

An uncommonly encountered perirenal mass: Robotic resection of renal vein leiomyosarcoma

Amanda F. Saltzman, MD,* Elizabeth T. Brown, MD;† Shams K. Halat, MD;§ Ryan C. Hedgepeth, MD‡

*Ochsner Clinic Foundation, Department of Urology; and Louisiana State University Health Sciences Center, Department of Urology, New Orleans, LA; †Vanderbilt University, Department of Urology, Nashville, TN; §Ochsner Clinic Foundation, Department of Pathology, New Orleans, LA; ‡Ochsner Clinic Foundation, Department of Urology, New Orleans, LA

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Abstract

Primary leiomyosarcoma (LMS) of the renal vein is a rare tumour and poorly described in the literature. Surgical resection, using open and laparoscopic approaches, is the mainstay of treatment. In this report, we describe a patient with left renal vein LMS, report the first robotic laparoscopic resection for this tumor, and review the typical presentation, imaging, pathology and treatment for this rare clinical entity.

Introduction

Primary leiomyosarcoma (LMS) of the renal vein is a rare tumour and poorly described in the literature.¹ The most common site of origin is the inferior vena cava (IVC) (>50% cases), with <35 cases of RVLMS reported.^{1,2} Surgical resection, using open and laparoscopic approaches, is the mainstay of treatment.^{3,4} We describe the first robotic en bloc excision and review the typical presentation, imaging, pathology, and treatment for this rare tumour.

Case report

A 73-year-old female was referred to urology with a perihilar tumour. She reported a 4-month history of worsening left flank pain and denied constitutional symptoms. Her medical history included morbid obesity (body mass index >40), hypertension, hyperlipidemia, diabetes, chronic renal insufficiency, and breast cancer status-post bilateral mastectomy. She denied smoking or occupational carcinogen exposure. Her family history was significant for breast cancer (BRCA negative). Physical exam revealed a morbidly obese woman without palpable abdominal masses or lymphadenopathy.

Lab work revealed an elevated creatinine (1.7 mg/dL, glomerular filtration rate 34 mL/min/1.73m²), but was otherwise unremarkable.

Computed tomography (CT) of the abdomen and pelvis revealed a left 4.8 × 4.3-cm perihilar mass (Fig. 1). Percutaneous biopsy was considered, but the central location and inability to discern the tumour as separate from the renal vein made this undesirable. Renal ultrasound was performed and was also unable to clarify the origin of the lesion (Fig. 2). Moreover, a magnetic resonance imaging (MRI) of the abdomen was performed and it was also unable to determine origin of the lesion (Fig. 2). The metastatic workup was negative.

After medical optimization she was taken to the operating room for resection. A robotic laparoscopic approach with a modified flank position was used. Ports were placed in the standard configuration for a robotic nephrectomy. The mass was anterior to the left renal vein and we could not grossly determine if the mass originated from the left kidney or the left renal vein. The tumour, kidney, and adrenal were removed en bloc and extracted through a Gibson muscle splitting incision. She recovered without incident and was discharged on postoperative day 1.

The pathology exam revealed a 4.3 × 4.5-cm high-grade malignant spindle cell tumour arising from small branches of the left renal vein, with focal renal vein thrombosis (Fig. 3). There was no necrosis and there were 2 mitotic figures/10 high-power fields (HPFs). There was no invasion into the kidney or adrenal (margins negative). Cells stained positive for smooth muscle actin (SMA) and desmin, and were consistent with smooth muscle origin. S100 stain was negative. These histologic features were consistent with a high-grade leiomyosarcoma. One lymph node was negative for metastasis.

The patient was seen during follow-up at 6 months with an MRI evaluation showing no evidence of recurrent or metastatic disease.

