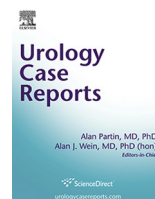




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Oncology

Leiomyosarcoma of the Inferior Vena Cava With Kidney Invasion

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ABSTRACT

Primary leiomyosarcomas of the inferior vena cava (IVC) are rare tumors associated with poor prognosis, and surgical resection with the goal of obtaining negative margins is the gold standard for initial treatment. Tumor characteristics of both extraluminal extension into renal parenchyma and intraluminal extension of the subdiaphragmatic IVC are even less common. The prognosis of vascular leiomyosarcomas is determined by the location and the size of the tumor, as these factors determine the risk of local recurrence and metastasis. We present a case of a 30-year old female incidentally found to have a 14 cm right renal mass and IVC thrombus.

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Introduction

Primary leiomyosarcomas of the inferior vena cava (IVC) are rare tumors associated with a poor prognosis. Surgical resection with the goal of obtaining negative margins is now the gold standard for initial treatment given evidence of improved survival.¹ It is thought that the tumors originate from the endothelial smooth muscle of the intima, and can have intra- and extraluminal extension with invasion into adjacent structures. Published literature shows a wide array of cases, however it is thought that inferior vena cava leiomyosarcomas (IVCL) have a female predominance with a median age of 54 years old.² Patients with IVCL often present with nonspecific symptoms which may include weight loss, nausea, vomiting, fever, malaise, and abdominal pain.² We present a case of a 30-year old female found to have a 14 cm right renal mass and IVC thrombus with leiomyosarcoma tumor revealed on final pathology.

Case presentation

A 30-year old female with a past medical history of obesity, diabetes mellitus type II and ovarian cysts presented with fevers, chills, nausea and vomiting. Workup for her nausea and vomiting, and to initially rule-out appendicitis, a CT abdomen and pelvis with

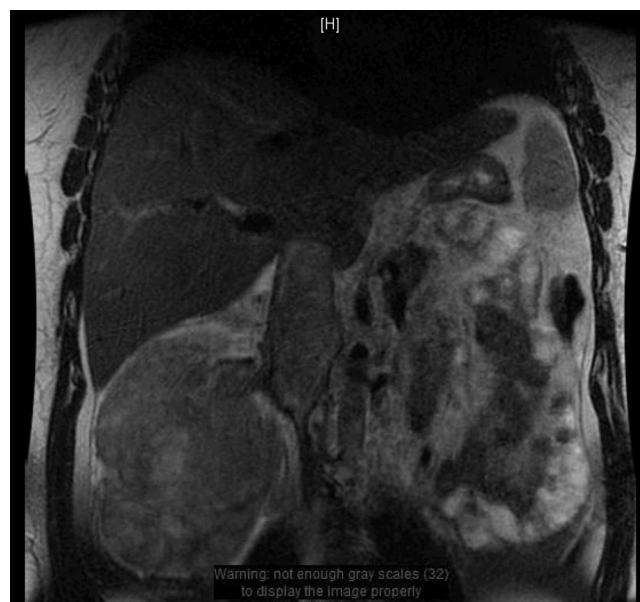


Figure 1. MRI abdomen and pelvis w/without IV contrast (T2 coronal image): 1. A 14 cm solid right renal mass with invasion of the inferior collecting system and extension into Gerota's fascia. Right renal vein and infra-diaphragmatic IVC tumor thrombus. 2. Inflammatory changes in left pelvis with a 7 x 6 cm complex fluid-filled para-ovarian cystic mass. 4.4 x 3.6 x 2.3 cm enhancing collection anterior to right ovary (not shown).

Consent: retained and available by author.

