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Case Report
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RECURRENCE OF A HUGE RENAL SOLITARY FIBROUS TUMOR: CASE REPORT AND REVIEW OF LITERATUR.

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Abstract

Solitary fibrous tumor (SFT) is a spindle cell neoplasm of mesenchymal origin. First reported in 1931, the pleura is the most common localization of SFT, and it's exceptionally rare in the kidney. Overall, it represents less than 2% of all soft tissue tumors. In most cases, renal SFT (rSFT) presents with hematuria, flank pain, and a palpable mass. To our knowledge, less than 112 cases of rSFT have been reported. We report a case of rSFT of a 30-year-old male thought to be a renal cell carcinoma (RCC). Radical nephrectomy (RN) was done to remove a large right-sided mass invading the inferior vena cava. Immunohistochemistry confirmed the diagnosis of SFT showing positivity for CD34, CD99, and Bcl-2 protein, with no staining for cytokeratin. A post-operative CT (15 months) showed tumor recurrence in the renal compartment with huge inferior vena cava thrombus extending to the external iliac veins. With this case, we illustrate and highlight the importance of this diagnosis because of the uncertain biological behavior and prognosis of these tumors.

Key words
CD34, Immunohistochemistry, Kidney cancer, Solitary fibrous tumor, Spindle cells,

Introduction

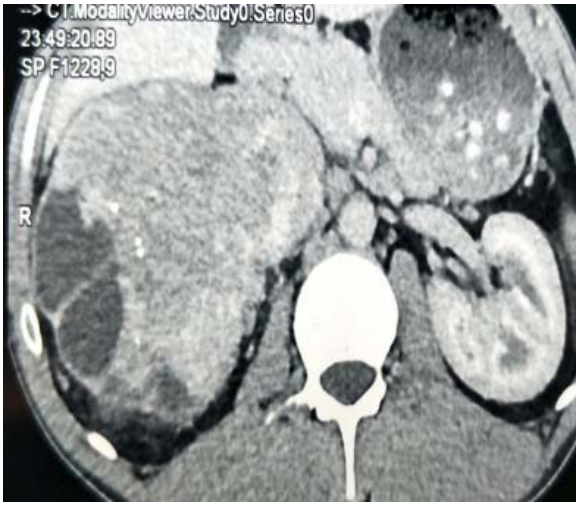
Solitary fibrous tumor (SFT) is a rare spindle cell neoplasm of mesenchymal origin representing < 2% of all soft tissue tumors[1]. .First reported in 1931[2-3], the pleura is the most common localization of SFT. Extra-pleural solitary fibrous tumor can be found anywhere in the human body, especially in the deep soft tissues of the extremities, head and neck region (principally the orbit), chest wall, mediastinum, pericardium, retroperitoneum, abdominal cavity, meninges, spinal cord, and periosteum. It is very rare in the urogenital tract[2] . To our knowledge, the first case of SFT in the kidney was reported in 1996 by Gelb et al.[4] . Only 111 cases of renal SFT (rSFT) have been published to date[5].

We report a large rSFT with an 15-months follow-up, a rare site of recurrence, and discuss the importance of the diagnosis because of the uncertain biological behavior and prognosis.

Case Report

A 30-year-old man presented to the primary care physician because of painless hematuria. Ultrasonography (US) showed a 17cm solid heterogeneous mass in the right kidney. CT scan confirmed a large right-sided renal mass of soft tissue density, discreetly enhanced, heterodense by the presence of irregular calcifications, and with invariably appearing hypodense areas after bolus injection of contrast agent. The tumor measured

17x12x13cm. It compressed the right renal vein and inferior cavernous vein, but without a vascular thrombus. The tumor was in intimate contact with the liver and the psoas muscle. The contralateral kidney appeared normal. Note was made of two hepatic angiomyolipomas (Fig. 1 A&B).



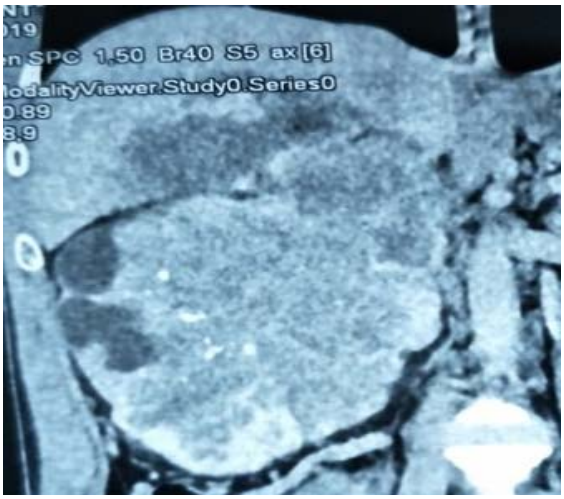


Fig. 1 A&B: Preoperative CT images showing a large 17,6 ×12,3×13,5 cm heterogeneous solid mass in the right kidneywith intimate contact to the cavernous vein.

