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Lipoesclerosing Myxofibrous Tumor: When The Site It Is Important

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1. Abstract

Liposclerosing Myxofibrous Tumor (LSMFT) is a benign fibro-osseous lesion with distinct clinical and radiological features, characterized by a complex mixture of histologic elements, including its fibrous dysplasia-like features, lipoma, myxofibrous tissue, ischemic ossification, xantoma features and pseudo- Paget's bone patterns. Most lesions are discovered incidentally, but patients can present with bone pain or fracture.

This lesion is considered by some researchers as a variant of fibrous dysplasia or as the non-specific result of degenerative change, while it is considered by others as a definite clinicopathologic entity. A small risk of malignant transformation has been described in these lesions that include fibrosarcoma, malignant fibrous histiocytoma and other soft tissue malignancies like osteosarcomas. Patients with asymptomatic LSMFT discovered incidentally usually may not require theraphy, however symptomatic lesions are treated with curettage, bone grafting and fixation with a favourable prognosis. We report a case of LSMFT located in the intertrochanteric region of the femur in a 42year-old-female.

2. Introduction

Liposclerosing Myxofibrous Tumor (LSMFT) is a benign fibroosseous lesion that has a marked predilection for the intertrochanteric region of the proximal femur [1, 2, 3]. The age range of the affected patients varies from the 2nd to the 7th decade of life with the mean age being about 40 years and an equal incidence of these lesion in both sexes [4]. Pain at the site of the lesion is the most common presenting clinical symptom, which varies markedly in duration with also asymptomatic patients reported [5]. Although rare, some patients may present with pathological fracture, following gradual enlargement of the lesion [5, 10].

The diagnosis of LSMFT relies on the combination of clinical presentation, radiographic findings and the complex mixture of histologic elements that include lipoma, fibroxanthoma, myxofibroma, fibrous dysplasia-like features, cyst formation, fat necrosis, ischemic ossification, and rarely cartilage [6, 7]. Characteristic findings to LSMFT if seen can be very helpful in suggesting the diagnosis; these include the location and the relatively characteristic radiologic appearence. Radiographs typically show a well-defined geographic lytic lesion with a patchy sclerotic rim, reflecting an indolent pattern of grown [3]. The bone contours are normal or show expansion of cortical and soft tissue mass component was usually not identified.

Computed Tomography (CT) provides further assessment of the matrix which, although of relatively decreased attenuation secondary to the myxoid component [6], is not grossly fatty, thus excluding an intraosseous lipoma [11].

The origin of this bone tumor is unclear and remains a source of discussion. Some authors consider LSMFT a clinicopathologic entity, while others regard it as a variant of fibrous dysplasia or as a nonspecific result of degenerative changes in other benign bone lesions [1, 6, 8]. Additionally, its similarity in location and histology to Fibrous Displasia (FD) has led many to interpret this lesion as a variant of FD, possibly through trauma [1]. Moreover, an activating point mutation in the alpha subunit of a G protein (GNAS) has been identified in cases of McCune-Albright syndrome as well as in some LSMFT samples. This mutation is present in nearly all

cases of monostotic and polyostotic fibrous dysplasia providing additional evidence for this relationship [9, 13-15]. However, other authors have suggested that this lesion is an involutional ischemic variant of intraosseous lipoma [10].

Because of his histologic heterogeneity, this tumor can be mistaken for a variety of lesions with potentially overlapping features, that include fibrous dysplasia (FD), intraosseous lipoma, nonossifying fibroma, cartilaginous neoplasms, osteoblastoma and bone infarct. A small risk (estimated between 10 and 16%) [5, 7] of malignant transformation has prompted close monitoring of this entity, but some authors have questioned this, attributing this alleged malignant behavior to mimicry of the LSMFT morphology by other, truly malignant lesions [12].

3. Case Report

The patient, a 42year-old female, with a history of uterine leiomyomata and plantar fasciitis, was referred to the traumatology clinic after the incidental finding of a right femoral bone lesion in an abdominopelvic MRI study that had been requested by her gynecologist. The lesion was in the intertrochanteric region, measured 6.8 x 3 x 3 cm, and was lytic with a heterogeneous matrix and a thick sclerotic rim. On interrogation, the patient complained of a diffuse and dull pain in her proximal right thigh, that was more intense in the trochanteric and gluteal region. No mass was felt on physical examination.

