Case Report
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A Solitary Fibrous Tumor with Myxoid Change and Giant Cells of the Kidney

Cinzia Fasano, Stefania Vadrucci, Antonella Maglietta, Maria Arcangela Cascaranoe and Giovanni Serio*

Department of Surgical Pathology, Hospital Vito Fazzi, Lecce, Italy

Abstract

Solitary fibrous tumor of the kidney is exceptionally rare and limited knowledge regarding its behavior makes the prognosis of this tumor difficult. We report a case of solitary fibrous tumor with diffuse myxoid change and numerous giant cells affecting the left kidney of 69 years of age patient. We discuss the pathologic features, the immunohistochemical profile and the differential diagnosis.

Keywords

Solitary fibrous tumor • Spindle cells renal tumor • Immunohistochemistry

Introduction

Solitary Fibrous Tumor (SFT) is a mesenchymal spindle cell tumor which may arise in any part of the human body but it is uncommon in the kidney, in which it has similar histologic features and biologic behavior as the SFTs found elsewhere [1].

Just over a hundred cases of SFT of the kidney have been reported so far in the literature. In general, they are slow-growing tumors with a favorable prognosis, although there have been some malignant cases [2]. The case here reported describes a SFT arising in the kidney, the final diagnosis of which was made by immunohistochemical study. We report a case of renal SFT with extensive mixoid changes, the immunohistochemical findings and summary of the clinicopathologic features of the previously reported cases.

Case Report

A 69-year-old patient with no prior surgical history presented for diffuse lumbar pain. The abdominal ultrasound revealed the presence of a large retroperitoneal entirely destroying the left kidney. Computed tomography confirmed a 12 × 9 × 8 cm mass involving the left kidney in the absence of other lesions. The patient was submitted to surgery and the tumor was resected en bloc with left radical nephrectomy. The postoperative course was uneventful and the patient was discharged on the sixth postoperative day.

Macroscopic examination of the resected left kidney showed a grayish to white, firm with gelatinous areas tumor occupying the entire kidney. The mass measured 11 × 7 × 7 cm and did not extend to the capsular adipose tissue (Figure 1).

Figure 1. Gross morphology: the tumor was firm and showed a yellowish white to tran-gray, myxoid and lobulated cut surface with hemorrhage and necrosis.

Microscopic examination revealed a mesenchymal neoplasm surrounded by fibrous tissue occasionally separated by strip-like bands of collagen. The proliferation was composed of long spindle cells with acidophilic cytoplasm and vesicular nuclei with bland atypias, organized in a pattern less architecture with alternating hypo cellular and hyper cellular areas separated from each other by thick bands of hyalinized collagen. Hypo cellular areas showed extensive myxoid changes with numerous floret-like giant cells. There were numerous branching, hemangiopericytoma-like blood vessels present throughout the tumor. There was no evidence of necrosis and the mitotic count was <1/2 mm^2 . Immunohistochemical study showed a diffuse positivity of the tumor cells for vimentin, CD34, BCL2, STAT6 and CD99 (Figure 2).

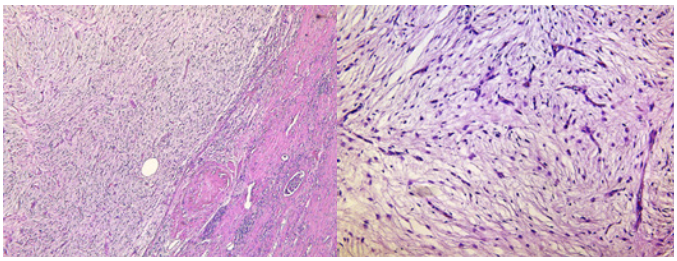

Figure 2. Microscopic findings: intrarenal spindle cell neoplasm (A, H&E x 100) with prominent myxoid change (B, H&E x 200).

Stainings for cytokeratins, S-100, Melan A, HMB45, a- smooth muscle actin, Desmin and CD117 were negative.

***Address for Correspondence:** Giovanni Serio, Department of Surgical Pathology, Hospital Vito Fazzi, Lecce, Italy; E-mail: gserio.anapat.le@gmail.com*

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Based on the histologic and immunohistochemical features, a diagnosis of SFT was established.

The patient is now currently free of disease at the 6th month of follow-up.

Discussion

Extra pleural SFTs are common and they occur at any anatomical site, including superficial and deep soft tissues and within visceral organs and bones. SFT arising from the genitourinary system is rare and renal SFT is especially uncommon. The age range of patients with renal SFT reported in the literature during the past 15 years is 31years-76 years. Men and women are equally affected and the tumors appear more often in the right kidney. Patients are mostly asymptomatic or have pain secondary to a mass effect due to the size of the tumor. The origin of the tumor is difficult to determine. Most reported cases of SFTs of the kidney were reported to have originated from the renal capsule or renal pelvis [3]. In our case the tumor showed an intra-renal growth pattern without connection to the renal capsule or renal pelvis.

Macroscopically, renal SFTs range from 2 cm to 25 cm in maximum diameter and most of the lesions were described as pseudo-encapsulated, lobulated or firm masses with a homogeneous gray or tan-white cut surface [1]. The case reported here showed translucent areas and extensive myxoid features.

