

## CASE REPORT

### Primary renal synovial sarcoma

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#### ABSTRACT

Primary renal synovial sarcoma is a rare tumor, with a poor prognosis which must be differentiated from other renal tumors. We report a new case of renal synovial sarcoma in a 60 year-old man. Histologic examination showed a proliferation of monomorphic spindle cells arranged in fascicles with hemangiopericytomatous pattern. Tumor cells were immunoreactive for bcl2, CD99, cytokeratin and epithelial membrane antigen. The diagnosis of spindle cell type monophasic synovial sarcoma was proposed and confirmed by fluorescence in situ hybridization. Patient was treated with chemotherapy. He developed bone and liver metastasis and died 7 months after nephrectomy. The diagnosis of renal synovial sarcoma is based on histological and immunohistochemical studies and must be confirmed by molecular techniques which showed the translocation t (X; 18).

**Key words:** kidney, synovial sarcoma, SYT-SSX gene fusion.

#### INTRODUCTION

The synovial sarcoma (SS) account for about 5 to 10 % of soft tissue sarcomas. It occurs mainly in the limbs of young adults.<sup>1,2</sup> Other sites are rare represented by the heart, lungs, esophagus, prostate and central nervous system.<sup>1</sup> Renal localization is extremely rare, first described by Argani et al. in 2000.<sup>3</sup>

We report a new case of renal SS confirmed by fluorescence in situ hybridization (FISH). Through this observation and the literature review, we discuss the

morphologic and immunohistochemical features, differential diagnosis and prognosis of this rare tumor.

#### CASE REPORT

A 60 year old man presented with a 1 month history of right flank pain. The computed tomography scan showed a right renal mass measuring 17 cm with heterogeneous enhancement (Fig. 1). Right nephrectomy was performed.



**Figure 1,** Computed tomography scan showed a renal mass with heterogeneous enhancement.

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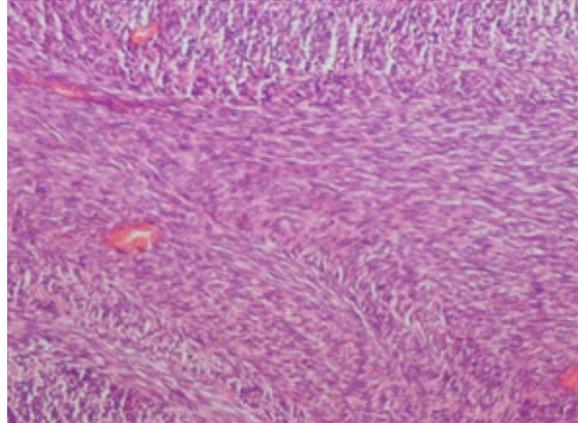
On gross examination, the mass was grey-white with focal hemorrhage and necrosis (Fig. 2).



