Case Report

Primary synovial sarcoma of kidney: A rare tumor with an atypical presentation

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ABSTRACT

Synovial sarcoma (SS) is a tumor of the soft tissues with a unique chromosomal translocation t(X;18)(p11.2;q11.2) that can be detected by polymerase chain reaction in tissue homogenates. Here we present a case of a 20-year-old female presenting PSS of the left kidney with caval thrombus. The diagnosis was corroborated by reverse transcription polymerase chain reaction (RT-PCR). Similar cases of PSS of kidney with tumor extension in the inferior vena cava are extremely rare and to date, approximately three cases have been reported in the literature.

Key words: Renal cell carcinoma, synovial sarcoma, chromosomal translocation

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INTRODUCTION

Synovial sarcoma (SS) occurs primarily in the soft tissues, mostly in para-articular regions of the extremities. These tumors have been described in other unusual locations,[1] including the pleura, lung, mediastinum, and kidney. They are specifically associated with a unique chromosomal translocation (t(X;18)(p11.2;q11.2) that results in the fusion of SYT gene on chromosome 18 with an SSX family gene on chromosome X(SSX1,SSX2, or SSX4).

Primary synovial sarcoma (PSS) of the kidney is a recently described entity; to date, a total of approximately 33 cases have been reported.[2] This tumor presents a diagnostic dilemma because it is quite difficult to differentiate it from other renal neoplasms, such as metastatic sarcoma, renal cell carcinoma with sarcomatoid differentiation, which may have similar histological features.

CASE REPORT

A 20-year-old female was presented with a mild left flank pain of one-week duration, with no associated history of hematuria or any other systemic symptoms. Physical examination revealed a large nontender lump involving left lumbar and left hypochondriac region.

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Computed tomography of the abdomen and pelvis showed a large heterogeneously enhancing mass of 14.3 × 9.4 × 8.5 cm over the mid-region and lower pole of the left kidney with areas of necrosis within it. An iso-to-hypodense heterogeneously enhancing thrombus was noted in the left renal vein and adjacent portion of the inferior vena cava [Figures 1 and 2].

Renal cell carcinoma was suspected pre-operatively. Intraoperatively, a tumour mass was seen replacing the whole kidney. Left radical nephrectomy was performed and left renal vein ligated flush with the IVC after milking the thrombus into the left renal vein. Macroscopic examination of the specimen revealed a tumor of size 12.8 × 11 × 4.5 cm, replacing the entire renal parenchyma, involving pelvi-calyceal system and medulla with thin rim of cortex seen

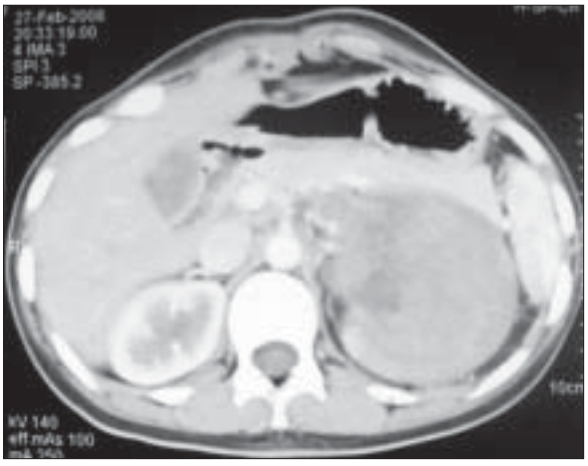

Figure 1: CT Image showing tumor invading almost whole of kidney and involving IVC

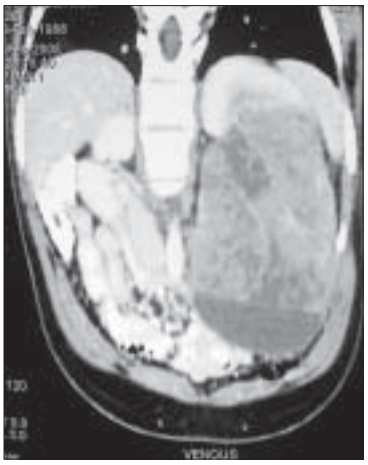

Figure 2: CT Image (coronal)

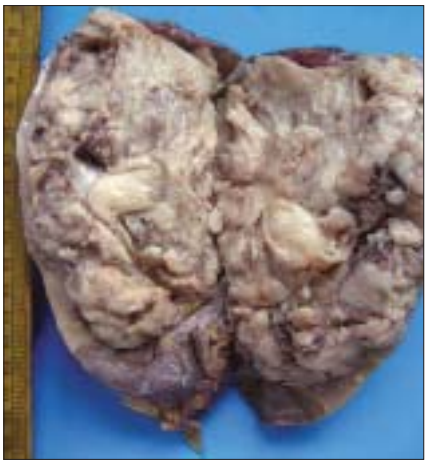

Figure 3: Macroscopically, tumour seen replacing whole of kidney with tumour thrombus in left renal vein

