no patients had metastasis or recurrence. In conclusion, the LSMFT is a benign lesion of the bone that was characterized by complex combination of several histological elements. Most lesions were incidentally discovered in femur. Radiographs show a well-defined lytic lesion, often with thick sclerotic margins. Cortical destruction and soft-tissue extension are aggressive characteristics that may indicate malignancy. These characteristics suggest that the lesion may represent a variant of fibrous dysplasia, with a high risk of malignant transformation.

Conflict of interest

The authors declare to have no commercial interests (e.g. consultancies, equity interest, stock ownership, or patent/licensing arrangements). The authors also declare to have no conflict of interest in this article.

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