Case report

Primary Renal Synovial Sarcoma: A Rare Tumor with an Atypical Presentation

Ankita Majumder•[1], Soumit Dey MD[1], Binny Khandakar MD[1], Sripurna Medda MD[1], Prabir Chandra Paul MD[1]

Abstract

Primary renal synovial sarcoma (PRSS) is a very rare tumor, first described by Argani, et al. The exact incidence of PRSS is not yet known. Here we present a case of PRSS diagnosed by histopathology, supplemented with immunohistochemistry.

Keywords: Atypical presentation, immunohistochemistry, synovial sarcoma

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Introduction

Synovial sarcoma is a clinically and morphologically well-defined entity, occurring primarily in the para-articular regions of the extremities, usually in close association with tendon sheaths, bursa, and joint capsules. These tumors are rarely diagnosed in unexpected sites, including the visceral organs, lungs, and kidneys. PRSS is rare, presents like any other common renal neoplasms. The scope of a definite pre-operative diagnosis is limited. The diagnosis is established by morphological examination with supplementary immunohistochemistry and/or cytogenetic analysis. A characteristic and consistent translocation, t (X; 18) (p11; q11) is seen in the majority of patients with synovial sarcomas. The resulting chimeric SYT-SSX transcript is a signature molecular marker of this neoplasm. A panel of immunohistochemistry including CD 99, SMA, CD 34, EMA, CK, S 100, and BCL 2 is often required to rule out other morphological mimickers.[1]

Case Report

A 46-year-old female patient presented with a 12-week history of fullness in the right flank and hematuria. Per-abdominal examination revealed a mass palpable in the right lumbar region. An initial ultrasonography and computerized tomography scans of the whole abdomen revealed a large retroperitoneal homogeneous mass involving the right kidney. A pre-operative ultrasound-guided fine needle aspiration cytology (FNAC) was done. The cytological aspirate was fairly cellular and revealed loosely cohesive clusters of spindle cells with oval hyperchromatic nuclei showing mild to moderate pleomorphism with minimal fat and blood vessels (Figures 1c and 1d). A provisional diagnosis of primary renal spindle cell sarcoma was made. Angiomyolipoma was kept as a differential. The patient subsequently underwent radical right nephrectomy, and the specimen was sent for histopathological examination.

Grossly, the tumor was well circumscribed, solid, homogeneous, greyish white (18 × 10 × 8 cm) with focal myxoid areas, originating from the upper pole of the kidney (Figures 1a and 1b). Normal renal parenchyma was identified in the periphery of the mass. There was no capsular breach or extra renal extension. The renal sinus and the hilar vessels were grossly free of tumor. Sections examined showed tumor cells arranged in intersecting fascicles, interspersed with a few antler-like blood vessels. The individual tumor cells displayed indistinct cytoplasmic borders, scant to moderate pale eosinophilic cytoplasm, oval to spindle hyperchromatic nuclei with moderate pleomorphism (Figures 2a and 2b). An occasional bizarre tumor cell and mitosis was also identified. The following morphological differentials were considered-leiomyosarcoma, malignant peripheral nerve sheath tumor, fibrosarcoma, solitary fibrous tumor and hemangiopericytoma, as well as PRSS and angiomyolipoma (epithelioid variant). Thorough sampling did not reveal any epithelial component. Reticulin stain was performed, only highlighted the individual tumor cells. A panel of IHC was performed – CK, SMA, S100, CD 34, BCL2, CD 99, and HMB 45. The tumor cells were diffusely immune-positive for cytoplasmic CD 99 (Figure 2c) and BCL2 while immune-negative for HMB 45 (Figure 2d) and S 100. Based on the histomorphology and IHC findings, a final diagnosis of PRSS was made. Post-operative period was uneventful. After 2 months the patient came for follow up and she was doing well.

