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Primary Leiomyosarcoma Of The Femoral Vein : A Case Report And Review Of The Literature

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Abstract

Primary leiomyosarcoma of vascular origin is an exceptionally rare malignant tumor arising from the smooth muscle cells of the vessel wall. It represents less than 2% of all leiomyosarcomas and most commonly involves the inferior vena cava. Peripheral venous involvement, particularly of the femoral vein, is extremely uncommon and frequently mimics deep vein thrombosis, leading to diagnostic delay. We report the case of a 50-year-old woman with primary leiomyosarcoma of the left femoral vein initially misdiagnosed as deep vein thrombosis, who subsequently developed local recurrence and pulmonary metastatic progression. This case illustrates the diagnostic pitfalls, aggressive behavior, and therapeutic challenges associated with vascular leiomyosarcoma.

Keywords

Leiomyosarcoma; Femoral vein; Vascular sarcoma; Deep vein thrombosis mimic; Case report

1. Introduction

Leiomyosarcoma is a malignant mesenchymal tumor originating from smooth muscle cells and accounts for approximately 5–10% of all soft tissue sarcomas [1]. Vascular leiomyosarcomas are particularly rare, representing less than 2% of cases, and most frequently arise from the inferior vena cava [2]. Involvement of peripheral veins, such as the femoral vein, is exceptional and has been reported mainly in isolated case reports and small retrospective series [3,4].

Because these tumors grow within the vessel lumen, they often present with signs of venous obstruction, closely mimicking deep vein thrombosis and leading to inappropriate initial management [5]. Vascular leiomyosarcomas are aggressive tumors with a high risk of local recurrence and distant metastases, particularly to the lungs [6]. We report a rare case of primary femoral vein leiomyosarcoma with early local recurrence and dissociated metastatic progression under systemic therapy.

2. Case Presentation

A 50-year-old woman with a medical history limited to well-controlled arterial hypertension presented with progressive pain and swelling of the left lower limb. Initial clinical evaluation and Doppler ultrasound findings were suggestive of deep vein thrombosis, and anticoagulant therapy was initiated. Due to the absence of clinical improvement and the persistence of atypical imaging features, surgical exploration was performed in the vascular surgery department.

Intraoperatively, an intraluminal mass arising from the left femoral vein was identified and resected. Histopathological examination revealed a malignant spindle cell proliferation arranged in intersecting fascicles. Tumor cells exhibited elongated hyperchromatic nuclei and eosinophilic cytoplasm. Immunohistochemical analysis demonstrated diffuse positivity for smooth muscle actin, desmin, and h-caldesmon, confirming smooth muscle differentiation, while endothelial and neural markers were negative. The tumor was classified as **grade 2 according to the FNCLCC grading system**, with a Ki-67 proliferation index of approximately 18%, consistent with an intermediate-grade leiomyosarcoma [1].

Histopathological examination revealed a malignant spindle cell proliferation with smooth muscle differentiation on immunohistochemistry (Figure 1).

Nine months after surgery, routine surveillance with thoraco-abdomino-pelvic computed tomography revealed multiple pulmonary nodules suspicious for metastatic disease, a common dissemination pattern in vascular leiomyosarcoma [6]. An angio-computed tomography scan of the lower limbs demonstrated a locally recurrent endovascular tumoral process involving the left femoral vein, extending along the femoral canal with infiltration of adjacent vascular structures. Complete surgical resection was considered impossible because of extensive vascular involvement and the high risk of acute limb ischemia.

A positron emission tomography scan using 18F-fluorodeoxyglucose showed intense hypermetabolic activity confined to the recurrent femoral vein lesion, with no other hypermetabolic foci.

Treatment and Outcome

Given the unresectable nature of the local recurrence and the presence of pulmonary lesions, systemic chemotherapy was initiated. The patient received doxorubicin as first-line treatment, in accordance with international recommendations for advanced soft tissue sarcomas [7]. After three cycles, radiological reassessment demonstrated a significant local response, with a 42% reduction in the size of the endovascular femoral vein tumor. However, despite this favorable local response, there was a concomitant increase in both the size and number of pulmonary nodules, indicating metastatic progression.

This dissociated response pattern led to multidisciplinary discussion. In line with published evidence supporting alternative agents after anthracycline failure in leiomyosarcoma [8,9], the disease was considered progressive, and a decision was made to switch to gemcitabine as second-line systemic therapy.

3. Discussion

Primary leiomyosarcoma of the femoral vein is an exceptionally rare entity. Large historical series have shown that the majority of vascular leiomyosarcomas arise from central veins, while peripheral venous involvement remains uncommon [3,4].

When present, femoral vein tumors typically manifest with limb swelling and pain, closely resembling venous thromboembolic disease, as observed in the present case [5].

Previously reported cases and series of primary venous leiomyosarcoma involving peripheral veins are summarized in **Table 1.**

Imaging plays a central role in diagnosis and staging. While Doppler ultrasound is often the first-line investigation, it is limited in differentiating tumor from thrombus. Cross-sectional imaging with contrast-enhanced computed tomography or magnetic resonance imaging is essential to characterize the lesion, assess vessel wall involvement, and evaluate local extension [10]. Positron emission tomography can provide complementary information regarding metabolic activity and may help exclude distant disease.

