Case Report

A case of solitary fibrous tumor of the kidney

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Abstract

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 to our knowledge, only 39 cases of SFT of the kidney have been described. Although SFT of the kidney is extremely rare, this tumor must be included in the differential diagnosis, whenever a renal tumor consisting of mesenchymal elements is encountered. We report a case of a large SFT of the right kidney which was clinically and radiologically thought to be renal cell carcinoma and a final diagnosis of SFT was made only after immunohistochemical study.

Key Words: Immunohistochemical study, kidney, solitary fibrous tumor, spindle cells

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INTRODUCTON

A solitary fibrous tumor (SFT) is an unusual spindle cell neoplasm that usually occurs in the pleura and rarely in extrapleural sites like upper respiratory tract, lung, nasal cavity, paranasal sinuses, orbits, mediastinum, major salivary glands, breast, meninges, liver, and urogenital organs. Morphologically, SFT is characterized by spindle cell proliferation with a patternless architecture, and a final diagnosis is made only after immunohistochemical study.[1] We report a case of SFT which was radiologically thought to be a malignant lesion.

CASE REPORT

A 52-year-old woman presented with pain abdomen and right renal mass. Computed tomography scan showed a well-delineated mass arising from upper pole of right kidney measuring 10 × 10 cm with contrast enhancement [Figure 1a and b]. She underwent right radical nephrectomy with a presumptive diagnosis of renal cell carcinoma (RCC). Intraoperatively, there was renal vein thrombus extending to inferior vena cava (IVC) just proximal to renal vein ostium. Thrombectomy was also done.

Macroscopically, the size of the kidney was 17 × 10 × 10 cm with nodular surface [Figure 2]. Cut section showed a circumscribed pale brown mass measuring 18 × 9 × 10 cm involving most of the renal parenchyma. Tumor was nodular with focal cystic areas and homogeneous pale brown areas [Figure 3]. Microscopy showed a neoplasm characterized by proliferation of bland spindle cells which had a patternless architecture with a combination of alternating hypocellular and hypercellular areas. Ectatic blood vessels were seen. Areas of myxoid degeneration and hyalinization were present. The surrounding renal parenchyma was compressed and showed interstitial fibrosis and inflammation. Section from the vein showed bland thrombus. There was no evidence of malignancy. These findings were suggestive of benign spindle cell neoplasm suggestive of SFT [Figure 4a and b]. On immunohistochemistry, neoplastic cells were positive for Vimentin, CD 34, Factor VIII, CD-99, Bcl-2 and negative for SMA, S-100, and CD-68. Ki-67 proliferative index was low. These features were suggestive of benign SFT of kidney [Figure 5].

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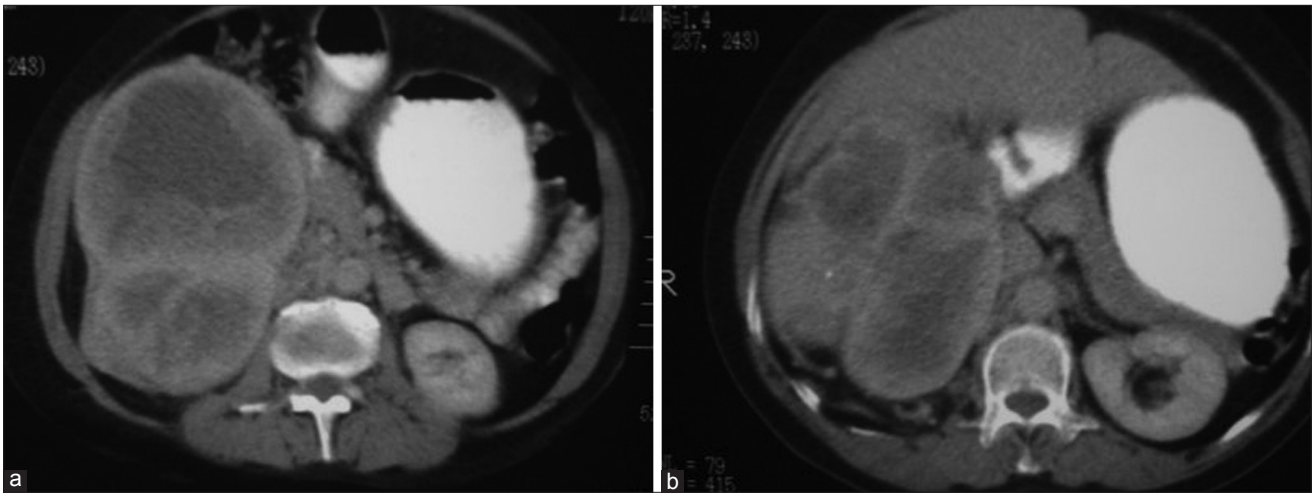

