The Korean Journal of Pathology 2010; 44: 420-5
DOI: 10.4132/KoreanJPathol.2010.44.4.420

Solitary Fibrous Tumor of the Kidney
– A Report of Two Cases with Review of Literature –

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Received : April 15, 2009
Accepted : August 12, 2009

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Solitary fibrous tumor (SFT) is a benign mesenchymal neoplasm usually occurring in the pleura. Kidney is one of the rarest sites for SFT. We report here on two cases of renal SFT found in 30-year-old and 33-year-old men with review of the literatures. Both cases manifested as well-enhanced solid masses in kidney and radical nephrectomies were done. The tumors consisted of bland-looking spindle cells arranged in short, ill-defined fascicles and storiform pattern with characteristic hemangiopericytoma-like blood vessels. The tumor cells were strongly positive for CD34 and CD99, focally positive for bcl-2, and negative for cytokeratin and human melanoma black-45 on immunohistochemical stainings. Possibility of SFT should be considered in the differential diagnosis of a renal mass which consists of benign-looking spindle cells and hemangiopericytomatous blood vessels. Immunohistochemical staining for CD34 is essential to confirm the renal solitary fibrous tumor.

Key Words : Solitary fibrous tumor; Kidney; Immunohistochemistry

Solitary fibrous tumor (SFT) is a relatively rare spindle cell neoplasm usually occurring in the pleura. However, SFTs may occur at various sites other than the pleura, including the central nervous system, orbit, oral cavity, nasopharynx, salivary gland, thyroid gland, mediastinum, peritoneum, retroperitoneum, liver, pancreas, bone, bladder, prostate, skin and vulva. To date, only 23 cases of SFT of the kidney have been reported in the Korean and English literatures (Table 1).[1-19] We report here on two cases of benign SFT in the kidney with immunohistochemical findings and summary of the clinicopathologic features of the previously reported cases.

CASE REPORTS

Case 1

A 30-year-old man was referred to our institute for evaluation and management of the renal mass which was found incidentally at outside hospital. Computed tomography (CT) revealed a 4 cm-sized well-circumscribed mass in the left kidney. The mass showed homogeneous density in non-enhanced image and strong early enhancement and rapid washout pattern in contrast-enhanced abdominal CT (Fig. 1A). Laboratory tests for blood and urine were normal except for mild elevation of alanine transaminase (49 IU/L; normal range, up to 40 IU/L). Further workup did not reveal any evidence of metastasis. The patient underwent left radical nephrectomy. The postoperative course has been unremarkable without tumor recurrence for four months.

Case 2

A 33-year-old man presented with a renal mass which was incidentally found on his regular medical check-up. Laboratory findings were unremarkable. Abdominal CT revealed a well-circumscribed mass in the right kidney. The mass showed isodensity with surrounding renal parenchyma in non-enhanced CT scan, well enhancement in intravenous contrast injection scan, and prolonged enhancement in delayed image (Fig. 1B). A Tc-99m diethylenetriamine-pentacetic acid renal scan revealed pelvocalyceal retention at the right upper pole, probably due to mass effect. No evidence of metastasis was revealed during further workup. He underwent right radical nephrectomy with no complication. The patient is alive and well without tumor recur-