Discussion

The PRSS is rare, originally described by Argani, et al., in 2000.[2] Till now around 40 cases have been published in the literature. Synovial sarcoma accounts for 5%–10% of all soft tissue sarcomas.[3] Synovial sarcoma has three distinct morphological variants: monophasic, biphasic, and poorly differentiated.[1] The biphasic variant, with epithelial and spindle cell component, is easier to diagnose. The monophasic variant often causes potential difficulty in differentiating from the other spindle cell sarcomas namely leiomyosarcoma, fibrosarcoma, malignant peripheral nerve

Authors' affiliation: [1]Department of Pathology, Nilratan Sircar Medical College, Kolkata, India.
•**Corresponding author and reprints:** Ankita Majumder, 303 Jodhpur Park, Kolkata 700068, West Bengal, India. Tel: +91-947-643-9911,
E-mail: ptpecha@gmail.com.
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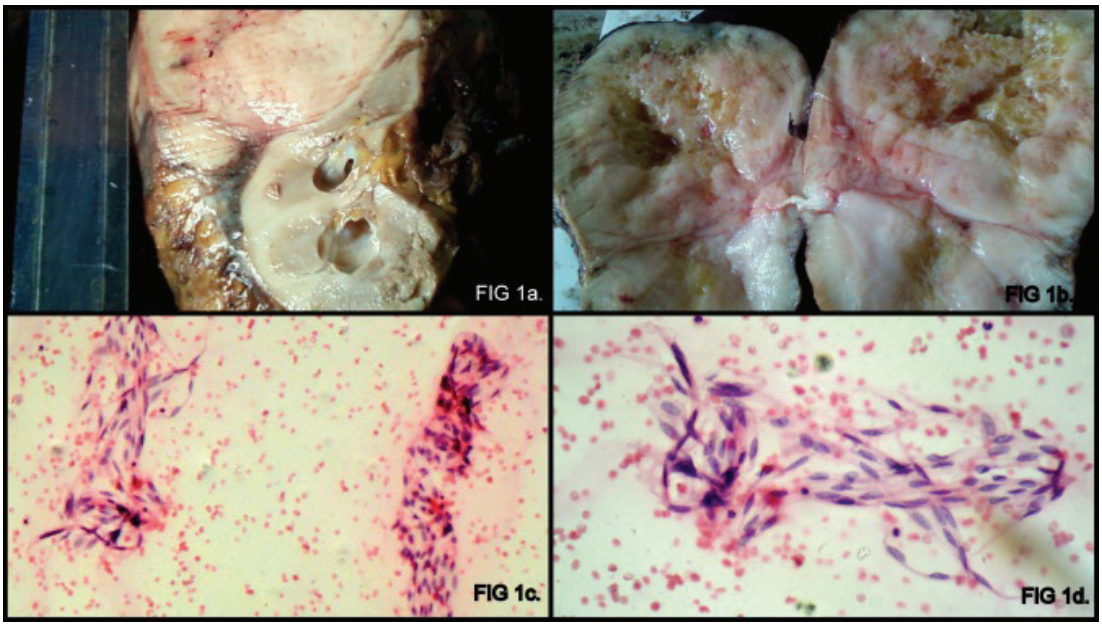

Figure 1. a) This photograph shows a mass originating from the upper pole of kidney. **b)** This photograph shows the cut surface of the renal mass revealing a solid, homogenous, greyish-white appearance with focal myxoid areas. **c)** This photomicrograph (100×) of the FNAC smear (H and E stained) shows loosely cohesive clusters of spindle cells with minimal fat and blood vessels. **d)** This photomicrograph (400×) of the FNAC smear (H and E stained) shows spindle cells with oval hyperchromatic nuclei showing mild to moderate pleomorphism.

