

## Case Report

# Metastatic renal synovial sarcoma: a case report and review literature

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**Abstract:** We present the case of a 49-year-old patient presenting a metastatic synovial sarcoma originating in the kidney. The patient underwent left armpit neoplasm resection in 2001, 2002, 2003 and 2008, respectively. Postoperative pathology revealed synovial sarcoma. An enhanced abdominal computed tomography (CT) scan revealed a mass in the right kidney in 2013. A right radical nephrectomy was performed using an intraperitoneal approach through an anterior subcostal incision. However, only 6 months after surgery, the patient died from multiple organs failure due to multiple organs metastasis. This case report is the second report of metastatic synovial sarcoma in the kidney. Primary renal synovial sarcoma is very rare with aggressive behavior and poor prognosis. Immunohistochemical staining is helpful to the diagnosis, but a final diagnosis of renal synovial sarcoma can be gained basing on genetic analysis. It can be treated with a multidisciplinary approach and that radical surgery for the primary tumor, if possible, remains the standard of care up-to-date.

**Keywords:** Synovial sarcoma, kidney, metastatic, diagnosis, treatment

### Introduction

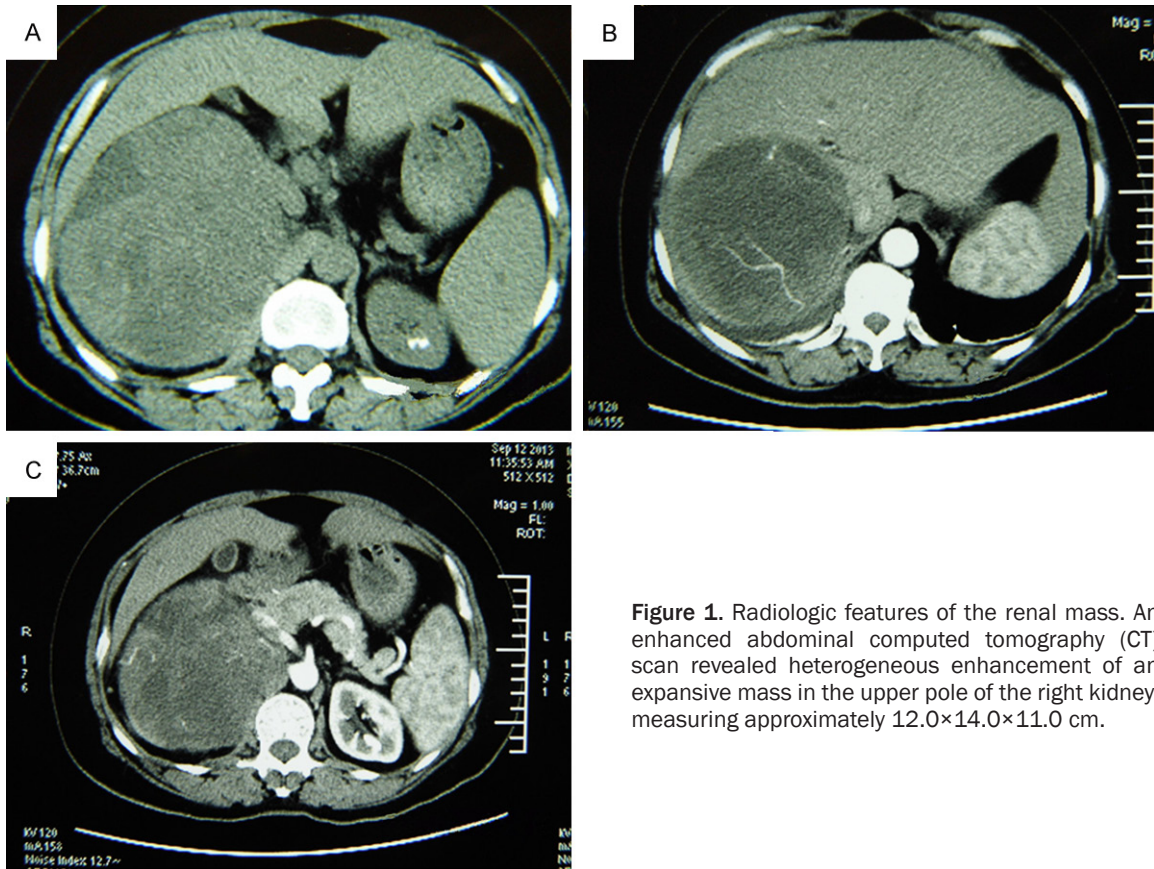
Synovial sarcomas are a group of soft tissue sarcomas of uncertain histogenesis and affect mainly proximal limbs of young adults. But it can also involve other sites (head and neck, heart, duodenum, lung, mediastinum, abdominal wall, kidney, prostate and so on) [1]. Primary renal synovial sarcomas are rare tumors of the kidney, firstly described in 1999 and published in 2000 by Argani [2]. However, metastatic renal synovial sarcoma is an extremely rare entity first reported in 2005 by Stage [3]. To date, there are only two cases of synovial sarcoma described in the literature.

