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Leiomyosarcoma Arising from the Great Saphenous Vein

AUTHORS:
Kelly Fairbairn, DO; Mrugesh Shah, MD; Javier Romero, MD, FACS; and Shawn Steen, MD, FACS

CORRESPONDENCE AUTHOR:
Kelly Fairbairn, DO
Ventura County Medical Center
300 Hillmont Ave
Building 340, Suite 401
Ventura, CA 93003-3099
Email: KFairbairn@cmhshealth.org
Phone: 805-652-5672

Background	This is a case presentation and recent literature review of a 43-year-old male presenting with a great vessel leiomyosarcoma arising from his great saphenous vein.
Summary	Our patient, a 43-year-old male, presented to the surgical clinic seven months after initial presentation to the emergency department with painless thigh swelling. MRI revealed a soft tissue mass. Surgical en bloc resection containing the great saphenous vein allowed for a tissue diagnosis of well differentiated, Grade 1B leiomyosarcoma. The patient is undergoing adjuvant external beam radiation treatment and chemotherapy with gemcitabine and docetaxel. Great vessel leiomyosarcoma is a rare malignancy most commonly arising from the IVC or lower extremity venous system. Less than 40 cases arising from the great saphenous vein have been reported in the literature. It can present with indolent symptoms. Surgical resection is the mainstay of treatment and prognosis is based on primary site and tumor grade. Distant metastases are possible, and most commonly appear in the lungs. Radiation and chemotherapy guidelines are not well established due to its rarity. Close monitoring after treatment is recommended for at least three years.
Conclusion	Leiomyosarcoma of the great saphenous vein is an extremely rare tumor with often indolent symptoms leading to late diagnosis. We present a case of a 43-year-old male with such malignancy after successful resection, and a brief review of the recent literature.
Keywords	Leiomyosarcoma, saphenous vein

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The authors have no conflicts to disclose.

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Case Description

The patient is a 43-year-old male who presented to the emergency room with reported sudden onset of painless swelling in his left upper thigh after a minor trauma to the area. He denied any associated neurologic symptoms including numbness, tingling, changes in strength, or changes in sensation. An ultrasound demonstrated a large hypoechoic avascular collection around the greater saphenous vein measuring 8.8 x 3.5 x 8.8 cm, with a patent greater saphenous vein, superficial femoral artery, and femoral vein. The mass was thought to be a hematoma, and the patient was sent home with instructions to follow up if the swelling did not improve.

Seven months later, the patient presented to his PCP with an unchanged mass of the left thigh. An MRI was completed which showed a large, slightly lobulated, enhancing mass in the subcutaneous fat of the medial upper thigh that does not appear to invade the adjacent muscle, compatible with a primary soft tissue tumor measuring 6.2 x 9.1 x 10 cm (Figure 1). A staging CT chest/abdomen/pelvis did not demonstrate any metastatic disease or regional lymphadenopathy.

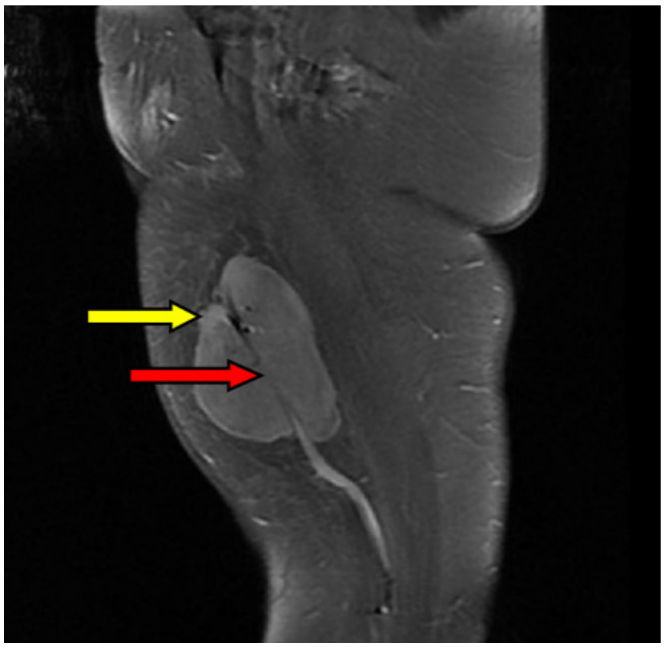

Figure 1 A. Sagittal MRI view of the tumor with the Great Saphenous Vein. Red arrow marks great saphenous vein, yellow arrow marks tumor margin.

