CASE REPORT

# *A solitary fibrous tumor of the kidney in a 26-year-old man*

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***Background:*** *Solitary fibrous tumors (SFTs) are rare spindle cell neoplasms that typically arise in the pleura. There are several reports of cases that arose from a variety of sites. SFT of the kidney is rare and, to our knowledge, only 25 cases have been published in the literature to date. SFTs in the kidney have similar morphologic and immunologic features and biologic behaviors as SFTs found elsewhere. In general, patients with SFTs of the kidney have a favorable prognosis.*

***Case:*** *We report a case of SFT of the right kidney in a 26-year-old man. The tumor was localized in the upper and mid pole of the kidney. A nephrectomy was performed. The tumor was a well-circumscribed, solid mass attached to the renal capsule without necrosis or hemorrhage. Microscopically, a spindle cell neoplasm with alternating hypo- and hypercellular areas, storiform, fascilular and hemangiopericytoma-like growth pattern and less cellular dense collagen deposition was observed. Some glomeruli and renal tubules were entrapped by the tumor cells. There were no mitotic figures. Immunohistochemically, the tumor cells were consistently positive for CD34, CD99, and bcl-2. There was no evidence of recurrence after 6 months of follow-up.*

***Discussion:*** *Although morphology is most important in formulating the initial differential diagnosis, the addition of immunohistochemistry is vital in arriving at the correct classification of renal spindle cell tumors.*

**Key Words:** renal tumor, CD34, solitary fibrous tumor, soft tissue

## Background

Solitary fibrous tumors (SFTs) are rare spindle cell neoplasms found principally in the pleural cavity, hence the nomenclature of "fibrous mesothelioma" or "benign fibrous tumor of the pleura."[1,2] SFTs have been described in many extrapleural locations. These include the meninges, eyelid, orbit, neck, nasal cavity, mediastinum, retroperitoneum (attached to the bladder, ureter, ascending colon, or adrenal gland), liver, spleen, pelvic cavity (attached to the uterus or ovary), abdominal wall, paratesticular adnexa, soft tissue of the chest, and in the extremities.[3-5] In general, SFTs are slow growing with a favorable prognosis, although there have been reports of malignant cases.[2,6] The histogenesis of SFTs has been debated for years, with studies suggesting a mesothelial or mesenchymal origin. These lesions have been reported to arise from

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the renal capsule, renal pelvis, and peripelvic soft tissue. However, modern ultrastructural and immunohistochemical studies strongly point to a fibroblastic/primitive mesenchymal cell origin.[7] The SFTs arising in the kidney are a relatively recent addition to this group of neoplasms, with, to our knowledge, only 25 cases previously reported in the literature.[3,5,8-26] We report a case of SFT of the kidney in 26-year-old man. The diagnosis of SFT was established by light microscopy as well as by immunohistochemistry.

## Case

A 26-year-old man was admitted to our hospital following a car accident. He complained of pain in his right abdomen and had no hematuria or other symptoms. Until then he had been asymptomatic. A physical examination revealed a mild tenderness in his right abdomen. No lymphadenopathy was found. The patient's blood pressure and pulse rate were normal. Laboratory tests revealed no abnormalities. A contrast-enhanced abdominal computed tomography (CT) scan showed a 7 cm x 5.8 cm x 4.5 cm well-circumscribed tumor at the upper and mid pole of the right kidney. The mass consisted of a hypodense, heterogeneous area. The mass had enhanced areas in the corticomedullary phase, and this enhancement was maintained in the nephrographic phase, Figure 1. A chest x-ray, chest CT scans, and bone scans were all negative for metastasis.

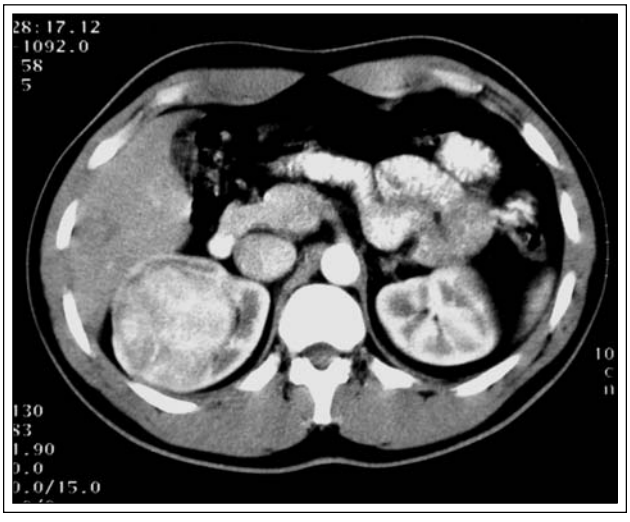


**Figure 1.** Contrast-enhanced computed tomography (CT) revealed a 7 cm x 5.8 cm x 4.5 cm well-circumscribed tumor at the upper and mid pole of the right kidney that consisted of hypodense, heterogeneous, and enhanced areas.