**Figure 2**, Gross examination of a heterogeneous renal mass with areas of hemorrhage and necrosis.

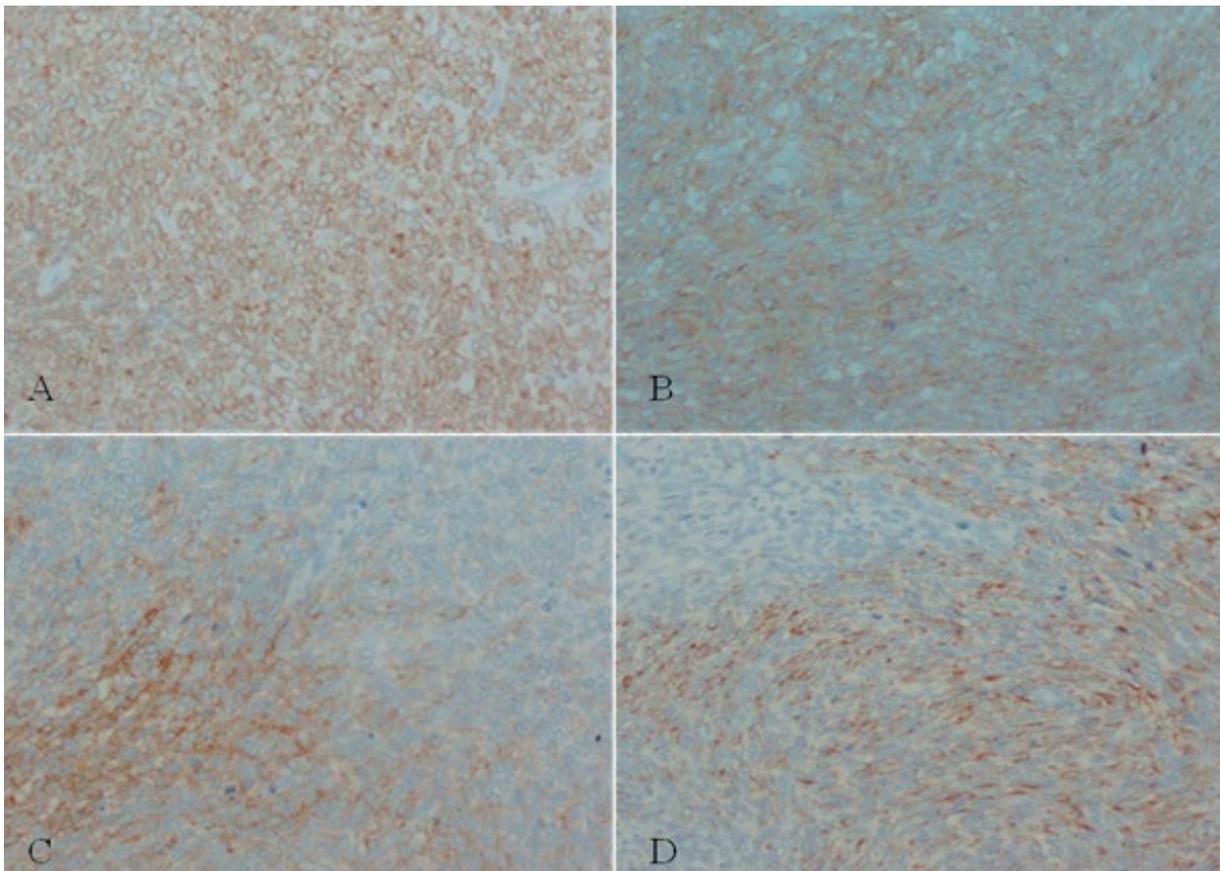
Cells had scant cytoplasm and ovoid pale-staining nuclei. Mitotic rate was 36 mitoses per 10 high power fields. A hemangiopericytomatous pattern was focally present. The tumor infiltrated the perinephric fat. There was no renal vein thrombosis.

Histologically, the tumor consisted of a monomorphic, plump spindle cells growing in fascicles (Fig.3).



**Figure 3**, Monomorphic plump spindle cells arranged in fascicles (Hematoxylin-eosin X 200).

On immunohistochemical stains, diffuse expression of bcl2 and focal expression of epithelial membrane antigen (EMA), cytokeratin and CD99 were detected in the tumor cells (Fig. 4).



**Figure 3**, Immunohistochemical study: diffuse positivity of tumor cells for bcl2 (A) and focal immunoreactivity for CD99 (B), EMA(C) and cytokeratin (D) (X 200).

We diagnosed the tumor as spindle cell type monophasic synovial sarcoma of the kidney.

A study by FISH performed at the Institut Curie (France), on tissue section using a dual color break-apart probe (probe Vysis) showed a rearrangement of SYT gene, confirming the diagnosis of synovial

sarcoma.

The patient received chemotherapy with Adriamycin. The evolution was marked by the occurrence of bone and liver metastases and the patient died 7 months after nephrectomy.

## DISCUSSION

Renal SS is a rare tumor that usually occurs in young adults.<sup>1,4</sup> However, age ranges between 15 and 78 years.<sup>1,5</sup> This tumor is characterized by a slight male predominance.<sup>5,6</sup> There are no clinical or imaging characteristics that can allow preoperative diagnosis. The diagnosis is based on histological examination and confirmed by molecular analysis.<sup>7</sup> Synovial sarcoma has 4 histologic subtypes: biphasic, monophasic spindle cells, monophasic epithelial cells and poorly differentiated.<sup>1,2</sup> Most renal SS correspond to the monophasic spindle cell variant. Histologically, this variant of SS consists of plump spindle cells with minimal cytoplasm, arranged in short intersecting fascicles. Rich and rarefied areas of cells are mixed and a hemangiopericytomatous vascular pattern is frequent. Cysts are commonly present and are lined by cells with eosinophilic cytoplasm and apical nuclei that create a hob nail appearance. These cysts correspond to entrapped and dilated renal tubules.<sup>1,2,6</sup> SS usually stain positively for vimentin, transducing-like enhancer protein1 (TLE 1), bcl2, CD99, cytokeratin and EMA. Desmin, smooth muscle actin and CD34 are not expressed.<sup>1,2,5,6,8</sup>

The diagnosis of renal SS is difficult, especially when it is, as in our case, a monophasic spindle cell variant. Primary renal SS must to be differentiated from renal sarcomatoid carcinoma. For this it is necessary to multiply the samples in search of conventional renal cell carcinoma areas. The tumor cells of renal cell carcinoma express vimentin and epithelial markers and do not express bcl2 and CD99. Other differential diagnoses of renal SS are: solitary fibrous tumor, malignant peripheral nerve sheath tumor, leiomyosarcoma and fibrosarcoma.<sup>9,10</sup> Solitary fibrous tumor is also characterized by hemangiopericytomatous vascular pattern. However, tumor cells express CD 34 in solitary fibrous tumor and no epithelial markers are detected. In malignant peripheral nerve sheath tumor, the cells express S100 protein. Leiomyosarcoma and fibrosarcoma do not express bcl2 and CD99.

