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CASE REPORT

Solitary fibrous tumor of the renal pelvis: A case report

Min Liu, Chao Zheng, Jin Wang, Ji-Xue Wang, Liang He

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Abstract

BACKGROUND

Solitary fibrous tumor (SFT) is a rare mesenchymal neoplasm. SFT derived from the renal pelvis is an exceedingly rare entity. In this study, we report a case of renal pelvis SFT and review the relevant literature on this rare tumor.

CASE SUMMARY

A 76-year-old man was hospitalized due to right lumbar and abdominal pain. Abdominal computed tomography showed a hypervascular space-occupying renal lesion, sized 2.3 cm × 1.8 cm. Based on the computed tomography findings, the patient was diagnosed with right renal pelvis tumor and underwent nephrectomy. Postoperative immunohistochemical results confirmed the diagnosis. As of the 3-year follow-up, there were no signs of recurrence, and the patient has recovered well.

CONCLUSION

We report a rare case of SFT derived from the renal pelvis and discuss the imaging and histopathological features that distinguish renal pelvis SFT from other renal pelvis tumors.

Key Words: Renal pelvis; Solitary fibrous tumor; Surgery; Computed tomography; Case report

Core Tip: Extrapleural solitary fibrous tumor is an extremely rare mesenchymal neoplasm, especially in cases of origination from the renal pelvis. We report a rare case of renal pelvis solitary fibrous tumor and discuss the detailed radiologic and pathologic findings. The differential diagnosis of renal pelvis solitary fibrous tumor is discussed based on literature review.

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INTRODUCTION

Solitary fibrous tumors (SFTs) were reported for the first time by Wagner *et al*[1] in 1870. As hemangiopericytomas, SFTs are usually benign and arise from mesenchymal spindle cells, typically of the pleura. Extrapleural SFTs are relatively rare[2]. Although several cases of SFT have been reported, the cause of SFT remains unknown[3]. Most of the SFTs are benign lesions with slow progress, but some have malignant potential[4]. Renal pelvis SFT is an extremely rare entity, with only 11 cases reported in the published literature[5]. Renal pelvis SFT is liable to be misdiagnosed as renal cell carcinoma (RCC), upper-tract urothelial cancers (UTUCs) or renal angiomyolipoma (RAML). The diagnosis and treatment of renal pelvis SFT is challenging. In this work, we report a rare case of renal pelvis SFT. In addition, we review the relevant literature to facilitate accurate diagnosis and treatment of renal pelvis SFT.

CASE PRESENTATION

Chief complaints

A 76-year-old man (height: 167 cm; weight: 70 kg) presented in our hospital for persistent right lumbar and abdominal pain.

History of present illness

The right lumbar and abdominal pain had persisted for more than 3 years and gradually aggravated in the last 3 mo. There was no associated nausea or vomiting.

History of past illness

The illness history of the patient was unremarkable.

Personal and family history

The patient had no specific personal or family history of illnesses.

Physical examination

The vital parameters of the patient on the day of admission were as follows: body temperature, 36.3 °C; heart rate, 80 beats per min; respiratory rate, 18 breaths per min; blood pressure, 125/80 mmHg; and oxygen saturation in room air, 98%. There was mild tenderness in the left lumbar and abdominal area.

Laboratory examinations

Urinalysis, routine blood tests, coagulation indices, blood urea nitrogen, and liver function were normal. Urinary cytology revealed no heterocytes.

Imaging examinations

Abdominal contrast-enhanced computed tomography (CT) showed a right renal intrapelvic hypervascular space-occupying lesion sized 2.3 cm × 1.8 cm (Figure 1).

Histopathological findings identified the tumor as renal pelvis SFT (Figure 2). The size of the tumor was 2.7 cm × 2.5 cm × 1.6 cm. The mitotic image of the tumor was higher than 4/10 high power field. There were no signs of neurovascular invasion by tumor cells.

Immunohistochemical results were as follows: CD34 (+), desmin (-), H-caldesmon (-), SMA (+), STAT6 (+), vimentin (+), Bcl-2 (+), CD117 (-), Dog-1 (-), HMB45 (-), S-100 (-), Ki-67 (+10%), CD99 (+) and EMA (-). The immunohistochemical results confirmed the diagnosis of renal pelvis SFT (Figure 3).

