Case Report: Primary Synovial Sarcoma of Kidney

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Citation

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Abstract

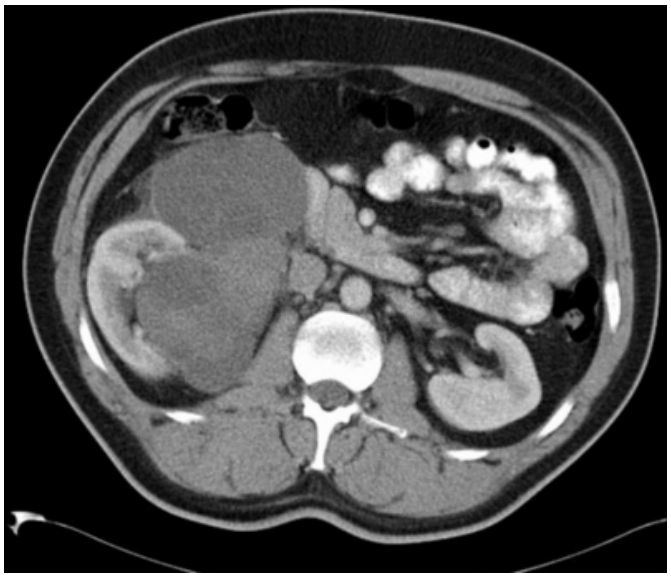
Primary renal cell synovial sarcoma is a rare entity and less than 30 cases are reported in literature. We report a case of 35-year-old male with primary synovial sarcoma of kidney, which was confirmed by SYT-SSX translocation.

CASE REPORT

A 35-year-old male presented with sudden onset right flank pain with haematuria. He was a non- smoker with unremarkable past medical and family history. He was apyrexial with tenderness in the right lumbar region. Urine dip was positive for blood and protein and abdominal X- ray was inconclusive.

A CT of the abdomen showed a large heterogeneous well defined mass measuring 15.4x13.8x9.5 cm. with cystic and solid components arising from the right kidney and centred around the renal hilum with antero-posterior extension abutting the psoas posteriorly (Fig.1).

Figure 1



Mild hydronephrosis involving the upper and lower poles was seen due to mass effect but no invasive feature. There was no lymphadenopathy or intra abdominal metastasis.

The patient was operated and a large cystic kidney was found for which a nephrectomy was performed. The cyst was ruptured during operation.

Macroscopically, the specimens weighed 979 g and the largest specimen measured 15x10x7.5 cm. It was largely occupied by partially solid and partially cystic lesion with haemorrhagic and shiny grey solid content.

Microscopically, the tumour showed the presence of monomorphic spindle cell proliferation growing in short intersecting fascicles or in solid sheets with high mitotic activity. The tumour also showed cysts of variable size lined by inactive polygonal eosinophilic cells with apically located nuclei (hobnail appearance). (Fig. 2, 3, 4)

Figure 2

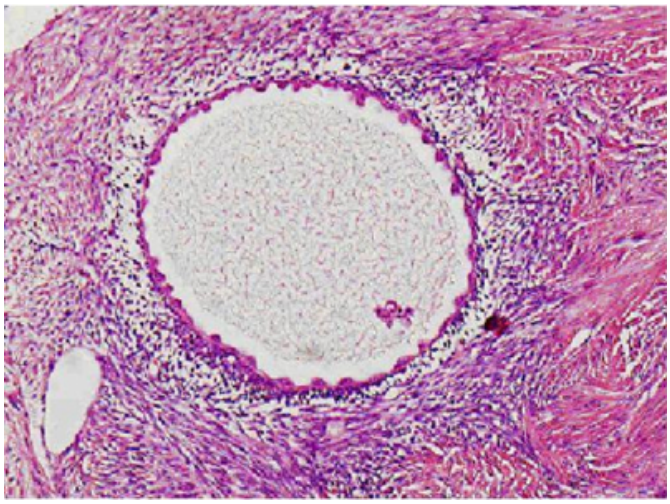


Figure 3

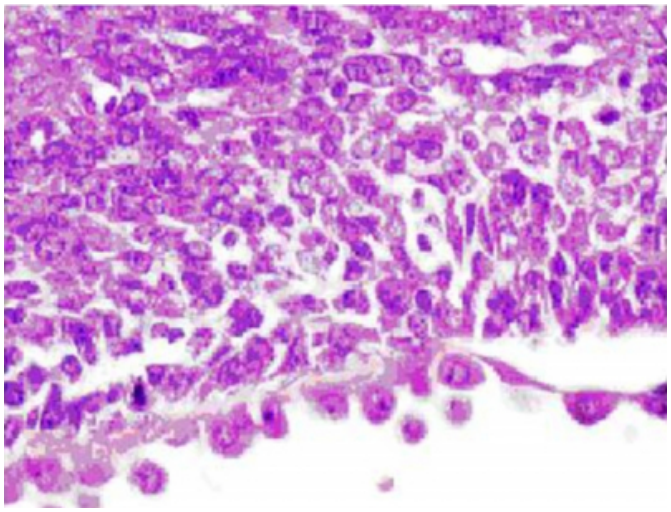
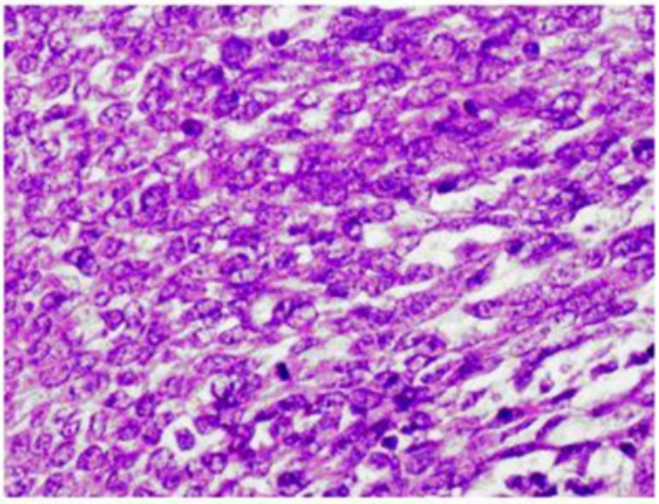