**Figure 2.** Macroscopy revealed a well-circumscribed mass, measuring 7 cm x 5.6 cm x 4 cm that consisted of firm, white-tan tissue, principally involving the renal parenchyma compressing the renal pelvis.

The patient underwent a right nephrectomy with no complications. The tumor consisted of a 7 cm x 5.6 cm x 4 cm, well-circumscribed, solid mass attached to the renal capsule. The cut surfaces were white-tan and firm without necrosis or hemorrhage, Figure 2. The mass principally involved the renal parenchyma and compressed the renal pelvis. Microscopic examination revealed a spindle cell neoplasm with alternating hypo- and hypercellular areas, storiform, fascilular and hemangiopericitoma-like growth patterns, Figure 3, and smaller areas of dense collagen

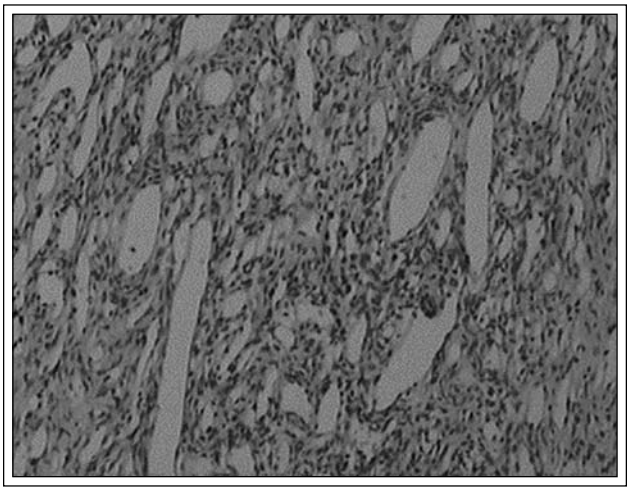

**Figure 3.** Microscopic features of the solitary fibrous tumor showed a hemangiopericytoma-like growth pattern.

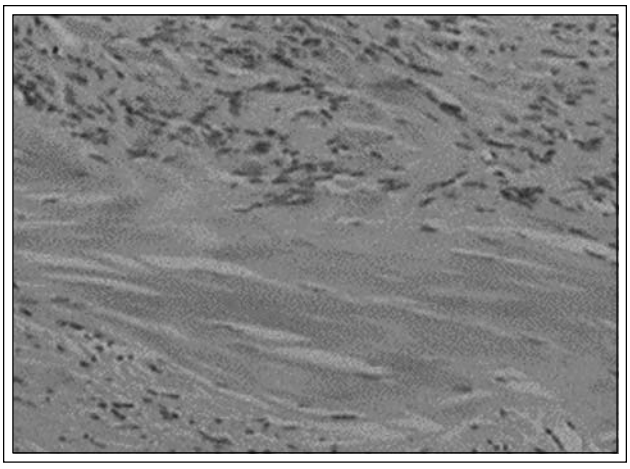

**Figure 4.** Microscopic features of the solitary fibrous tumor showing dense keloid-type collagen (hematoxylin-eosin, magnification x100).

deposition in cells**,** Figure 4. No invasion of the renal capsule, pelvis, or blood vessels was found. Some glomeruli and renal tubules were entrapped. The cells lacked cytological atypia, and mitoses were rare (< 1-2 mitoses per 10 high power fields). The immunohistochemical tests showed that the tumor cells were consistently positive for CD34, Figure 5, bcl-2, Figure 6, and CD99, but they were negative for CD117(c-kit), HMB45, and desmin antibodies. The findings were consistent with a SFT of the kidney. At a follow-up 6 months after the nephrectomy, there was no evidence of disease recurrence or metastatic disease.

