

Biphasic renal synovial sarcoma with extensive venous tumor thrombosis: A rare presentation

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Abstract

Primary renal synovial sarcoma (SS) is a very rare soft-tissue tumor arising from the kidney. These tumors histologically closely resemble other sarcoma variants. Immunohistochemical and cytogenetic techniques remain the cornerstone in achieving the correct diagnosis. Radical surgical excision is the primary treatment for local tumor control and alleviation of symptoms. Adjuvant chemotherapy, at present, has only a limited role. We report a unique case of a 21-year-old young male with the diagnosis of a biphasic renal SS with extensive venous system thrombosis involving the inferior vena cava, bilateral iliac, and femoral vessels which was managed by radical nephrectomy and tumor thrombectomy followed by adjuvant chemotherapy.

Keywords: Cytogenetics, immunohistochemistry, renal synovial sarcoma, venous tumor thrombosis

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INTRODUCTION

Synovial sarcoma (SS) is a clinically and morphologically distinct tumor entity predominantly arising from the extremities and rarely can involve other organ systems.^[1] Faria *et al.* reported the first case of SS of the kidney in 1999.^[2] Till date, approximately fewer than 70 cases of SS have been reported in literature.^[3] These tumors are characterized by unique chromosomal translocation t (X; 18) (p11.2;q11.2) in most of the cases, that results in the fusion of SYT gene on chromosome 18 with an SSX family gene on chromosome X (SSX1, SSX2, or SSX4).^[4] These tumors share similar histologic and immunohistochemical (IHC) characteristics with sarcomatoid renal cell carcinoma, adult Wilms' tumor, and congenital mesoblastic nephroma leading to a diagnostic dilemma.^[5] We report an atypical case of biphasic renal SS with extensive tumor thrombus involving the renal vein,

inferior vena cava (IVC), and bilateral iliac and femoral veins.

CASE REPORT

A 21-year-old young man presented to our outpatient clinic with complaints of dull aching right flank pain and low-grade fever of 1-week duration. Clinical examination revealed a nonpitting pedal edema over the left shin and nonreducing left varicocele. His blood counts and renal function tests were normal. Ultrasonography revealed a 6 cm × 6 cm hyperechoic mass with anechoic central gross internal debris involving lower pole of the right kidney. Contrast-enhanced computed tomography showed an enhancing right lower pole renal mass of 6.5 cm × 6.4 cm with enhancing tumor thrombus extending superiorly to suprahepatic part of IVC and inferiorly involving bilateral

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iliac and femoral veins [Figure 1]. The metastatic workup such as chest radiography and liver function tests found to be normal. The patient was clinically staged as T_{3c}N₀M₀ and underwent open right radical nephrectomy with IVC and bilateral femoral vein thrombectomy. The postoperative period was uneventful.

Histopathology revealed a renal neoplasm demonstrating spindle cells with marked nuclear atypia [Figure 2]. Immunohistochemistry showed positivity for cytokeratin, CD99, S100, and Bcl-2 indicating features of biphasic SS. The patient was offered adjuvant chemotherapy with adriamycin and ifosfamide.

DISCUSSION

Primary SS of the kidney is a very rare neoplasm and constitutes <1% of all malignant kidney tumors.^[1] It is subclassified into monophasic, biphasic, and poorly differentiated variants.^[6] The primary biphasic SS contains both glandular elements and spindle epithelial cells. The monophasic variant is composed of only spindle cells which makes it difficult to diagnose on histopathology and IHC alone and requires further confirmation by cytogenetic analysis.^[4] Among the three histologic variants, the poorly differentiated sarcoma has the worst prognosis.^[6] Renal SS are typically positive on IHC for Bcl-2, CD99, CD56, vimentin and focally for epithelial membrane antigen. Making an accurate diagnosis on histology is difficult on most occasions, since renal SS is usually difficult to differentiate from other soft-tissue sarcomas. The current gold standard for renal SS is demonstration of the t (X; 18) (p11.2, q11.2) translocation using reverse transcriptase polymerase chain reaction, involving

fusion of the SYT (Synonyms: SS18-SS translocation, chromosome 18) gene on chromosome 18 to either the SXX1 (SS, X breakpoint 1) or the SXX2 (SS, X breakpoint 2) gene on chromosome Xp11.^[7]

SS affects primarily young adults with a mean age of 37 years with no sex predilection.^[8] The common presenting symptoms include flank pain, hematuria, and rarely palpable mass, fever and hypertension.^[3] Although there are no specific imaging characteristics, Lalwani *et al.* have described “triple sign” on T2-weighted magnetic resonance imaging representing areas of hemorrhage, calcification, and air-fluid levels which can raise a suspicion for a preoperative diagnosis of these tumors.^[9] Renal SS can present rarely at an advanced stage with caval thrombus and/or metastasis.^[10] To the best of our knowledge, approximately five cases of SS with the caval thrombus have been reported previously [Table 1].

