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CASE REPORT
Reconstructive

Solitary Fibrous Tumor of the Lower Leg: A Rare and Difficult Diagnosis

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Summary: Solitary fibrous tumor (SFT) is a rare neoplasm that commonly 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, also as a consequence of inappropriate treatment. In this article, we report the case of an SFT of the lower leg in a 37-year-old man. Leg SFT is a rare occurrence, and differential diagnosis may be difficult because they can mimic a variety of benign and malignant mesenchymal tumors; immunohistochemical analysis for CD34, CD99, vimentin, and Bcl-2 is necessary. Misdiagnosis carries a significant risk of inadequate removal with subsequent increased risk of recurrence and distant metastases. *(Plast Reconstr Surg Glob Open 2015;3:e528; doi: 10.1097/GOX.0000000000000501; Published online 1 October 2015.)*

Solitary fibrous tumors (SFTs) are rare tumors, first described as pleural tumors of mesothelial origin. Extrapleural SFTs deriving from fibroblast-like cells[1] are an even more unusual variant.

We report a case of leg SFT and provide some clues for preoperative diagnosis and treatment plan.

CASE REPORT

A 37-year-old man presented with 2 painful lumps on the distal third of his left leg, which had appeared 2 years before and had grown progressively. One was on the anterolateral surface (8.5×7 cm), the other in the medial supramalleolar region (6×5 cm); both had regular ill-defined margins, hard consistency, and were fixed to the deep structures while gliding on the skin. Clinical examination showed a pulse synchronous with the peripheral pulses and a corresponding systolic murmur, confirmed with an 8-MHz Doppler probe, that allowed to trace the anterior (ATV) and posterior tibial vessels (PTV) entering the mass and exiting from it distally, mimicking an arteriovenous malformation. A preoperative ultrasound was suggestive for liquid content of the mass.

The magnetic resonance imaging (MRI) scan showed an hourglass-shaped mass that seemed to involve the peroneal artery, 6.6-cm diameter, fed by branches of the ATV and PTV. The mass seemed to have clear-defined margins and expanding growth. Angiography showed the peroneal artery entering the mass and branches from the anterior and posterior tibial arteries nourishing it (Fig.1).

The preoperative diagnosis was of a tumoral mass of vascular origin. Transperoneal removal was planned through 3 accesses: lateral, demolitive transperoneal approach for interosseous space and peroneal and tibial vessels exposure; anterior, to expose the anterior portion of the mass and the ATV; extended medial retromalleolar approach, to expose the mass and the PTV (Fig. 2).

During surgery, antibiotic prophylaxis was performed[2] and a white, hard mass was exposed

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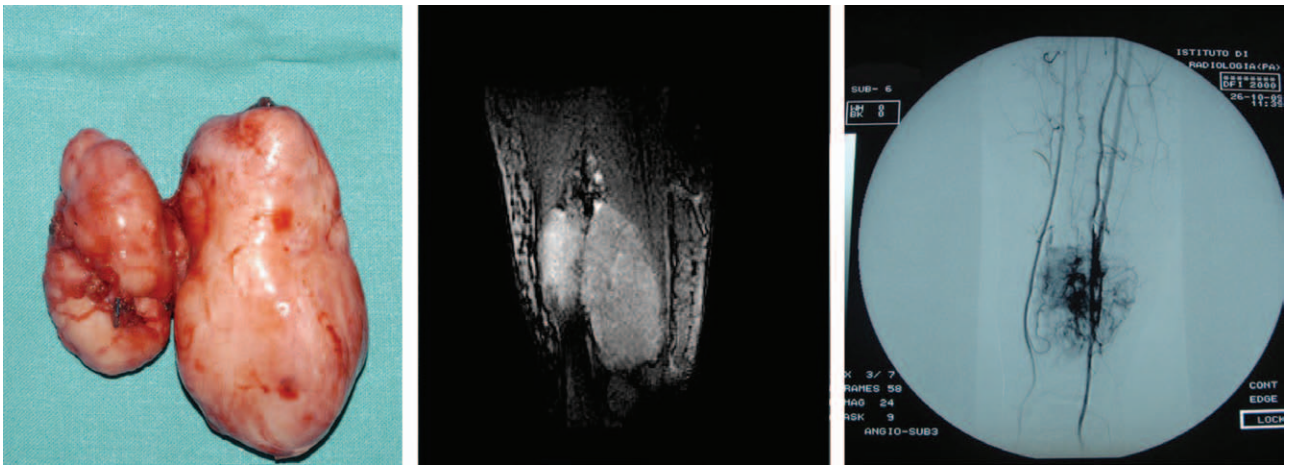

Fig. 1. This figure compares the resected specimen (A) with the MRI (B) and arteriography (C). The hourglass shape, due to the footprint of the interosseous membrane, is clearly visible in both A and B. The MRI shows a well-defined, homogeneously intense mass, findings suggestive of a tumoral lesion with expansile growth. The arteriography shows a peroneal artery that seems to end in the mass and the branches from the PTV and ATV that nourish the mass.

