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Solitary Fibrous Tumor of the Leg: About A Case

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Abstract **Case Report**

Solitary fibrous tumor (SFT) is a rare tumor that usually originates in the pleura. Extrapleural locations are rare and, for this reason, sometimes difficult to diagnose. Malignant forms with local recurrence or distant metastases have been reported, often the result of inappropriate treatment. In this article, we report the case of SFT of the leg in a 45-year-old man. SFT of the leg is a rare occurrence, and the differential diagnosis can be difficult because it can mimic a variety of benign and malignant mesenchymal tumors; immunohistochemical analysis for CD34, CD99, vimentin and Bcl-2 is required.

Keywords: Solitary Fibrous Tumor, Leg Tumor.

INTRODUCTION

Solitary fibrous tumor (SFT) is an uncommon tumor, first described in 1931 as arising from the pleura. Subsequent studies have shown that SFT is a ubiquitous neoplasia, arising from intrathoracic organs as well as extra-thoracic regions, including soft tissues [1–3].

The symptoms of extrapleural SFT are nonspecific (painless, smooth, slow-growing mass) and are related to the presence of a mass in the deep soft tissues.

Biologically, SFT is currently classified as a "borderline" neoplasia, meaning that the lesion can reappear locally without almost ever metastasizing. To date, only a few cases of SFT originating from the limbs have been reported in the literature [1-4].

The current study describes the case of a young patient who presented with a solitary fibrous tumor of the leg. Radiological, histological and immunohistochemical findings are discussed as well as differential diagnosis, treatment and outcomes.

CASE REPORT

This is a 45-year-old man, a chronic smoker who presents a painless mass on the postero-internal aspect of the distal quarter of the leg which has gradually increased in volume and evolved over a period of 3 years. The clinical examination reveals a hard mass, well defined and immobile in relation to the deep plane without any abnormality of the adjacent skin. The vascular-nervous examination was normal (figure 1).

A standard x-ray plus magnetic resonance image was initially performed the x-ray does not show any bone lesion, however the MRI showed in T2 a hyper intense heterogeneous mass with areas of mixed signal in T1 a mass iso-intense in relation to the muscles (figure 2).

The pathological study of the biopsy reveals a tumor proliferation of a connective nature made of spindle cells devoid of atypia or mitotic figures and equipped with elongated nuclei with fine chromatin and scant cytoplasm; arranged within a fibrohyaline stroma.The immunohistochemical study confirmed the diagnosis of SFT by showing cell expression of CD34 and negativity for AE1/AE3, Desmin, PS100 and AML antibodies (figure 3).

The surgical procedure was carried out under locoregional anesthesia with the installation of a pneumatic tourniquet inflated to 350 mmHg which consisted of a resection of the entire tumor (figure 4).

