

# Monophasic Primary Renal Synovial Sarcoma Accompanied With a Hemorrhagic Cyst

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Keywords: *synovial sarcoma, kidney neoplasms, cysts, diagnosis*

*Urol J. 2011;8:244-7. www.uj.unrc.ir*

## INTRODUCTION

Synovial sarcoma is a soft tissue sarcoma with uncertain histogenesis, which is most prevalent in young adolescents. Primary synovial sarcoma rarely originates from the renal parenchyma. When this entity occurs, origin of this unusual tumor type has been the subject of discussion in the literature, with a suggestion that some previously reported cases might be more correctly described as renal cell carcinoma with sarcomatoid dedifferentiation. Renal synovial sarcoma (RSS) and sarcomatoid renal cell carcinoma may be hard to be distinguished only on histopathologic and immunohistochemical examination, but these tumors contain distinctly different sets of chromosomal abnormalities.<sup>(1)</sup>

Renal synovial sarcoma produces three types of fusion gene formed in part by SS18 (SYT) from chromosome 18 and by SSX1, SSX2 or, rarely, SSX4 from the X chromosome.<sup>(1)</sup> The SYT-SSX fusions do not seem to occur in other tumor types. Of 45 cases reported as RSS previously, only 29 are available, whose diagnosis was validated by fluorescence in situ hybridization (FISH) or reverse transcription polymerase chain

reaction, which demonstrated SYT-SSX translocation, a characteristic chromosomal abnormality for synovial sarcoma. Hence, proper molecular analysis, in addition to conventional immunohistochemical analysis, should be undertaken to establish a proper diagnosis.<sup>(2)</sup> We present a case of primary RSS validated by FISH analysis.

## CASE REPORT

A 63-year-old woman presented with dysuria and gross hematuria. She had been diagnosed and treated for a presumed hypertension. Laboratory study was within normal limit.

Cystoscopy revealed bleeding from the right ureter. Ultrasonography showed a complex renal mass of 5 cm in the greatest dimension. Axial contrast-enhanced computed tomography scan demonstrated a 5-cm complex cystic mass that contained mildly thickened septation, along with a slightly contrast-enhancing region (Figure 1). No calcification or tumor capsule was observed. On magnetic resonance imaging, the solid components of the tumor showed slightly high signal intensity as the renal medulla on T1-weighted and slightly low signal intensity in T2-weighted

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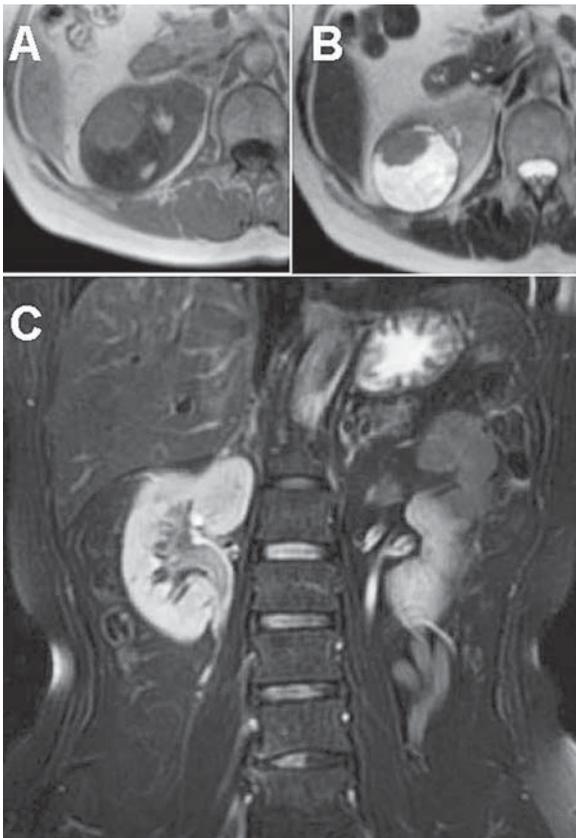
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images (Figure 2). On coronal images using T1-weighted fat-saturated technique, the tumor was present on the upper pole of the kidney with pre-operative diagnosis of renal cell carcinoma, stage cT1cN0cM0, the patient underwent a radical



**Figure 1.** Computed tomography scan showing a right renal tumor (2.5 × 1.5 cm) with multilocular hemorrhagic cysts. Enhanced solid component is seen between the cysts on contrast-enhancement.