Herein, we report a metastatic synovial sarcoma, originating in the kidney in a 49-year-old woman. Patient follow-up and literature review are also presented.

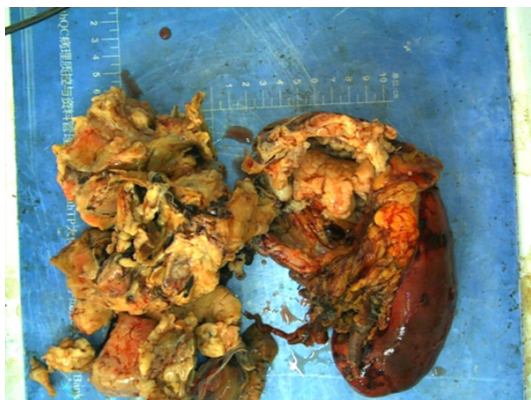
### Case report

A 49-year-old female patient with normal blood hemogram and biochemistry data complained

of pain in right lumbar region, accompanied with gross hematuria for 18 h. The patient had undergone left armpit neoplasm resection in 2001, 2002, 2003 and 2008, respectively. Postoperative pathology revealed synovial sarcoma which was confirmed by the result of SYT-SSX fusion gene mRNA expression using RT-PCR detection. The patient was a nonsmoker and had no history of exposure to chemicals. An enhanced abdominal computed tomography (CT) scan revealed heterogeneous enhancement of an expansive mass in the upper pole of the right kidney, measuring approximately 12.0×14.0×11.0 cm (**Figure 1**). Further evaluation including bone scan and chest CT scan did not demonstrate any evidence of metastasis. A right radical nephrectomy was performed using an intraperitoneal approach through an anterior subcostal incision. By intraoperative observation the tumor was approximately 15 cm, and originated from the upper pole of the right kidney and adhered to the inferior vena cava. No gross invasion of the adjacent structures nor regional lymphadenopathy or venous thrombus



**Figure 1.** Radiologic features of the renal mass. An enhanced abdominal computed tomography (CT) scan revealed heterogeneous enhancement of an expansive mass in the upper pole of the right kidney, measuring approximately 12.0×14.0×11.0 cm.