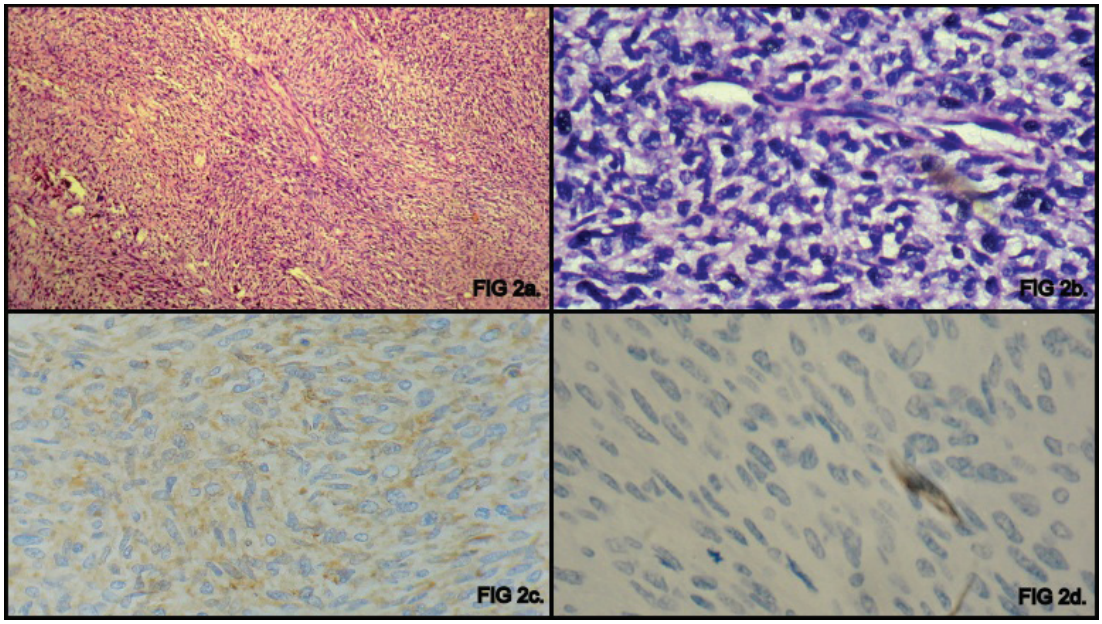

Figure 2. a) This H and E stained photomicrograph (40×) shows spindle shaped cells arranged in intersecting fascicles, interspersed with a few antler-like blood vessels. **b)** This H and E stained photomicrograph (400×) shows individual tumor cells having indistinct cytoplasmic borders, scant to moderate pale eosinophilic cytoplasm, oval to spindle hyperchromatic nuclei with moderate pleomorphism. **c)** This photomicrograph (400×) shows immunostain with CD 99 showing strong and diffuse cytoplasmic positivity. **d)** This photomicrograph (400×) shows negative immunostaining with HMB 45.

sheath tumors, primitive neuroectodermal tumors, solitary fibrous tumors and hemangiopericytoma.[1] As synovial sarcoma comes under the spectrum of spindle cell tumors, morphology alone is not sufficient for a definite diagnosis. In many instances a reliable diagnosis is not possible without ancillary diagnostic techniques such as IHC with/without cytogenetic studies.

Hemangiopericytoma, and solitary fibrous tumor, are both rare in the kidney. Hemangiopericytoma often shows staghorn-shaped vascular arrangement. Synovial sarcoma may occasionally show hemangiopericytoma like areas. However in synovial sarcoma, pleomorphism and mitotic activity is much more compared to hemangiopericytoma and solitary fibrous tumor. Immunohistochemistry can be quite helpful, as hemangiopericytoma and solitary fibrous tumor both express strong and diffuse positivity for CD34 while they are negative for epithelial markers.[4,5]

Malignant peripheral nerve sheath tumors, especially the ad-

enoid variant, histologically mimicks synovial sarcoma. About 70% of malignant peripheral nerve sheath tumors express strong and diffuse cytoplasmic S100,[6] for which synovial sarcoma is usually negative. Although a few malignant peripheral nerve sheath tumors focally express CD56 and EMA, they are generally negative for CK and CD 99, markers of synovial sarcoma.

Mixed epithelial and mesenchymal tumor as well as cystic nephroma both are composed of stroma (like ovarian stroma) and occasional cysts, lined by cuboidal cells. These above mentioned tumors with less cystic areas may be included in the differential for SS. SS with its much higher degree of pleomorphism and mitotic activity, is different from the benign mesenchymal component of mixed epithelial and mesenchymal tumor as well as cystic nephroma. Moreover the ovarian stroma of these tumors express inhibin, ER and PR.[7]

Renal sarcomatoid carcinoma expresses epithelial and mesenchymal markers,[8] therefore it may cause a dilemma in differentiation from Synovial sarcoma, especially the biphasic variant. Renal sarcomatoid carcinoma shows typical carcinomatous areas, which are very different from the small benign looking tubules found in synovial sarcoma. In addition, the sarcomatoid areas of renal sarcomatoid carcinoma are immunonegative for CD99 and CD56.

The management of renal synovial sarcoma comprises of surgical resection with post-opertaive ifosfamide or doxorubicin based chemotherapy.[9] The exact prognosis of PRSS is unclear due to limited number of reported cases.

In conclusion, in spite of its rarity and non-specific presentation, clinicians should consider synovial sarcoma in the differential diagnosis of renal masses composed of spindle cells. Since morphological delineation from other tumors may be tricky, additional diagnostic techniques like immunohistochemistry, cytogenetics, and advanced molecular analyses needs to be employed.

Competing interests

The authors declare that they do not have competing interests.

Acknowledgment

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