Case Report

Primary renal synovial sarcoma: Report of a rare case

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ABSTRACT

Primary renal synovial sarcoma is a relatively recently described uncommon tumor of kidney, with only 34 cases described till date. We report a case of PRSS in a 19-year-old female who presented with a lump in right lumber region, along with brief review of literature.

Key words: Renal neoplasm, sarcoma, synovial

INTRODUCTION

Synovial sarcoma is a clinically and genetically defined morphologic defined entity of uncertain histogenesis.[3] Primary renal cell sarcoma are rare tumors and primary renal synovial sarcomas (PRSSs) even more so. Formerly designated as embryonal sarcoma of kidney, PRSS was first mentioned by[1] and further described as unique entity in August 2003.[2,3] This tumor poses a diagnostic dilemma because it is quite difficult to differentiate it from metastatic sarcoma, sarcomatoid renal cell carcinoma, and hemangiopericytoma which may have similar histological features.[4] The tumor grows very slowly mimicking benign lesions, thus often delaying right diagnosis and treatment. Being sensitive to chemotherapy,[2,5] it is distinct from other sarcomas of kidney. Therefore, it should be a diagnostic consideration, especially in young adults with malignant spindle cell neoplasm of the kidney.

CASE REPORT

A 19-year-old female presented with lump in the right lumber region since 3 months. There was no history of hematuria or pain in abdomen. There was no significant previous medical history. On examination, right kidney was palpable bimanually and ballotable. Biplanar radiography revealed a cyst with excrescences. Ultrasonography abdomen and pelvis showed evidence of a well-defined heterogeneous mass lesion of 3 × 7 cm with solid and cystic components, replacing middle part of right kidney, and therefore a diagnosis of renal cyst was kept. On computed tomography (CT) scan, features suggestive of a well-defined nonenhancing fluid density lesion with soft tissue component were present. Patient subsequently underwent nephrectomy.

Access this article online	
Quick Response Code:	Website: www.ccij-online.org
	DOI: 10.4103/2278-0513.121546

PATHOLOGICAL FINDINGS

Gross

Specimen of kidney of 9 × 7 × 6 cm showing tumor mass in renal pelvis compressing the kidney tissue. On cut-section, there was a tan white solid tumor mass of 4 × 3 cm with focal areas of hemorrhages occupying renal pelvis and pushing the kidney tissue [Figure 1].

Microscopic examination

Showed circumscribed cellular lesion composed of fascicles of plump spindle cells with hyperchromatic elongated nuclei and indistinct cytoplasm. Mitotic figures were 4-5/10HPF. Large areas of necrosis were seen. Compressed normal kidney was seen at the periphery [Figures 2 and 3].

Immunohistochemical studies revealed that the tumor cells were immunoreactive for vimentin, epithelial membrane antigen (EMA) and bcl-2, whereas the tumor cells were

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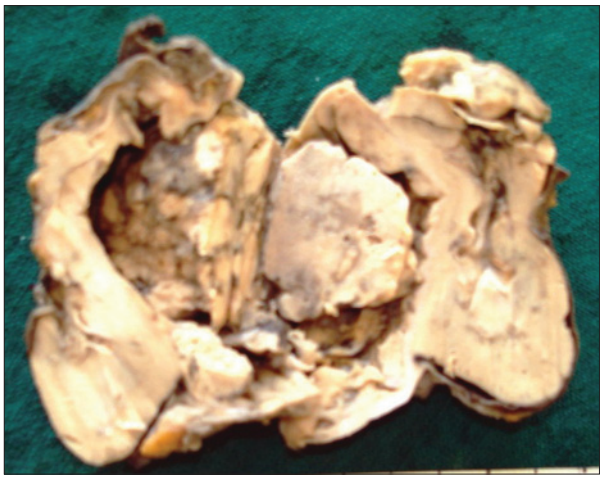

Figure 1: Gross specimen of kidney of 9 × 7 × 6 cm showing tumor mass in renal pelvis compressing the kidney tissue

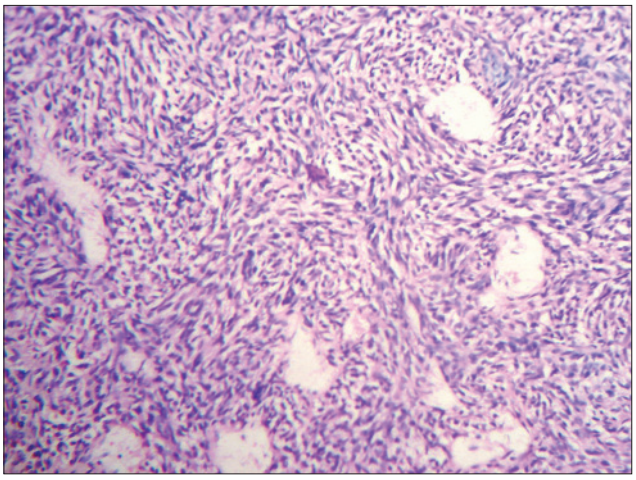

Figure 2: Circumscribed cellular lesion composed of fascicles of plump spindle cells with hyperchromatic elongated nuclei and indistinct cytoplasm