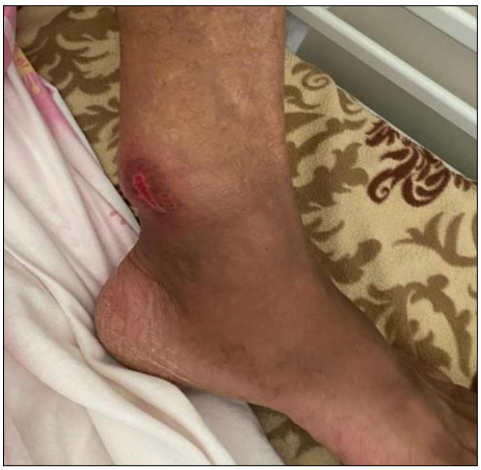


Figure 1

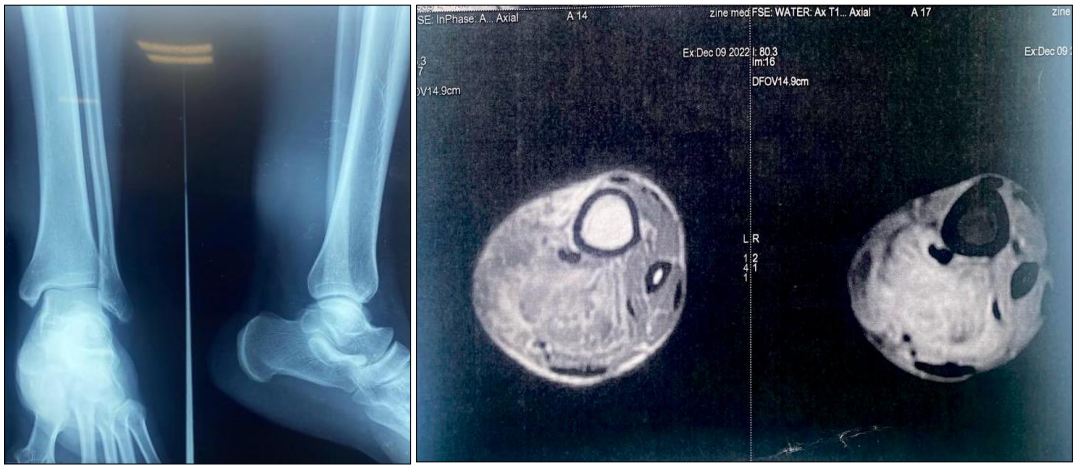


Figure 2

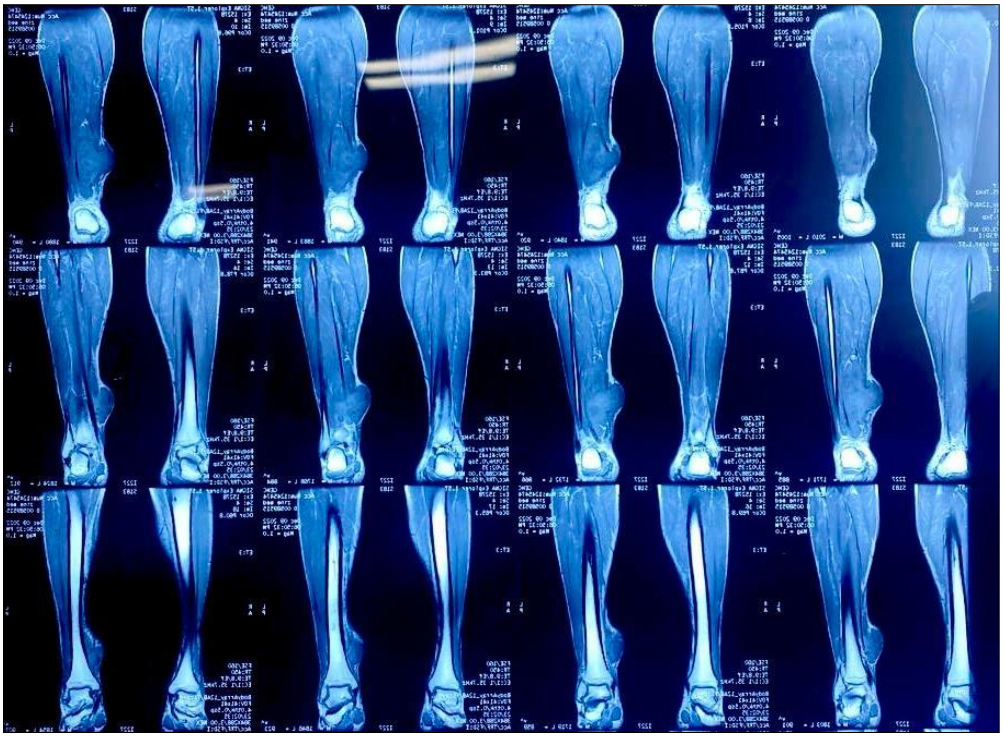


Figure 3

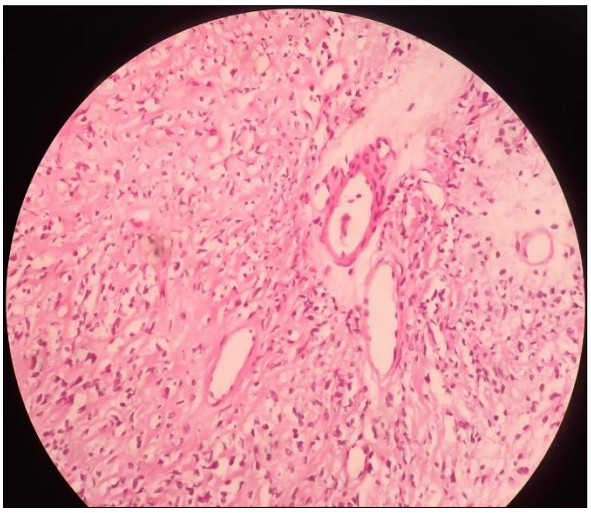
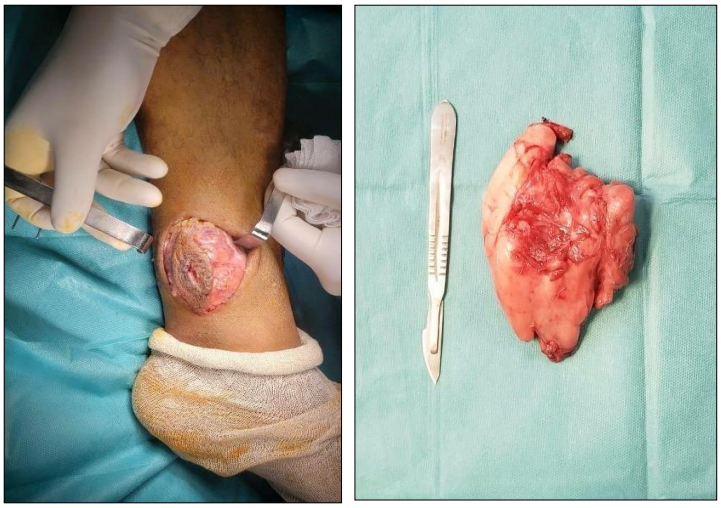


Figure 4



DISCUSSION

The reported case is that of a fibrous tumor of the leg in a young subject; a rare form that can mimic more common lesions; it may go unnoticed, making surgical planning inappropriate.

Symptoms related to extrapleural SFT are relatively nonspecific and simply refer to the presence of a mass: painless, smooth, slow growing; usually in deep soft tissues [2–6].

SFTs do not present specific radiological characteristics making their diagnosis more difficult: on standard radiography, they are radiopaque; on ultrasound, they appear as a nodule with well-defined contours and a homogeneous echostructure. MRI shows intermediate signal intensity on T1 and heterogeneous hyposignal with empty flow on T2 and enhancement after injection of contrast material. The absence of enhancement of the central areas and the long and tortuous vessels support the diagnosis.

The definitive diagnosis of SFT can only be obtained through histological and immunohistochemical studies. The histological features of soft tissue SFTs are often referred to as "patternless", characterized by numerous thin-walled branched blood vessels, with a partial "antler" configuration, with areas of spindle cells arranged in fascicles, or randomly with areas of hyalinization.

CD34 immunoreactivity is strong and is also positive for Bcl-2 and CD99. Negative immunohistochemical stains for vimentin, cytokeratins, smooth muscle actin, epithelial membrane antigen, desmin, c-kit (CD117), and S-100 protein are sometimes useful in the differential diagnosis of FTS versus tumors of muscular, epithelial or nervous origin, namely monophasic synovial sarcoma, malignant histiocytoma, fibrosarcoma and desmoid tumor.

