

# Leiomyosarcoma of the Femoral Vein Mimicking a Peripheral Nerve Sheath Tumor: A Case Report <sup>1</sup>

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Primary vascular leiomyosarcoma is a rare malignant tumor arising from the smooth muscle cells of the media on the vessel wall. We describe here the ultrasonography (US) and magnetic resonance (MR) findings, as well as pathologic correlation, of a leiomyosarcoma of the femoral vein in a 42-year-old woman. US revealed a spindle-shaped mass with a connection to the thrombosed femoral vein. We initially misdiagnosed this mass as a peripheral nerve sheath tumor due to the misinterpretation of the thrombosed vein as a nerve bundle.

**Index words :** Leiomyosarcoma  
Soft tissue, neoplasms  
Veins, femoral

Primary vascular leiomyosarcoma is a rare malignant tumor originating from the smooth muscle cells of the vessel wall media. 210 cases of primary venous leiomyosarcomas were found in a review of the literature in 1992 by Dzsini et al. (1). Vascular leiomyosarcomas arise from the large veins rather than the arteries, and the inferior vena cava is the most common location: it represents about 50 percent of all locations (2, 3). One third of these tumors involve the veins of the lower extremity, which are saphenous, femoral, iliac and popliteal veins in the decreasing order of tumor incidence (2, 4). We describe here a case of leiomyosarcoma of the femoral vein in a 42-year-old woman that was initially misdiagnosed as a peripheral nerve sheath tumor such as a neurofibroma or neurilemmoma.

## Case Report

A 42-year-old woman presented to us with a painful mass in the left thigh. The mass was first found 4 months earlier and it has shown no significant increase in size before her hospital admission. She had a 6-year history of intermittent thigh pain that was aggravated with walking. The physical examination revealed a firm, non-tender and non-pulsatile mass in the antero-medial aspect of the mid-portion of the left thigh. There was no edema of the lower extremities. The laboratory findings were all within the normal ranges.

Ultrasonography (US) was done on the left thigh, and it revealed a well-circumscribed homogeneously hypoechoic mass that measured 4 × 3 × 3 cm, and it was adjacent to the femoral artery (Fig. 1). The mass was spindle shaped and it had a distal connection to the tubular structure, and this was retrospectively confirmed as the thrombosed femoral vein. We initially misinterpreted the thrombosed vein as a nerve bundle, and so we misdiagnosed the mass as a peripheral nerve sheath tumor.

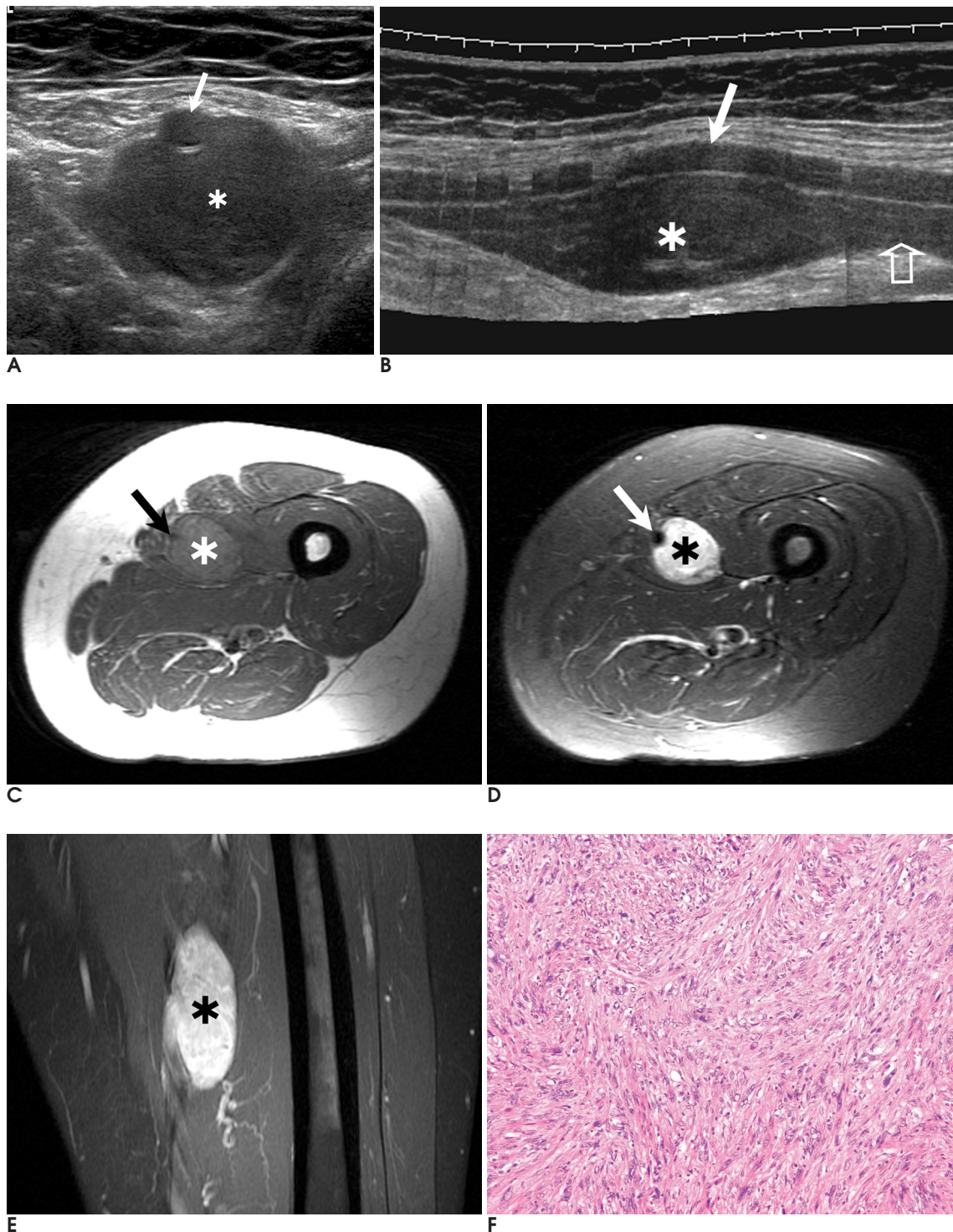
Magnetic resonance (MR) imaging demonstrated a