Further studies with plain radiography and magnetic resonance (RNM) revealed an intramedullary well defined lytic lesion in the right intertrochanteric region with sclerotic margins. No disruption of the cortex or periosteal reaction was seen (Figure 1). Although the overall appearance suggested a non-aggresive entity, the lesion was considered to have a significant risk of pathological fracture, and a surgical management with curettage of the tumor was decided.

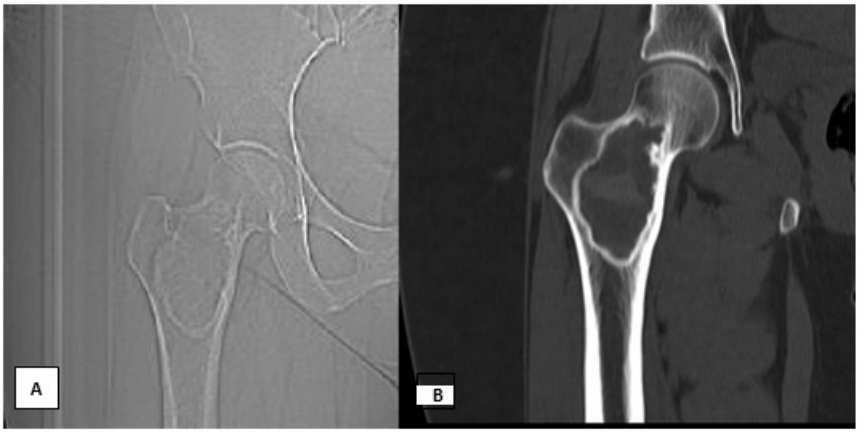

Figure 1: A) Radiographic findings described a well-defined lesion with geographic margins and a sclerotic rim without extension to soft tissue. B) RNM show with more detail the integrity of the cortical.

Histopathological examination of the curettage specimen showed an heterogenous lesion with two distinct components. One of them was composed of small curvilinear trabeculae of woven bone without osteoblastic rimming in a fibrous hypocellular stroma reminiscent of FD and the second one showed fibrous tissue with no bone formation and low cellularity. Tumor cells in the fibrous areas were spindled, resembling fibroblasts, and lacked any significant atypia. Mitotic figures were rare. Interestingly, abundant foamy macrophages could be seen in the fibrous areas. No adipose tissue was seen (Figure 2).

4. Discussion and Differential Diagnosis

As we mentioned previously it is important to make de differential diagnosis with other lesions in the bone that can show the same histologic patterns described in LSMFT. The main differential diagnosis includes FD, intraosseus lipoma, bone infarct, nonossifying fibroma and conventional low-grade osteosarcoma. These differential diagnoses should be made with clinical, radiographic, and histological findings.

FD, this is a benign fibroosseous proliferation that replace the normal bone marrow. This is more frequent in childhood and commonly occurs in the proximal femur [16]. The histological findings show a homogeneous pattern characterized by curved, Chinese character-like bony trabeculae without osteoblastic rim. However, the most important difference is that LSMFT is composed of constellation of histological features like fibroxanthomatous areas, adipose tissue, or irregular woven bone. This entity is associated with a somatic mutation in the GNAS gen [9, 13-15] like those seen in LSMFT. Some authors suggests that LSMFT is a traumatized variant of FD in response of weight-bearing stress in the femoral neck [10,17].