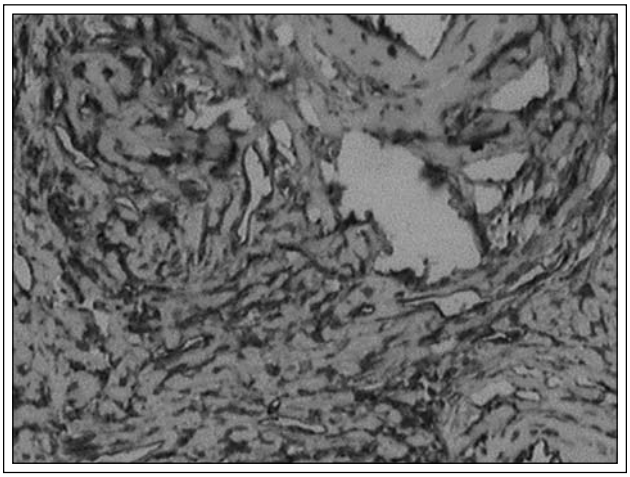

**Figure 5.** The tumor cells stained diffusely and strongly for CD34 (magnification x200).

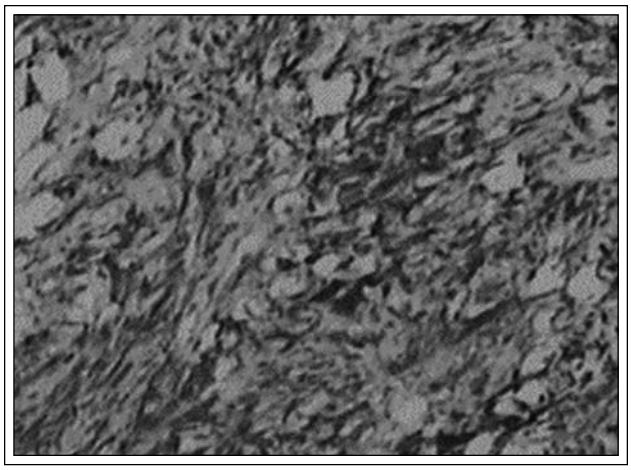

**Figure 6.** The tumor cells stained diffusely for bcl-2 protein (magnification x100).

## Discussion

SFTs may occur at any site, but they have been most frequently described in the pleura. An SFT of the kidney is rare, and, to our knowledge, only 25 cases have been reported to date in the literature, Table 1.[3,5,8-26] To our knowledge, our patient is the youngest reported case. Cortes-Gutierrez et al reported a case of SFT of the kidney in a 28-year-old woman.[8] The previously reported patients with SFT comprised 11 men and 14 women, aged 28 to 85 years. Their tumors measured between 2.0 cm and 25.0 cm in the greatest dimension. Fourteen tumors were located in the right kidneys; ten were in the left kidneys and one patient had bilateral lesions.[18] The clinical results after nephrectomy were available for 21 of the 25 patients, and showed that 19 patients had no evidence of disease, 1 patient died but not from kidney disease,[10] and 1 patient developed pulmonary metastases.[24]

In general, SFTs of the kidney have blunt-ended spindle cells upon histological examination and have a benign clinical course.[3,5,8-23,25,26] The criteria of malignancy include the presence of necrosis, hemorrhage, high mitotic rate (more than 4 per 10 high-power fields) and atypical mitosis or p53 expression.[2,6] By these criteria, we can clearly classify our case as histologically benign. Clinically, the mass in our study was considered to be malignant (renal cell carcinoma) because of its size and the radiographic study findings. Recently, Fine et al reported the first case of a malignant SFT of the kidney.[24] Cytologic malignancy in an SFT does not always portend an unfavorable clinical course.[2] Conversely, there are a few cases with benign histologic