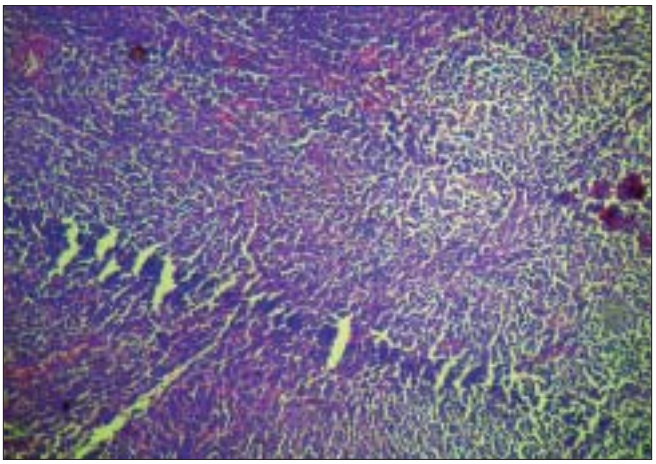

Figure 4: Tumour composed of spindle cells arranged in intersecting fascicles alternating with hypocellular areas

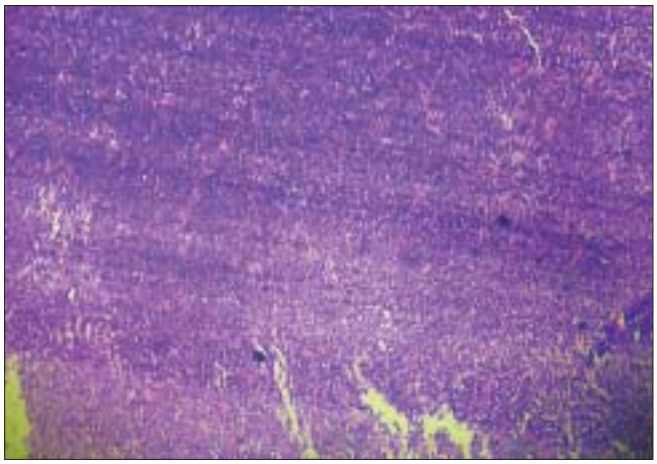

Figure 5: Microscopic image showing spindle cells

all around. Tumor thrombus was seen within the lumen of left renal vein [Figure 3].

Microscopically, the tumor was composed of spindle cells arranged in the intersecting fascicles, alternating with hypocellular areas, suggestive of monophasic SS [Figures 4 and 5]. On immunohistochemistry, tumor cells expressed bcl-2, calponin, and EMA. Both Mic-2 and CK were focally positive. Molecular analysis revealed a translocation between the SYT gene on chromosome 18 and SSX on chromosome X, which is consistent with the diagnosis of SS.

DISCUSSION

Primary synovial cell sarcoma of kidney is a rare entity. It was first reported in 1999 by Faria *et al*.[3] To the best of our knowledge, approximately 33 cases have been reported till date. Synovial sarcoma usually involves adolescents and young adults; however, the age at presentation ranges from 17 to 61 years. Diagnosis is difficult due to the rarity of the tumor and its similar presentations as compared to other renal tumors. Differential diagnosis includes Adult Wilms tumor, transitional cell carcinoma, renal cell carcinoma and hemangiopericytoma, congenital mesoblastic nephroma, and primitive neuroectodermal tumor.

Primary synovial sarcoma occurs in two forms: Biphasic and Monophasic. The primary biphasic synovial sarcoma contains both glandular elements and spindle epithelial cells. The primary monophasic synovial sarcoma is composed of only spindle cells. The monophasic variant is particularly difficult to diagnose only on histopathological examination and requires confirmation by molecular analysis. The characteristic chromosomal translocation seen is t(x;18) (p11.2;q11.2). This translocation results in fusion of SYT gene located on chromosome 18 with SSX gene located on chromosome X. Five variants of SSX gene have been identified; however, only SSX1 and 2 have been shown to fuse with SYT gene.[4]

The patients commonly present with flank pain and/or hematuria. No clinical feature or imaging modality is diagnostic. The CT scan usually reveals a heterogeneously enhancing renal mass and the confirmation of diagnosis is by molecular and cytogenetic analysis. Rarely is the presentation at an advanced stage with caval thrombus and/or metastasis. To the best of our knowledge, approximately three cases of SS with the caval thrombus have been reported previously. In 2007, Tornkvist *et al*.,[5] reported six cases of metastatic disease.

There are no established guidelines regarding management of this tumor given the limited number of cases reported. Primary surgical treatment is considered to be the treatment of choice; prognosis is poor with this treatment alone. The value of chemotherapy is yet to be proven but SS may be sensitive to high doses of Ifosamide-based regimens. Park *et al*.,[6] has reported complete remission of metastatic lung lesion using Ifosamide and doxorubicin during the four-week course.

In conclusion, we have described a case of primary SS of the kidney with caval thrombus in a young female. This rare tumor is likely to be confused with other spindle cell tumors of the kidney. An accurate diagnosis, including cytogenetic and molecular studies is imperative. Primary SS should be included in differential diagnosis when dealing with the spindle cell tumors of kidney.

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