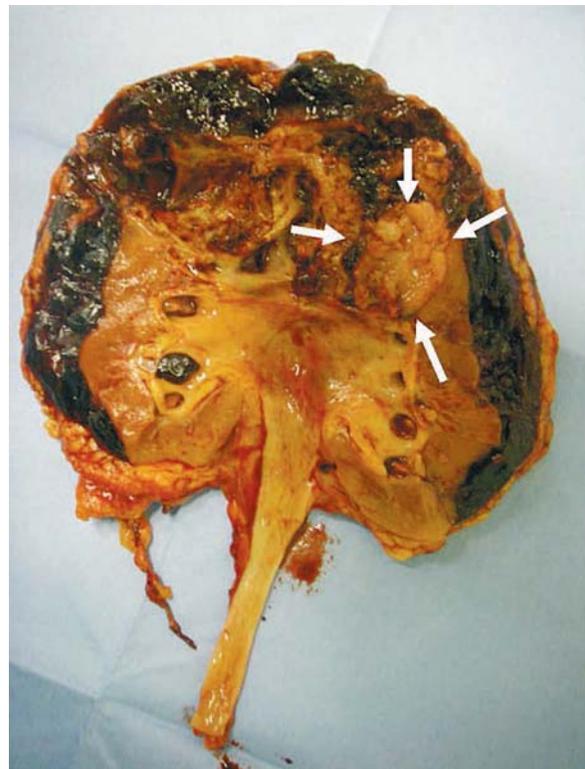


**Figure 2.** On magnetic resonance imaging, cysts in the tumor are shown as low intensity,

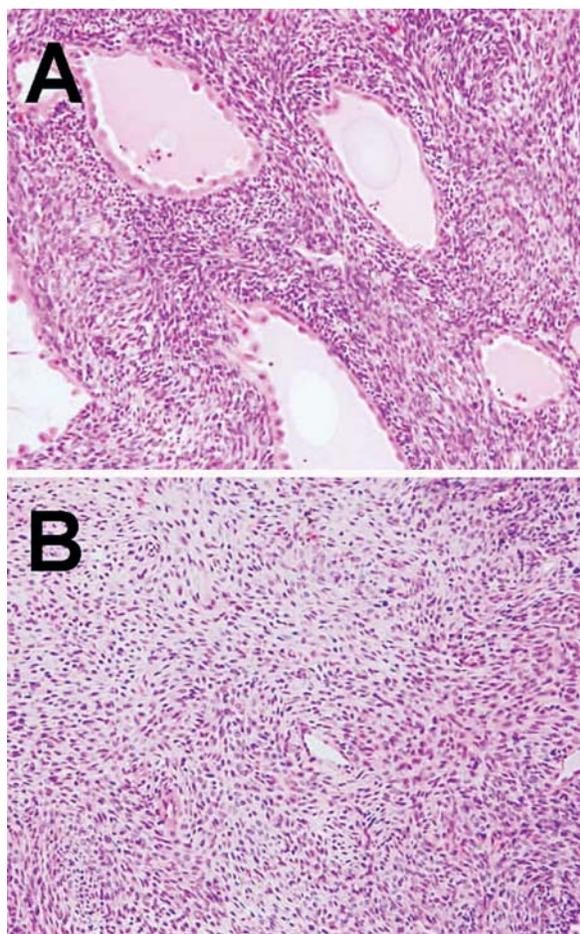
nephrectomy. Macroscopically, the lateral side of the upper pole of the kidney has been replaced by a multiloculated cystic mass accompanied with a solid component. A massive hematoma, formed by intra-operative bleeding, was seen between the kidney and extra-renal connective tissue (Figure 3).