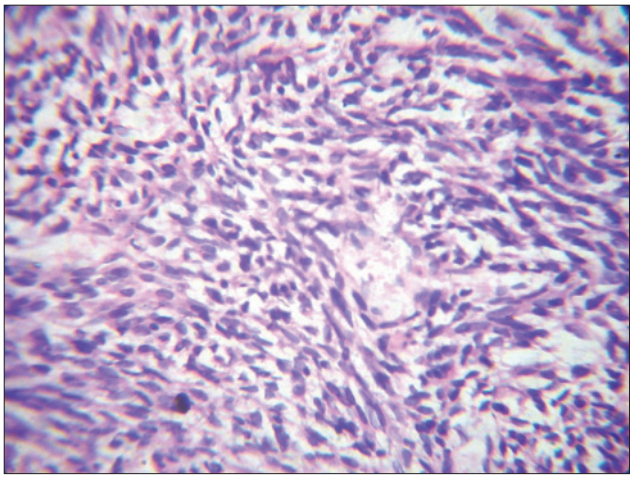

Figure 3: Microscopy picture showing mitotic figures were 4-5/10HPF. Large areas of necrosis were seen. Compressed normal kidney was seen at the periphery

negative for cytokeratin (CK), smooth muscle actin (SMA), desmin, myogenin, HMB-45, and S-100 protein.

This histological appearance and immunoprofile was suggestive of primary synovial sarcoma of kidney.

DISCUSSION

Synovial sarcomas account for approximately 10% of all soft tissue sarcomas.[4] It is clinically and histologically well-defined entity which occurs predominantly in the para-articular, deep soft tissues of the extremities in adolescents and young adults in 25-40 years of age group. But it has also been found at various other sites unrelated to joints/synovial tissues and has been described recently in kidney by Faria in 1999.[1,3]

Grossly, PRSS are usually large, often tan, and rubbery with cystic and necrotic areas.[2] Histologically, PRSS is divided into biphasic, monophasic, and poorly differentiated variants. Biphasic type shows both epithelial and spindle cell components while monophasic type shows only spindle cell component.[3] Monophasic variant of PRSS is more common and has a better prognosis than the biphasic variant. Microscopically mitotically active, monomorphic highly cellular neoplasm composed of plump spindle cells with indistinct cell border growing in short intersecting fascicles is seen.[2] The tumor cells are strongly immunoreactive for vimentin and shows focal positive immunostaining for epithelial markers EMA and CK.[2,3] These are also positive for bcl-2.[5] Cells are nonimmunoreactive for desmin, smooth muscle actin, S-100,[2,3] and CD34.[6] In the present case, necrotic spindle cell sarcoma of the renal pelvis was seen. The tumor cells express vimentin and bcl-2. EMA was focal. There was negativity for CK, SMA, desmin, myogenin, HMB-45, and S-100.

Marker study was done, but molecular analysis by reverse transcriptase polymerase chain reaction (RT-PCR) to detect SYT-SSX gene fusion was not performed in our case. Diagnosis can be confirmed by characteristic translocation t (X; 18) forming SYT-SSX fusion transcripts using reverse transcriptase polymerase chain reaction (RT-PCR) and fluorescent *in situ* hybridization (FISH).[5,7]

These tumors need to be distinguished from sarcomatoid renal cell carcinoma, cellular mesoblastic nephroma, fibrosarcoma, and malignant peripherial nerve sheath tumor (PNET), Sarcomatous transformation of multilocular cystic nephroma, hemangiopericytoma, and other sarcomas involving the kidney.[2,5,8] Immunostaining and cytogenetic analysis are helpful.

Prognosis of primary synovial sarcoma of kidney is uncertain, although relapses clearly occur of the 20 cases reported by in (2002)[7] have involved local or metastatic recurrence and two have resulted in death.[7] Synovial

sarcoma may be sensitive to high dose isophosphamide and adriamycin based regimen.[5,9] Surgical resection and ifosfamide-based chemotherapy are used in the management of renal synovial sarcoma.[10] As the number of cases of synovial sarcoma of the kidney is less due to its extreme rarity, no clear medical guidelines have yet been established.[10]

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Cite this article as: Bhatkule MA, Dhawle MS, Kokandakar HR, Bindu RS. Primary renal synovial sarcoma: Report of a rare case. Clin Cancer Investig J 2013;2:353-5.

Source of Support: Nil, **Conflict of Interest:** None declared.