A marked increase in cellularity, nuclear atypia, mitotic activity (>4 mitoses/10 fields at high magnification) and the presence of areas of necrosis should be considered as criteria for malignancy, and are associated with aggressive clinical behavior. SFTs can occur in all anatomical locations. Most case series show an almost equal distribution between male and female

patients, with ages ranging from the third to the eighth or ninth decades, with the peak incidence observed between the fifth and sixth decades.

SFTs have a slow growth rate and rarely tend to metastasize, behaving aggressively in 10-31% of cases. Recurrences range between 20 and 36% of cases. Sites of distant metastases included lung, liver, bone, mesentery, mediastinum and retroperitoneum.

According to the literature, two different concepts are known: complete surgical excision to obtain healthy margins or wide local resection. Depending on the involvement of adjacent soft tissues, a wider excision has been described [3-8].

Incomplete resection leads to local and distant recurrences. Although they may appear encapsulated, these lesions cannot simply be enucleated otherwise the risk of recurrence would be too high. To achieve a radical resection, a sufficiently wide exposure is necessary.

CONCLUSION

Most SFTs have a benign clinical course, but their behavior is unpredictable and there is no correlation between morphology and clinical behavior.

Radical surgical excision is considered the treatment of choice for SFT. However, close monitoring is necessary to detect recurrences.

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