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**Fig. 1.** 42-year-old woman with a leiomyosarcoma of the femoral vein.

**A.** Axial US shows an ovoid hypoechoic mass (asterisk) and the adjacent femoral artery (arrow).

**B.** Longitudinal extended-field-of-view US shows a spindle-shaped mass (asterisk) with a distal connection to the thrombosed femoral vein (open arrow), posterior to the femoral artery (arrow).

**C-E.** The mass (asterisk) is seen posteromedial to the femoral artery (arrow), and it shows intermediate to low signal intensity on the T1-weighted axial MR image (**C**), and intermediate to high signal intensity on the fat-suppressed T2-weighted MR image (**D**). The coronal fat-suppressed T1-weighted MR images with contrast enhancement (**E**) reveal diffuse enhancement of the mass (asterisk) without any areas of necrosis.

**F.** Photomicrograph shows interlacing bundles of spindle cells with a palisading or fascicular arrangement. (H & E stain,  $\times 100$ ) The individual tumor cells are pleomorphic and anaplastic with abundant cytoplasm, and this is accompanied by tripolar mitotic figures (not demonstrated).

lobulated mass that was continuous with the femoral vein (Fig. 2). The mass was located posteromedial to the section of the femoral artery that was between the adductor longus, adductor brevis and vastus intermedius muscles. The mass showed an intermediate to low signal intensity on the T1-weighted images, and it was of near-uniform intermediate to high signal intensity on the fat-suppressed T2-weighted images. On the fat-suppressed T1-weighted MR images with contrast enhancement, the mass demonstrated diffuse enhancement without any areas of necrosis. The femoral vein was dilated and engorged distal to the mass, and collateral veins were seen.

The mass was excised, and it easily dissected away from the muscle and the femoral artery during surgery. The mass was confirmed to arise from the femoral vein and it showed a predominant intraluminal growth. The femoral vein was completely occluded by the mass.

The surgical specimen consisted of a multilobulated solid tumor that measured 4×3×3 cm, and it was brown to pink in color. The light microscopic examination revealed that the tumor was mainly composed of spindle cells that were arranged in bundles and interlaced with collagen fibers. The diagnosis of leiomyosarcoma was confirmed by the positive immunostaining with desmin and smooth muscle specific actin (Fig. 3).

## Discussion

Leiomyosarcoma is a highly malignant tumor that originates from the smooth muscle cells. This tumor represents approximately 9 percent of all the soft tissue sarcomas and it is the third most common tumor following malignant fibrous histiocytoma and liposarcoma (2). Vascular leiomyosarcoma is a rare tumor that represents only 2 percent of all leiomyosarcomas, and it involves veins five times more often than arteries (2, 3). The inferior vena cava is the most common location of vascular leiomyosarcomas, which represents about 50 per cent of all the locations (2, 3, 5). The great saphenous vein is next most common location with the remainder of the tumors arising from the femoral, internal jugular and iliac veins, in their decreasing order of incidence (2, 4 - 7).