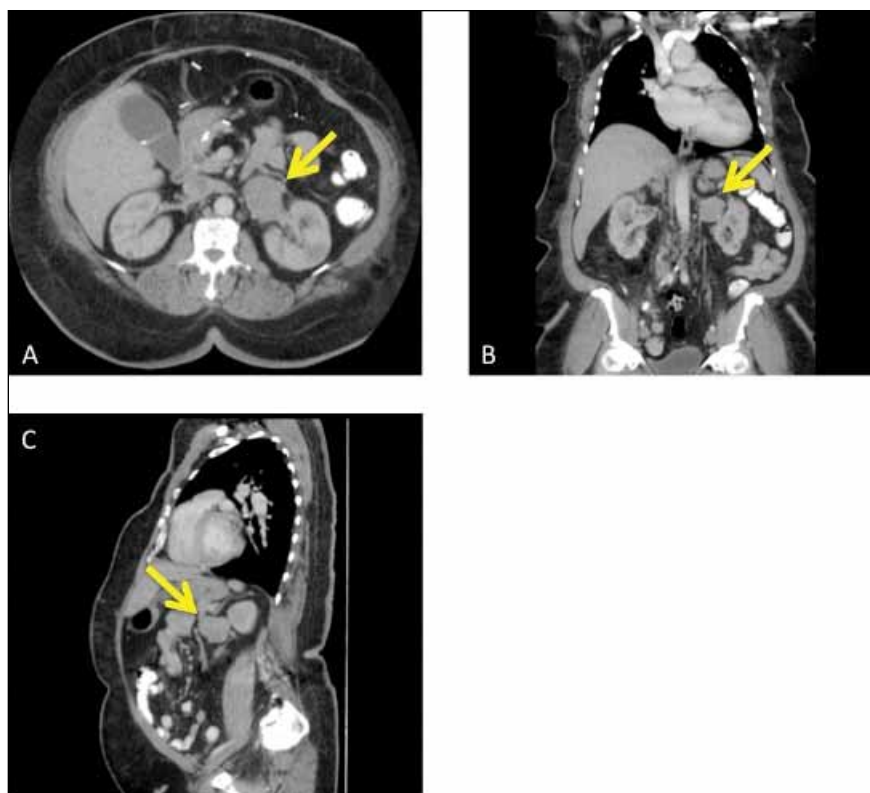


Fig. 1. Enhancing mass at left renal hilum (arrow) in the axial (A), coronal (B), and sagittal (C) planes.

Discussion

Primary LMS of the renal vein is an exceedingly rare tumour, with less than 35 reported cases.⁵ Overall, LMS tumours occur predominately on the left side of the body and in women ages 50 to 69 years.^{1,2} Symptoms, as in our case,

are usually abdominal pain; LMS is also usually incidentally discovered. Gross hematuria and palpable masses are rare, unlike in renal cell carcinoma (RCC) and urothelial cell carcinoma.⁶ Genetic predisposition may play a role in development of primary LMS of the renal vein, with a case reported in a patient with retinoblastoma.⁵ Our patient had



Fig. 2. Further imaging to characterize the mass (arrow), ultrasound with flow into and out of kidney closely associated with mass (A) and magnetic resonance imaging identifying a mass in hilum without further clarification as to whether lesion is intrinsic or extrinsic to the left renal vein (B).