On histological examination, spindle cells with dark-staining nuclei and indistinct cytoplasm proliferated densely in the solid component. Spindle cells were arranged in bundle, fascicular, or storiform pattern (Figure 4). Immunohistochemical studies revealed that vimentin was positive for both spindle cells of the solid component and epithelial cells lining cysts. CD99, CD56, BCL2, and focal cytoplasmic staining for c-Kit were positive in spindle cells, but negative in epithelial cells. Pancytokeratin, cytokeratin-7, CD10, and beta-2 microglobulin were positive in epithelial cells, but negative in spindle cells.

Given these findings, the cysts were thought



**Figure 3.** Formalin-fixed kidney demonstrating an ill-defined, pale, 2-cm medullary mass with central necrosis (arrow).



**Figure 4.** Results of some fluorescence in situ hybridization experiments: SSX1 in red, SSX2 in violet. **(A)** Distribution of signals in a tetraploid nucleus of the present case carrying a fusion gene. **(B)** Control distribution pattern shown in merged image in yellow.

to be entrapped dilated renal tubules, but not an epithelial component of biphasic synovial sarcoma; hence, monophasic synovial sarcoma was suspected. Result of the FISH analysis using a break-apart style probe was consistent with synovial sarcoma (Figure 4). After 1-year follow-up period, this patient is still free of recurrence.

## DISCUSSION

Primary RSS is a rare tumor first described in 1999 and further elaborated upon by two separate studies in 2000.<sup>(3)</sup> This rare tumor is distinct from other more common forms of sarcoma originating in the kidney. It is difficult for a pathologist to differentiate between synovial sarcoma and congenital mesoblastic nephroma, adults Wilms

tumor, or clear cell sarcoma of the kidney due to similarities in histological appearance and the absence of specific immunohistochemical markers.<sup>(4)</sup> No clinical or imaging characteristics can indicate the diagnosis. The clinical symptoms most frequently observed, including abdominal pain and hematuria, do not differ from those present in other malignant renal tumors.

The main histologic subtypes of RSS are biphasic synovial sarcoma, monophasic spindle synovial sarcoma, and monophasic epithelial synovial sarcoma.<sup>(5)</sup> When a tumor with epithelial and stromal components is diagnosed as synovial sarcoma in the kidney, the characteristic of epithelial cells should be determined. If the epithelial cells are entrapped renal tubules, the tumor is defined as monophasic synovial sarcoma.<sup>(5)</sup> If the cells are neoplastic, it is defined as biphasic synovial sarcoma. In the present patient, the epithelial cells are identified as entrapped renal tubules by the following results. Epithelial cells indicated positive immunostaining for CD10, beta-2 microglobulin, and antibodies to renal tubules, and negative for N-CAM and BCL-2, antibodies often positive in synovial sarcomas.<sup>(4,5)</sup> Therefore, the diagnosis of monophasic synovial sarcoma was established. Furthermore, a morphologic transition from epithelial cells to spindle cells, often seen in biphasic synovial sarcomas, was not observed in the present subject.

The translocation  $t(X;18)(p11.2;q11.2)$  is specific for synovial sarcoma regardless of location or type and grade of differentiation.<sup>(5,6)</sup> The translocation results in the fusion of the 5' part of the SS18 gene and the 3' part of SSX1, SSX2, SSX4 gene, or rarely the splice variant SSX4v.<sup>(5,6)</sup>

Polymerase chain reaction testing has greatly aided in confirming the diagnosis of RSS by detecting the SYT-SSX fusion gene that results from the translocation of the SYT gene on chromosome 18 with the SSX gene on the X chromosome.<sup>(5,6)</sup> A variety of diagnostic methods exist to detect translocation, including cytogenetic analysis, reverse transcription polymerase chain reaction, and FISH. Various FISH methods have been described to allow the visualization of structural chromosomal abnormalities in archival

tissue. Dual-color break-apart probe FISH assays employ probes that flank the translocation breakpoint and are separated by the translocation event, and have been optimized for use on paraffin-embedded tissues.

Complementary to conventional histopathologic and immunohistochemical analysis, in characterizing the status of the SSX and SYT genes, FISH study is important in the differential diagnosis when dealing with a renal tumor with epithelial and stromal components.

#### CONFLICT OF INTEREST

None declared.

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