Case Report

Primary renal synovial sarcoma

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Abstract

Primary synovial sarcoma (SS) of kidney is very rare and difficult to diagnose. Here, we present a case of a 21-year-old female clinically diagnosed as renal cell carcinoma. Right nephrectomy specimen showed a cystic tumor in the upper pole of kidney with areas of hemorrhage and solid growth. Histologically, it showed poorly differentiated cells with hemangiopericytoma-like vascular pattern. Morphologic and immunohistochemical features were compatible with the diagnosis of poorly differentiated SS of kidney. Primary renal SS is a recently described entity. To the best of our knowledge, approximately 34 cases have been reported till date and this is the eighth documented case of poorly differentiated variant. Most of the time, poorly differentiated SS of kidney exhibits hemangiopericytoma like histology. Reverse transcriptase-polymerase chain reaction analysis to demonstrate SYT–SSX fusion gene transcript helps to confirm the diagnosis.

Key Words: Kidney, poorly differentiated, synovial sarcoma

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INTRODUCTION

Synovial sarcoma (SS) is a clinically and genetically defined morphologic entity of uncertain histogenesis. It accounts for 5–10% of adult soft tissue sarcomas. SSs occur predominantly (80%) in para-articular regions of extremities but can involve almost any other site.[1,2] Primary renal SS is a rare tumor, first described by Faria *et al.* in 1999.[2] This tumor poses 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. The tumor grows slowly and insidiously, mimicking benign lesions, thus often delaying right diagnosis and treatment.[3]

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CASE REPORT

A 21-year-old female presented with a history of fever, hematuria and right flank pain since 1 month. Abdominal computerized tomography (CT) revealed a heterogenous, well-marginated soft tissue mass arising in the upper pole of right kidney with solid, necrotic components and heterogenous enhancement. There was no evidence of renal vein, inferior vena cava or right atrial thrombosis. No local invasion or lymphadenopathy was identified [Figure 1a and b]. Radiological and clinical diagnosis was consistent with renal cell carcinoma.

Right nephrectomy specimen weighed 220 g and measured 12×6×3.5 cm. Cut section showed a cystic tumor with soft, solid growth having yellow-brown tan color in the upper pole, measuring 4.5×4.4×4 cm. Areas of hemorrhage and necrosis were seen [Figure 2].

Histopathology revealed neoplasm composed of solid monomorphic sheets of round, plump cells showing high nucleocytoplasmic ratio, clear to eosinophilic cytoplasm with central vesicular nuclei and prominent nucleoli. Stroma showed

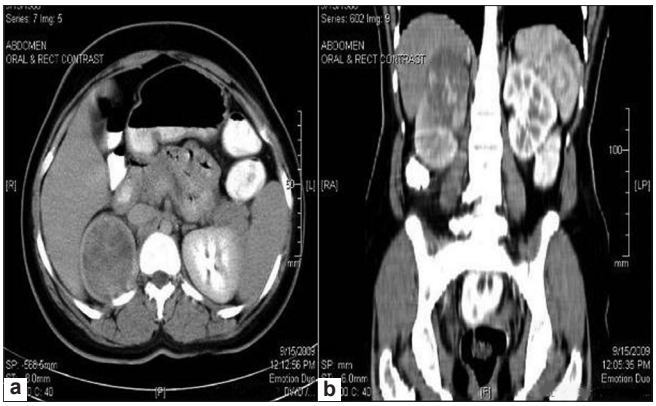

Figure 1: (a and b) Abdominal CT showing well marginated soft tissue mass arising in the upper pole of right kidney

rich vascularity with vessels showing hemangiopericytoma-like pattern. Five mitoses per ten high-power fields were noted. Areas of necrosis and hemorrhage were also observed [Figures 3 and 4]. Adjacent renal parenchyma showed multiple cortical scars and hyalinization. Extension of tumor into renal calyces was seen. Ureter and renal vessel showed no evidence of thrombosis. Overall, the features strongly suggested malignant hemangiopericytoma.