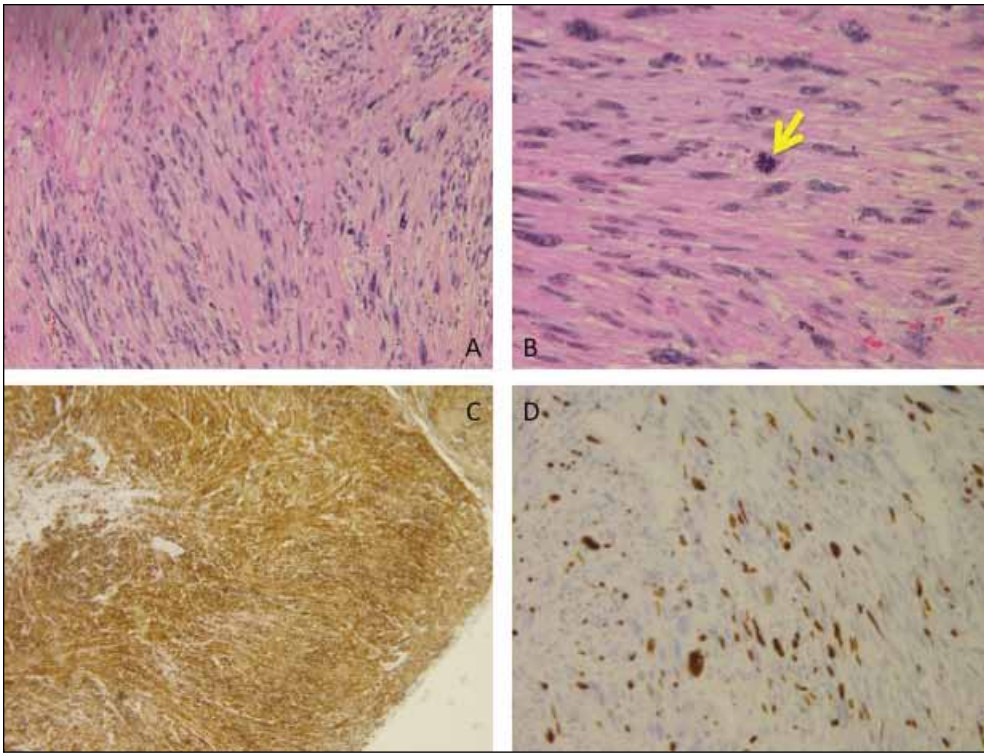


Fig. 3. (A) Hematoxylin & eosin stain section showing fascicles of spindle cells with marked nuclear atypia and pleomorphism, (B) High power view showing atypical nuclei and a mitotic figure (arrow), (C) Immunohistochemical stain for desmin showing strong positivity, (D) Ki67 stain showing high proliferative index (10%).

a very strong personal and family history of breast cancer and, although BRCA negative, it certainly raises the question of genetic susceptibility.

As described in our report, imaging studies are often inconclusive when assessing tumour origin and do not allow preoperative distinction from other tumours that can have renal vein extension.^{6,7} RCC involving the renal vein typically measures >8 cm, with <5% of patients having a primary tumour <4 cm. We have found that 88% of RCC with IVC extension are >7 cm. Primary LMS of the renal vein are typically much smaller, lying mostly or entirely outside the kidney (RCC is typically more intrarenal).⁷ Upper tract urothelial carcinoma (UTUC) very rarely invades the renal vein, with <12 reported cases. When LMS or UTUC are possible diagnoses, percutaneous biopsy is avoided to prevent tract seeding.⁷

CT imaging usually reveals a homogenous, solid, well-circumscribed mass at the renal hilum, with minimal contrast enhancement.¹ MRI reveals a well-defined perihilar mass with intermediate- to low-signal intensity on T1 and intermediate- to high-signal intensity on T2.⁸ As in our case, it is very difficult to determine if the mass abuts is associated with or arises from the renal vein. In addition to LMS, RCC and UTUC, differential diagnosis includes renal vein thrombus, lymphoma, granulomatous disease, and metastatic lesion in a patient with history of malignancy.

Grossly, LMS are well-circumscribed, tan-grey, firm tumours with a whorled cut surface. Necrosis, hemorrhage or cystic change can be present. Microscopically, there are sheets and fascicles of spindle cells with elongated, blunt-ended nuclei. Mitotic figures vary from 0-50/10 HPFs. LMS stains positive for desmin, vimentin, and occasionally actin (smooth muscle origin) and negative for S100.¹

En bloc surgical resection is the best treatment for this rare entity.¹ This surgery has been described using open and laparoscopic approaches.^{3,4} Our case report represents the first resection described using a robotic laparoscopic approach.

Current treatment guidelines recommend adjuvant chemotherapy and/or radiation for patients with large, high-grade or partially resected tumours, because these factors are strong predictors for metastases and recurrence.⁹ There have been isolated case reports of good outcomes without adjuvant therapies, but this is not the current standard of care.^{10,11} In cases of metastatic disease, chemotherapy with doxorubicin and ifosfamide has been reported.¹²

Primary LMS of the renal vein is a dangerous tumour. The 5-year survival rate of the largest case series of non-IVC venous LMS is 25%.² IVC LMS 5-year survival has been reported between 33% and 53%.² There is a trend towards distant metastases rather than lymph node involvement or local recurrence.¹ Spread is primarily hematogenous (lungs, liver, bones).^{1,6} About half of cases are metastatic at diag-

nosis.⁶ The major prognostic factor improving survival is surgical resection with negative margins.^{6,13}

Conclusion

Primary LMS of the renal vein is a rare entity that is difficult to diagnose on preoperative imaging. En bloc resection is the mainstay of treatment. This report describes the feasibility and first report of en bloc resection of a RVLMS using a robotic laparoscopic approach.

Competing interests: The authors declare no competing financial or personal interests.

This paper has been peer-reviewed.

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Correspondence: Dr. Amanda F. Saltzman, Ochsner Clinic Foundation, Department of Urology, New Orleans, LA; afsaltzman@gmail.com