

## Original Article

# Primary synovial sarcoma of the kidney: a rare and easily misinterpreted renal entity

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**Abstract:** Primary synovial sarcomas of kidney are very rare, present a diagnostic dilemma. In this study, we reported clinicopathologic, immunohistochemical, and genetic characteristics of 12 cases of primary renal synovial sarcoma (SS). The ages of the 12 patients ranged from 21-52 years. Microscopically, 8 cases were monophasic type and 4 cases were biphasic type. The epithelial components of biphasic type SS were positive for AE1/AE3 (4/4) and EMA (4/4). Monophasic synovial sarcomas were focally positive for AE1/AE3 (3/8) and EMA (5/8). In all 12 cases, the spindle cell components were diffusely positive for vimentin (100.0%), BCL2 (83.3%), and CD99 (33.3%). CD10, CD34, SMA, and desmin were negative in all cases. The SS18 gene was rearranged in 10 cases. The SS18-SSX fusion gene was identified in another 2 cases. Clinical outcome data were available for 8/12 (66.7%) patients. Follow-up revealed that 5 patients (62.5%) died of the disease within 11 to 37 months after diagnosis (mean, 20.6 mo). Three patients (37.5%) were alive without evidence of disease 1 to 23 months after diagnosis (mean, 9 mo). We concluded that primary renal synovial sarcoma is a rare, aggressive tumor with poor prognosis that occurs over a wide age range but primarily in young adults and is morphologically similar to embryonal and mesenchymal neoplasms of the kidney. Adequate sampling, immunohistochemical staining, and specific fusion gene detection are essential to establish an accurate diagnosis.

**Keywords:** Kidney, synovial sarcoma, chromosomal translocation

## Introduction

Primary synovial sarcomas (SS) occur primarily in soft tissues, generally near the large joints of the extremities. SS can occur in unusual locations or organs, including the orbit, vascular lumen, pleura, lung, esophagus, larynx, mesentery, and mediastinum [1-5]. SS of the kidney are rare; only case reports or small series have been published since its first description in 2000 [6-10].

SS of the kidney present a diagnostic dilemma because it occur in a relatively wide age range from 15 to 61 years [11]. Within this age range, other types of biphasic and spindle cell neoplasms of both children and adults are more common than synovial sarcomas of the kidney, such as sarcomatoid renal cell carcinoma, Wilms' tumor, and solitary fibrous tumor. Additionally, synovial sarcomas are frequently asso-

ciated with extensive cystic change, creating a differential diagnosis of cystic renal neoplasms, such as sarcomas arising in cystic nephroma [12]. Therefore, differentiating SS from other renal neoplasms with similar histological features, which have very different clinical prognosis, is clinically important.

Most SS are associated with a specific reciprocal chromosome translocation  $t(X;18)(p11.2;q11.2)$  that results in fusion of the SS18 gene on chromosome 18 with the SSX gene on chromosome X [13-17]. Five variants of the SSX gene have been identified, however, only SSX1 and SSX2 have been shown to fuse with the SS18 gene [13]. These unique chromosome translocations are considered the best markers for the diagnosis of SS [18]. In 2000, Argani et al reported 15 cases of SS of the kidney, with genetic analysis of 4 cases [8]. In 2014, Schoolmeester et al reported a series of SS of

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the kidney, comprising 16 cases [9]. In these two studies, all cases were monophasic type synovial sarcoma (MSS). Here, we report an additional 12 genetically confirmed new cases with follow-up, including 4 biphasic synovial sarcomas (BSS), and emphasized the differential diagnosis, to further characterize this rare tumor of the kidney.

### Materials and methods

#### *Patients and clinicopathological data*

This study analyzed 12 cases of primary renal SS at Fudan University, Shanghai Cancer Center, the First Affiliated Hospital of Xinjiang Medical University, Huadong Hospital of Fudan University and the People's Hospital of Xinjiang. The relevant clinical data were collected by retrospective review of patient files. Follow-up information was updated through October 2014 by reviewing medical records and telephone follow-up. Special information with regard to the clinical stage was limited given the consultative nature of the cases. The histopathological diagnoses were based on World Health Organization criteria. The Institutional Review Board approved the use of the patients' tissue blocks and chart reviews.

#### *Immunohistochemical analysis*

Tissue slides were immunostained according to the manufacturer's protocol (Biocare Medical, Concord, CA). Briefly, antigen retrieval was performed, and slides were washed and incubated in 3% hydrogen peroxide. Slides were stained overnight at 4°C with primary antibodies, followed by application of secondary antibodies and avidin-biotin complex. Immunostaining was evaluated by two senior pathologists (B.C. and X.Y). Anti-AE1/AE3 (clone AE1/AE3, 1:50), EMA (clone E29, 1:100), BCL2 (clone 124, 1:50), CD99 (clone 12E7, 1:50), CD34 (clone QBEnd10, 1:50), SMA (clone 1A4, 1:500), Desmin (clone D33, 1:100) antibodies (Dako, Carpinteria, CA); CD10 (clone SP67, 1:100), Ki-67 (clone 30-9, 1:200) antibodies (Roche, AZ, USA); vimentin (1:200) (ChangDao, Ltd., Shanghai, China), and PAX8 (clone 10336-1-AP, 1:200) (Proteintech Group Inc, Chicago, USA) were used for immunohistochemical staining. Negative control staining was performed by replacing the primary antibodies with phosphate buffered saline.