In all the reported cases of renal SFTs, final diagnosis was made by histologic examination and immunohistochemical stainings were necessary to the differential diagnoses [4]. All tumors were characterized by spindle cell proliferation showing a patternless architecture with alternating hypo cellular and hyper cellular areas separated each other by thick bands of hyalinized collagen and branching hemangiopericytoma-like vessels. SFTs are highly variable in appearance, depending on the relative proportion of cells and fibrous stroma. Myxoid change is common and, when extreme, may produce an appearance similar to myxoid liposarcoma. Fat-forming [5], giant cell-rich [6] and dedifferentiated tumors [7] have also been described.

By immunohistochemistry, tumor cells show diffuse and strong expression of CD34 present in up to 95% of cases. Although it is not specific for SFT, strong CD34 reactivity is currently regarded as characteristic and an indispensable finding in the diagnosis of SFT. 70% of SFTs express CD99 and BCL2; only 20% to 35% are variably positive for epithelial membrane antigen and smooth muscle actin.

Mesenchymal tumors that should be differentiated from renal SFT include sarcomatoid renal cell carcinoma, angiomyolipoma, fibroma, fibrosarcoma, leiomyoma, leiomyosarcoma, hemangioma, angiosarcoma and gastrointestinal stromal tumor. Appropriate immunohistochemical stains allow a differential diagnosis. 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 summarized in Figures 3A-3E.

The genetic hallmark of SFT is a paracentric inversion involving chromosome 12q, resulting in the fusion of the NAB2 and STAT6 genes. STAT6 stain has proved to be a good surrogate marker for the genetic alteration (NAB2-STAT6 gene fusion) in solitary fibrous tumor [8]. To date, STAT6 immunoreactivity has been reported only in few cases of SFT of the kidney [9] and the prognostic role of NAB2-STAT6 fusion deserves further investigations [10].

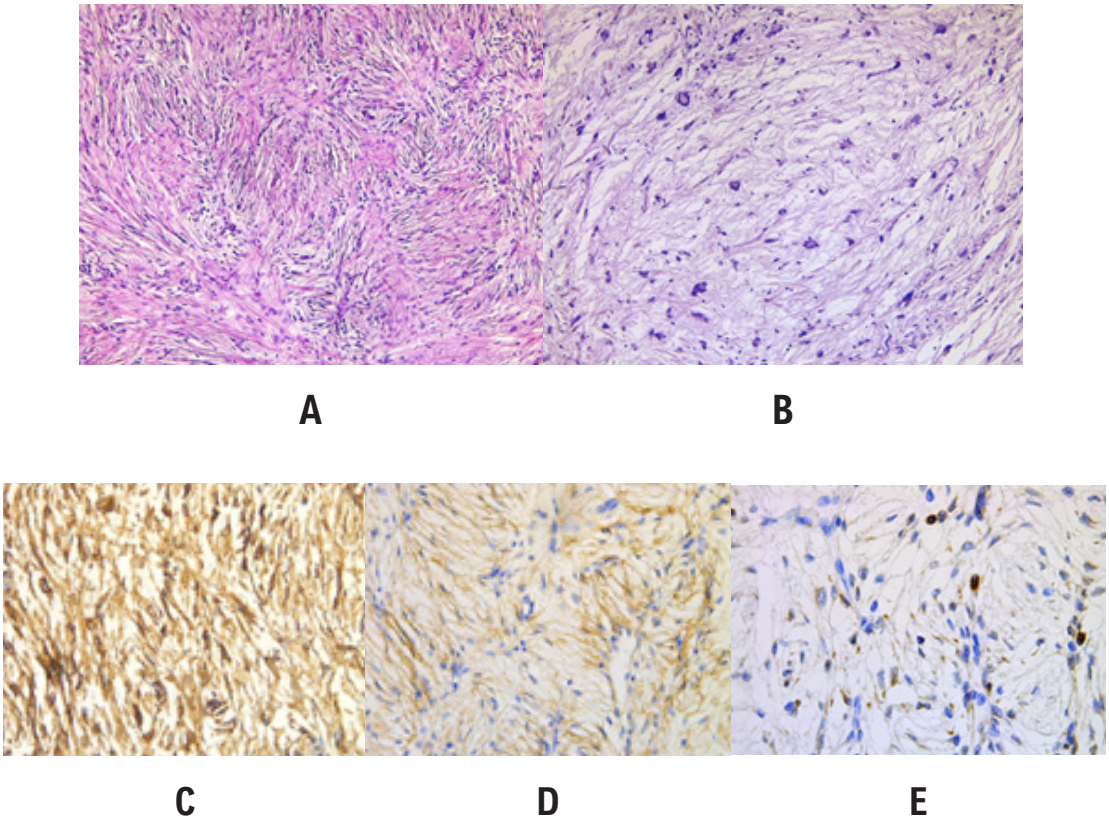

Figure 3. (A): Storiform arrangements of spindle cells in fibrous stroma containing dense collagen fibers (× 400 original magnification); 3. (B); Haphazard arrangements of multinucleated floret-like cells in a loose myxoid stroma (× 400 original magnification). 3. (C-E): CD34, CD99 and STAT6 stains respectively.

Conclusion

Although most cases of renal SFTs are benign, the behavior is unpredictable. About 10% of these tumors behave aggressively. Increased cellularity, pleomorphism, mitotic activity>4 mitoses/10 high power fields, necrosis, hemorrhage are histological features related to malignant outcome. There is currently little information on the clinical behavior of extra pleural SFTs and follow-up is required.

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Consent

Written informed consent was obtained from the patient for the publication of this report and any accompanying images.

Competing interests

The authors declare that they have no competing interests

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