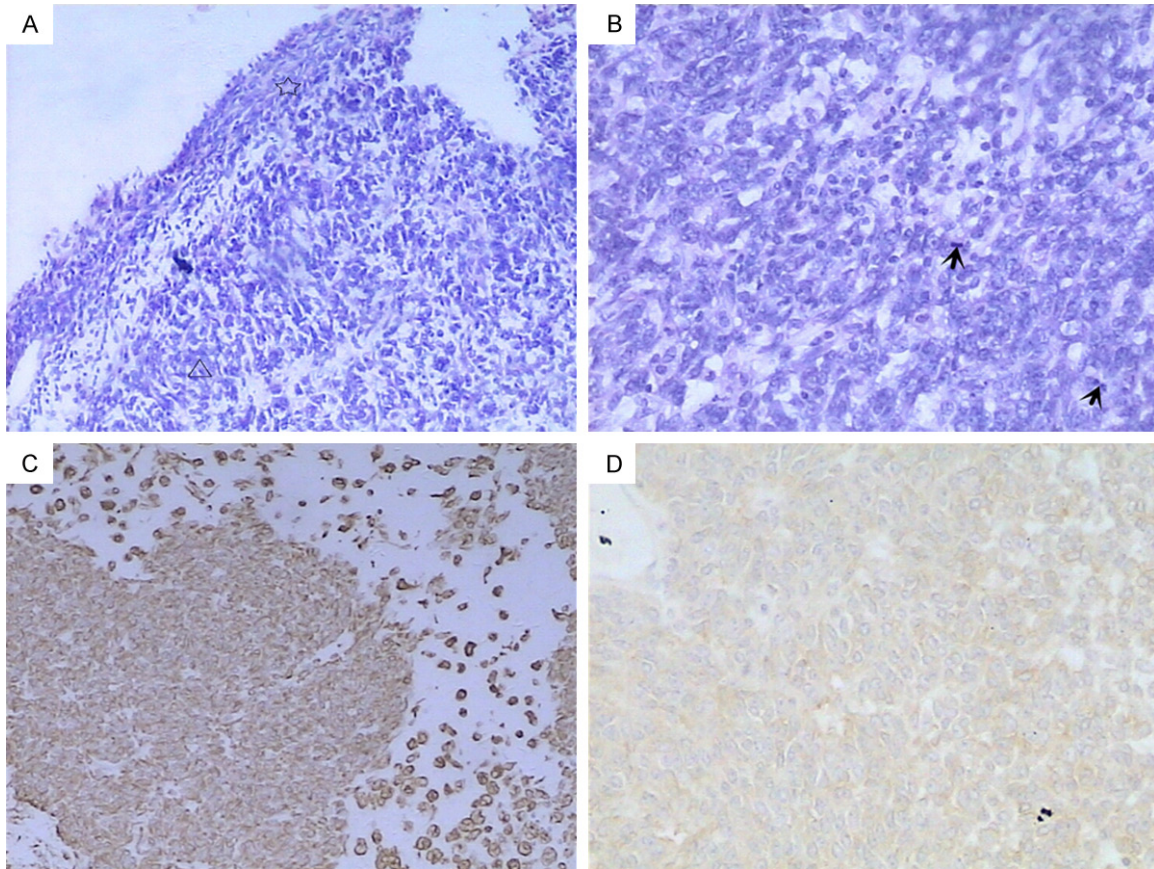
**Figure 2.** The resected tumor appeared irregular in shape, with the size of 12.8×11.2×11.0 cm.

was found during the operation. The resected tumor appeared irregular in shape, with the size of 12.8×11.2×11.0 cm (**Figure 2**). Histological evaluation by high power magnification field of the biopsy specimen from the right renal mass revealed that it was composed of monomorphic spindle cells with non-uniformly bounded cytoplasm in large areas and fascicles with cystic structures settled among them (**Figure 3A**).

There was prominent mitotic activity (**Figure 3B**). The tumor also exhibited necrotic and hemorrhagic areas. Immunohistochemically, the tumor cells showed positive for vimentin and CD 99 (**Figure 3C** and **3D**) but negative for other markers such as CGA, CD34, CD117, Desmin, CK and NSE.

Based on morphological and immunohistochemical features, the final pathological diagnosis of this case was renal synovial sarcoma. Combined with synovial sarcoma in patient with a history of multiple surgeries and the result of SYT-SSX fusion gene mRNA expression by RT-PCR detection, the patient was finally diagnosed as metastatic renal synovial sarcoma.

The patient underwent 2 courses of chemotherapy, each of which was carried out for five days. The chemotherapy regimens were 2,500 mg/m<sup>2</sup> ifosfamide at 1-5 days and 60 mg/m<sup>2</sup> doxorubicin at 1-5 days. No serious chemotherapy-related side effects were observed. However, six months after surgery, the patient died from multiple organs failure due to multiple organs metastasis.



**Figure 3.** A. Histological evaluation by high power magnification field of the biopsy specimen from the right renal mass revealed that it was composed of monomorphic spindle cells (HE  $\times 200$ ). B. There was prominent mitotic activity (HE  $\times 400$ ). C. Immunohistochemically the tumor cells stained positive for vimentin ( $\times 200$ ). D. Immunohistochemically the tumor cells stained positive for CD99 ( $\times 400$ ).

### Discussion

Synovial sarcomas account for only 6% to 10% of soft-tissue sarcomas and primarily occur in the limbs of young individuals. Synovial sarcomas originating from the kidney is extremely rare, with the incidence age ranging from 20 and 72 years old with median age of 35 years. The limited number of cases reported has shown a gender ratio close to one [4].

