Surgery for Leiomyosarcoma of the Inferior Vena Cava

Endocrine and Oncologic surgeons at Penn Medicine are performing resection and reconstruction for leiomyosarcoma of vascular origin.

Leiomyosarcomas make up between 5% - 10% of soft-tissue sarcomas, which together comprise fewer than 1% of all cancers. These rare tumors arise from smooth muscle cells of the intima and viscera, and occur most often at the retroperitoneum and uterus. Rarer still are the minority of leiomyosarcomas that originate at a major blood vessel, typically the inferior vena cava, where they present as masses or intraluminal obstructions.

Generally, leiomyosarcomas of vascular origin have a poor prognosis, and carry a significant risk of metastasis and recurrence. Sarcomas are generally aggressive and difficult to treat, and leiomyosarcomas were long considered inoperable. With advances in technique and technology, however, resection has become the mainstay of treatment in recent years for sarcoid tumors and is now considered the only therapy with the potential for cure.

The Division of Endocrine and Oncologic Surgery at Penn Medicine currently provides surgical, radiological and medical options for leiomyosarcoma and the majority of the more than 50 subtypes of soft-tissue sarcoma, with the object of tumor control and optimal recovery.

In patients with leiomyosarcoma of vascular origin, moreover, surgery has the specific goal of avoiding recurrence by achieving negative margins, thereby improving survival. This can be accomplished via en bloc resection of both the tumor and IVC segment followed by graft placement and reconstruction of the vena cava. Surgery for leimyosarcomas at Penn Medicine has the advantage of venovenous extracorporeal bypass (ECMO) through a collaboration with the surgeons of the Divisions of Cardiac and Vascular Surgery.

CASE STUDY

Mr. W, a 70-year-old man, came to Penn Endocrine and Oncologic Surgery for a second opinion after receiving a diagnosis of unresectable leiomyosarcoma and a referral for palliative care at another institution.

Mr. W had a past medical history significant for hypertension, chronic renal insufficiency and worsening lower back pain of six months duration radiating to his lower extremities. An MRI of the spine to identify the source of his back pain found an 8 cm mass involving the IVC adjacent to the right kidney, with severe compression of the middle segment of the vessel (Figure 1, arrow).

On review of his imaging at Penn Medicine, there was concern that extensive involvement of the right renal vein would make reconstruction following a kidney-sparing resection very challenging. A split renal scan was then performed to determine whether the right kidney was functionally compromised, which would influence the decision to perform a nephrectomy.

Figure 1: Abdominal CT scan demonstrating the presence of a large leiomyosarcoma at the right inferior vena cava.

In consultation with Penn Vascular Surgery, the decision was made to proceed with resection, with the goal of preserving the right kidney.

After proximal and distal control of the IVC was obtained, Mr. W was placed on venovenous bypass (ECMO), and the tumor was resected with part of the adjacent right adrenal gland, a portion of the right renal vein, and approximately half the circumference of the IVC for a length of 5 cm. The defect was then reconstructed with a bovine pericardial patch. Postoperative surgical pathology demonstrated a 10 cm low grade leiomyosarcoma with negative margins.

Following his surgery, Mr. W was taken to the intensive care unit for close monitoring, and rapidly transitioned to the floor. He was discharged to home on postoperative day seven in good condition. Twenty months after his surgery, he has no evidence of disease.

References

1. Wachtel H, Jackson BM, Bartlett EK, Karakousis GC, Roses RE, Bavaria JE, Fraker DL. Resection of Primary Leiomyosaroma of the Inferior Vena Cava (IVC) with Reconstruction: A Case Series and Review of the Literature. *J Surg Oncol*. 2015;111:328–333.

FACULTY TEAM

The Endocrine and Oncologic Surgery (EOS) program at Penn Medicine offers surgical expertise by nationally recognized and skilled surgeons for patients with a wide variety of malignant and non-malignant tumors. Comprised of experts from various specialties, our multidisciplinary approach achieves outstanding outcomes earning us the reputation as one of the most respected and talented surgical oncology departments in the country.

Performing Surgery for Vascular Leiomyosarcoma at Penn Medicine

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FY19-21669 03.19