Vascular Leiomyosarcoma of the Thigh: A Rare Presentation

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ABSTRACT
High-grade mesenchymal soft tissue tumors are rare neoplastic lesions that occur uncommonly in the extremities and in other sites of the body. Herein, we report a case of high-grade vascular leiomyosarcoma of the thigh. The patient was a 72-year-old male with a 4-day history of swelling in his right thigh. Excision biopsy of the tumor showed highly pleomorphic tumor cells arranged typically in perivascular fashion with large areas of hemorrhage and necrosis. Immunohistochemistry was positive for vimentin, smooth muscle actin, and showed a 90% proliferation index on ki-67 labeling. Vascular leiomyosarcomas comprise a group of very infrequent tumors with varied presentation and can occur at unusual sites.

Keywords: Leiomyosarcoma, Malignant, Mesenchymal tumor, Vascular.

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INTRODUCTION
Soft tissue sarcomas are malignant tumors that arise in any of the mesodermal tissues of the extremities (50%), trunk and retroperitoneum (40%), or head and neck (10%).1 The reported international incidence rates range from 1.8 to 5 per 100,000 per year.2 Histopathological examination along with ancillary immunohistochemical stains remains a gold standard approach to classify these tumors to determine the cell of origin, and assess their behavior, prognosis, and recurrence. The American Joint Committee on Cancer (AJCC) staging system for soft tissue sarcomas is based on histologic grade, tumor size and depth, and the presence of distant or nodal metastases. Despite improvements in local control rates with wide local resections and radiation therapy, metastasis and death remain a significant problem in 50% of patients who present with high-risk soft tissue sarcomas. Herein, we report an unusual case of a high-grade leiomyosarcoma.

CASE REPORT
A 72-year-old male presented to the surgical outpatient department with a right-sided thigh swelling, which he had noticed only 4 days back. The swelling was painless, nontender, firm to hard in consistency, and occupied the lateral aspect of the thigh. The surgeon carried out wide excision of the swelling. We received several morcellated, grey-white, soft to firm tissue pieces of the tumor, all aggregating 15 x 10 x 4 cm, with areas of hemorrhage and necrosis. Histopathological examination revealed a tumor comprising of tumor cells arranged in predominantly perivascular and alveolar pattern, vague fascicular, and solid sheets separated by confluent areas of necrosis. Individual tumor cells were round to oval, occasionally spindled, showing high N:C ratio, and marked nuclear pleomorphism with hyperchromic nuclei and eosinophilic to vacuolated cytoplasm giving an epithelioid-visá- vis rhabdoid morphology. Multiple tumor giant cells, atypical mitoses, and areas of calcification were noted. There was no evidence of myxoid, osseous, cartilaginous areas, or any lipoblasts or pigment in the tumor cells (Figs 1A and B). According to the French Federation of Cancer Centers Sarcoma Group (FNCLCC), a score of 3 + 2 + 2 = 7, inferring grade 3 histologically was reported. A preliminary diagnosis on morphology was that of high-grade malignant neoplasm favoring sarcoma.

On immunohistochemistry, the tumor cells were strongly positive for vimentin and smooth muscle actin, and negative for pancytokeratin, desmin, CD34, HMB-45, leukocyte common antigen, myogenin, and s100. Ki-67 and Bcl-2 positivity was seen in 90 and 30% of tumor cells respectively (Figs 2A to D). Thus, a diagnosis of leiomyosarcoma was rendered.

DISCUSSION
Leiomyosarcoma is a malignant neoplasm arising from smooth muscle cells which affects individuals of all ages, especially between the 5th and 7th decade of life. Leiomyosarcomas of vascular origin comprise a seemingly rare group of tumors with only a few hundred cases reported in the literature and only isolated instances are recorded in several large autopsy series.3 The morbidity
and mortality associated with these tumors are primarily a result of direct extension of the tumor along vessels, compromising the circulation. On the contrary, epithelioid leiomyosarcoma in the external deep soft tissue is extremely rare. Site-specific superficial leiomyosarcomas are divided into cutaneous (or dermal) and subcutaneous leiomyosarcomas.

In general, leiomyosarcomas are uncommon tumors and thought to have poor long-term prognosis. Svarvar et al. reported on 225 patients with leiomyosarcoma of all types from the Scandinavian Sarcoma Group with a cumulative survival of 49% at 10 years.

Various authors have reported the behavior of vascular leiomyosarcomas. Leu and Makek reported good

![Figures 1A and B: Tumor cells with notable perivascular arrangement and vague fascicles with marked nuclear pleomorphism (A: ×10, B: ×40, H&E stain).](image)

![Figures 2A to D: (A) Positivity for vimentin; (B) smooth muscle actin; (C) Ki-67 proliferation of 90%; and (D) Bcl-2 index of 30%.](image)
prognosis on 5 cases of intramural venous leiomyosarco-
mas, and Hadju\(^8\) reported low metastatic potential. On
the contrary, all the cases reported by Berlin et al\(^9\) had
metastases with 5 cases dying from metastatic disease
within 5 years. The results of one series showed a very
poor outcome in vascular leiomyosarcoma with 75% of
patients dying of metastatic disease within the first 3 years
diagnosis, although good local control by surgery and
radiotherapy was achieved. This is very similar to the
series of Berlin et al.\(^9\) In their experience about half of
the patients with leiomyosarcomas of vascular origin had
metastatic disease at diagnosis, which indicated either
a very aggressive course of the disease or due to delay
in diagnosis, as most patients had been misdiagnosed
as deep venous thrombosis. Of the 9 patients without
metastases at the time of diagnosis, 5 patients developed
metastases within 36 months of the disease, indicating
that the tumors had an aggressive clinical course.\(^{10}\)

Our patient died after 5 months of follow-up. Progress
in the molecular characteristics of these tumors should
in the near future translate into molecular based therapi-
es that can be incorporated into standard treatment
strategies.

**CONCLUSION**

Leiomyosarcomas, being rare, should be kept in the dif-
fferential diagnosis of high-grade tumors occurring in the
extremities. Immunohistochemistry plays an important
role in diagnosing such high-grade tumors with an
unusual morphology. Thorough sampling, patient history,
and clinical correlation with further ancillary testing are
mandatory and the need of the hour.

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