

Oncology

Primary Renal Synovial Sarcoma: An Oncologic Surprise[☆]

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ABSTRACT

Primary renal synovial sarcoma is a rare tumor having a specific chromosomal translocation t(X; 18) (p11.2; q11.2). The clinical features of this tumor and radiologic appearances are quite similar to those of renal cell carcinoma. Confirmatory diagnosis requires fluorescent in situ hybridization or reverse transcriptase polymerase chain reaction validation for differentiating the tumors from sarcomatoid renal cell carcinoma. We present a case of primary renal synovial sarcoma that was diagnosed in a middle-aged man.

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Introduction

Synovial Sarcoma is the fourth most common type of soft tissue sarcoma primarily affecting the extremities in young adults.¹ However, primary renal synovial sarcomas very rarely occur in the kidney, with only <50 cases reported in literature.² We present a case of primary renal synovial sarcoma in a middle-aged man treated by us.

Case presentation

A 46-year-old man, known to be diabetic and on oral hypoglycemic agents since the last 5 years, presented to us with dull aching left flank pain of few weeks duration. Clinically, there was a palpable lump in the left hypochondrium and lumbar region. His renal function test reports were normal. Ultrasonography scan of the abdomen showed mixed echogenic left renal mass of size 12 × 11 cm suspicious of renal cell carcinoma. Computed tomography scan of the abdomen showed a heterogeneously enhancing mass lesion of size 12.8 × 11.7 cm (Fig. 1). The patient underwent left radical nephrectomy.

Histopathology (Fig. 2) revealed renal tissue to be infiltrated by a tumor composed of monomorphic spindle shaped cells with

indistinct cell borders and scant cytoplasm. Cells were arranged in short intersecting fascicles and sheets. Tumor showed high mitotic activity with cystic structures lined by polygonal epithelial cells with hobnailing of nuclei and occasional mitotic figures in the lining epithelial cells.

On immunohistochemistry analysis, the glandular component showed positive reactivity for pancytokeratin and epithelial membrane antigen, whereas the spindle cell component was reactive for Bcl2. Tumor cells were nonreactive for CD34. This overall profile was suggestive of synovial sarcoma.

To confirm the diagnosis of synovial sarcoma, FISH (fluorescent in situ hybridisation) and RT-PCR (reverse transcriptase polymerase chain reaction) were done. FISH was positive for SYT (18q11.2) rearrangement (Fig. 3) and RT-PCR was positive for SYT-SSX2 translocation.

Discussion

Primary renal synovial sarcoma was first described in 1999 by Faria et al.³ Histologically, renal synovial sarcoma is classified as biphasic synovial sarcoma, monophasic spindle synovial sarcoma, and monophasic epithelial synovial sarcoma.

It is difficult to differentiate this tumor from sarcomatoid renal cell carcinoma, adult Wilms tumor, and congenital mesoblastic nephroma because of histologic similarities and nonspecific immunohistochemistry markers.⁴ Metastatic sarcoma and invasion from retroperitoneal sarcoma also should be ruled out.^{5,6} This entity lacks any specific imaging or clinical findings. However, t(X; 18)

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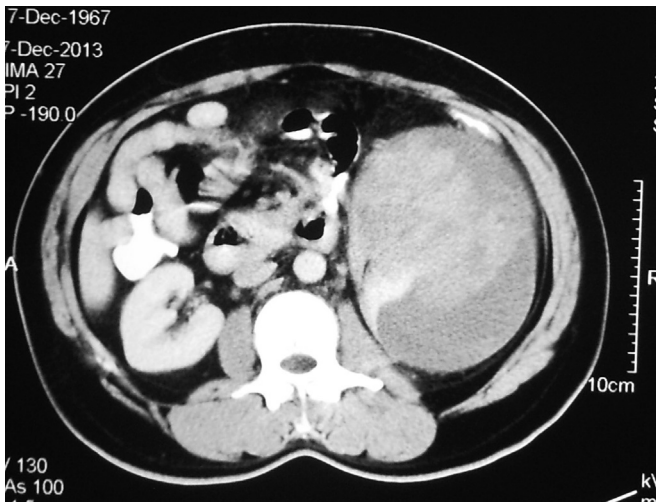


Figure 1. Computed tomographic appearance of the renal tumor.

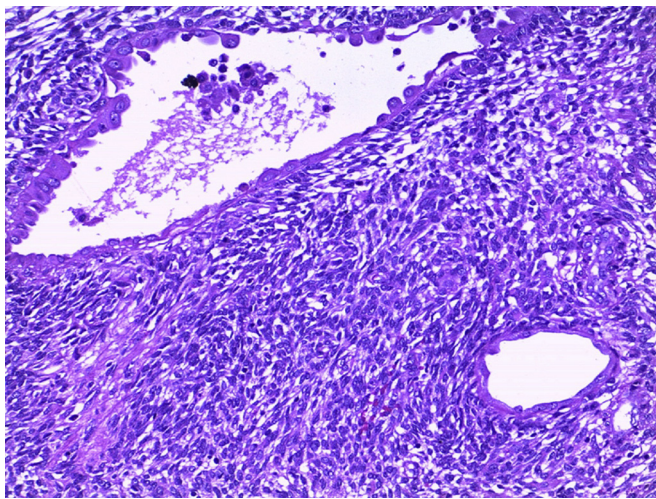


Figure 2. Histopathology of the renal tumor.

(p11.2; q11.2) translocation is characteristic of synovial sarcoma, and methods to detect this translocation include cytogenetic analysis, RT-PCR, and FISH.⁷ Three types of fusion defects, mainly *SYT-SSX1*, *SYT-SSX2*, and rarely *SYT-SSX4* are demonstrated by renal synovial sarcoma.⁸ These findings were also observed in the tumor studied in our patient.

Surgical resection is the mainstay, although surgery alone has poor prognosis. Reports of sensitivity of this tumor to ifosfamide- and doxorubicin-based chemotherapy do exist,⁹ but no clear

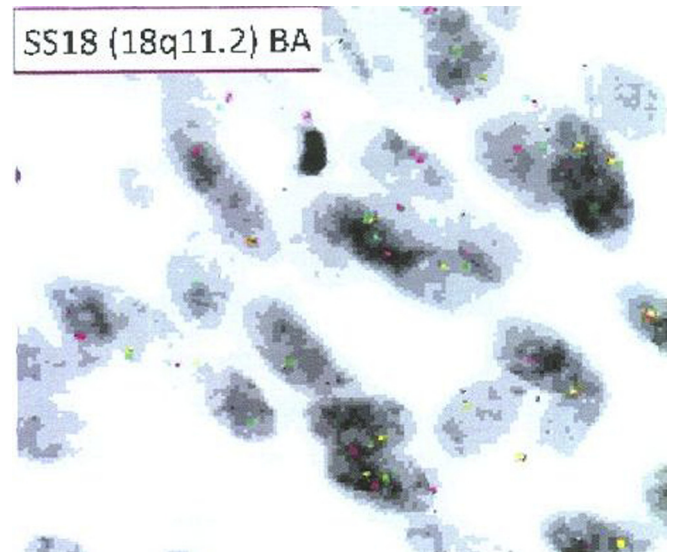


Figure 3. Fluorescent in situ hybridization of the renal tumor.

guidelines are available regarding adequate treatment of this rare entity.

Conclusion

Primary Renal Synovial Sarcoma is a very rare tumour and correct diagnosis requires specific pathological tests.

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