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Journal of Clinical Imaging Science

Genitourinary and Gynecologic Imaging *Case Report*

Smooth Muscle Tumor of Uncertain Malignant Potential Arising from Renal Vein

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Received : 05 February 2021
Accepted : 02 April 2021
Published : 19 April 2021

DOI
10.25259/JCIS_24_2021

Quick Response Code:

ABSTRACT

Smooth muscle tumors (SMTs) typically fall under either benign leiomyoma or malignant leiomyosarcomas based on histologic characteristics. SMTs that cannot be diagnosed as benign or malignant are then labeled as smooth muscle tumors of uncertain malignant potential (STUMP). A 31-year-old male presented with the right abdominal pain for 6 months. Imaging showed an enhancing soft-tissue mass arising from the right renal vein. A pre-operative biopsy confirmed STUMP of renal vein, and the patient underwent surgical resection of the mass with partial nephrectomy. Imaging and pathologic findings play a crucial role in an accurate pre-procedural diagnosis of unusual SMTs.

Keywords: Smooth muscle tumor of uncertain malignant potential, Smooth muscle tumors, Renal tumors, Renal vein

INTRODUCTION

Smooth muscle tumors (SMTs) typically fall under either benign leiomyoma or malignant leiomyosarcomas, and this classification is based on histologic characteristics.[1] These features include a mitotic index, cytological atypia, and presence or absence of tumor cell necrosis.[2] SMTs that cannot be diagnosed as either benign or malignant are then labeled as smooth muscle tumors of uncertain malignant potential (STUMP).[3] Nearly all reported cases of STUMPs are uterine tumors.[4] In this study, we present, to the best of our knowledge, the first case of a STUMP arising from the right renal vein with a brief review of the current literature.

CLINICAL CASE PRESENTATION

A 31-year-old male presented to our institution's surgical outpatient clinic with a 6-month history of the right-side abdominal pain. The pain was intermittent with no relieving or exacerbating factors and was not associated with nausea, vomiting, or bowel habits changes. The patient denied any unexpected weight loss or night sweats. The patient had a history of nephrolithiasis, obesity, and hypertension and a family history of gastric cancer. The physical examination was unremarkable at the initial presentation. The abdominal pain was further assessed with an abdominal ultrasound that showed a 4.3 × 4.5 cm hypoechoic mass located lateral to the pancreatic head and inferior vena cava and medial to the gallbladder right kidney [Figure 1]. The right renal vein appeared partially open. No gallstones or gallbladder thickening was seen; neither was any

hydronephrosis in the right kidney nor any free fluid in the abdomen. Computed tomography (CT) of the abdomen without contrast confirmed a homogenous soft-tissue mass in the right upper quadrant anterior to the right renal vein. The lesion abutted the right kidney's upper pole and was separate from the liver, right adrenal gland, and pancreas [Figure 2]. Magnetic resonance imaging (MRI) of the abdomen with intravascular contrast showed the right upper quadrant mass abutted the right kidney, duodenum, liver, and right renal vein. The right renal vein appeared partially open without any collaterals. The mass was relatively isointense to muscle on T1- and T2-weighted images and demonstrated mild enhancement on postcontrast images [Figure 3]. Chest CT was performed and was negative for lung metastasis. The differential diagnoses based on the imaging findings included renal cell carcinoma (RCC) with renal vein invasion, adrenal carcinoma, renal or perirenal lymphoma, and stromal tumor likely arising from the right renal vein.

Upper gastrointestinal endoscopy was performed, which was normal. The mass was subsequently biopsied under ultrasound guidance from a transhepatic approach. Pathologic examination revealed a cellular spindle cell neoplasm with a fascicular growth pattern. The specimen was positive for smooth muscle antigen and desmin, supporting a diagnosis of SMTs, such as leiomyoma versus leiomyosarcoma. Immunohistochemistry staining showed Ki-67 reactivity in 5–10% of spindle cells, caused by sarcoma.

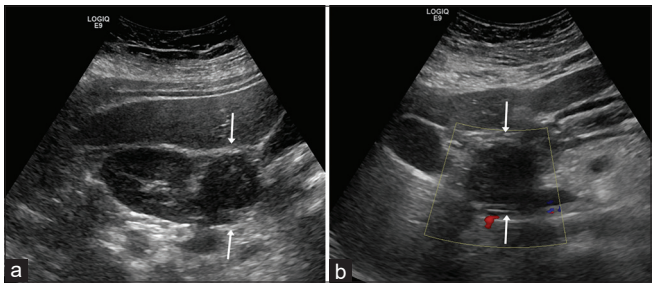

Figure 1: A 31-year-old male presented with the right-sided abdominal pain. (a) The gray-scale ultrasound images of the right upper abdomen demonstrated a 4.3 × 4.5 cm hypoechoic mass (arrows) inferior to the liver and medial to the right kidney. (b) Color Doppler ultrasound image shows no vascularity of the mass lesion.