TABLE 1. **Previously reported cases of Renal SFTs**

| No of cases | Source | Year | Sex | Age (years) | Side | Location | Size (cm) | Outcome |
|---|---|---|---|---|---|---|---|---|
| 3 | Fain et al[11] | 1996 | Female | 46 | Right | Kidney | 7.2 | NED |
| | | | Female | 45 | Right | Kidney | 6.0 | NED |
| | | | Male | 51 | Left | Kidney | 4.5 | NED |
| 1 | Gelb et al[10] | 1996 | Female | 48 | Right | Kidney | 3.0 | DNOD |
| 2 | Fukunaga et al[9] | 1997 | Female | 33 | Right | Renal pelvis | 3.5 | NED |
| | | | Female | 36 | Left | Renal pelvis | 2.0 | NED |
| 1 | Hasegawa et al[3] | 1999 | Male | 64 | Right | Kidney | 4.5 | NED |
| 1 | Leroy et al[12] | 2000 | Female | 66 | Right | Kidney | 9.0 | NA |
| 1 | Morimitsu et al[5] | 2000 | Female | 72 | Left | Kidney | 8.0 | NED |
| 1 | Yazaki et al[19] | 2001 | Male | 70 | Right | Renal pelvis | 6.0 | NA |
| 2 | Wang et al[13] | 2001 | Male | 41 | Left | Kidney | 14.0 | NED |
| | | | Male | 72 | Right | Kidney | 13.0 | NED |
| 1 | Cortes-Gutierrez et al[8] | 2001 | Female | 28 | Left | Kidney | 15.0 | NED |
| 1 | Magro et al[14] | 2002 | Female | 31 | Right | Kidney | 8.6 | NA |
| 1 | Durand et al[16] | 2003 | Male | 35 | Right | Kidney | 17.0 | NED |
| 1 | LIarena Ibarguren et al[18] | 2003 | Female | 51 | Left | Kidney | 25.0 | NA |
| | | | | | Right | Kidney | 2.0 | NA |
| 1 | Bugel et al[15] | 2003 | Female | 60 | Right | Kidney | 11.0 | NED |
| 1 | Gres et al[17] | 2004 | Male | 83 | Right | Kidney | 9.0 | NED |
| 1 | Yamada et al[20] | 2004 | Male | 59 | Left | Renal capsule | 6.8 | NED |
| 1 | Johnson et al[21] | 2005 | Female | 51 | Right | Kidney | 11.0 | NED |
| 1 | Kohl et al[22] | 2006 | Female | 85 | Left | Renal hilar | 4.5 | NED |
| 1 | Alvarez et al[23] | 2006 | Male | 36 | Right | Kidney | - | NED |
| 1 | Fine et al[24] | 2006 | Male | 76 | Left | Kidney | 12.0 | PM |
| 1 | Bozkurt et al[25] | 2007 | Female | 51 | Left | Kidney | 4.0 | NED |
| 1 | Znati et al[26] | 2007 | Male | 70 | Left | Kidney | 15.0 | NED |
| 1 | Current report | 2007 | Male | 26 | Right | Kidney | 7.0 | NED |

NED= no evidence of disease, DNOD= died not of disease, PM= pulmonary metastases, NA= not available.

findings that have invaded the wall of the renal vein.[12] These criteria, therefore, may be helpful in categorizing cases of SFT, but definite behavior cannot be predicted based on histologic criteria alone. As such, complete resection with long-term follow-up and surveillance is recommended.

SFTs are typically immunoreactive for CD34 and CD99 and most are negative for desmin, cytokeratin, S100 and α smooth muscle actin.[5,8,13,20,22-26] Immunoreactivity for bcl-2, when performed, was positive in all of the cases of SFT of the kidney,[5,13,20,22-26] suggesting that it may also be a sensitive marker for SFT. Immunoreactivity for CD34 and vimentin were identified in all of the reported cases of SFT of the kidney, including ours.

The differential diagnosis of SFT arising in the kidney includes fibroma, fibrosarcoma, hemangioma, angiosarcoma, leiomyoma, leiomyosarcoma, hemangiopericytoma, schwannoma, malignant peripheral nerve sheath tumor, fibroepithelial polyp of renal pelvis, renal sarcoma, sarcomatoid renal cell carcinoma, inflammatory myofibroblastic tumor (pseudotumor), benign and malignant fibrous histiocytoma, synovial sarcoma, angiomyolipoma, and gastrointestinal stromal tumor.

In conclusion, we present a case of SFT of the kidney

in a 26-year-old man, which was found incidentally in a CT scan. The outcome for patients with renal SFT is generally favorable, but complete excision of the mass and long-term follow-up is warranted because of its unpredictable behavior. SFTs should be included in the differential diagnosis for well-circumscribed renal spindle cell tumors (RSCTs). Although morphology is most important in formulating the initial differential diagnosis, the addition of immunohistochemistry is vital in arriving at the correct classification of RSCTs. Immunohistochemical studies with CD34, CD99 and bcl-2 are helpful in the diagnosis of SFT of the kidney. □

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