Table 1. Clinicopathologic features of 24 renal solitary fibrous tumors

Case	Authors	Age (yr)	Gender	Size (cm)	Location	Surgery	Follow-up
1	Gelb *et al.*[1]	48	F	3 × 2.5 × 1.5	Right renal capsule	Radical nephrectomy	3 days, died of acute respiratory event
2	Fain *et al.*[2]	45	M	6 × 5 × 3.5	Right kidney	Radical nephrectomy	8 mo, NED
3	Fain *et al.*[2]	46	F	7.2 × 6 × 5.5	Right kidney	Radical nephrectomy	33 mo, NED
4	Fain *et al.*[2]	51	M	4.5 × 4 × 2.5	Left kidney	Radical nephrectomy	2 mo, NED
5	Fukunaga and Nikaido[3]	33	F	3 × 2.5 × 2.5	Right renal peripelvis	Nephrectomy	90 mo, NED
6	Fukunaga and Nikaido[3]	36	F	2 × 1.5 × 1.5	Left renal peripelvis	Nephrectomy	12 mo, NED
7	Hasegawa *et al.*[4]	64	M	4.5	Right kidney	Radical nephrectomy	8 mo, NED
8	Leroy *et al.*[5]	66	F	9 × 7 × 6	Right kidney	Radical nephrectomy	9 mo, NED
9	Yazaki *et al.*[6]	70	M	6 × 4.5 × 4	Right renal pelvis	Radical nephrectomy	60 mo, NED
10	Cortés-Gutierrez *et al.*[7]	28	F	15 × 11	Left renal capsule	Radical nephrectomy	1 yr, NED
11	Morimitsu *et al.*[8]	72	F	8	Left kidney	Radical nephrectomy	10 mo, NED
12	Wang *et al.*[9]	41	M	14 × 12 × 7	Left renal parenchyma	Nephrectomy	4 yr, NED
13	Wang *et al.*[9]	72	M	13 × 9 × 7	Right renal parenchyma	Nephrectomy	5 mo, NED
14	Magro *et al.*[10]	31	F	8.6	Right renal parenchyma	Radical nephrectomy	8 mo, NED
15	Kunieda *et al.*[11]	53	M	14 × 13 × 10	Right renal capsule	Mass extirpation	3 yr, NED
16	Yamada *et al.*[12]	59	M	6.8 × 4.4	Left renal capsule	Nephrectomy	4 yr, NED
17	Yamaguchi *et al.*[13]	51	F	10 × 10 × 5	Left kidney	Nephrectomy	NS
18	Johnson *et al.*[14]	51	F	13	Right renal capsule	Radical nephrectomy	NS
19	Fine *et al.*[15]	76	M	12 × 10 × 7.5	Left kidney	Radical nephrectomy	4 mo, metastasis
20	Znati *et al.*[16]	70	M	15 × 12 × 4	Left renal parenchyma	Radical nephrectomy	6 mo, NED
21	Bozkurt *et al.*[17]	51	F	4 × 3.5 × 3.5	Left renal parenchyma	Radical nephrectomy	10 mo, NED
22	Lee *et al.*[18]	71	F	3	Left renal pelvis	Radical nephrectomy	1 yr, NED
23	Constantinidis *et al.*[19]	26	M	7 × 5.8 × 4.5	Right kidney	Nephrectomy	6 mo, NED
24	Present case 1	30	M	3.5 × 3.5 × 2	Left renal peripelvis	Radical nephrectomy	4 mo, NED
25	Present case 2	33	M	5.2 × 4 × 4	Right renal peripelvis	Radical nephrectomy	4 yr, NED

M, male; F, female; NED, no evidence of disease; NS, not specified.

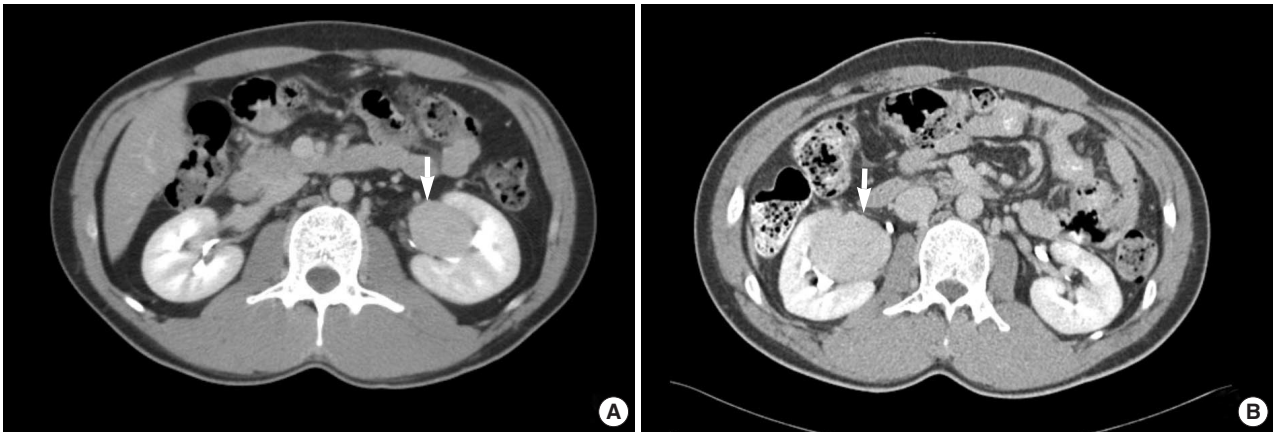

Fig. 1. Abdominal computed tomography scan in delayed phase. (A) Well-circumscribed mass with homogeneous density is located in the left kidney in case 1 and (B) in the right kidney in case 2.

rence or metastasis at 48 months postoperatively.