Since the malignant etiology of the mass could not be ruled out, the patient finally underwent surgical resection, which showed that the mass was arising from the right renal vein. Therefore, a partial right nephrectomy was performed. The surgical pathology evaluation demonstrated no more than four mitoses in 10 high-power fields (HPFs), absence of coagulative necrosis, and lack of destructive growth, which ruled out leiomyosarcoma. The presence of moderate-to-severe nuclear pleomorphism ruled out leiomyoma. Immunohistochemical study was again positive for smooth muscle antigen and desmin. Altogether, these findings supported the diagnosis of STUMP arising from the renal vein [Figure 4].

DISCUSSION

Demographics and presentation

STUMPs are very rare tumors of smooth muscle origin, much rarer than leiomyomas and leiomyosarcomas.[5] They almost always involve women with the mean age of 43 years[5] and present as uterine tumors. Still, there have been cases reported in the retroperitoneum and other parts of the genitourinary tract such as the bladder.[4,6] SMTs may be asymptomatic, and they can be discovered incidentally on imaging. The retroperitoneal SMTs may present with chronic or acute onset abdominal or flank pain, which can be relieved, aggravated, or not impacted by eating.[7] Our patient had chronic pain that was not affected by meals. There have been multiple reported cases of leiomyomas and leiomyosarcomas arising from the renal vein,[8-11] although, to the best of our knowledge, there are no reported cases of STUMP originating from the renal vein.

Imaging findings and differential diagnoses

CT and MRI are the primary imaging modalities for the diagnosis of SMTs. Hypo- to isointense signal relative to

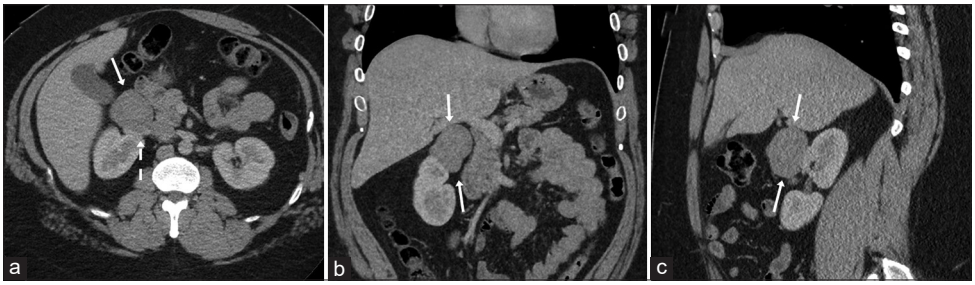

Figure 2: A 31-year-old male presented with the right-sided abdominal pain. (a) Axial, (b) coronal, and (c) sagittal unenhanced CT of the abdomen demonstrate a homogenous soft-tissue mass (solid arrows). The lesion abuts the anterior right renal vein (dashed arrow, a) and upper pole of the right kidney and is separate from the liver and pancreas.