Immunohistochemical study showed strong positivity for Bcl-2, vimentin, CD99/MIC-2, calponin, CD56, focal positivity for epithelial membrane antigen (EMA), cytokeratin (CK) and negativity for muscle specific desmin, actin, CD117, CD34 and CD31 [Figure 5]. Morphologic and immunohistochemical features were compatible with the diagnosis of poorly differentiated SS of kidney.

Postoperatively, the patient is free of local recurrence or metastasis 6 months after surgery.

DISCUSSION

SS is a clinically and morphologically well-defined, but uncommon, entity which is known to occur at unusual sites.[2] Histologically, it is subclassified into biphasic (BSS), monophasic spindle cell (MSSS) and poorly differentiated variants. Amongst these, poorly differentiated synovial sarcoma (PDSS) comprises approximately 20% of cases and shows poorest prognosis. PDSS shows three histologic variants – large cell, small cell and high grade spindle cell variant – and is composed of sheets of undifferentiated round cells with hyperchromatic nuclei and frequent mitoses. Such tumors often have a richly vascular pattern with dilated, thin-walled vascular spaces resembling hemangiopericytoma.[1-3]

Primary renal sarcomas are rare neoplasms that account for 1% of malignant renal tumors. Leiomyosarcoma is the most frequent type comprising 40–60%, followed by rhabdomyosarcoma, chondrosarcoma, osteosarcoma, liposarcoma, angiosarcoma and hemangiopericytoma. In the kidney, 25 cases of MSSS, 2 cases of BSS and 7 cases of PDSS have been reported. To the best of our knowledge, approximately only 34 cases have been reported till date and this is the eighth documented case of poorly differentiated variant.[1,4] It affects young individuals between 20 and 50 years. There is no clinical or imaging characteristic that can indicate the diagnosis. The diagnosis is difficult due to rarity of tumor and its similar presentations as compared to other renal tumors.[4,5] Most of these neoplasms were interpreted radiologically as renal cell carcinoma. Grossly, they varied from case to case but were generally well-defined, irregular large masses ranging from 5 to 20 cm. Extension into inferior vena cava was reported in three cases. Grossly identifiable smooth-walled cysts were identified in eight cases.[1,6,7] The present case also showed a cystic component. A subset of PDSS exhibits a hemangiopericytoma-like morphology and many have been misinterpreted as hemangiopericytoma at diagnosis.[3]

PDSS, primitive neuroectodermal tumor and malignant peripheral nerve sheath tumor may show immunophenotypic overlap. So, these three entities may be differentiated using a panel of antibodies. Immunohistochemical studies of primary renal SS cases have consistently shown positivity for Bcl2, CD99/Mic2, CD56, Vimentin and focal positivity for EMA. They do not stain for desmin, actin, CD34, CD31.[1-10] Although EMA is not a specific marker of SS, its presence in poorly differentiated sarcoma can suggest SS. The simultaneous use of antibodies to both CD99 and CD56 may be useful in the differential diagnosis of PDSS because SS will almost always be positive with both, whereas PNETs will be CD99 positive and CD56 negative and MPNSTs will be CD99 negative but CD56 positive.[10]

Reverse trancriptase-polymerase chain reaction (RT-PCR) analysis of SS shows a unique chromosomal translocation t(X;18)(p11.2;q11.2) that results in the fusion of *SYT* gene on chromosome 18 with *SSX* family gene on chromosome X and helps to confirm the diagnosis.[1-12]

Differential diagnosis includes Wilms' tumor, mixed epithelial–stromal tumor, sarcomatoid renal cell carcinoma, congenital mesoblastic nephroma, PNET, MPNST and hemangiopericytoma. To arrive at a diagnosis of renal SS, the possibilities of distant metastasis and secondary extension of retroperitoneal SS must be ruled out. Hemangiopericytoma differs from renal SS by its CD34 positivity and CK negativity.[1-3,12]