Histopathological examination with immunohistochemical analysis remains the cornerstone of diagnosis. Expression of smooth muscle markers such as smooth muscle actin, desmin, and h-caldesmon confirms smooth muscle differentiation and helps distinguish leiomyosarcoma from other intravascular malignancies. The FNCLCC grading system remains a reliable prognostic tool, with higher grades associated with increased risks of recurrence and metastasis [1].

Complete surgical resection with negative margins remains the cornerstone of treatment and offers the best chance for local control [11]. However, in peripheral vascular locations, achieving adequate margins is often challenging due to anatomical constraints. Consequently, local recurrence rates are high, particularly in cases with close or positive margins. Systemic chemotherapy follows the principles applied to soft tissue sarcomas, with doxorubicin as standard first-line therapy and gemcitabine-based regimens representing validated options in subsequent lines [7–9].

The dissociated response observed in our patient, characterized by significant local tumor regression but simultaneous metastatic progression, highlights the biological heterogeneity and aggressive nature of vascular leiomyosarcomas.

Clinical Implications

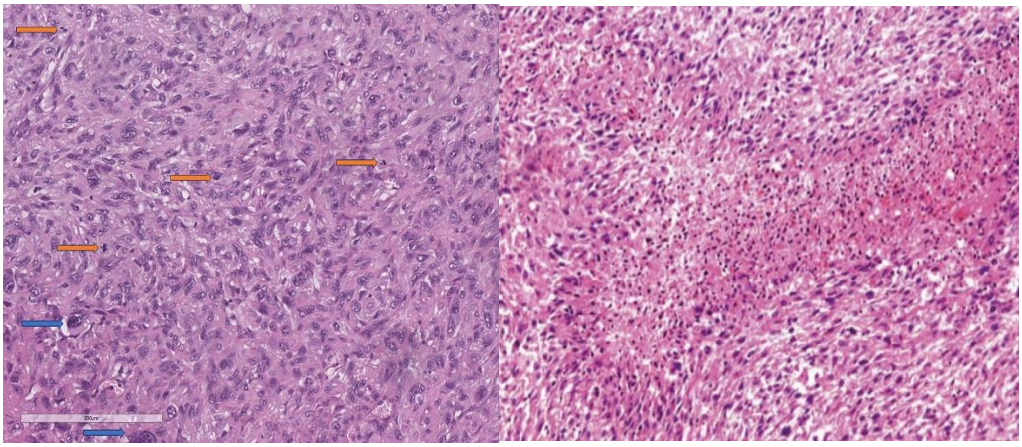
This case underscores the importance of considering vascular leiomyosarcoma in patients presenting with presumed deep vein thrombosis that fails to respond to adequate anticoagulation. Early suspicion and timely use of cross-sectional imaging may reduce diagnostic delay and improve management. The case also illustrates the need for close radiological monitoring during systemic therapy, as dissociated responses may occur. Finally, management should be centralized in specialized sarcoma centers, where

multidisciplinary expertise allows individualized treatment strategies and rapid adaptation of therapy in the event of disease progression.

4. Conclusion

Leiomyosarcoma of the femoral vein is a rare and aggressive malignancy frequently misdiagnosed as deep vein thrombosis. Early recognition, appropriate imaging, and histopathological confirmation are essential. Despite surgical resection and systemic therapy, the risks of local recurrence and metastatic progression remain high, emphasizing the need for multidisciplinary management and close follow-up.

Figure 1. Histopathological and immunohistochemical features of femoral vein leiomyosarcoma



Histopathological examination shows a malignant spindle cell proliferation arranged in intersecting fascicles. Tumor cells display elongated nuclei with eosinophilic cytoplasm, consistent with leiomyosarcoma

Table 1. Published cases and series of primary venous leiomyosarcoma involving peripheral veins

Author (Year)	Tumor Location	No. of Patients	Initial Presentation	Treatment	Outcome
Kevorkian & Cento (1973) [4]	Peripheral veins	12	Limb swelling	Surgery ± RT	High local recurrence
Dzsinich et al. (1992) [3]	Major & peripheral veins	34	DVT-like symptoms	Surgery ± CT	Poor long-term survival
Hines et al. (1999) [6]	IVC vs other veins	57	Venous obstruction	Surgery ± CT	High metastatic risk

Author (Year)	Tumor Location	No. of Patients	Initial Presentation	Treatment	Outcome
Hong et al. (2016) [11]	Femoral vein	1	Suspected DVT	Surgery	Local recurrence
Narayanan et al. (2019) [5]	Femoral vein	1	DVT-like presentation	Surgery + CT	Pulmonary metastases
Sheaffer et al. (2022) [11]	Venous LMS	16	Limb edema	Surgery ± reconstruction	Frequent recurrence
Present case (2026)	Femoral vein	1	Misdiagnosed DVT	Surgery + CT	Local recurrence and lung progression

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