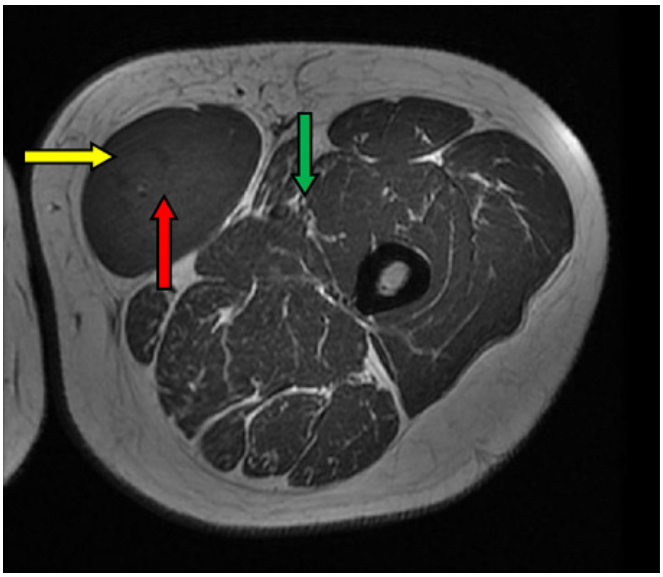

Figure 1 B. Axial MRI view of the tumor. Yellow arrow marks tumor margin, green arrow marks neurovascular bundle approximately 1.5-2 cm from the tumor margin, red arrow marks the great saphenous vein encased in tumor.

Given the relatively stable size of the mass, and without close proximity to major neurovascular structures, it was felt that neoadjuvant treatment would be unlikely to change the surgical approach. As such, a radical excision with appropriate oncologic margins was performed without further biopsy. During excision, the saphenous vein was found to be running through the mass, so it was ligated and resected en bloc. Two to five-centimeter margins, including the muscle fascia were resected with the mass. The neurovascular bundle was noted two centimeters from the mass medially, which restricted our margins in this direction. A closed suction drain was left in the remaining cavity. The patient has had no neurologic compromise since surgery and can ambulate normally.

Pathologic evaluation revealed the mass to be a well differentiated, T2NX, Grade 1, Stage 1B, extremity leiomyosarcoma arising from the saphenous vein, with the closest margin at 1 cm from the neurovascular bundle, and all other margins over 2 cm in distance (Figure 2). It stained strongly positive for desmin and caldesmon, and weakly for PgR. There were 14 mitosis per 10 high power fields (Figure 3). The specimen was sent to an outside laboratory for second opinion which confirmed the diagnosis. Multi-disciplinary discussion recommended external beam radiation and adjuvant gemcitabine and docetaxel. Adjuvant therapies are ongoing at the time of this report.

