A Curious Case of Leiomyosarcoma

SYED BUKHARI, MD; DEBORAH HORNACEK, MD

An 18 year-old-man with a two-year history of right lower extremity recurrent iliofemoral deep venous thrombosis (DVT) presented for evaluation of worsening right lower extremity swelling. He was previously diagnosed with post-thrombotic syndrome, and was advised compression stockings. On examination, he had significant thigh swelling with sparing of the lower leg, a pattern inconsistent with post-thrombotic syndrome. A venous duplex was initially performed, showing proximal chronic iliofemoral DVT, with additional finding of a vascularized lesion abutting the common femoral and femoral veins (**Figure A**). He subsequently underwent magnetic resonance imaging of pelvis and right lower extremity that showed an extensive aggressive-appearing mass involving the pelvis and upper leg, intimately associated with the venous vasculature, extending from the right common iliac vein to the distal thigh, suspicious for sarcoma (**Figure B**).

Subsequently, the patient underwent whole body positron emission tomography that demonstrated a hypermetabolic soft tissue masses infiltrating the right thigh musculature, and tumor thrombosis of the right thigh venous system with extensive collaterals (**Figure C**). In addition, a hypermetabolic left perihilar and left lower lung nodules and right tibial marrow lesion were detected, concerning for metastases. Finally, right thigh mass pathology showed densely packed spindle cells with abundant fibrillar eosinophilic cytoplasm and aligned in a palisade pattern, suggestive of leiomyosarcoma **(Figure D)**. Immunohistochemistry was positive for smooth muscle actin and H-caldesmon, suggestive of smooth-muscle origin. His case was discussed in multidisciplinary sarcoma tumor board and systemic chemotherapy was initiated upfront. At six-month follow-up, his tumor burden was reduced, and he had not experienced any recurrence of DVT.

Leiomyosarcoma is a rare disease that occurs more commonly in the abdomen, retroperitoneum, large blood vessels, and uterus. It is rarely described as originating from vascular tissue in the limbs as in this patient's case.[1] Recurrent iliofemoral DVT due to tumor compression is a rare initial presentation. Histologically, leiomyosarcoma of any origin reveals intersecting, sharply marginated fascicles of spindle cells with elongated, hyperchromatic nuclei and abundant eosinophilic cytoplasm.[1] Immunohistochemical stains are

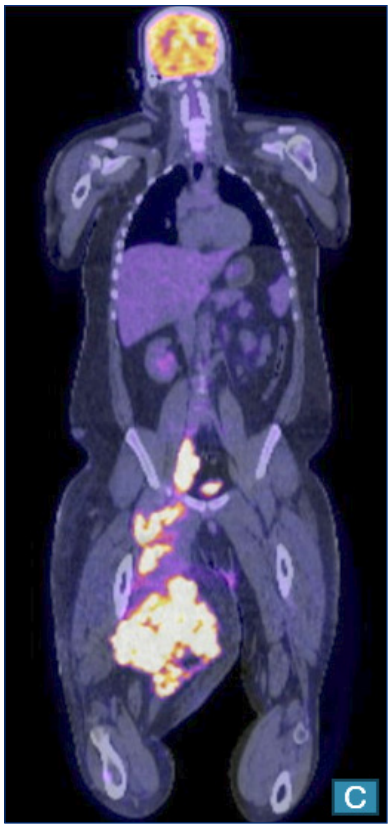


Figure C. Whole body positron emission tomography revealing hypermetabolic soft tissue lesion infiltrating right thigh musculature and associated venous tumor thrombosis.

Figure A. Venous duplex showing vascularized lesion surrounding the common femoral vein.

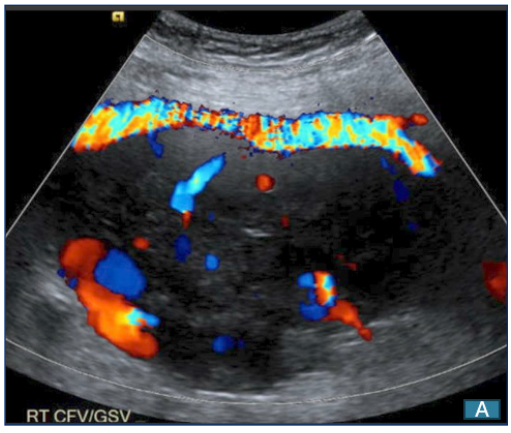


Figure B. Magnetic resonance imaging of pelvis and lower extremities - white arrow indicating mass in the right thigh.

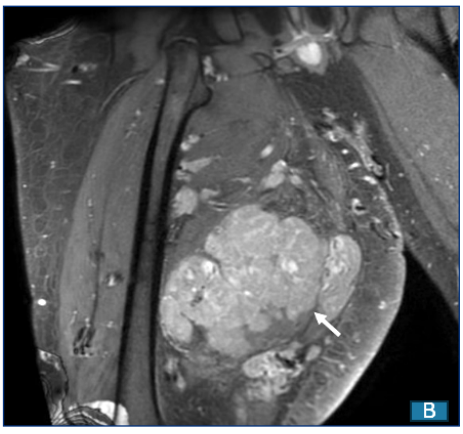
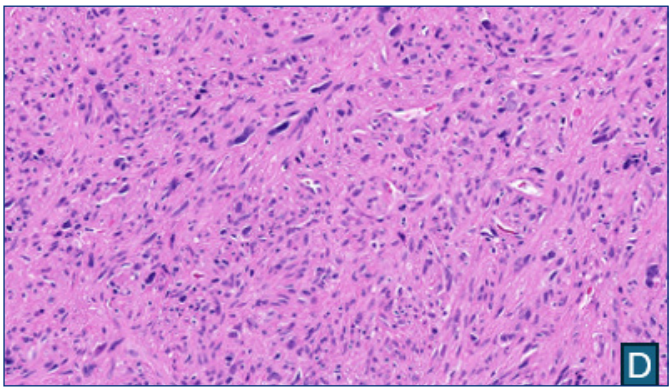


Figure D. Histological evidence of densely packed spindle cells with abundant fibrillar eosinophilic cytoplasm, consistent with leiomyosarcoma.



used for confirmation or in highly undifferentiated tumors. Treatment is individualized, as heterogeneity of its biological origin, clinical behavior, and responsiveness to chemotherapy make the treatment of leiomyosarcoma especially challenging.[2] Standard first-line chemotherapy consists of doxorubicin or gemcitabine based regimens, while later-line therapeutic regimen includes trabectedin and pazopanib. There are ongoing studies that are exploring the role of novel targeted therapies aimed at DNA repair pathways and tyrosine kinase receptors.[3,4] Finally, newer approaches to the treatment of leiomyosarcoma have involved targeting aberrant metabolic processes associated with oncogenesis.[5]

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Authors

Syed Bukhari, MD, Department of Cardiovascular Medicine, Section of Vascular Medicine, Cleveland Clinic, Ohio.

Deborah Hornacek, MD, Department of Cardiovascular Medicine, Section of Vascular Medicine, Cleveland Clinic, Ohio.

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Disclosures

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Correspondence

Syed Bukhari, MD
718-674-5378
mshahbukhari@yahoo.com