The patient underwent open radical nephrectomy (RN). To secure negative surgical margins, a patch of vena cava was resected together with the tumor (Fig. 2).

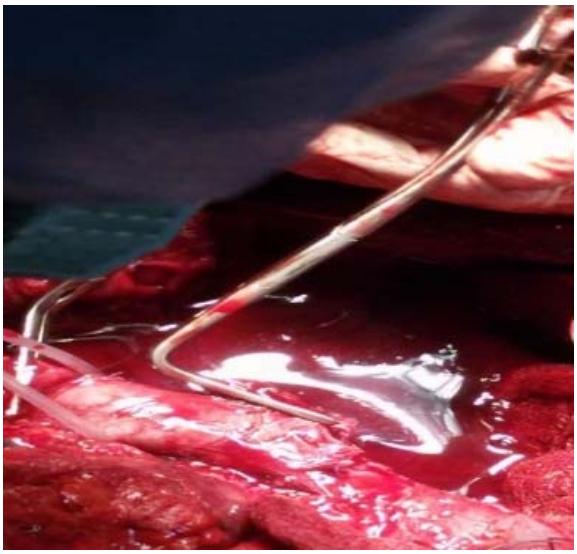


Fig.2: opened cavernous vein

The patient tolerated the procedure well. Blood loss was 500 ml. He was discharged on the 7th post-operative day without any complications.

Macroscopically, the mass was encapsulated with irregular surface and firm consistency. It weighed 1575 grams and measures 19.5 × 11 × 11 cm (Fig. 3).

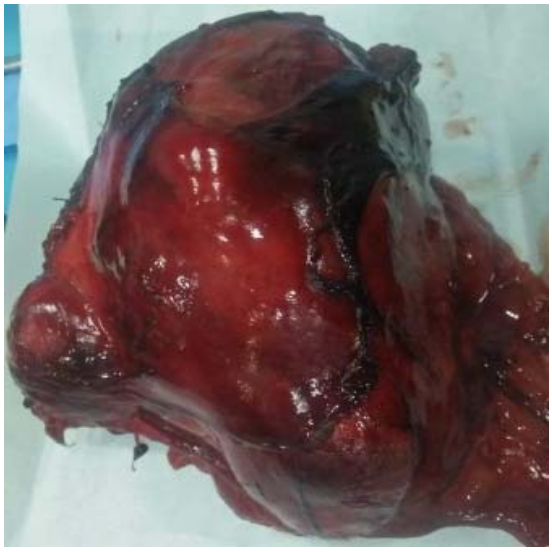


Fig.3: large irregular pseudo-encapsulated mass

On cut section, the tumor had a whitish appearance with areas of cystic degeneration. The cystic component had a citrine yellow liquid content.
Histological examination of the specimen showed renal parenchyma with a fuso-cellular tumor of variable density infiltrating adjacent fat (Fig. 4A). Tumor cells had indistinct cytoplasmic borders with a pale eosinophilic cytoplasm. The nucleus is vesicular with fine nucleoli. Mitosis were rare (< 3/ 10 HPF). These cells formed short tangled bundles surrounded by reticular fibers at reticular stain (Fig. 4B).

The diagnosis was confirmed by immunohistochemical staining. The tumor cells are diffusely positive for CD34 (Fig. 4C), CD99 (Fig. 4D), and Bcl-2 (Fig. 4E). However, they were negative for cytokeratin (AE1-AE3) (Fig. 4F), epithelial membrane Antigen (EMA), PAX2, PAX8, CD117, HMB45, Melan A and desmine.