Although primary surgical resection is the treatment of choice for SS, the prognosis is poor with this treatment alone. SS may

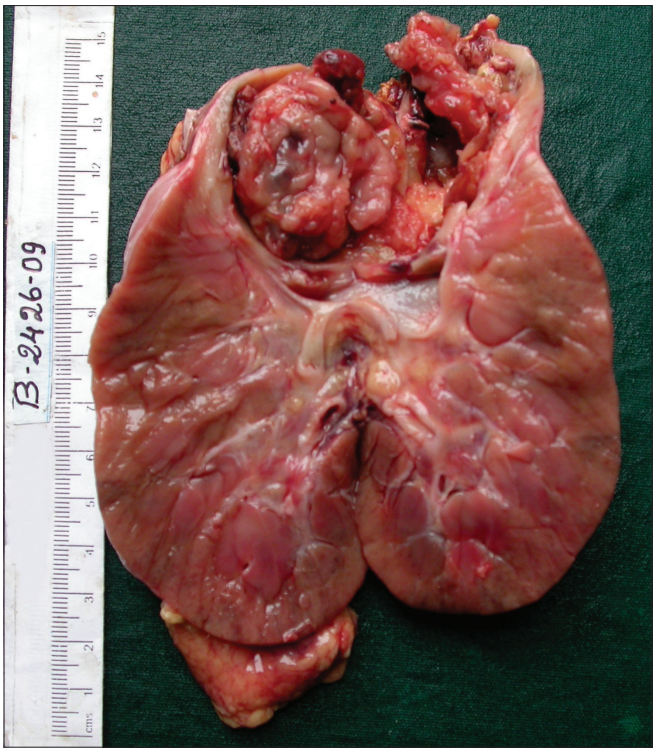

Figure 2: Grossly, yellow-brown tan mass in the upper pole in well-defined cystic space

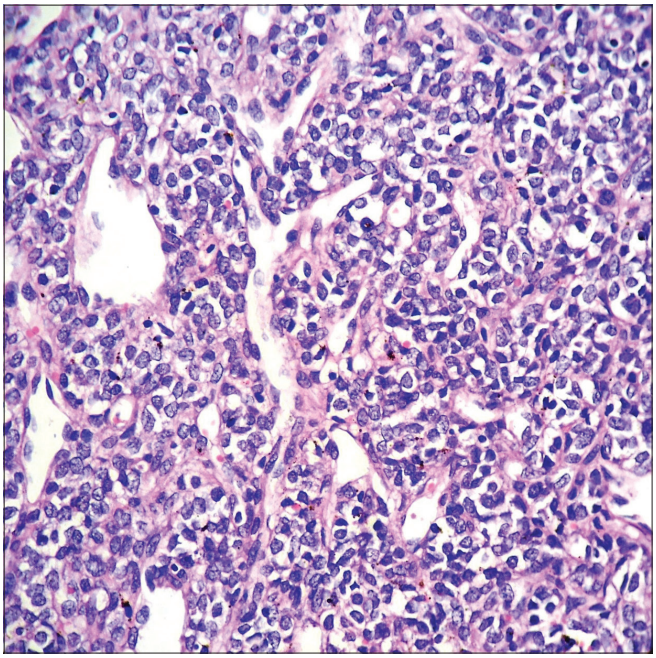

Figure 4: Tumor showing hemangiopericytoma-like vascular pattern (H and E, ×40)

be sensitive to high dose isophosphamide and adriamycin based regimen.[4,5] As the number of cases of SS of the kidney is less due to its extreme rarity, no clear medical guidelines have yet been established.[11]