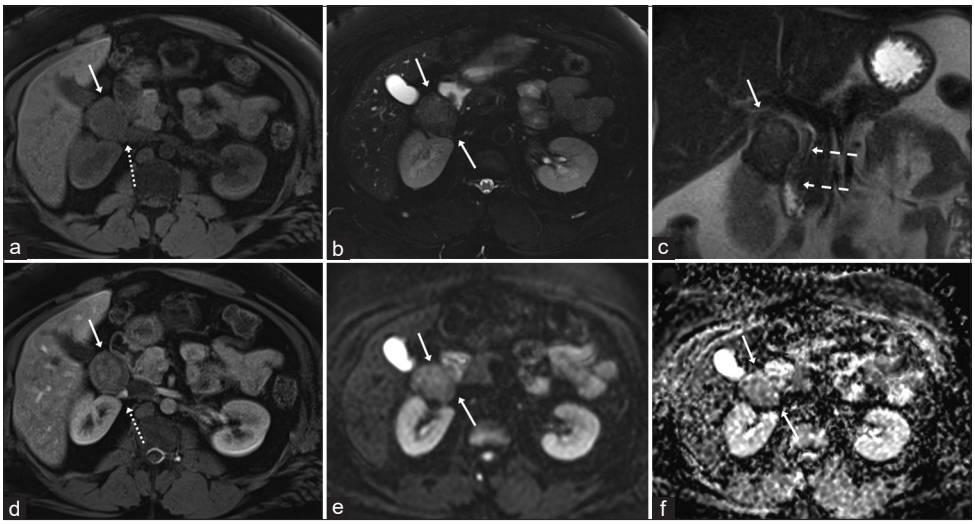

Figure 3: A 31-year-old male presented with the right-sided abdominal pain. (a) Axial T1-weighted, (b) axial and (c) coronal T2-weighted pre-contrast MRI images, and (d) axial T-1 fast-saturated post-contrast MRI images demonstrate a mildly enhancing right upper quadrant mass (solid arrows) abutting the right kidney, liver, duodenum (dashed arrows, c), and right renal vein (dotted arrows, a and d). (e) Diffusion-weighted imaging MRI image demonstrates heterogeneous signal of the mass and (f) apparent diffusion coefficient map shows no definite restricted diffusion.

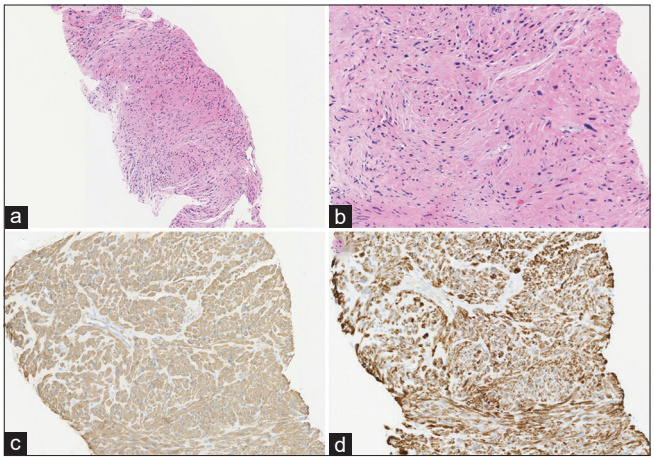

Figure 4: A 31-year-old male presented with the right-sided abdominal pain. (a) H and E ×100 showing spindle-shaped cells with eosinophilic cytoplasm. (b) H and E ×200 showing moderate pleomorphism in the spindle-shaped cells. (c) Immunohistochemical stain ×200 showing the positive smooth muscle actin membranous staining of the neoplasm. (d) Immunohistochemical stain ×200 showing the positive desmin membranous staining of the neoplasm.

normal myometrium on T2-weighted images on MRI is an important clue to suggest SMTs. It may be difficult to differentiate uterine STUMP from leiomyoma and leiomyosarcoma.[12] The STUMP may present as a large mass with heterogeneous signal on T1 and T2-weighted MRI images.[13,14] Uterine leiomyosarcomas commonly manifest as large infiltrating myometrial mass with irregular and ill-defined margins, heterogeneous hypointensity on T1-weighted images, central hyperintensity on T2-weighted image indicative of extensive necrosis, and early heterogeneous enhancement on post-contrast images.[13] They may have hemorrhage and foci of calcifications. Malignant lesions can present as hyperintense areas on diffusion-weighted imaging (DWI), and combined DWI and apparent diffusion coefficient values are useful to differentiate between uterine leiomyosarcoma, STUMP, and benign leiomyoma.[14]

The same rule may apply to SMTs arising in the renal vein, although little is known about renal vein SMTs. MRI of the abdomen in our patient with renal vein STUMP showed a mass of isointense on T1- and T2-weighted images with low-level enhancement and heterogeneous signal on DWI without a definite restricted diffusion. A study analyzing five renal vein leiomyosarcomas cases showed that the tumor size ranged from 4 to 8 cm in the renal vein. Four of the five patients had masses on the left side, unlike our patient, who presented with a right-sided tumor. Contrast enhancement varied across the five subjects, with three subjects having homogenous low-level enhancement, one subject with streaky enhancement throughout the entire mass; and one subject with peripheral enhancement only. Three of the five patients had renal hilum involvement.[10] The renal vein leiomyomas may present as a well-circumscribed non-enhancing to slightly enhancing lesion centered in the renal vein.[11] They may rarely contain central cystic changes which may represent necrosis[9] or degeneration similar to uterine degenerated leiomyomas. There are no clear data that how commonly the renal vein SMTs can be intraluminal and extraluminal. Other differential diagnoses for renal vein/hilar mass include RCC with renal vein invasion, renal lymphoma, adrenal carcinoma, and

bland thrombosis. Table 1 summarizes the imaging findings of the differential diagnoses of lesions involving renal vein in general.[15-19]