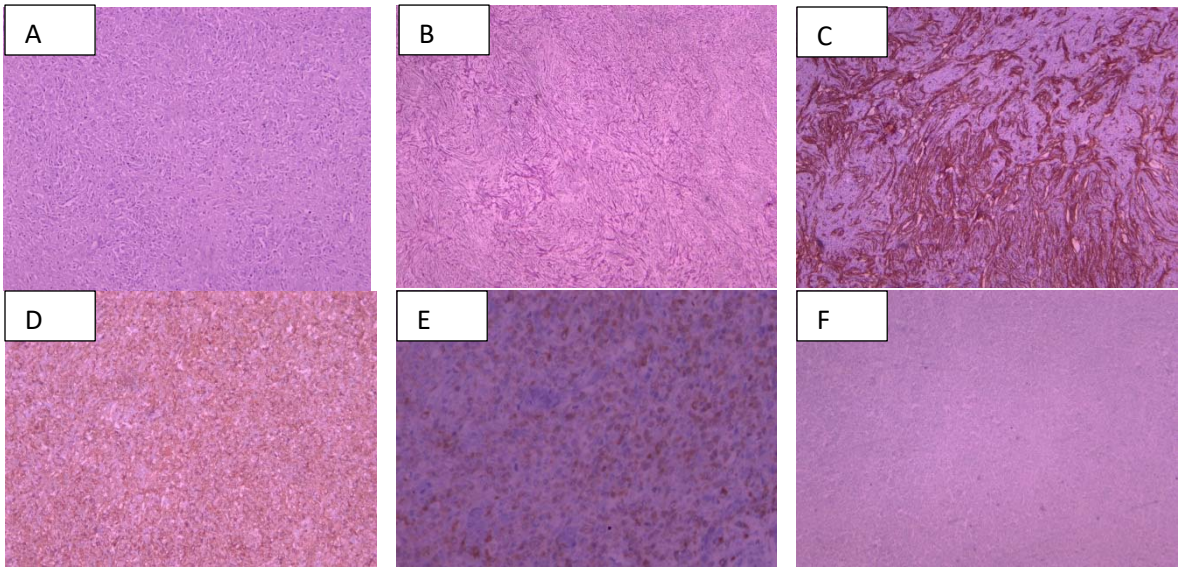


Fig. 4: **(A)** Fusocellular tumor with high cell density (HEX 250), **(B)** the cells are surrounded by a reticular fiber (X 250). Immunostaining shows a positivity for **(C)** CD34 (X 250), **(D)** CD99 (x250), and **(E)** bcl2 (X 400) . **(F)** The cells are negative for CK (AE1-AE3) (X100)

There were hyperplastic hilar lymph nodes without infiltration and a normal adrenal gland.
At 6-months follow-up, there was no evidence of disease recurrence, nevertheless at 15-months, the patient came to a urology consultation for bilateral lower extremity edema.

Imaging CT showed tumor recurrence in the renal compartment with huge inferior vena cava thrombus extending to the external iliac veins (Fig.5A,B,C,D). The patient have not been seen since this time. (No answer of our calls)

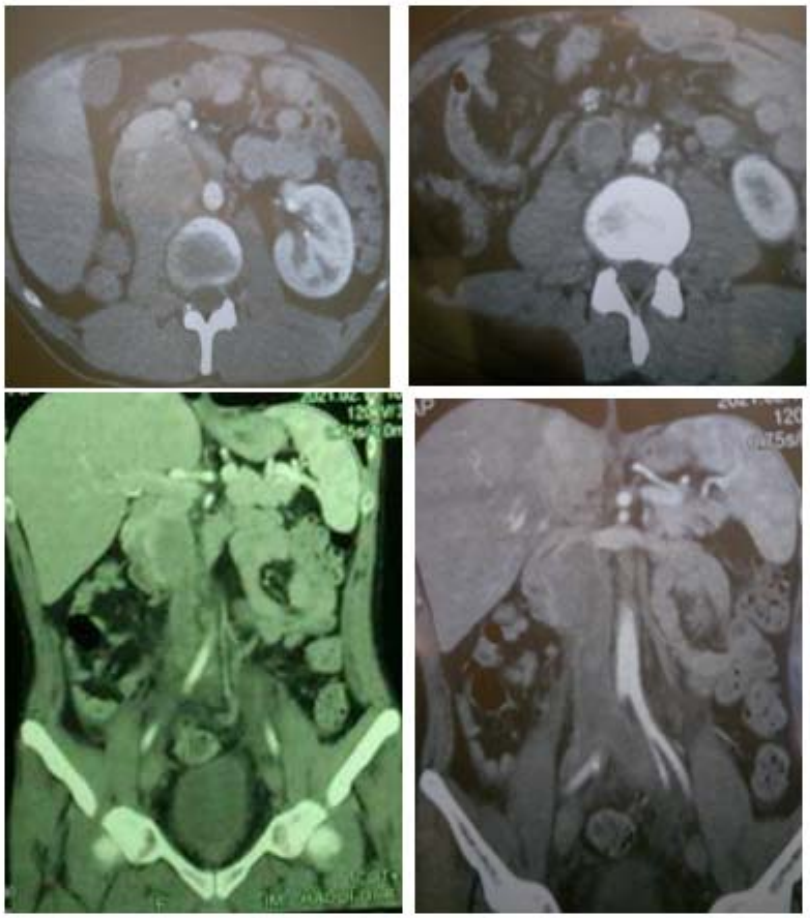


Fig. 5 A&B&C&D: Postoperative CT images showing Tumor recurrence in the renal compartment with huge inferior vena cava thrombus extending to the external iliac veins

Discussion

SFT is an uncommon, soft tissue mesenchymal neoplasm that has been first reported in the pleura. This entity was initially considered as a variant of hemangiopericytomas (HPC)[6]. Nowadays, the term SFT expanded to include both, pleural SFT and most tumors previously branded as HPC[7]. About 10%–15% of SFT show malignant behavior [3]. The urogenital localization of SFT is rare[2&5].