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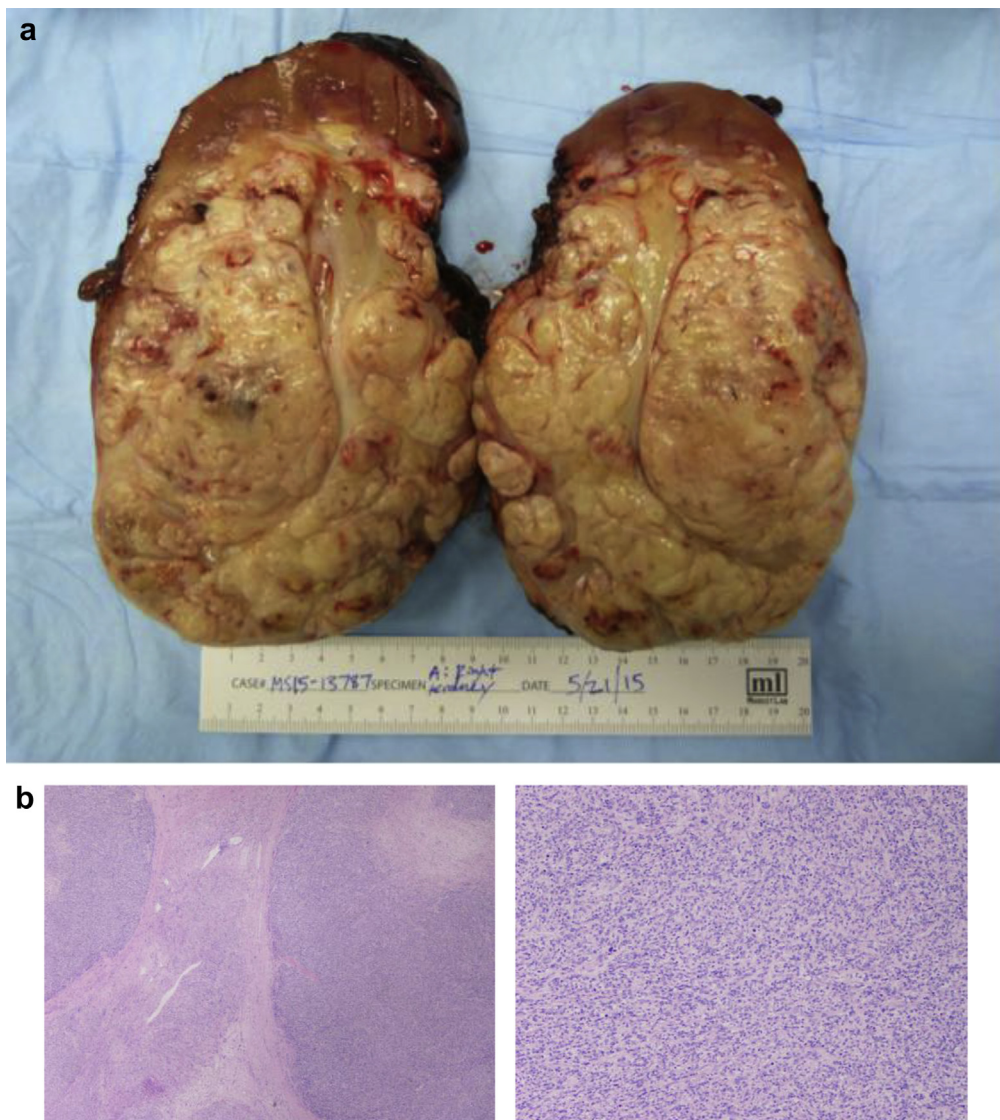


Figure 2. Specimen from right radical nephrectomy, (a) macroscopic and (b) microscopic.

and without IV contrast was performed. Incidentally, CT scan revealed a 14 cm right renal mass and right renal vein thrombus extending into the infrahepatic IVC, with possible bland thrombus. She was also found to have left para-ovarian abscess.