Pathologic findings

In the literature, there are no standard pathological characteristics of STUMPs, but they have been described in various ways. STUMPs are tumors that cannot be classified as either leiomyoma or leiomyosarcoma.[5] Studies have shown that molecular and cytological markers have shown that Ki-67 and p53 can be useful, significantly when differentiating STUMPs from leiomyosarcomas.[3,20] The positive p53 and Ki-67 markers tend to be linked to leiomyosarcoma.[20] Uterine SMTs that are positive for estrogen and progesterone receptors may respond to hormone therapy.[21] Our patient's

Table 1: Imaging findings of the differential diagnoses of lesions involving renal vein.[15-19]

Differential diagnoses	Ultrasound findings	CT/MRI findings
RCC with renal vein invasion	• The epicenter is the kidney • May show color flow on Doppler ultrasound	• Usually >8 cm at presentation • Presence of enhancement is the most specific sign • The renal vein may be expanded • Diffusion restriction of the renal vein on DWI[19] • May show FDG avidity on PET-CT
Renal vein leiomyoma	• Homogenous to slightly heterogeneous solid mass • Centered in the renal hilum	• Non-enhancing to slightly heterogeneous enhancing[8,11] • Rarely have central cystic and necrotic changes[9]
Renal vein STUMP	• Homogenous to slightly heterogeneous • Difficult to differentiate from other SMTs	• Well-defined solid soft-tissue mass on CT • Large mass with heterogeneous signal on T1 and T2
Renal vein leiomyosarcoma	• The epicenter is the renal vein • Heterogeneous ill-defined mass	• Usually 3–5 cm at presentation • Large infiltrating myometrial mass with irregular and ill-defined margins • Heterogeneous, hypointense on T1 • Central hyperintensity on T2 indicative of extensive necrosis • Early heterogeneous enhancement on post-contrast images[13] • May have hemorrhage and foci of calcifications • Hyperintense areas on DWI[14]
Transitional cell carcinoma	• Solid, hypoechoic mass (es) located within the renal pelvis or a dilated calyx • Difficult to differentiate from hydronephrosis	• Soft-tissue mass with only mild enhancement • Significantly less enhancing than renal parenchyma or renal cell carcinomas • Maintain a normal renal shape • CT urography is the modality of choice for diagnosis[20]
Adrenocortical carcinoma with venous invasion	• Suprarenal well-defined mass • Larger lesions can be heterogeneous	• large (>6 cm) irregular ill-defined mass • Central necrosis and hemorrhage • Calcification in up to 30% • <40% relative wash-out on delayed contrast-enhanced CT[21] • Hyperintense on T2
Renal lymphoma	Hypoechoic lesion (s) within the kidney	• Usually presents as multiple renal masses • Hypointense on T1 • Iso- to hyperintense on T2 • Poor enhancement compared to renal parenchyma • Restricted diffusion on DWI[22] • Encases rather than invasion of renal vein
Bland thrombus	• Hyperechoic filling defect	• No enhancement • Acute thrombus: High signal intensity on T1 and T2 • Chronic thrombus: Low signal on T2 or appears as a flow voids[23]

CT: Computed tomography, MRI: Magnetic resonance imaging, RCC: Renal cell carcinoma, FDG: F-18-fluorodeoxyglucose, PET: Positron emission tomography, T1: T1-weighted images, T2: T2-weighted images, DWI: Diffusion-weighted imaging

pathology result showed moderate atypia, <4 mitosis/10 HPF, and no coagulative necrosis, most compatible with a diagnosis of STUMP.

Treatment and outcome

Since STUMPs are sporadic tumors, there is not a set protocol on how to manage them.[6] Resection is commonly practiced, especially if they are symptomatic. This is not followed by chemotherapy or radiotherapy. However, the recurrence risk for STUMPs is not low, as it ranges up to 36% and can present as another STUMP or even a leiomyosarcoma.[5,22] It has been shown that STUMPs with a high level of coagulative necrosis are more likely to recur in STUMPs with >10 mitosis/10 HPF or atypia.[23] The patients with STUMPs should be monitored with cross-sectional imaging every 6 months for at least 5 years for leiomyosarcoma risk.[22]

CONCLUSION

An infrequent case of STUMP arising from the right renal vein in a young male is described in this study. STUMPs are a rare subset of SMTs usually arising as uterine tumors in middle-aged women. Imaging and pathologic findings play a key role in an accurate pre-procedural diagnosis.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Kord A, Memon A, Bui JT, Xie KL. Smooth muscle tumor of uncertain malignant potential arising from renal vein. J Clin Imaging Sci 2021;11:23.