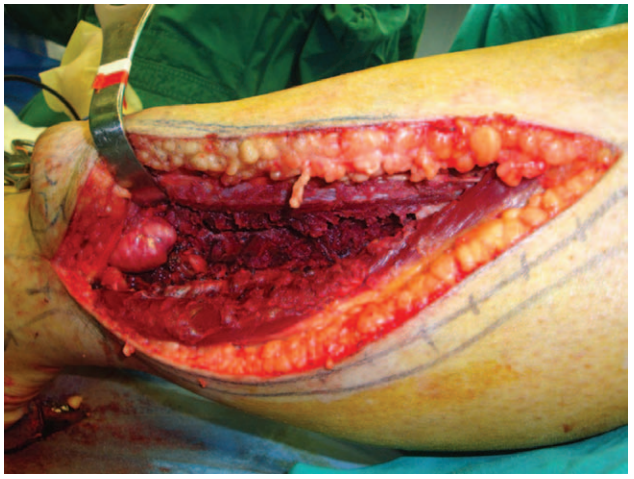

Fig. 2. Lateral view after fibula removal. This access provides wide exposure of the interosseous space and full control on the ATV and PTV and peroneal vessels, together with the tibial nerve. The mass is visible on the distal part of the field close to the tip of the retractor that holds the peroneal and anterior tibial muscles and the anterior neurovascular bundle. The planned peroneal vessels resection excluded preservation of the devascularized fibular segment removed for exposure because a 12-cm nonvascularised graft would have carried a high risk of failure and pseudoarthrosis.

(Figs. 1, 2). Frozen sections revealed a tumoral lesion with a vascular component. The mass was radically removed and sent for definitive pathology. The patient was immobilized for 5 days with gradual resumption of load.

The final pathology report revealed an SFT of mixed cellularity with clear margins and the following immunohistochemistry profile: CD34+, Bcl-2+, CD99±, CD31–, CK7–, Ki67 <5%. The patient underwent postoperative radiotherapy (54 Gy). At a 5-year follow-up, the patient is alive and free from disease; he complains only of a slight functional limitation and a limb swelling and a feeling of heaviness after a prolonged period of standing. His postoperative Lower Extremity Functional Scale score is 72/80[3].

DISCUSSION

The case reported is a rare disease that can mimic more common lesions; it might pass unsuspected, making surgical planning inappropriate. The purpose of this manuscript is to raise awareness about SFT and provide clues for diagnosis and treatment in such difficult anatomic location. SFTs lack specific radiological features: on x-ray, they are radiopaque; on ultrasound, they seem as a nodule with well-defined edges and a homogenous echostructure.[4] The case reported shows how clinical and imaging data might be misleading. MRI shows an intermediate signal intensity in T1 and a heterogeneous low signal with empty flow in T2 and contrast enhancement. Nonenhancing central areas and long, tortuous vessels support diagnosis. MRI allows differential diagnosis with vascular malformations (VMs): Low-flow VMs show heightened signal intensity on T2; high-flow VMs have an empty signal. A definitive diagnosis of SFT can only be achieved through histological and immunohistochemical studies. Histological features of soft tissue SFTs are frequently referred to as "patternless," characterized by numerous thin-walled ramifying blood vessels, with a partial "staghorn" configuration, with spindle cell areas arranged in short, ill-defined fascicles, or randomly with areas of striking hyalinization with cells surrounding ramifying vascular network. CD34 immunoreactivity is strong

and diffuse as well as positive for Bcl-2 and CD99. Negative immunohistochemical stainings for vimentin, cytokeratins, smooth muscle actin, epithelial antigen membrane, desmin, c-kit (CD117), and S-100 protein are sometimes useful for differential diagnosis of SFT from tumors of muscle, epithelial, or neural origin.[1] The lack of a specific growth pattern in an SFT lesion makes the differential diagnosis often challenging. Due to the similar growth pattern, SFT and hemangiopericytomas are considered the same entity by Gengler and Guillou, Fletcher, and Fletcher et al,[1,5,6] and the term hemangiopericytoma has been abandoned. SFTs are "typical" or "malignant" based on number of mitoses, cellular atypia, presence of necrosis, and hypercellularity. SFTs can occur in every anatomic location. Most case series showed almost equal distribution for male and female patients, with ages ranging from the third to the eighth or ninth decade with a maximum incidence in the fifth to sixth decade.[1,7]

SFTs have a slow growth rate and rarely tend to metastatize,[7] behaving aggressively in 10–31% of cases. Recurrences range from 20–36%.[8] Incomplete resection leads to local and distant recurrences. Although they might seem encapsulated, these lesions cannot be just enucleated or the risk of recurrence will be too high.[9] For a radical resection to be obtained, an appropriately wide exposure is mandatory. In this case, 3 incisions and removal of the fibula were necessary to remove the mass without disrupting it and causing potential spread or sacrificing radicality of resection. Furthermore, this exposure facilitates vessels exposure and control.

CONCLUSIONS

Leg SFTs are a rare occurrence and can mimic VM. These tumors must be taken into account in cases of hard masses of likely vascular origin that show uncertain features on MRI. In the interosseous region of the lower leg, wide exposure using a conservative transperoneal access and multiple incisions when needed allow to achieve good control of the mass.

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