Figure 1 (a and b): CT scan showed a well-delineated mass arising from upper pole of right kidney measuring 10 x 10 cm with contrast enhancement

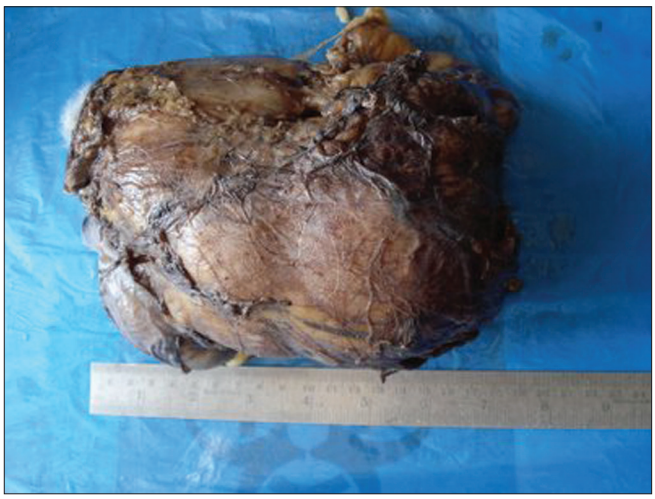

Figure 2: Macroscopically, kidney was 17 × 10 × 10 cm, with nodular surface

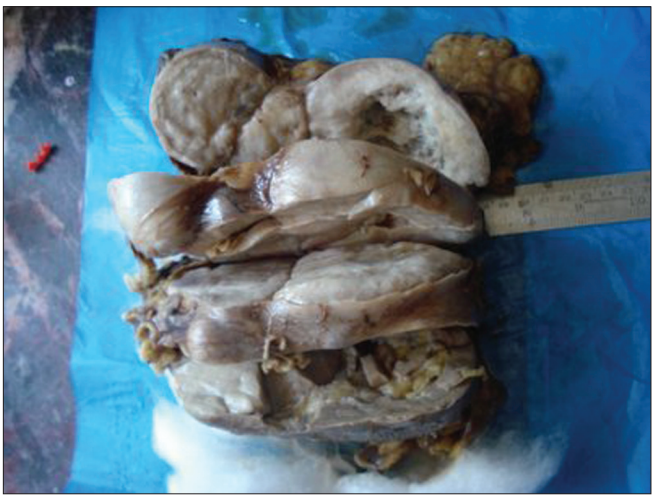

Figure 3: Tumor was nodular with focal cystic areas and homogeneous pale brown areas

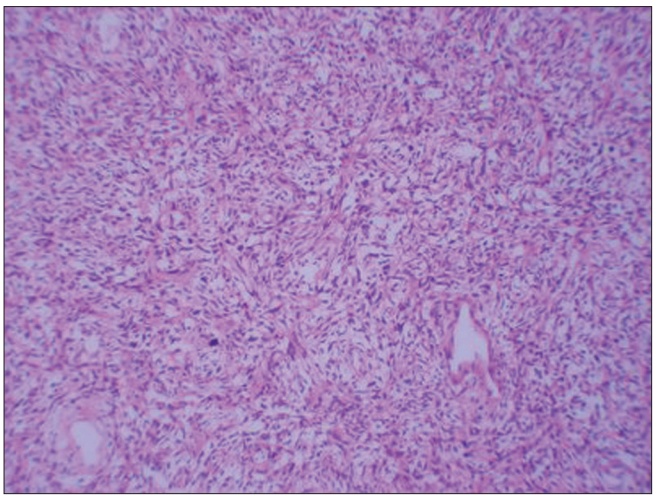

Figure 4a: Low power field view (×10) showing proliferation of bland spindle cells with ectatic blood vessels