Pathologic findings

On gross examination, the resected kidney of case 1 showed a 3.5 × 3.5 × 2.0 cm sized well-defined solid mass located at mid- to lower portion of kidney and displacing renal pelvis upward. The mass merged with renal sinus fat. The cut surface of the mass was homogeneously grayish yellow, solid and fibrous with no hemorrhage or necrosis (Fig. 2A). In case 2, a 5.2 × 4.0 × 4.0 cm-sized well-circumscribed, bulging mass compressing the renal pelvis was present at the mid-portion of the kidney. The cut surface of the mass was pinkish gray, solid, and slightly trabeculated with no hemorrhage or necrosis (Fig. 2B).

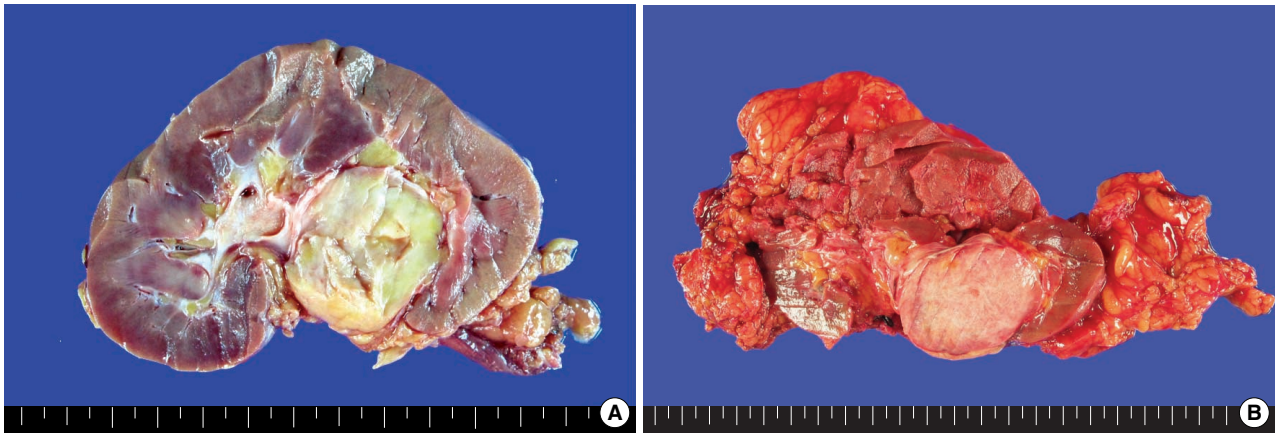

Fig. 2. Gross features of the renal masses. (A) Case 1. The tumor is well-defined and solid with homogeneously grayish yellow and fibrous cut surface. (B) Case 2. A well-defined bulging contoured mass occupies the mid-portion of the kidney. The cut surface is pinkish gray and solid with fascicles of fibrous tissue.