Figure 4



Immunohisto-chemistry showed tumour cells to be positive for Vimentin, BCL2 and CD99. Tumour cells were negative with Desmin, SMA, S100, CD34, CK, CD31, Factor8, Melan A, CD117, LCA, EMA, HMCK, CD10 and MyoD1.

These appearances were in keeping with biphasic synovial sarcoma with focal high grade areas.

Cytogenetic analysis later confirmed the diagnosis of synovial sarcoma. RT-PCR analysis showed SYT/SSX translocation, a characteristic feature of the condition.

The patient is currently being treated with VIDE (Vincristine, Ifosfamide, Doxorubicin and Etoposide) chemotherapy.

DISCUSSION

Sarcomas originating in kidneys of adults are rare.

Lieomyosarcoma is the most common renal sarcoma, which accounts for 40-60% followed by rhabdomyosarcoma, angiosarcoma, haemangiopericytoma, liposarcoma, chondrosarcoma and osteosarcoma. ([1]). Primary renal sarcoma in childhood is mainly confined to clear cell sarcoma of kidney and malignant rhabdoid tumour ([2]).

Synovial sarcoma is the fourth most common sarcoma, which accounts for 5-10% of all soft tissue sarcoma cases ([3]). Synovial sarcoma primarily occurs in extremities of young adult but has been reported in variety of unusual locations in which there is no synovial structure as detailed in Table 1.

Figure 5

Table 1: Unusual sites of Synovial Sarcoma ()

Head & Neck	Chest Wall
Orofacial	Intrathoracic
Tonsil	Pleura
Larynx	Lungs
Trachea	Mediastinum
Intracranial	Heart
Third Ventricle	Oesophagus
Abdominal wall	Intraosseous
Vulva	Intravascular
Intra-abdominal	Intraneural
Mesentry	
Retroperitoneum	

Primary renal synovial sarcoma is extremely rare and was first described by Faria et al in 1999 ([5]). It has been reported in the age group between 19-61. A M:F ratio of 1.7:1 has been established from review of first 19 cases ([6]). Patients presented with either flank pain or haematuria or both. The tumours at the time of diagnosis ranged from 5-19 cm with 67% having grossly identifiable cyst and 82% having cyst on microscopy ([6]). Histologically, tumours are typically mitotically active with monomorphic plump spindle cells and indistinct cell borders growing in short, intersecting fascicles or in solid sheets. Cysts are lined by mitotically inactive polygonal eosinophilic cells with apically located nuclei (“hobnailed epithelium”), and appear to be entrapped native renal tubules, which may be extensively dilated. Areas of solid aggregation or fascicles of the tumour cells alternating with hypocellular myxoid tissues, together with areas displaying a prominent haemangipericytoma-like pattern, may be found. Histologically, the tumour is classified as Monophasic, Biphasic (epithelial cells and

spindle cells) and Poorly differentiated.
Immunohistochemical markers are helpful in differential diagnosis of spindle cell neoplasm. However, none of these markers so far available is specific enough to make the diagnosis of Synovoial Sarcoma ([3]). Table 2 details the immunohistochemistry of synovial sarcoma

Figure 6

Table 2: Immunohistochemistry of Synovial Sarcoma()

Marker	Biphasic	Monophasic	All Cases
Cytokeratin	100%	68%	81%
EMA	100%	73%	84%
CEA	37%	8%	20%
LEU7	50%	33%	40%
S100	42%	27%	30%
CD99	67%	61%	62%
Bcl-2	100%	100%	100%

Despite the Histopathology and Immunohistochemistry differentiating between synovial sarcoma and congenital mesoblastic nephroma or clear cell sarcoma can be difficult. Cytogenetics, Chromosomal study and Molecular analysis has greatly aided in diagnosis of Synovial sarcoma. A characteristic translocation t(X;18) is found in 90% of the patients of synovial sarcoma ([7]). Crew et al demonstrated that the t(X;18)(p11.2;q11.2) results in the fusion of chromosome 18 SYT gene to either of the two homologous genes SSX1 or SSX2([7]).The prognosis of patients with SYT-SSX2 translocation is better than SYT-SSX1 translocation ([8]).

The treatment of primary renal synovial sarcoma involves surgery and adjuvant chemotherapy. Synovial sarcomas are chemosensitive to ifosphamide and doxorubicin based chemotherapy with a response rate of 24% ([9]). The prognosis despite treatment is poor with a 5-year survival rate of synovial sarcoma being 42-89%([8]). Factors predicting worse prognosis for patients with synovial sarcoma include tumour size (>5cm), male gender, older age (>20 years), extensive tumour necrosis, high grade, large number of mitotic figures (>10 per 10 high powered fields), neurovascular invasion and, recently, the SYT-SSX1 variant ([10]).

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