Venous leiomyosarcoma is a relatively slow growing tumor and its size is usually variable. The tumor growth may be intraluminal or extraluminal, and usually both components are present (2, 6). Intraluminal tumor growth usually follows the direction of the blood flow (2). The site of tumor origin within the vein may be

broad or narrow (6).

The clinical presentation of a venous leiomyosarcoma is variable according to the location and pattern of the tumor growth, and a patient usually presents with no clinical manifestations before the tumor occludes the vascular lumen (5). In the lower extremities, the main clinical findings are palpable mass, edema and ill-defined pain, and these symptoms may mimic the symptoms of deep vein thrombosis (4, 5, 7). When the tumor arises from the superficial veins, such as a greater saphenous vein or a common femoral vein, diagnosis is generally achieved by palpation at a smaller sized tumor (5). The patient typically experiences lower extremity edema when the tumor produces venous obstruction, but the patients are asymptomatic in most cases since the collateral circulation is sufficient to compensate for this venous obstruction (5). Although the common causes of lower extremity swelling include deep venous thrombosis, mechanical causes, arteriovenous fistula, varicose vein and lymphedema, vascular tumor also must be considered as an infrequent cause of leg pain and swelling (7). Vascular tumor can lead to secondary thrombus formation without total venous obstruction (7). Pain rarely occurs unless a palpable mass is present (7).

The early and correct diagnosis of venous leiomyosarcoma is quite important: US, MR imaging, computed tomography (CT) and angio-CT are useful imaging techniques for uncovering the nature of the lesion and for performing the vascular mapping of the lower limb. US was the first diagnostic method used for patients having a palpable soft tissue mass. US of a venous leiomyosarcoma demonstrated a nonspecific well-defined mass with variable echogenicity (6). Color Doppler US may allow identification of the vascular origin of a mass, but it is difficult to distinguish a intraluminal tumor from a blood clot with this modality (6). In our case, US showed a spindle-shaped mass with a long distal tubular structure, and we initially misdiagnosed this lesion as a peripheral nerve sheath tumor due to the misinterpretation of the thrombosed vein as a nerve bundle.

MR imaging has become the method of first choice for the detection and staging of soft tissue masses due to its high soft tissue contrast and its ability to produce multiplanar sectional images (4). MR imaging is also useful for evaluating the extraluminal soft tissue invasion and the intraluminal extension of a venous leiomyosarcoma (6). The MR signal intensity of the venous leiomyosarcoma is nonspecific: there is low to intermediate signal in-

tensity on the T1-weighted images and high signal intensity on the T2-weighted images (4 - 6). Blood clots can be distinguished from tumor by their high signal intensity on the T1-weighted images (6).

CT is useful for showing the intraluminal and extraluminal extensions of venous leiomyosarcoma, but it is not so easy to distinguish a tumor from a blood clot with this modality (6). CT angiography is particularly helpful in evaluating the full intraluminal extent of the tumor (4).

Angiography has been used to diagnosis vascular tumor by demonstrating the intraluminal tumor and the narrowing of the vascular lumen. At present, phlebography is used for mapping the collateral circulation and the patent vessels when venous reconstruction is contemplated (5).

Management of venous leiomyosarcoma of the lower extremity is difficult because the incidence of this tumor is very low and most reports have been single-case studies. The best treatment seems to be complete surgical excision or when this is not possible, at least partial surgical excision accompanied by aggressive radiotherapy (3). The prognosis for these neoplasia is uncertain because only a few cases have been reported on. From the results of the reports by Dzinich *et al.* (1) and Berlin *et al.* (8), the prognosis seems to be poor with a median survival of only 31 months, and this poor prognosis is due to the early occurrence of metastasis. Metastasis is the main cause of death and it usually occurs within 2 years after treatment of the initial tumor.

US including color Doppler US, MR imaging and CT can be helpful modalities for the preoperative diagnosis

of a venous leiomyosarcoma involving the extremities. Even though this is a rare tumor, the physicians' awareness about the imaging findings for venous leiomyosarcoma is important for the early detection and a correct diagnosis. We initially misdiagnosed this tumor as a peripheral nerve sheath tumor due to our misinterpretation of the thrombosed vein as a nerve bundle. We report here on a case of venous leiomyosarcoma of the femoral vein and we have included the US and MR imaging findings.

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