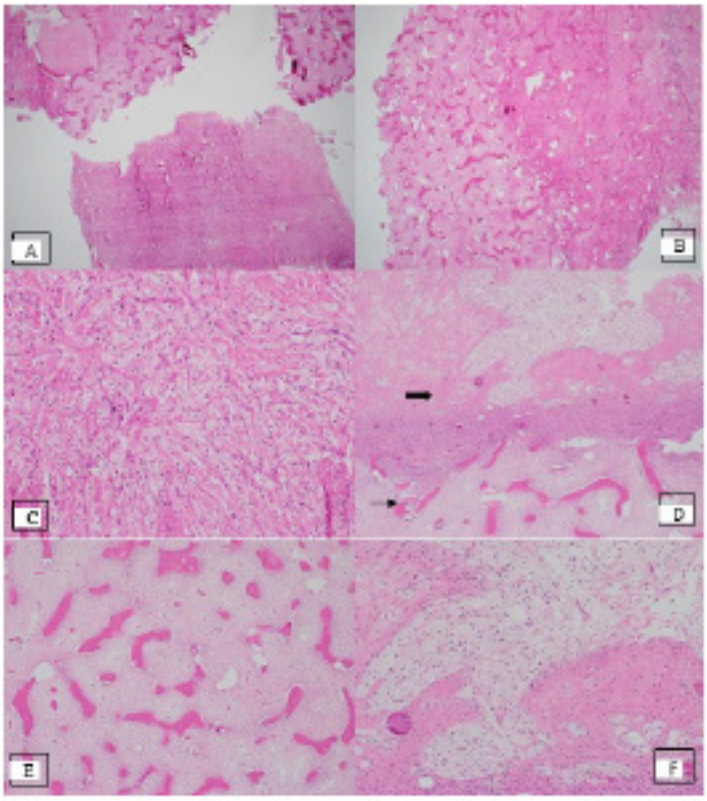

Figure 2: A) Histological section of LSMFT stained with hematoxylin and eosin (H&E) show different histological patterns. B) Other zones of the histological preparation show combination of hypocellular areas of the tumor with others with appearance of fibrous displasia-like bone trabeculae. C) High magnification of the stroma in this hypocellular areas show foamy histiocytes and myxofibrous stroma. D) Transition areas between areas of fibrous dysplasia-like woven bone and fibrous stroma with foamy histiocytes. E) Fibrous dysplasia-like woven bone trabeculae without osteoblastic rimming are seen. F) Aggregates of foamy histiocytes adjacent to areas of fibrous displasia-like woven.

Intraosseus lipoma is another lesion that has been included in the LSMFT entity. This is a rare bone tumor (<0,1% of all primary bone tumours) mostly discovered incidentally or after report of pain [18]. They have a wide age predominantly in the fifth decade. Predominantly affects the calcaneous and metaphysis of long tubular bones. The histological appearance consists of lobules of mature fat with delicate trabeculae of woven and pre-existing lamellar bone. This lesion may demonstrate fat necrosis, histiocytes and fibrosis but lacks the characteristic constellation of features seen in LSMFT [12, 16]. Fat necrosis can be seen in higher-stage lesions of intraosseous lipoma. Radiographically, presents a well-defined lytic mass with a sclerotic rim and cystic areas [19]. In our case we did not find any adipose tissue.

Bone infarct affects the femoral head and may also occur in other anatomical locations (e.g., shoulder, knee, and ankle). However, bone infarct does not affect the inter-subtrochanteric region of the femur [10]. The histological aspects are the same of osteonecrosis in other location. Nonossifying fibroma is one of the most common benign fibrous lesions that mainly affects young adults and could present with pain or asymptomatic. The most common region is the metaphysis of distal femur/proximal tibia. Histologically, consists of fibrous tissue with storiform pattern, often with scattered osteoclast-type giant cells. Radiographic findings typically are lytic, lobulated, well demarcated lesion with sclerotic rim. These images are similar to those seen in LSMFT, but its localization in the distal femur and in young adults distinguishes it from LSMFT and histological examination not include the wide variety of components seen in LSMFT [20].

Low grade osteosarcoma is the most important tumor to exclude because the risk and the prognosis of these entity. Supports approximately 1-2% of all osteosarcomas of the bone, with a peak of incidence in the third decade of life. These osteosarcomas are located predominately in distal femur and proximal tibia. The lack of permeation margins and the presence of sclerotic border in imaging studies exclude conventional osteosarcoma. The most useful histological findings that support low grade osteosarcoma include permeation of pre-existing bone and soft tissue extension [18].

5. Conclusion

In summary, LSMFT is a rare entity not universally recognized for all authors and presents heterogeneous histological pattern, radiological features, and location characteristic with an unespecific symptons. Unless most of the differential diagnosis of these entity are benign, it is important to recognize this tumor, not describe in many textbooks specialized in bone pathology. Although, radiological features are characteristic of these lesions, the heterogeneous

microscopic appareance support a challenge for the pathologists. It is important to know and to keep in mind these kinds of lesions to make a correct differential diagnosis with their principal mimics. We encouraged to discuss these cases with multidisciplinary comites for a better management. Finally, although it is thought that the risk of malignancy is think to be overestimated, we believe that it is not negligible, so we proposed a follow-up of LSMFT.

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