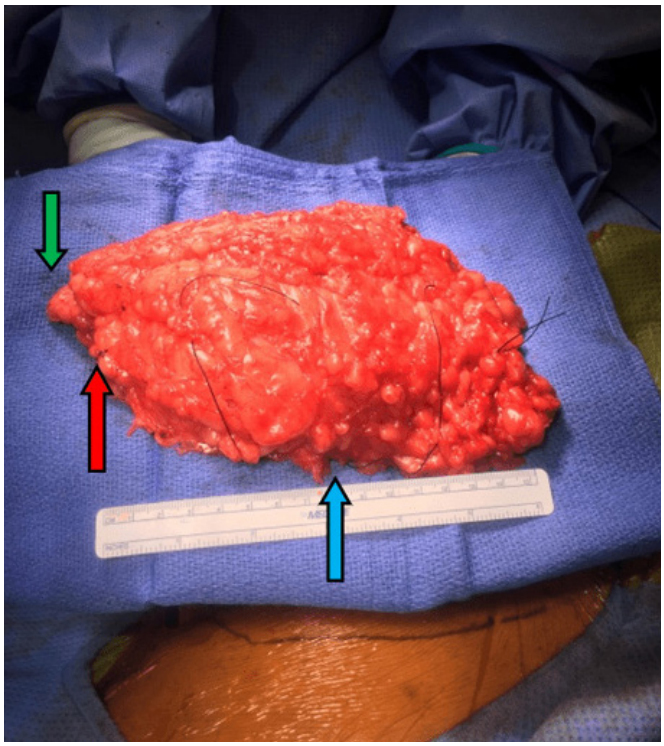

Figure 2 A. Excised tumor specimen en bloc with surrounding fat and muscle fascia. The great saphenous vein is encased in the tumor and the inferior tied edge of the vein is marked by the red arrow. Green arrow marks inferior resection margin, blue arrow marks lateral margin.

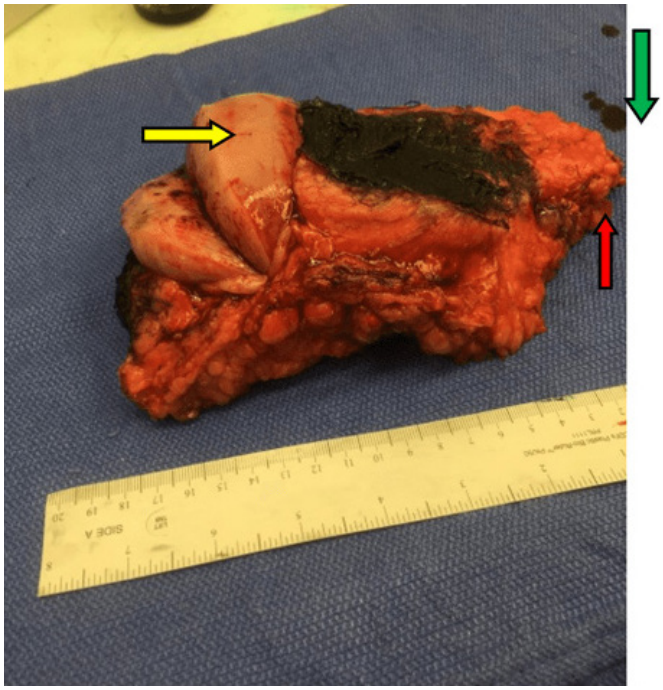

Figure 2 B. Incised tumor with inked, deep margin (muscle fascia). The sarcoma is well encapsulated, smooth, and lighter in color than the surrounding lobulated yellow fat. Yellow arrow marks incised tumor capsule with sarcoma bulging, green arrow marks inferior resection margin, red arrow marks inferior tied edge of the great saphenous vein.

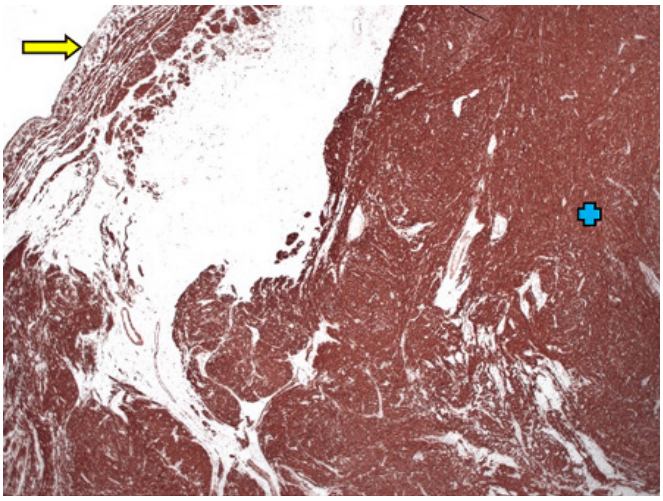

Figure 3 A. Histology of the leiomyosarcoma arising from the great saphenous vein staining positive for desmin (pictured) and caldesmin. Vessel wall marked with yellow arrow with the sarcoma extending radially and marked by blue +.