Further workup with an MRI confirmed the above findings and a level II IVC thrombus (Fig. 1). Further workup was negative for clinical metastasis. The patient underwent a right radical nephrectomy, IVC thrombus extraction and reconstruction, and left oophorectomy. A midline laparotomy was performed to obtain access to the IVC and kidney. A 6 cm incision was made to traverse the IVC in a cephalad direction at the right renal vein junction. The cava was cleared of thrombus and flushed with heparinized saline. Venoplasty was performed using 5-0 prolene suture. Immediately postoperatively in recovery room, a bilateral lower extremity Doppler ultrasound was obtained which was negative for deep vein thrombosis.

Final pathologic diagnosis demonstrated a poorly differentiated 15 cm leiomyosarcoma right kidney mass, with extensive venous invasion and tumor necrosis (Fig. 2). The tumor thrombus was also consistent with leiomyosarcoma and measured 7 cm (Fig. 3). The patient has been disease free for 14 months.

Discussion

Most often, as in this case, leiomyosarcomas of the IVC are detected as an incidental finding on an imaging study. Given the slow-growing nature of this malignant tumor, tumor metastasis is uncommon but are seen preferentially in the liver, lungs, lymph nodes, or bones.¹ Occasionally, patients will present with symptomatic flank pain. Various imaging studies, including but not limited to CT scan (+/- angiography), MRI (+/- angiography), ultrasonography, transthoracic echocardiogram or transesophageal echocardiogram may be useful for operative planning. In this case, given the renal involvement of the tumor, primary renal cell carcinoma with IVC thrombus was also in the differential diagnosis prior to surgery. With the use of histopathologic and immunohistochemical methods a definitive diagnosis of leiomyosarcoma can be made. Findings of spindle-shaped tumor cells that stain positive for markers such as smooth muscle cells, vimentin, muscle actin, alpha-smooth muscle actin and desmin are pathognomonic for leiomyosarcoma tumors.³