PDSS constitutes, by definition, a problem at diagnosis because

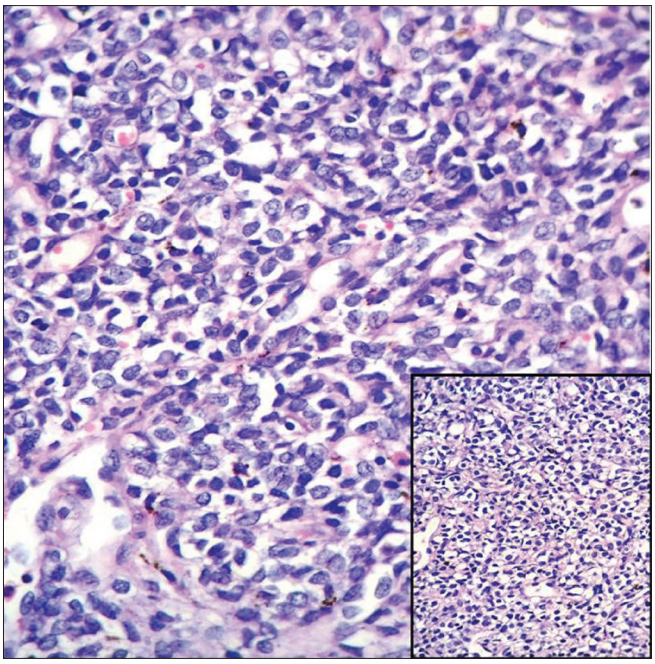

Figure 3: The tumor shows solid sheets of plump spindle cells with high N:C ratio, pleomorphism and frequent mitoses. The inset shows the clear cell change in tumor cells (H and E, ×40)

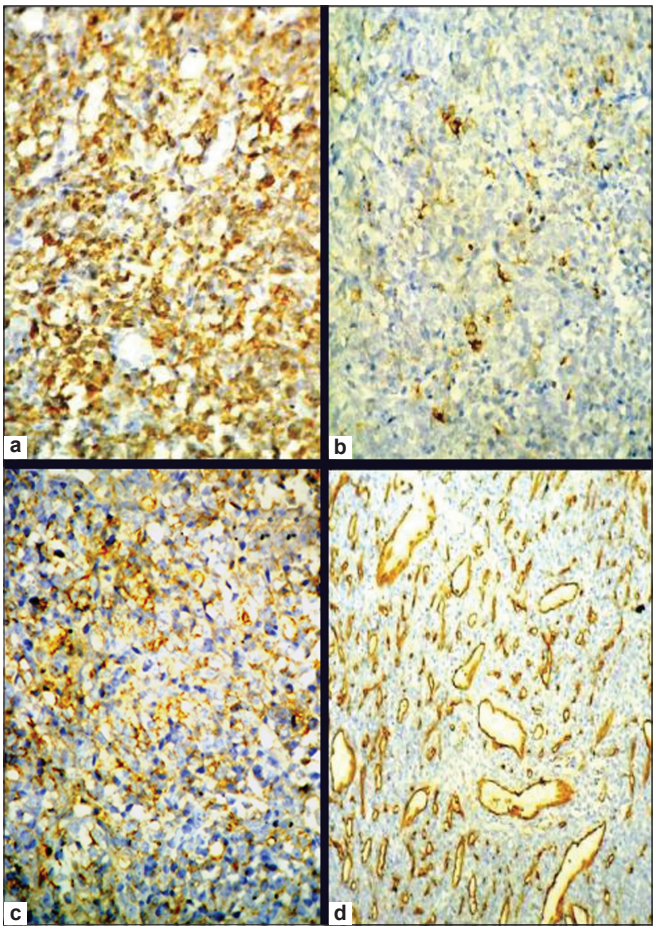

Figure 5: Immunohistochemical study of tumor cells showing (a) diffuse positivity for BCl-2; (b) focal positivity for EMA; (c) strong positivity for CD99; and (d) negativity for CD34

of its unclear and variable histomorphological appearance. However, correct diagnosis is crucial as these tumors are associated with more aggressive behavior and metastasis and thus, with less favorable prognosis.[3]

Hence, SS should be included in differential diagnosis while dealing with renal sarcomas.

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