



## Case Report

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



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## ARTICLE INFO

## Article history:

Received 24 September 2015

Received in revised form 12 February 2016

Accepted 6 March 2016

Available online 26 March 2016

## Keywords:

Bone tumors

Liposclerosing myxofibrous tumor

Series of cases

## 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.<sup>1,2</sup> Despite its typical characteristics, it is considered an extremely rare tumor.<sup>3</sup> 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.<sup>4</sup> Genetic analysis suggested that the lesion may represent a variant of fibrous dysplasia.<sup>5</sup> 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.<sup>4,6</sup> 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.<sup>2</sup>

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).

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