Fu et al.[8] reported in their single institution study a total of 10 cases (1,42 %) of rSFT out of 702 patients with renal tumors treated over a span of 16 years. To our knowledge, to date 111 cases of rSFT have been reported in the English literature[5]. The male to female ratio is 1:1.5 [2].

Because imaging features are not specific for rSFT, they are usually clinically diagnosed and managed as renal cell carcinoma (RCC), most often by RN[2,7&8] They should be considered in the differential diagnosis of renal tumors, along with angiomyolipoma, fibroma, and fibrosarcoma.

It has been suggested that the origin of most rSFT is the renal capsule [2&4], and indeed many of these tumors exhibit an external growth. Cases invading the renal pelvis have also been reported[9]. Bilateral occurrence of primary rSFT is rare[10]. rSFT can grow quite large, and the rSFT reported in the literature range from 2 to up to 25 centimeters in diameter [2&11].

Based on histopathological features, rSFT can be classified as cellular or fibrous [6].

Macroscopically, rSFT are well-circumscribed, pseudo encapsulated, lobulated, grayish to tan-white, firm and heterogenous, sometimes with cystic and necrotic changes[2,7,8&11]Calcifications are noted in some cases, too.

The key to diagnosis is immunohistochemistry. SFT typically show strong and diffuse positivity for CD34, which is characteristic and pathognomonic for the diagnosis. CD34 positivity has been reported in 95 to 100% of cases [6&12]. 70% of SFT express CD99 and Bcl-2 [12]. Reticulin staining may show reticular fibers surrounding the tumor cells [13].

In turn, a double negative CD34 and Bcl-2 makes the presence of SFT highly unlikely[6,7&12]. Often, there is a negative expression for epithelial markers (cytokeratin AE1/AE3, and /or EMA) [14]. Other differentiating markers (SMA, S-100, CD31) are negative. So is CD-117, which is one of the best markers for the diagnosis of gastrointestinal stromal tumors excluding SFT. The histopathological profile in our case corresponds nicely with the above literature evidence.

In addition, a recent study suggested that some SFT express either PAX8 or PAX2 [15]. We did not find positivity for these markers in our case.

NAB2-STAT6 gene fusion has been recently identified as the genetic hallmark of most SFTs in both, benign and malignant entities. This may be helpful in cases with diagnostic uncertainties[8&12].

With the increasing reported evidence on these rare tumors over the last 25 years, it seems that the prognosis is favorable[3,4,11&14], and up to 90% of cases can be considered as benign[2,11,&14]. Yet, rSFT can show malignant behavior with a risk of local recurrence or distant metastasis[17-20]. In fact, there is an ongoing controversy regarding its potential malignancy compared to thoracic SFT. Some authors suggest that rSFT have a similar prognosis[18,20&21], whilst others reported better oncological outcomes[22]. The diagnostic criteria for malignant SFT were proposed by England et al. in 1989 [23]:

(1) mitotic count >4/10 HPF,
(2) diversity of shapes of cells,
(3) many focal points in cells,
(4) necrosis of some cells.

These criteria were still in use in the latest classification of extra-pleural malignant SFT formulated by the WHO in 2013. This WHO classification of bones and soft tissue neoplasms terms them as fibroblastic/ myofibroblastic neoplasms with intermediate, rarely metastasizing biological [behavior6].

The optimal follow-up schedule is unknown due to the rarity of the condition, but complete surgical excision and long-term surveillance are recommended for these patients[18&19].

Overall recurrence rate is up to 10% for all extra-

pleural localizations[20&22]. Two mechanisms of development of malignant SFT are described by Yokoi et al.[24]. The first is a primarily aggressive malignant behavior of SFT, growing rapidly and metastasizing quickly. The other is the de-differentiation of a pre-existing SFT with an initial benign long-lasting clinical behavior.

We did not find in the English literature recurrence as we have described in this case, even if Almohammad and Bakour reported recurrence with thrombus extension into the inferior vena cava above renal veins level up to the right atrium [25].

In conclusion, only about a hundred cases of rSFT are reported in the literature. These rare mesenchymal neoplasms are most often diagnosed as renal cell carcinoma on imaging exams, and they have the potential for aggressive behavior. The mainstay of management is surgical treatment, even in the absence of evidence-based guidelines. The diagnosis of SFT is based on immuno-histochemical analysis. We reported what is, to our knowledge, the first case of rSFT recurrence as a thrombus into the subhepatic inferior vena cava extending to the external iliac veins. Hus, SFT need regular follow-up due to the uncertainty of prognosis.

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