Solitary Fibrous Tumor of the Kidney: A Case Report and Review of the Literature

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A solitary fibrous tumor (SFT) is an unusual spindle cell neoplasm that usually occurs in the pleura but has recently been described in diverse extrapleural sites. Urogenital localization is rare, and only 19 cases of SFT of the kidney have been described. We report a case of a large SFT clinically thought to be renal cell carcinoma arising in the kidney of a 70-year-old man. The tumor was well circumscribed and composed of a mixture of spindle cells and dense collagenous bands, with areas of necrosis or cystic changes noted macroscopically and microscopically. Immunohistochemical studies revealed reactivity for CD34, CD99, and Bcl-2 protein, with no staining for keratin, S-100 protein, or muscle markers, confirming the diagnosis of SFT. This tumor is benign in up to 90% of cases. The immunohistochemical study is the key to diagnosis.
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Key words: Immunohistochemical study • Kidney • Renal neoplasm • Solitary fibrous tumor • Spindle cells

A solitary fibrous tumor (SFT) is an unusual spindle cell neoplasm of adults that was first described in 1931.[1] This type of tumor usually occurs in the pleura but has recently been described in diverse extrapleural sites. Urogenital localization is rare. To our knowledge, only 19 cases of SFT of the kidney have been reported (Table 1), and the origin of these tumors remains controversial.[2-16] In general, they are slow-growing tumors with a favorable prognosis, although there have been some malignant cases. This case study describes an SFT

Table 1
Clinicopathologic Findings of 19 Renal Solitary Fibrous Tumors

Case No.	Year	Authors	Age of Patient (y)	Sex of Patient	Site	Size (cm)	Treatment	Follow-up
1	1996	Gelb AB et al[2]	45	M	R kidney	3 × 2.5 × 1.5	Rad Np	Died (3 mo)
2	1996	Fain JS et al[3]	45	M	R kidney	6 × 5 × 3.5	Rad Np	8 mo; NED
3	1996	Fain JS et al[3]	46	F	R kidney	7.2 × 6 × 5.5	Rad Np	33 mo; NED
4	1996	Fain JS et al[3]	51	M	L kidney	4.5 × 4 × 2.5	Rad Np	2 mo; NED
5	1997	Fukunaga M and Nikaido T[4]	33	F	R renal peripelvis	3 × 2.5 × 2.5	R Np	2 mo; NED
6	1997	Fukunaga M and Nikaido T[4]	36	F	L renal peripelvis	2 × 1.5 × 1.5	L Np	12 mo; NED
7	1999	Hasegawa T et al[5]	64	M	R kidney	4.5	Rad Np	8 mo; NED
8	2001	Yazaki T et al[6]	70	M	R renal pelvis	6 × 4.5 × 4	Rad Np	60 mo; NED
9	2000	Morimitsu Y et al[7]	72	F	L kidney	8	Rad Np	10 mo; NED
10	2001	Wang J et al[8]	41	M	L kidney	14 × 12 × 7	L Np	4 y; NED
11	2001	Wang J et al[8]	72	M	R kidney	13 × 9 × 7	R Np	5 mo; NED
12	2002	Magro G et al[9]	31	F	R kidney	8.6	Rad Np	8 mo; NED
13	2003	Llarena Ibarguren R et al[10]	51	F	Bilateral	25 (L) 2 (R)	Resection	NED
14	2004	Kunieda K et al[11]	53	M	R kidney	14 × 13 × 10	R Np	NED
15	2004	Yamada H et al[12]	59	M	Renal capsule	6.8 × 4.4	L Np	4 y; NED
16	2005	Yamaguchi T et al[13]	51	F	L kidney	10 × 5 × 10	L Np	NED
17	2005	Johnson TR et al[14]	51	F	R kidney	11	Rad Np	NED
18	2006	Alvarez Mugica M et al[15]	36	M	R kidney		Rad Np	NED
19	2006	Fine SW et al[16]	76	M	L kidney	12	Rad Np	Malignant
Our case	2006	Znati K et al	70	M	L kidney	15 × 12 × 4	Rad Np	6 mo; NED

NED, no evidence of disease; Np, nephrectomy; Rad, radical; R, right.

arising in the kidney, the final diagnosis of which was made by immunohistochemical study. We discuss the clinicopathologic features of SFTs, the differential diagnosis, and prognosis of renal spindle cell neoplasms.

Case Report

A 70-year-old man complained of pain in his left lower back and hematuria of 1-month duration. The clinical examination revealed a left renal tumor. Laboratory data revealed no abnormalities. Computed tomography demonstrated a well-delineated, encapsulated tumor of the left kidney that exhibited enhancement with contrast medium and measured approximately 15 cm in diameter (Figure 1). The tumor involved the renal cortex and extended extensively into the perirenal adipose tissue. A magnetic resonance imaging (MRI) scan showed the tumor to be of low intensity on T1-weighted images and of irregular high intensity on T2-weighted images. No renal vein or inferior vena cava thrombosis was seen. No enlarged lymph nodes were seen in the abdomen. The patient underwent radical nephrectomy without complications. He is alive and has no evidence of disease 6 months after the surgery.