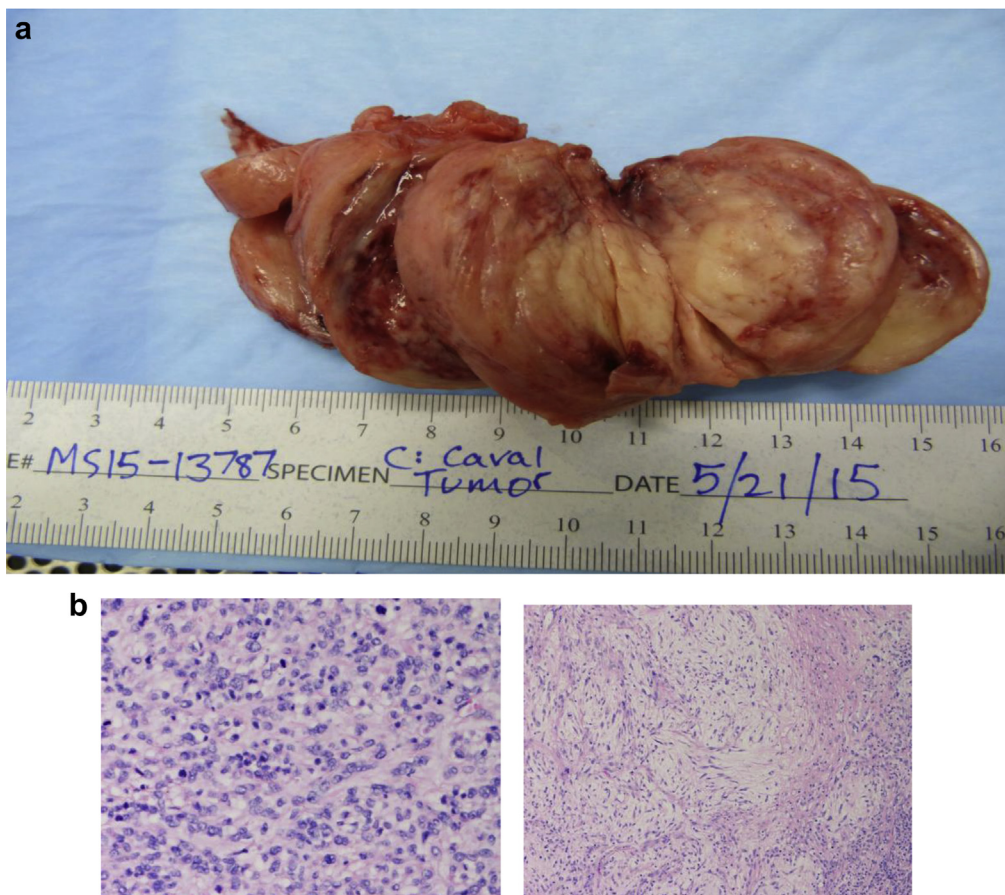


Figure 3. Specimen from IVC thrombus extraction, (a) macroscopic and (b) microscopic.

IVC leiomyosarcomas arise from the tunica media of the caval wall. They demonstrate 3 growth patterns: extraluminal, intraluminal, or both. As previously described in the literature, tumor involvement is classified into 3 groups according to the level of extension in the IVC: segment I: infrarenal; segment II: inter- and supra-renal up to but not including the main suprahepatic veins; and segment III: suprahepatic with possible intracardiac extension. Hines et al⁴ and Hollenbeck¹ each describe predominance of segment II and III. However, these tumors are particularly challenging for surgical resection. The preferred approach is open abdominal by laparotomy or by right subcostal incision and optimizing exposure with a slightly lateral decubitus position. However, if the tumor extends to the retrohepatic or suprahepatic part of the IVC, a thoracoabdominal approach is required. Kieffer et al⁵ suggests combining sternotomy with laparotomy for optimal exposure. This incision provides good conditions for establishing cardiopulmonary bypass if intracardiac extension is present. Furthermore, sternotomy with laparotomy is better tolerated compared to combined thoracophrenotomy with laparotomy from a respiratory standpoint. In the former technique, extensive phrenotomy is not performed and sternotomy is often limited to the caudal end.

Current literature demonstrates that surgical resection is the best initial treatment modality for these tumors. Of note, IVCL may have a 4% perioperative mortality rate and up to 42% recurrence rate.⁵ Evidence also shows that a combination of

surgery with adjuvant chemotherapy and/or radiation may be beneficial.^{1,5} In our case, we were able to resect the tumor in its entirety and the vena cava was primarily repaired. However, if the tumor is not amenable to complete resection, surgery is a good option for initial treatment followed by radiation to the surgical bed.⁴ A pre-operative chemotherapy regimen can be given to reduce the tumor size and increase feasibility for surgical resection.

Interestingly, our patient's tumor likely originated from the IVC with extension both cephalad to the infra-diaphragmatic IVC and caudally to the renal parenchyma. Given the rarity of leiomyosarcoma of venous origin invading into the renal parenchyma, it is possible that the tumor spread extra-luminally. The prognosis of vascular leiomyosarcomas is determined by the location and the size of the tumor (>3 cm); these factors will determine the risk of local recurrence and metastasis. Of the cases reported, mean tumor size was 11.5 cm (range 5–25 cm), which is consistent with our case. The 5-year cumulative survival rate has been reported as 53% for patients with IVC leiomyosarcoma, which suggests that radical surgical excision and adjuvant therapy likely offer the best outcome.^{1,4}

Conclusion

Leiomyosarcoma of the IVC is a rare and aggressive pathologic diagnosis. Tumor characteristics of both extraluminal extension

into renal parenchyma and intraluminal extension of the sub-diaphragmatic IVC are even less common. The gold standard treatment of vascular leiomyosarcomas is surgical resection. Adjuvant chemotherapy or radiation is growing increasingly popular given the poor prognosis of these patients.

Conflict of interest

None.

Acknowledgments

None.

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