Primary renal synovial sarcomas has highly aggressive course and its prognosis is poor. There are no specific clinical or imaging characteristics which can provide a definitive preoperative diagnosis. Synovial sarcoma has 3 histologic subtypes: monophasic (only spindle cell), biphasic (epithelial cell and spindle cell) and poorly differentiated [5]. Bidirectional synovial sarcoma is easy to diagnose due to the presence of epithelial cell and spindle cell component, but sometimes it is difficult to distin-

guish monophasic synovial sarcoma from other spindle cell sarcomas such as leiomyosarcoma, fibrosarcoma, sarcomatoid renal cell carcinoma, Wilms's tumors, mixed epithelial and mesenchymal tumors.

It is a clinical challenge to diagnose renal synovial sarcomas. Immunohistochemical staining is helpful to the diagnosis, but, unfortunately, it is not very accurate due to the low sensitivities and specificities of the currently available immunohistochemical markers. By reviewing the literature, we found many immunohistochemical markers such as Bcl-2, vimentin, CD99, EMA, CD56, desmin have been investigated in cases of RSS. Primary renal synovial sarcomas are typically positive for Bcl-2, CD99, CD56, vimentin and focally for EMA. However, this pattern can also be seen in other tumor types such as primitive neuroectodermal tumors and malignant peripheral nerve sheath tumors [6].

In the present case, we observed a high nucleus to cytoplasm ratio and an increased presence of spindle shaped tumor cells with non-uniform bounded cytoplasm. On immunohistochemical staining, tissues were positive for vimentin and CD 99, but negative for CGA, CD34, CD117, Desmin, CK and NSE. Renal synovial sarcomas were potentially diagnosed based on the pathological examination, but a final diagnosis could be made based on the SYT-SSX fusion gene.

The current gold standard for the diagnosis of synovial sarcoma is the t(X; 18) (p11.2, q11.2) translocation using RT-PCR or florescent in situ hybridization (FISH), involving fusion of the SYT-SSX. We emphasized the importance of an adequate pathological diagnosis to ensure optimized treatment.

As the tumor is very rare, no definite standard treatment guidelines are available at present. Treatment is usually based on the scattered cases published in the world literature. For renal synovial sarcomas, surgery, radiotherapy, and chemotherapy are the options that may be administered separately or in combination. Surgery is advisable. However, the clinical benefit of adjuvant chemotherapy for renal synovial sarcomas is still controversial. Initial studies, mostly based on anthracycline-only chemotherapy, did not show an improved survival [7, 8]. Later researches on anthracycline- and ifosfamide-based chemotherapy also revealed a small gain in survival, which could not be reproduced in a subsequent, large clinical trial [8]. Schaal [6] reported a patient who underwent a combination of chemotherapy with ifosfamide and doxorubicin. The volume of tumor was reduced by fifty percent. Lakshmaiah [9] reported a case in which the patient received 6 cycles of adjuvant therapy with ifosfamide and adriamycin post-operatively. The patient was asymptomatic on regular follow-up 2 years. Our patient underwent 2 courses of chemotherapy, each of which lasted for five days. The chemotherapy regimens were 2,500 mg/m<sup>2</sup> ifosfamide at 1-5 days and 60 mg/m<sup>2</sup> doxorubicin at 1-5 days.

The natural history of renal synovial sarcomas is local recurrence. Eventual metastasis is common, with the most common site in lung, up to 80% in some series, followed by dissemination to the bones and bone marrow [3]. In the present case, bone scan and chest CT scan did not demonstrate any evidence of metastasis.

To our knowledge, after a MEDLINE and PubMed search, this is the second report of metastatic synovial sarcoma to the kidney.

### Conclusion

Primary renal synovial sarcoma is very rare with aggressive behavior and poor prognosis. Immunohistochemical staining can be helpful, but the final diagnosis of renal synovial sarcoma should be based on genetic analysis. We conclude that renal synovial sarcoma is a disease that can be treated with a multidisciplinary approach and that radical surgery for the primary tumor, if possible, remains the standard of care up-to-date.

### Disclosure of conflict of interest

None.

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