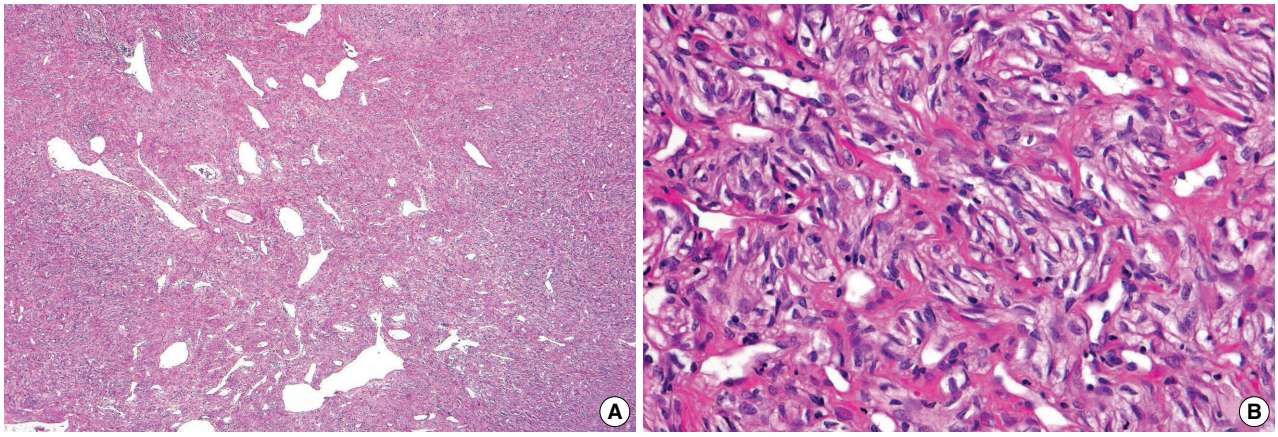

Fig. 3. Microscopic features. (A) The tumor consists of spindle cells in haphazard to storiform arrangement and hemangiopericytoma-like blood vessels. (B) Bland-looking spindle cells are admixed with interstitial collagen.

Microscopically, both tumors consisted of bland-looking spindle cells in haphazard to storiform arrangement and hemangiopericytoma-like blood vessels (Fig. 3A). The cellularity was variable and hypercellular and hypocellular areas alternated in the tumor. The tumor cells had elongated plump nuclei with dispersed chromatin and indistinct nucleoli and scanty cytoplasm. The cytoplasmic border of the tumor cells was indistinct and merged with collagen fibers (Fig. 3B). Mitotic figures were rare. The tumor was well demarcated from the renal parenchyma, but merged with renal sinus fat.

Immunohistochemical findings

In both cases, the tumor cells were positive for CD34 (1 : 500, Immunotech, Marseille, France) and CD99 (1 : 50, Dako, Glostrup, Denmark), focally positive fro bcl-2 (1 : 25, Dako), and negative for cytokeratin (CK; 1 : 250, Zymed, San Francisco, CA, USA) and human melanoma black (HMB)-45 (1 : 50, Dako) (Fig. 4). Positive reactivity for smooth muscle actin (SMA; 1 : 200, Dako) was identified only in case 1.

DISCUSSION

SFTs are usually slow-growing, benign spindle cell tumors arising in the pleura. Extrapleural SFTs may be found in a wide variety of sites, however, the preferential locations are soft tissues, the orbit, and the upper respiratory tract. SFTs are rare in the urinary tract including kidney.[1-19] Extrapleural SFTs usually arise in middle-aged adults, with no sex predilection. The

