Case Report

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

Eduardo Rinaldi Regado, Pedro Braga Linhares Garcia, Anabela Cunha Caruso, Ana Luzia Brito de Almeida, Ierece Lins Aymoré, Walter Meohas, Diego Pinheiro Aguiar *

Research Division, National Institute of Orthopedics and Traumatology/MS, Rio de Janeiro, RJ, Brazil

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.

Keywords: Bone tumors Liposclerosing myxofibrous tumor Series of cases

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.5,6 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%), pathological fracture (three patients – 33.3%) and local edema (two patients – 22%). The...
Bone foci of necrosis with fibrosis and amorphous calcifications myxoid matrix where there were also fat cells and histiocytes. It is 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).

Table 1
Location, size and treatment of liposclerosing myxofibrous tumor per patient.

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

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).

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).
دانلود مقاله

http://daneshyari.com/article/3251743

امکان دانلود نسخه تمام متن مقالات انگلیسی
امکان دانلود نسخه ترجمه شده مقالات
پذیرش سفارش ترجمه تخصصی
امکان جستجو در آرشیو جامعی از صدها موضوع و هزاران مقاله
امکان پرداخت اینترنتی با کلیه کارت های عضو شتاب
دانلود فوری مقاله پس از پرداخت آنلاین
پشتیبانی کامل خرید با بهره مندی از سیستم هوشمند رهگیری سفارشات