

# Primary Renal Synovial Sarcoma

## A Case Report

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### ABSTRACT

*Renal synovial sarcoma is a recently reported neoplasm rarely seen. We report a case of primary renal synovial sarcoma. The signs and symptoms are similar to any primary renal tumour; diagnosis is clinically difficult through general survey or multiple imaging modalities and requires specific molecular and genetic testing. Surgical resection and ifosfamide-based chemotherapy are the mainstay of management. The prognosis can be poor. Physicians should be aware that synovial sarcoma is one of the possibilities of malignancy in the kidney.*

Keywords: Renal tumour, synovial sarcoma

# Sarcoma Sinovial Renal Primario

## Un Reporte de Caso

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### RESUMEN

*El sarcoma sinovial renal es un neoplasma recientemente reportado raramente visto. Reportamos un caso de sarcoma sinovial renal primario. Los signos y síntomas son similares a los que se presentan en el caso de cualquier tumor renal primario. El diagnóstico es clínicamente difícil a través de un estudio general o las modalidades múltiples de la toma de imágenes, y requiere pruebas moleculares y genéticas específicas. La resección quirúrgica y la quimioterapia a base de ifosfamida constituyen el soporte principal de tratamiento. La prognosis puede ser pobre. Los médicos deben ser conscientes de que el sarcoma sinovial es una de las posibilidades de malignidad en el riñón.*

Palabras claves: Tumor renal, sarcoma sinovial

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### INTRODUCTION

Synovial sarcoma is a malignant soft-tissue neoplasm that usually arises in close association with the joints and generally carries a poor prognosis. Primary synovial sarcoma arising from the kidney is extremely rare; it was first reported and described in 1999 by Faria *et al* (1). Since then, cases have been sporadically reported. The knowledge on this rare malignancy is still limited. It is difficult to differentiate synovial sarcoma from other renal tumours; the diagnosis needs to be confirmed by molecular studies detecting SS18 (SYT)-SSX fusion transcripts. We report a case of primary renal

synovial sarcoma that was initially diagnosed as renal cell carcinoma.

### CASE REPORT

A 31-year old male merchant presented to hospital with a four-month history of gross haematuria and right flank pain. Physical examination did not reveal any palpable lymph nodes or abdominal mass. Ultrasound revealed a lower echogenic mass in the right kidney. Blood haemogram and biochemistry data were within normal limits. Enhanced computed tomography (CT) scan of the abdomen and pelvis demonstrated a 7.1 cm × 6.8 cm enhancing mass occupying the lower pole of the right kidney (Fig. 1). No local invasion or lymphadenopathy was identified. No venous thrombus was noted on abdominal vascular ultrasound. Bone scan and chest X-ray revealed no evidence of metastasis.

Right renal cell carcinoma was suspected preoperatively and right-sided radical nephrectomy was performed.

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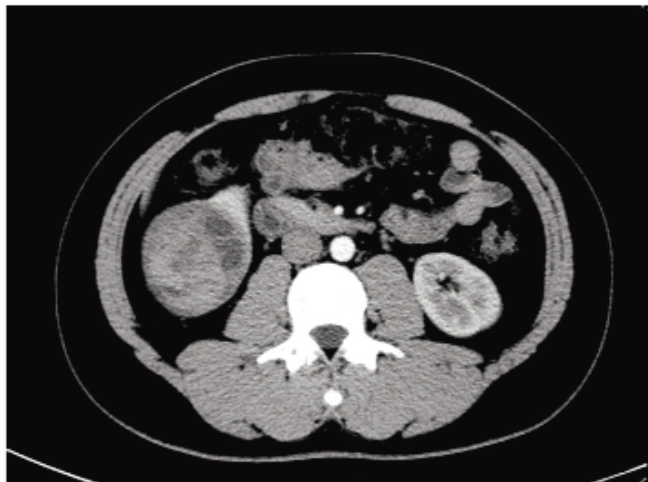


Fig.1: Contrast-enhanced computed tomography scan of abdomen showing enhancing mass occupying lower pole of right kidney.