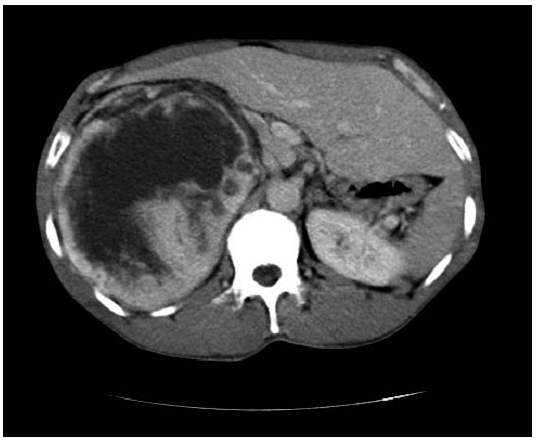


Figure 1. *Contrast computed tomography scan demonstrating an enhanced, well-demarcated tumor of the left kidney with multinodular configuration. The tumor involved the renal cortex and perirenal adipose tissue.*

Pathologic Findings

The cut section of the resected left kidney revealed a vaguely multinodular, grayish to white, firm, heterogenous tumor with cystic and necrotic changes. The tumor occupied the entire kidney and had slightly invaded the cortex and perirenal adipose tissue.

Microscopically, the mass consisted of bland, spindle-shaped cells with scant cytoplasm accompanied by prominent hyalinized collagenous tissue that was patternless or showed hemangiopericytomatous patterns (Figure 2). The cells lacked cytologic atypia and showed no mitosis. The tumor focally infiltrated the renal cortex, and some of the glomeruli and renal tubules were entrapped. The tumor was continuous to the renal capsule; however, the precise relationship between them was ambiguous. An immunohistochemical study was carried out with formalin-fixed, paraffin-embedded sections. The tumor cells were diffusely positive for CD34 (Figure 3), CD99, and Bcl-2. However, staining for cytokeratin, α-smooth muscle actin (α-SMA), S-100 protein, and p53 was negative for the tumor cells. The Ki67 index was also negative. Based on the histologic and immunohistochemical features, a diagnosis of SFT was established.

Discussion

The SFT is a rare but well-established neoplasm. Indeed, in 1942, Stout and Murray[17] introduced the concept that hemangiopericytoma originated from the pericytes of blood vessels. In 1994, Fletcher[18] proposed that hemangiopericytomas are heterogeneous in nature and may consist of SFTs and other distinctive soft tissue tumors, although the concept of hemangiopericytoma had been widely accepted. The new edition of the World Health Organization classification describes "hemangiopericytoma" as consisting of SFT and related conditions, including giant cell angiofibroma and lipomatous hemangiopericytoma.

Solitary fibrous tumors arise most frequently in the pleura; however, occurrences of these tumors at sites other than the pleura have been described in recent years. Extrapleural tumors have occurred in the upper respiratory tract, lung, nasal cavity, paranasal sinuses, orbits, mediastinum,

Figure 2. *Microscopic features of the solitary fibrous tumor. The tumor showed a hemangiopericytomatous growth pattern.*

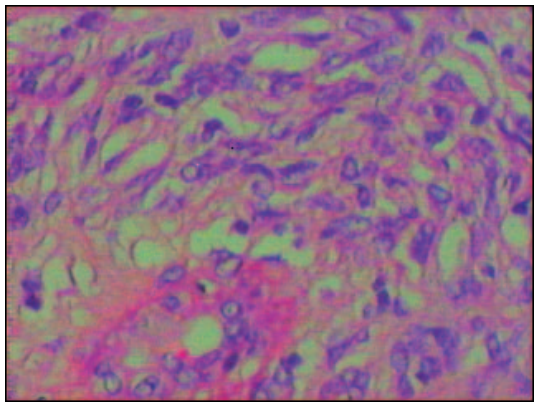
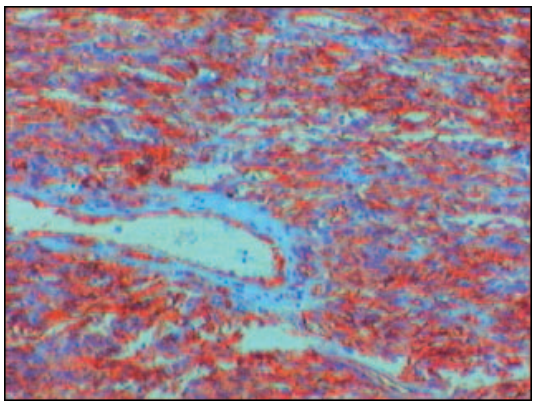


Figure 3. *The tumor cells stained diffusely and strongly for CD34. The vascular endothelium was also positive.*



major salivary glands, breast, meninges, liver, and urogenital organs. Of the renal SFTs reported in the literature during the past 10 years, more than 50% have occurred in patients older than 40 years (from 33 to 76 years, with an average age of 52 years). The male-to-female ratio appears to be almost equal (1:1.5).