Our case is unique in that there was an extensive renal, IVC, iliac, and femoral vein tumor thrombus with a biphasic SS histology.

Table 1: Summary of tumor thrombus extension in patients with Synovial Sarcoma reported in literature

Authors	Age of patient (years)	Extent of tumor thrombus
Dassi <i>et al.</i> , 2009 ^[10]	20	Renal vein, infrahepatic IVC
Chen <i>et al.</i> , 2003 ^[11]	19	Renal vein, suprahepatic IVC, right atrium
Trolliet <i>et al.</i> , 2014 ^[12]	66	Renal vein, infrahepatic IVC
Chandrasekaran <i>et al.</i> , 2016 ^[13]	44	Renal vein, infrahepatic IVC
Modi <i>et al.</i> , 2014 ^[14]	41	Renal vein, infrahepatic IVC
Our patient	21	Renal vein, suprahepatic IVC, bilateral iliac, and femoral veins

IVC: Inferior vena cava

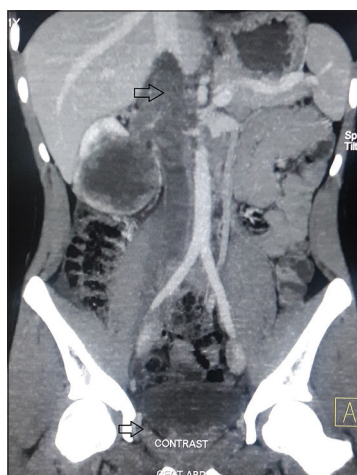


Figure 1: Contrast-enhanced computed tomography coronal section showing right kidney tumor with thrombus in IVC and bilateral femoral veins

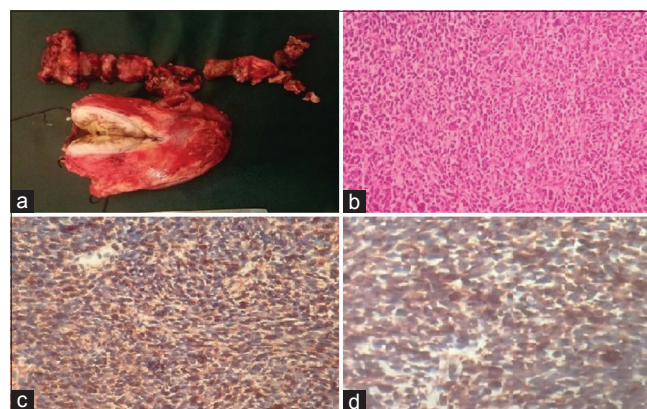


Figure 2: (a) Gross specimen of right renal tumor with thrombus. (b) tumor comprised of intersecting fascicles of tumor cells (H and E, x40). (c) Tumor cells showing strong positivity to CD99 (immunohistochemical, x40). (d) Tumor cells showing positivity to cytokeratin (immunohistochemical, x40)

Radical nephrectomy is primarily adopted for achieving local disease control as well as for alleviation of symptoms. About one-third of these patients can have local relapse or develop metastatic abdominal lymph nodes.^[3] The role of adjuvant chemotherapy is controversial, although few authors have reported favorable response in patients with metastatic disease.^[15] The most frequently used chemotherapy regime is a combination of anthracyclines and ifosfamide similar to protocols used in the treatment of other sarcomas. The median overall survival is approximately 48 months only.

Primary renal SS are rare neoplasms. Although diagnosis can be achieved by IHC it may also require molecular and cytogenetics techniques to establish an accurate diagnosis. These neoplasms should be considered in the differential diagnosis of renal masses with spindle cell components. Radical surgery is the best treatment modality available at present as these neoplasms mainly presents in younger population and chemotherapy has only a limited role. The overall prognosis of this tumor is extremely poor in spite of the presently available treatment modalities.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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