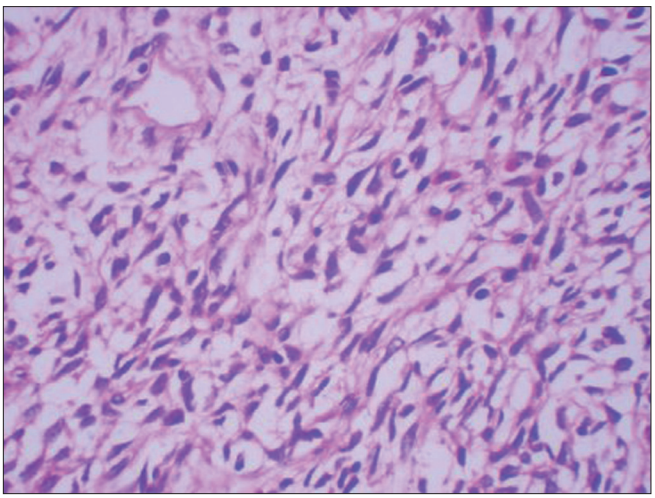

Figure 4b: High power field view (×40) showing proliferation of bland spindle cells with a patternless architecture

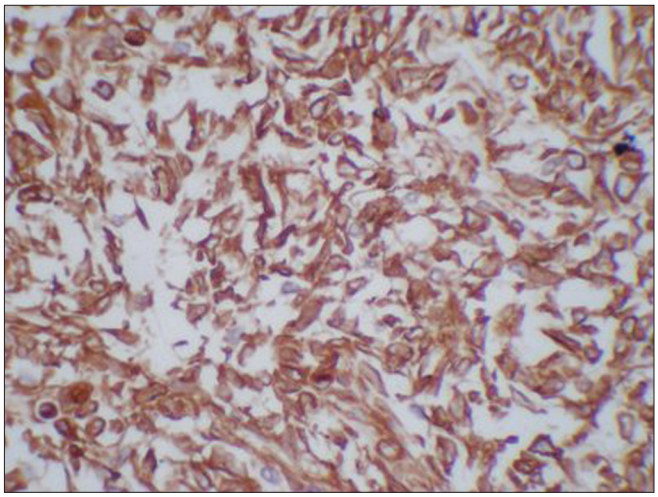

Figure 5: Immunohistochemistry, high-power view (×40) showing cells positive for Vimentin

She has no evidence of disease recurrence 6 months after the surgery.

DISCUSSION

SFTs are mesenchymal tumors, now considered a variant of hemangiopericytomas.[2] They 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, major salivary glands, breast, meninges, liver, and urogenital organs.[3] The origin of most cases of SFT of the kidney is difficult to determine and may originate from the renal capsule,[4] interstitial tissues, or peripelvic connective tissues.

Seventy percent of SFTs express CD 34, 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.[5] Diffuse positive expression of CD34, Bcl-2, and CD99 and negative expression of cytokeratin, α-SMA, S-100, CD31, and c-kit are useful in differentiating these entities. Electron microscopy shows fibroblast-like cells with well-developed rough endoplasmic reticulum, surrounded with collagen fibers.[5] Histopathological examination, immunohistochemical study, and ultrastructural study are the key to diagnosis for SFT. Strong CD34 reactivity is currently regarded as characteristic and an indispensable finding in the diagnosis of SFT.[5]

The differential diagnosis includes sarcomatoid renal cell carcinoma, angiomyolipoma, fibroma, and fibrosarcoma because these tumors typically show hemangiopericytomatous patterns.[1] SFT is a rare tumor mimicking renal cell carcinoma and must be included in the differential diagnosis when a renal tumors consisting of mesenchymal elements are encountered. Roughly 10 to 15% of these tumors behave aggressively.[1] 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, the clinical behavior cannot be predicted on histopathological basis with benign-appearing tumors exhibiting aggressive behavior and vice versa. Therefore, all patients with these tumors need to be on long-term follow-up.[1,3]

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