The origin of most cases of SFT of the kidney is difficult to determine. Some reported cases of solitary fibrous tumor of the kidney were reported to have originated from the renal capsule.[2,9,12] In our case study, the tumor exhibited a unique growth pattern as a primary tumor of the kidney. The tumor involved not only the renal cortex but also the extrarenal soft tissue, suggesting the possibility of a renal surface origin of the tumor.

Interestingly, one SFT of the kidney showed an intrarenal growth pattern without connection to the renal capsule or renal pelvis.

Interestingly, one SFT of the kidney showed an intrarenal growth pattern without connection to the renal capsule or renal pelvis.[8] Further research is necessary to clarify the pathogenesis of these rare tumors.

Grossly, the renal SFTs reported in the literature ranged from 2 to 25 cm (mean, 8.75 cm). Most of the lesions were described as well-circumscribed or pseudoencapsulated, lobulated, rubbery or firm masses with a homogeneous, gray or tan-white, whorled cut surface.[2-16] Except for those in case 19 and our case, no tumor contained areas of cystic change, hemorrhage, or foci of necrosis.[16]

In all the reported cases of SFT of the kidney, final diagnosis was made by means of pathology. All tumors were characterized by spindle cell proliferation showing a patternless architecture with a combination of alternating hypocellular and hypercellular areas separated from each other by thick bands of hyalinized, somewhat keloidal collagen and branching hemangiopericytoma-like vessels.[2-16]

Electron microscopy reveals fibroblast-like tumor cells with rough endoplasmic reticulum and scattered mitochondria embedded in a collagenous matrix. Therefore, because of the absence of characteristic findings on electron microscopy, it is not necessarily an indispensable technique. Instead, the immunohistochemical study is the key to diagnosis. CD34 immunoreactivity has been reportedly shown to be strongly and diffusely expressed in many cases of SFT, and although it is not specific for SFT, strong CD34 reactivity is currently regarded as characteristic and an indispensable finding in the diagnosis of SFT. Seventy percent of SFTs express CD99 and Bcl-2; only 20% to 35% are variably positive for epithelial membrane antigen and smooth muscle actin. Focal and limited reactivity of S-100 protein, cytokeratins, and/or desmin has also occasionally been reported.[8,19]

Mesenchymal tumors that should be differentiated from SFT include sarcomatoid renal cell carcinoma, angiomyolipoma, fibroma, fibrosarcoma, leiomyoma, leiomyosarcoma, schwannoma, malignant peripheral nerve sheath tumor, hemangioma, angiosarcoma, synovial sarcoma, and gastrointestinal stromal tumor because these tumors typically show hemangiopericytomatous patterns. Diffuse positive expression of CD34, Bcl-2, and CD99 and negative expression of cytokeratin, α-SMA, S-100, CD31, and c-kit are useful for their differential diagnosis.[9,19]

Although most cases are benign, the behavior of SFTs is unpredictable. Roughly 10% to 15% of these tumors behave aggressively; thus long-term follow-up is mandatory. The histopathologic features related to clinical malignancy include increased cellularity, pleomorphism, increased mitotic activity (> 4 mitoses/10 high-power fields), necrosis, hemorrhage, and atypical location (parietal pleura, pulmonary parenchyma). However, there is far less information regarding the behavior of extrathoracic SFTs.[19]

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To summarize, we report a case of an SFT arising in the kidney with characteristic histology of spindle-shaped cells distributed in a haphazard pattern. The tumor cells are CD34 and Bcl-2 positive and α-SMA and S-100 protein negative, consistent with the immunohistochemical profile for SFTs. Although SFTs in extrapleural sites remain uncommon, they should be considered in the differential diagnosis of spindle cell neoplasms. ■

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Main Points

- Solitary fibrous tumors (SFTs) are unusual spindle cell neoplasms of adults that typically occur in the pleura but have been described in diverse extrapleural sites. Urogenital localization is rare; to the authors' knowledge, only 19 cases of SFT of the kidney have been reported.
- The origin of SFTs remains controversial, and further research is necessary to clarify the pathogenesis of these tumors.
- In all the reported cases of SFT of the kidney, final diagnosis was made by means of pathology. All tumors were characterized by spindle cell proliferation showing a patternless architecture with a combination of alternating hypocellular and hypercellular areas separated from each other by thick bands of hyalinized, somewhat keloidal collagen and branching hemangiopericytoma-like vessels.
- Immunohistochemical study is the key to diagnosing SFTs. CD34 immunoreactivity has been shown to be strongly and diffusely expressed in many of these tumors, and although it is not specific for SFT, strong CD34 reactivity is currently regarded as characteristic and an indispensable finding in the diagnosis of SFT.
- Although most SFTs are benign, their behavior is unpredictable. Roughly 10% to 15% of these tumors behave aggressively; thus long-term follow-up is mandatory.