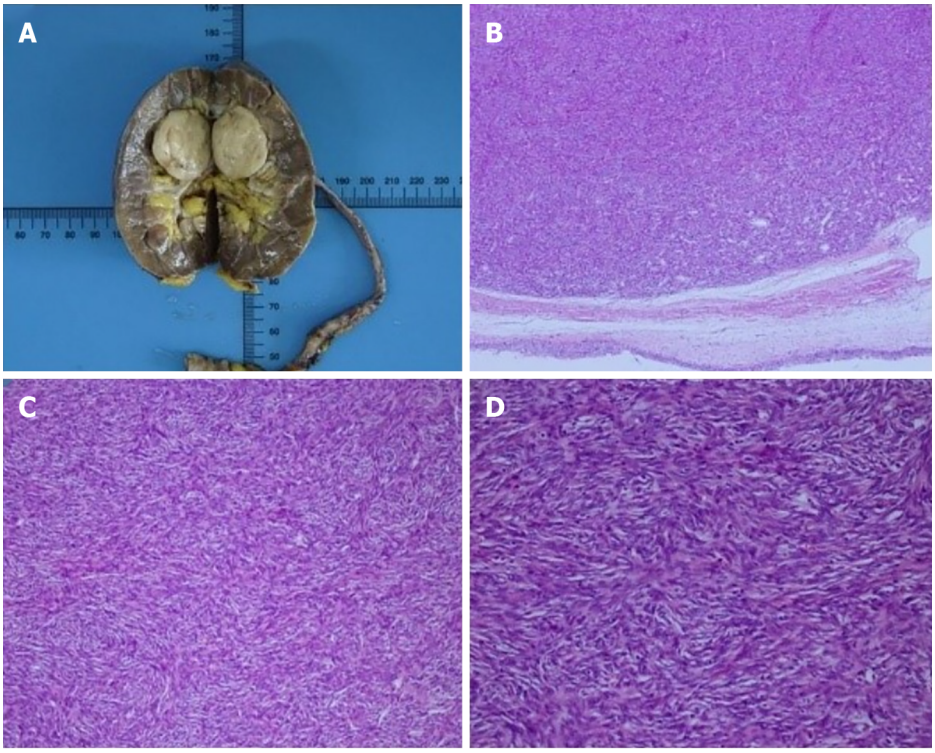

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Figure 1 Computed tomography images showed a mass in the right renal pelvis (diameter: 2.3 cm × 1.8 cm). Contrast-enhanced image showed a well delineated outline of the mass. A: Arterial phase computed tomography (CT); B: Cross-section CT (venous phase); C: Coronal contrastenhanced CT image; D: Sagittal surface contrastenhanced CT image. Red arrows indicate right renal pelvis solitary fibrous tumor lesion.

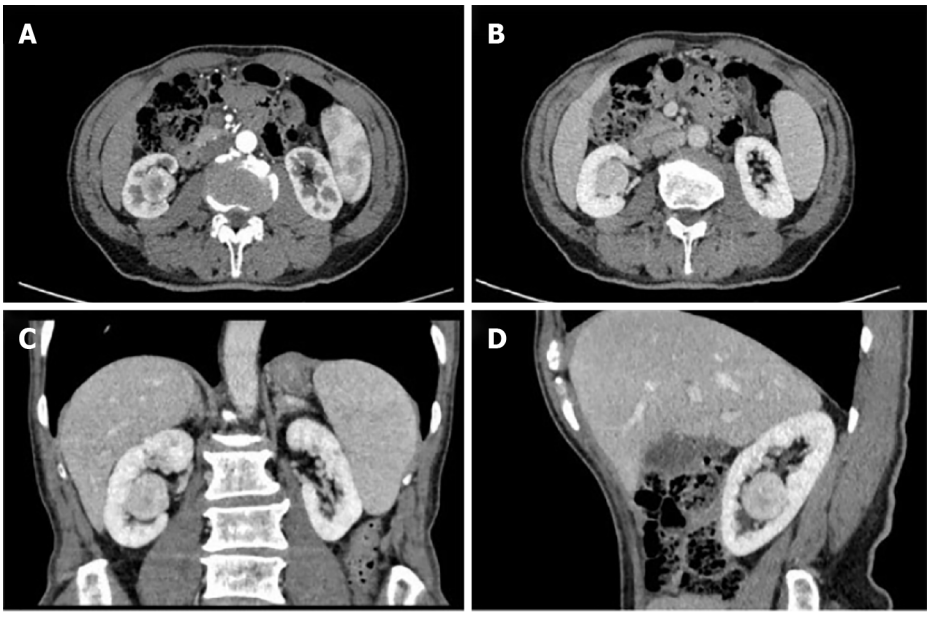

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Figure 2 Pathological specimen of the kidney and ureter. A: Photograph of the tumor; B: Hematoxylin and eosin (H&E)-stained section (magnification × 5); C: H&E-stained section (magnification × 40); D: H&E-stained section (magnification × 200).