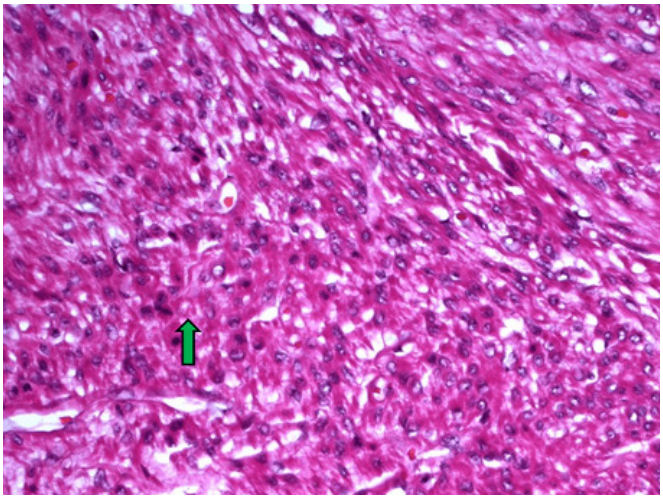

Figure 3 B. High powered field of the leiomyosarcoma displaying mitotic figures. Green arrow marks mitotic figure.

Discussion

Large vessel leiomyosarcoma is the most common sarcoma arising from blood vessels. It predominately stems from the media of veins rather than arteries, and the large vessel subtype accounts for approximately 2% of all leiomyosarcomas. It is most commonly associated with the inferior vena cava, followed by the venous system of the lower extremities.[1] Less than 40 cases of leiomyosarcoma arising from the greater saphenous vein have been reported in the literature. It has an equal female to male predominance and is most commonly diagnosed in the sixth and seventh decades of life, however the age range varies widely (ages 2 to 85 years).[2] The tumors tend to develop in an endovascu-

lar to exovascular direction. Distant metastasis is identified at the time of diagnosis in approximately 10 to 25% of patients.[3]

Large vessel leiomyosarcomas of the lower extremity often present late, as soft tissue swelling or mild pain, and can often be misdiagnosed as deep vein thrombosis.[4] MRI with gadolinium contrast is the imaging modality of choice for diagnosis. Biopsy techniques should be implored with consideration for standard sarcoma guidelines, including excisional biopsies for small tumors and image guided core needle biopsy or incisional biopsy in the axis of the extremity for larger tumors.[2] En bloc surgical resection is the mainstay of treatment and should include margins of at least two to three centimeters.[5]

The optimal chemotherapy and radiation treatment protocol is poorly studied due to the rare nature of these tumors. Many large vessel leiomyosarcomas are treated with adjunctive radiation therapy; however in tumors less than five centimeters in size it may not be necessary due to the relatively low rate of local recurrence.[6] Radiation and its role still largely depends on tumor grade, status of postsurgical margins, and institutional preferences.[2] Chemotherapy has not repeatedly shown a clear survival benefit in case series. However, a recent review of the literature reports several recommended regimens which include a combination of doxorubicin and dacarbazapine, or docetaxel and gemcitabine[7] all of which have been understudied. While large vessel leiomyosarcoma treatment is vastly understudied due to its uncommon nature, soft tissue sarcoma treatment guidelines have been well established and can be translated to this disease. The National Comprehensive Cancer Network publishes standardized guidelines and include consideration for postoperative external beam radiation and chemotherapy for low grade soft tissue sarcomas with negative resection margins. Multiple different chemotherapy regimens are offered including both previously mentioned.[8]

Overall, large vessel leiomyosarcomas have a five-year survival of 25 to 49%, however tumors arising from the IVC have the worst prognosis. Tumors arising from the great saphenous vein have an improved five-year survival of 80 to 90%.2 Recurrence typically occurs within two to three years after completion of treatment and can present as a local recurrence or distant metastasis. Pulmonary metastases are most common.[7] Close surveillance throughout this time is recommended, with clinical exams every three months and site-specific MRI and chest X ray recommended every six months.[2]

Conclusion

Leiomyosarcoma of the great saphenous vein is an extremely rare tumor arising from the media of the vessel with often indolent symptoms leading to late diagnosis. We present a case of a 43-year-old male with such malignancy after successful resection and a brief review of the recent literature.

Lessons Learned

Large vessel leiomyosarcoma is a rare malignancy most commonly arising from the IVC or veins of the lower extremities. Surgical resection with wide margins is the mainstay of treatment and the most appropriate chemotherapy and radiation protocol has not been well established, but standardized soft tissue sarcoma guidelines can be applied. Close surveillance throughout the first three years after completion of treatment is recommended.

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