During surgery, no gross invasion of the adjacent structures nor regional lymphadenopathy was noted. Grossly, the mass tumour was a well-circumscribed  $7 \times 7 \times 4$  cm renal neoplasm. The cut surface of the mass was tawny and firm with focal haemorrhage and necrosis. The renal capsule had not been invaded. Histologic examination showed the tumour composed of mitotically active typical plump spindle cells, monomorphic and with indistinct cell borders (Fig. 2A). Immunohistochemical analysis found that the tumour cells were positive for CD99, EMA, Ki-67, but negative for WT-1, desmin, PCK, LCA, S100, CK7, CK20, CD10, CD34. Fluorescence *in situ* hybridization (FISH) was used to detect the SYT-SSX fusion transcript produced by the  $t(X; 18)[p11.2; q11.2]$  using RNA extracted from the paraffin blocks. The case was positive for SSX18 group translocation (Fig. 2B).

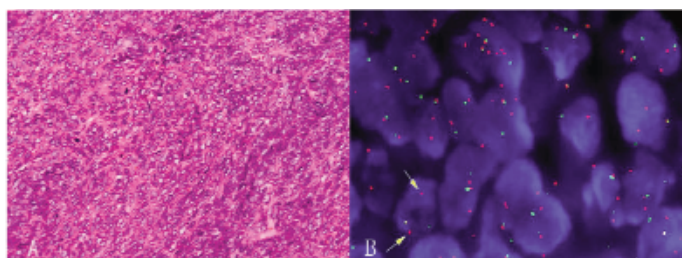


Fig. 2: (A) Plump, monomorphic neoplastic spindle cells and indistinct cell borders (Haematoxylin-eosin stain,  $40\times$ ).

(B) FISH picture shows that tumour cell with separate signal of red (5') and green (3'). The yellow arrow reveals the SSX18 chromosomal translocation.

The patient received chemotherapy after surgery for one month; he had received combination chemotherapy with 24-day cycles of pirarubicin 80 mg on day one plus dacarbazine 500 mg on days 1 to 3. A total of four cycles were given. The patient had no detectable evidence of recurrence after nine months of follow-up which continues.

## DISCUSSION

Synovial sarcomas account for 6% to 10% of soft-tissue sarcomas and occur primarily in the limbs in young adults (2). Primary synovial sarcoma arising from the kidney is an uncommon tumour (3). Over the last 10 years, less than 40 cases have been diagnosed and reported in the English literature. In a review, the average age of 19 cases was 38.5 years and the male-to-female ratio was 1.7:1 (4). In general, no clinical or imaging characteristics are diagnostic. The signs and symptoms are similar to those of any primary renal cancer, such as flank pain or haematuria. Computed tomography imaging studies have shown heterogeneous, enhancing masses with solid and cystic components.

The differential diagnosis should include other types of tumours, such as adult Wilms' tumour, spindle cell renal tumours, renal cell carcinoma and haemangiopericytoma *etc* (5). The diagnosis always requires pathologic confirmation. Synovial sarcomas are usually positive for CD99, vimentin, bcl-2, EMA, CD56 and are focally positive for EMA and Cytokeratin (6–8). However, no specific immunohistochemical markers for synovial sarcoma is available (9). Accumulated cytogenetic and molecular studies have shown that synovial sarcoma is specifically associated with a unique chromosomal translocation  $t(X; 18)[p11.2; q11.2]$  and the consequent fusion gene SYT-SSX (10). Primary renal synovial sarcoma can be confirmed by molecular detection of SYT-SSX fusion. Argani *et al* (6) reported 15 cases of primary renal synovial sarcoma. Only four of these were confirmed by molecular detection of SYT-SSX fusion transcripts. We use Fluorescence *in situ* hybridization (FISH) to detect  $t(X; 18)$  translocation on cytogenetic analysis; it is more accurate than PCR.

Currently, surgical resection and ifosfamide-based chemotherapy are the mainstay of management of synovial sarcomas. Radical nephrectomy has been performed. Although no specific chemotherapeutic regimens have been proposed for primary renal synovial sarcoma, some soft-tissue sarcomas have shown chemotherapeutic sensitivity and a response to high-dose ifosfamide-based protocols. Schaal *et al* (9) reported 50% tumour reduction using ifosfamide and adriamycin for a four-week period before nephrectomy. There has been no evidence of recurrence after one year of follow-up. From previously published data, renal synovial sarcomas have aggressive clinical courses and poor outcomes. In this case, the patient had received combination chemotherapy with pirarubicin plus dacarbazine.

In conclusion, primary renal synovial sarcoma is a rare malignancy. Preoperative diagnosis is very difficult. Accurate diagnosis includes cytogenetic and molecular studies. Surgical resection and chemotherapy have some curative effect. It has an aggressive course and poor prognosis. Physicians should be aware that synovial sarcoma is one of the possibilities of a renal malignancy.

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