FINAL DIAGNOSIS

The patient was diagnosed with left renal pelvis tumor.

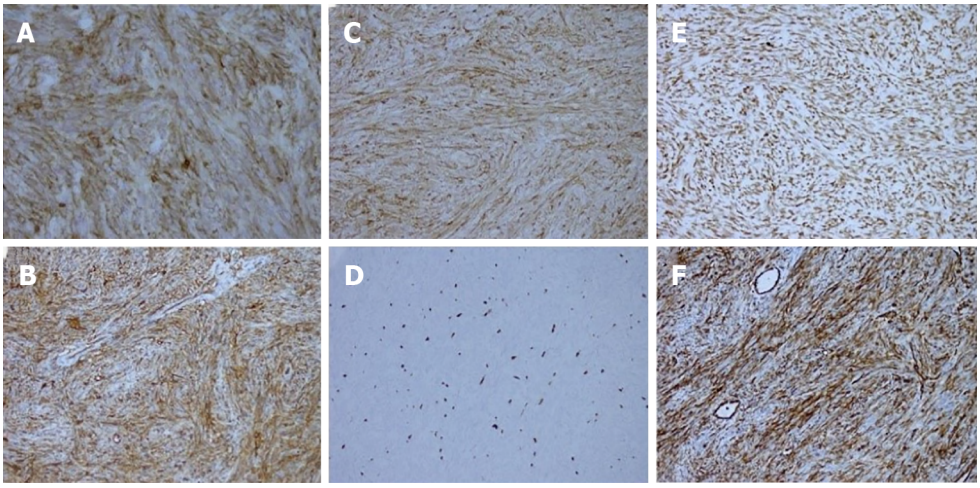

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Figure 3 Immunohistochemistry results (× 20). A: Bcl-2; B: CD34; C: CD99; D: Ki67; E: STAT6; F: Vimentin.

TREATMENT

After weighing different treatment options, the patient underwent laparoscopic right nephroureterectomy.

OUTCOME AND FOLLOW-UP

The patient recovered well after the operation and was discharged after 5 d. The outcome was satisfactory, and there were no signs of recurrence during the 3-year follow-up.

DISCUSSION

SFTs are rare mesenchymal tumors accounting for < 2% of all soft tissue tumors with an estimated annual incidence of 2 cases per million population[6]. The age of reported cases of SFT patients ranges between 4 years and 85 years, and there is no clear predilection for any particular sex[3]. SFTs mainly occur in the pleura. However, SFTs have also been reported at extrapulmonary sites, such as the liver, mediastinum, breast, lung, meninges, and urogenital organs[7]. According to previous reports, 15% of SFTs originating in the kidneys are located in the renal capsule, 6% are located around the pelvis, 3% are located in the renal pelvis, and 76% do not have a clear site of origin[8]. The first case of renal pelvis SFT was reported by Yazaki *et al*[9] in 2001. Until now, only 15 cases of renal pelvis SFTs have been reported. Renal pelvis SFT cases are summarized in Table 1.

Renal pelvis SFTs need to be differentiated from the more common renal pelvis tumors such as RCC, RAMLs, and UTUCs. Contrast-enhanced CT is the main method for the diagnosis of RCCs[10]. RCC is characterized by abundant blood supply, and tumor blood vessels and tumor staining can be observed on renal angiography. Contrast enhancement of renal pelvis SFTs is much lower than that of RCCs. UTUCs are another kind of common renal pelvis malignant tumor. Patients with UTUCs typically have a history of hematuria[11]. In contrast, symptoms of renal pelvis SFTs are mostly due to pressure effect of the lesion, and these patients rarely develop urinary symptoms. The UTUCs show signs of infiltrative growth in CT images. Compared with UTUCs, renal pelvis SFT appears as a well-defined, heterogeneous or homogeneous mass showing moderate to marked contrast-enhancement. RAML is the most common renal benign tumor. Most RAMLs exhibit mixed density on CT imaging, due to the complex fatty vascular components.

In addition to RCCs, UTUCs, and RAMLs, there are also some rare tumor types including hemangiopericytomas, renal pelvis fibroepithelial polyps, fibromas, renal leiomyoma and inflammatory myofibroblastic tumors[12]. The imaging characteristics of renal pelvis SFTs are usually indistinguishable from these rare tumors, and the differential diagnosis is based on immunohistochemistry[13].