The SS is characterized by the specific translocation t (X, 18) (p11.2; q11.2) found in 90 % of cases. This translocation leads to a fusion of the SYT gene on chromosome 18 to one of the homologous genes (SSX1, SSX2, and SSX4) of chromosome X. This translocation can be detected by reverse transcriptase polymerase chain reaction (RT-PCR) or by FISH using frozen or paraffin-embedded tissue.<sup>3,11,12</sup> Unlike soft tissue SS, primary renal SS is most often associated with a fusion of SYT- SSX2 genes. The latter being more common in monophasic spindle cells variant.<sup>6</sup> The mainstay of treatment of SS is surgical resection and chemotherapy. It is an aggressive tumor with a poor prognosis.<sup>13</sup> Most patients die within 1 to 2 years of tumor recurrence or metastasis.<sup>9,10</sup>

## CONCLUSION

Primary renal SS is a very rare tumor. In this loca-

tion, most tumors correspond to the monophasic spindle cell variant. After excluding sarcomatoid renal cell carcinoma, the diagnosis is based on histological examination associated with immunohistochemistry and must be confirmed by molecular analysis (RT-PCR and FISH) that highlight the translocation t (X, 18) characteristic of this tumor.

## REFERENCES

- Weiss SW, Goldblum JR. Synovial sarcoma. In: Enzinger and Weiss's Soft Tissue Tumor. Fourth edition. St.Louis, Mosby, 2001 .p.1483-509.
- Fisher C, de Bruijn DRH, Geurts van Kessel A. Synovial sarcoma. In: World Health Organization classification of tumours .Pathology and genetics of tumours of soft tissue and bone. Lyon: IARC Press; 2002. p. 200-4.
- Argani P, Faira PA, Epstein JI et al. Primary renal synovial sarcoma. Molecular and morphologic delineation of an entity previously included among embryonal sarcomas of the kidney. *Am J Surg Pathol* 2000; 24: 1087-96.
- Long JA, Dinia EM, Saada-Sebag G et al. Primary renal synovial sarcoma: a cystic tumor in young patients. *Prog Urol* 2009; 19: 474-80.
- Schoolmeester JK, Cheville JC, Folpe AL. Synovial sarcoma of the kidney: a clinicopathologic, immunohistochemical, and molecular genetic study of 16 cases. *Am J Surg Pathol* 2014 Jan; 38(1):60-5.
- Ro JY, Kim KR, Argani P. Synovial sarcoma of the kidney. In: World Health Organization classification of tumours .Pathology and genetics of tumours of the urinary system and male genital organs. Lyon: IARC Press; 2004.p.79-80.
- Chung SD, Huang KH, Chueh SC, Lai MK, Lin WC. Primary synovial sarcoma of the kidney. *J Formos Med Assoc* 2008; 107(4):344-7.
- Luo Yang, Kunjie Wang, Li Hong, Yan Wang, Xiang Li. The Value of Immunohistochemistry in diagnosing Primary Renal Synovial Sarcoma: A Case Report and Literature Review. *Int Surg* 2012;97 :177-181.
- Divetia M, Karpate A, Basak R, Desai SB. Synovial sarcoma of the kidney. *Ann Diagn Pathol* 2008; 12(5):333-9.
- Wang ZH, Wang XC, Xue M. Clinicopathologic analysis of 4 cases of primary renal synovial sarcoma. *Chin J Cancer* 2010; 29(2):212-6.
- Tvrđik D, Povysil C, Svatosova J , Dundr P. Molecular diagnosis of synovial sarcoma: RT-PCR detection of SYT-SSX1/2 fusion transcripts in paraffin-embedded tissue. *Med Sci Monit* 2005; 11(3):MT1-7.
- Kawauchi S, Fukuda T, Chochi Y, Kondo T, Oga A, Sasaki K. Reverse transcription-polymerase chain reaction in situ hybridization for SYT-SSX fusion gene transcripts in synovial sarcomas. *Int J Mol Med* 2005; 16(4):763-6.
- Iacovelli R, Altavilla A, Ciardi A et al. Clinical and pathological features of primary renal synovial