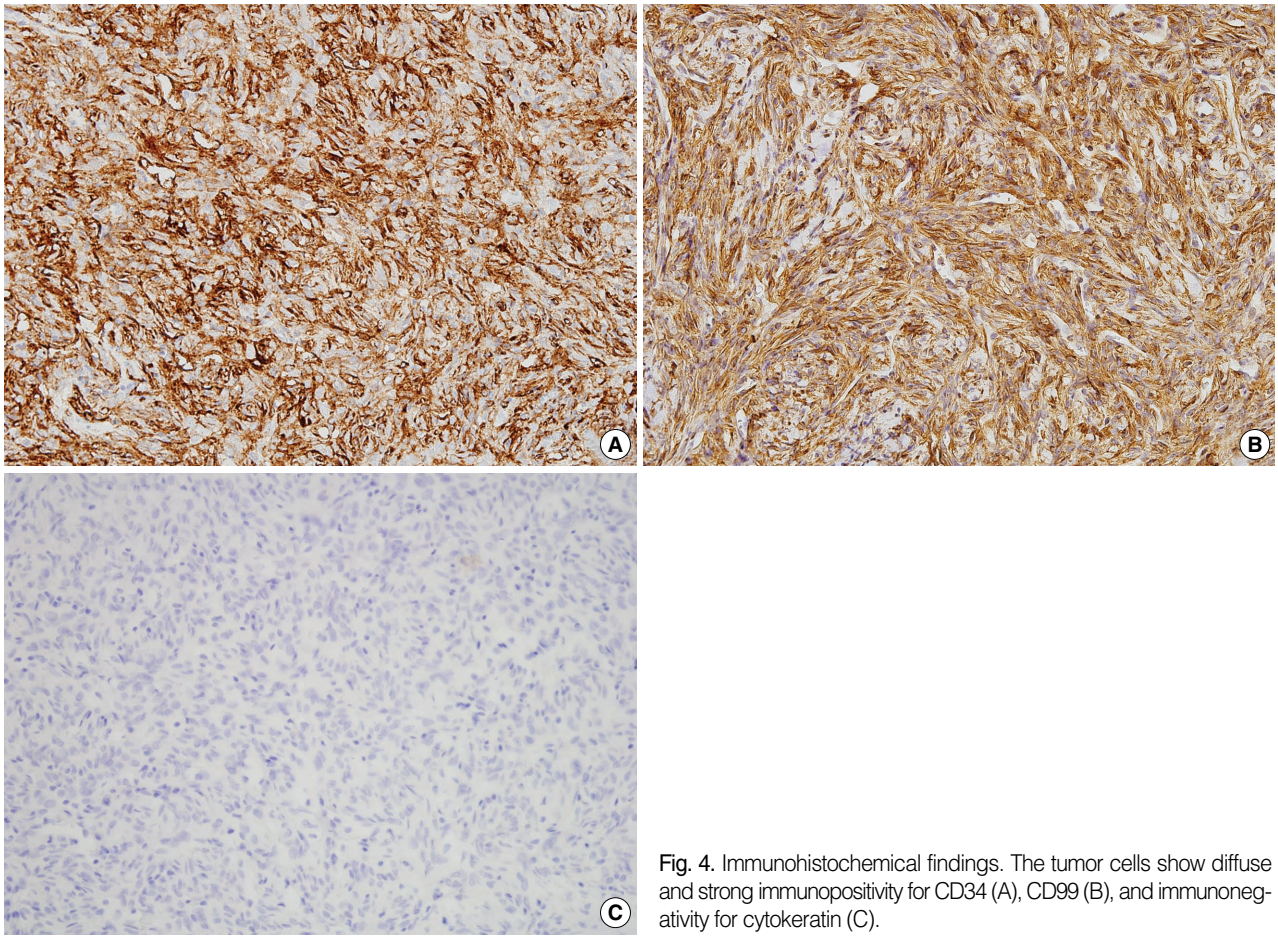

Fig. 4. Immunohistochemical findings. The tumor cells show diffuse and strong immunopositivity for CD34 (A), CD99 (B), and immunonegativity for cytokeratin (C).

histology of SFTs is characterized by a combination of alternating hypocellular and hypercellular areas and hemangiopericytomatous blood vessels. Cytogenetic changes specific for SFTs have not been unraveled yet.[20]

To date, only 22 cases and one case of renal SFT have been described in English and Korean literatures, respectively. The clinical features of the 25 renal SFTs including the present cases are summarized in Table 1. The age of the patients ranged from 26 to 71 years (median, 51 years) and the male-to-female ratio was almost equal (1 : 1.1). All cases except for one (case 15) underwent radical or simple nephrectomy. Most frequent presenting symptom was abdominal or back pain. Almost all tumors were well-circumscribed in spite of the large tumor size. However, one case (case 19) was poorly defined with gross invasion beyond the renal capsule in which the tumor consisted of pleomorphic, spindle cell sarcoma with a focal area of typical SFT, suggesting sarcomatous transformation from a benign SFT.[15] One case of renal SFT (case 8) showed renal vein invasion, suggesting an aggressive behavior.[5] The mean tumor size was 7.9 cm (range, 2 to 15 cm). Except for one case (case 19), the renal masses consisted of benign-looking spindle cells, which were arranged haphazardly or in a short storiform pattern with varying degrees of collagen deposition and hemangiopericytomatous blood vessels. In most cases, the postoperative course was uneventful excluding one case (case 19) metastasized to the lung at four months postoperatively.[15]

The histogenesis of renal SFTs has not been elucidated yet. However, some cases are presumed to originate from mesenchymal cells in the renal capsule[1,7,10,12] or in the peripelvic area,[3,6,9] because more than one-third of renal SFTs including the our ones is contiguous to the renal capsule or located in the peripelvic or pelvic area. Therefore, renal capsular or peripelvic connective tissue is considered the most possible origin of renal SFTs.