According to the literature, some renal pelvis SFTs show areas of calcification, cystic change or necrosis[14]. These changes indicate the aggressive nature of the tumor and poor prognosis.

In this case, the surgical method was laparoscopic right kidney and ureterectomy. There are currently no treatment guidelines for renal pelvis SFTs, but radical resection is generally chosen. Whether preoperative biopsy or nephron preservation surgery can improve the treatment efficacy and prognosis

Table 1 Previous case reports of renal pelvis solitary fibrous tumor

No	Ref.	Age/Sex/Side	Size	Treatment
1	Yazaki *et al*[9]	70/M/R	6 cm × 4.5 cm × 4 cm	Radical nephrectomy
2	Margo *et al*[20]	31/F/R	Approximately 8.6 cm	Radical nephrectomy
3	Marzi *et al*[21]	72/F/L	Approximately 19 cm	Radical nephrectomy
4	Sasaki *et al*[22]	48/M/R	28 cm × 18 cm × 10 cm	Radical nephrectomy
5	Usuba *et al*[16]	50/M/L	17 cm × 11 cm × 8 cm	Radical nephrectomy
6	Zhang *et al*[5]	45/F/L	4 cm × 2.5 cm × 2 cm	Radical nephrectomy
7	Hirano *et al*[23]	75/F/L	4.5 cm × 3.5 cm	Radical nephrectomy
8	Naveen *et al*[24]	52/F/R	17 cm × 10 cm × 10 cm	Radical nephrectomy
9	Dong *et al*[25]	71/F/L	4 cm × 3.5 cm × 4 cm	Radical nephrectomy
10	Mearini *et al*[26]	17/F/L	17 cm × 9.8 cm × 12 cm	Radical nephrectomy
11	Wang *et al*[27]	66/F/R	23 cm × 18 cm × 12 cm	Radical nephrectomy
12	Cheung *et al*[2]	49/F/L	19 cm × 12 cm × 10 cm	Radical nephrectomy
13	Fursevich *et al*[14]	66/F/L	9.3 cm × 7.9 cm × 9.4 cm	Radical nephrectomy
14	Bacalbasa *et al*[1]	49/M/R	15 cm × 15 cm × 10 cm	Radical nephrectomy
15	Luca *et al*[3]	52/F/R	7.4 cm × 6.3 cm × 5.8 cm	Radical nephrectomy

M: Male; F: Female; L: Left; R: Right.

is not clear. For most cases of SFT, due to the malignant potential of SFTs and lack of recurrence after radical nephrectomy, a complete removal is recommended[15].

Immunohistochemistry plays a key role in arriving at a definitive diagnosis. SFTs stain positive for CD34, CD99 and Bcl-2. STAT6 is also an antibody with high sensitivity for SFT diagnosis. These surface antigens are useful diagnostic markers of SFT[3]. It has been reported that CD34 and Bcl-2 negativity may indicate increased malignant potential[16]. Although STAT6 is expressed in most SFTs, SFTs located in the renal pelvis have not been evaluated. Whether STAT6-negative status increases the malignant potential of renal pelvis SFTs is unknown. In this case, tumor tissue stained positive for CD34, vimentin, Bcl-2, STAT6 and CD99. It was considered as a benign renal SFT, and there was no local or distant metastasis after 3 years of follow-up. However, benign renal pelvis SFT may also have the ability for distant metastasis. Therefore, renal pelvis SFT is considered to be a "moderately malignant tumor that rarely metastasizes." Metastasis may occur in the lungs, liver and bones[17]. There are also reports of retroperitoneal recurrence[18]. Rarely, the SFTs can also cause paraneoplastic syndromes such as Doege-Potter syndrome[19]. Hence, all renal pelvis SFT patients need long-term follow-up and regular review, such as abdominal and lung CT.

CONCLUSION

We reported a rare case of renal pelvis SFT. Compared to the previously reported renal pelvis SFT tumors, the tumor in our patient was small in size and localized in the renal pelvis. Clinicians should pay attention to clarifying the source of kidney SFT and differentiate it from other renal pelvis cancers, so as to reduce the occurrence of misdiagnosis.

FOOTNOTES

Author contributions: Liu M and Zheng C collected the information; Wang J conducted the follow-up; Wang JX validated the figures; Zheng C examined and photographed the pathological findings; He L conceptualized and organized the study, reviewed the literature and drafted the manuscript.

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