Immunohistochemical stainings are necessary to the diagnosis of SFTs. The tumor cells in SFTs show diffuse and strong CD34 positivity in 90% to 95% of cases.[20] Thus, CD34 immunoreactivity is a characteristic and indispensable finding for the diagnosis of SFTs.[1,3,6,8,20] Diffuse and strong immunoreactivity for CD34 suggests that SFTs are derived from CD34-positive subsets of fibroblast.[20] Occasional immunoreactivity for SMA and

desmin suggests possible myofibroblastic differentiation.[16,21] About two-thirds of SFTs show positivity for bcl-2 protein and CD99, however, SFTs are invariably negative for CK.[16,21] Therefore, immunohistochemical features of the present cases, i.e., immunoreactivity for CD34, CD99, bcl-2, and SMA (one case) and negativity for CK support the diagnosis of SFT.

The histologic differential diagnosis for renal SFT includes hemangiopericytoma, synovial sarcoma, sarcomatoid renal cell carcinoma, and other spindle cell neoplasms. Hemangiopericytoma is difficult to differentiate from SFT because of overlapping histologic features such as proliferation of fusiform spindle cells, staghorn vascular pattern and CD34 positivity. However, we prefer SFTs in the present cases on the basis of patternless architecture characterized by alternating hypocellular and hypercellular areas and unevenly distributed hemagiopericytoma-like vasculature. Monophasic synovial sarcomas can be confused with renal SFTs based on hemangiopericytoma-like vascular pattern and immunopositivity for bcl-2 protein and CD99. However, lack of morphologic features of malignancy such as high cellularity and frequent mitoses as well as immunopositivity for CD34 and immunonegativity for CK and/or epithelial membrane antigen would help to rule out synovial sarcoma.

Differentiation of renal SFTs from malignant spindle cell neoplasms such as fibrosarcomas, malignant peripheral nerve sheath tumors or sarcomatoid renal cell carcinomas might be difficult especially on the small biopsy sample.[2,10] However, benign SFTs can be distinguished from those tumors by the absence of pleomorphism, high cellularity and high mitotic rate in addition to diffuse positivity for CD34. Particularly, herringbone pattern of fibroblastic tumor cells in fibrosarcomas and CK expression in sarcomatoid renal cell carcinomas could be helpful to make a diagnosis.

Renal SFTs might be confused with angiomyolipomas especially when the tumor cells show diffuse SMA positivity or lipomatous change.[13] In that case, efforts to search hemangiopericytoma-like vascular pattern and immunohistochemical stainings for CD34 and HMB-45 could be helpful to reach the diagnosis. Histologic features favoring angiomyolipomas are thick-walled blood vessels and myoid differentiation of the tumor cells.

Other differential diagnoses include benign spindle cell tumors, such as fibromas and inflammatory myofibroblastic tumors.[4,21] Fibromas are typically small and lacks the typical growth patterns of SFT and CD34 immunoreactivity. Inflammatory myofibroblastic tumors are characterized by myofibroblastic cells arranged in myxoid or hyalinized stroma and infiltration of lymphocytes and plasma cells. Absence of significant lymphoplasma cell infiltration and CD34 positivity are features favoring SFTs over inflammatory myofibroblastic tumors.

Most cases of SFTs are benign, however, roughly 10-23% of these tumors are described to show aggressive behavior.[15,16,20] Malignant SFTs are usually hypercellular lesions, showing at least focal cytological atypia, tumor necrosis, numerous mitoses ($\geq$ 4 mitoses/10 high power fields) and/or infiltrative margins.[20] Among 25 renal SFTs (including the present cases), only one case showed malignant histologic features and two cases (8%) displayed aggressive clinical behavior. However, a long-term follow-up is needed to determine the biologic behaviors of the renal SFTs.

In summary, we present two cases of renal SFT with characteristic histologic findings and immunohistochemical features. It is important to consider the possibility of renal SFTs when we encounter renal spindle cell tumors, and immunohistochemical stainings for CD34, CK, CD99, and bcl-2 should be performed to confirm the diagnosis of SFT of the kidney.

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