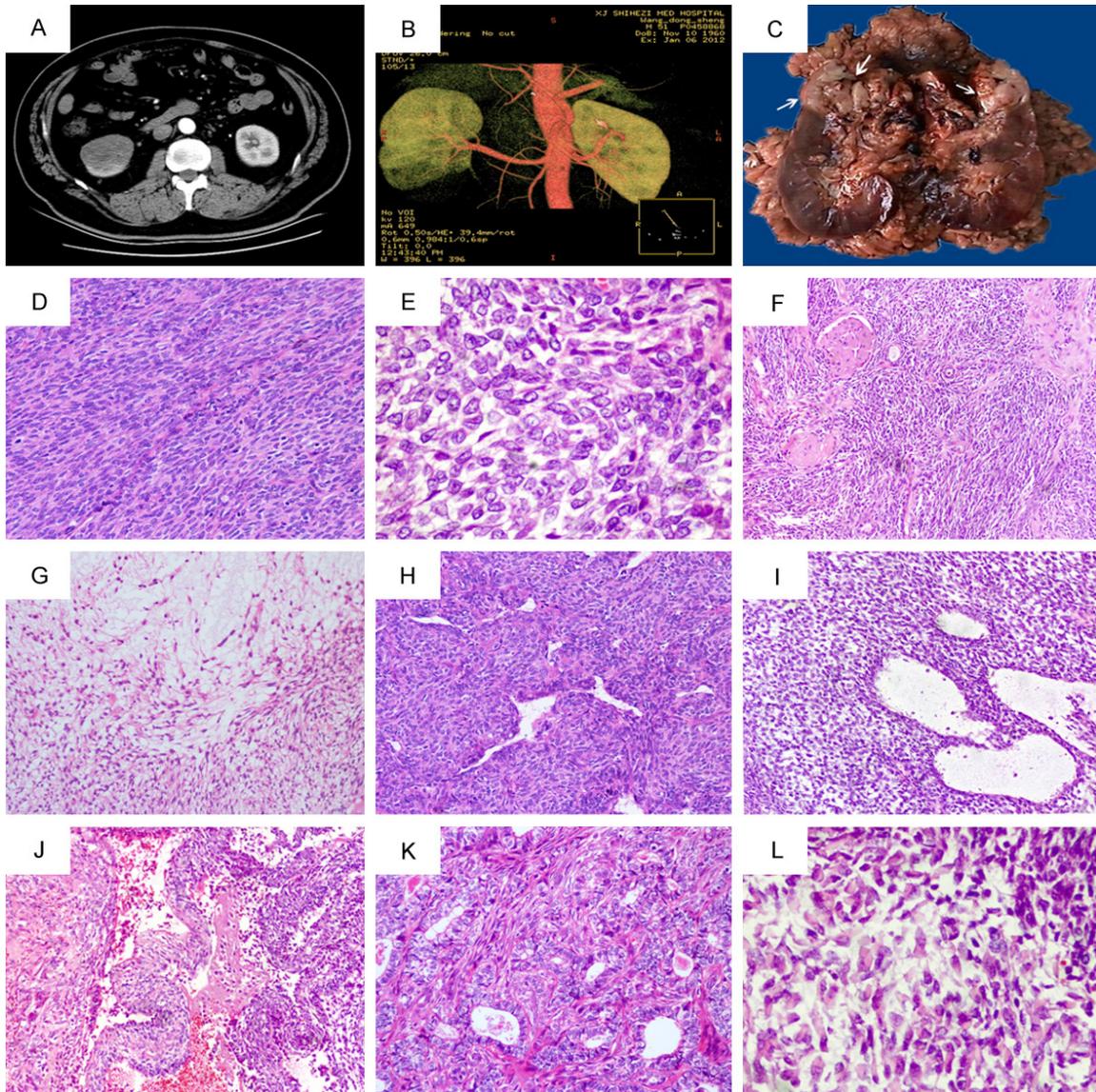
#### *Detection of SS18-SSX fusion gene by one-step reverse transcription polymerase chain reaction*

One-Step reverse transcription polymerase chain reaction (RT-PCR) was performed according to the manufacturer's instructions (QIAGEN, Venlo, the Netherlands) in a 25.0 µl volume containing 1 µg of total RNA, 5.0 µl of 5×RT-PCR buffer, 1.0 µl of 10 mM dNTP, 5 U of RNase inhibitor (Promega, Madison, WI), 0.6 µM forward and reverse primers and 1.0 µl of One-Step RT-PCR mixed enzyme [17]. A pair of consensus primers for SSX1 and SSX2 was used in our study to detect both SS18-SSX1 and SS18-SSX2 fusion genes. The sequences of the primers were SS18: 5'CCAGCAGAGGCCTTATGGATA-3' and SSX: 5'TTTGTGGGCCAGATGCTTC-3' (synthesized by Sangon Biological Engineering Technology and Services, Shanghai, China). RT-PCR was performed as follows: reverse-transcription at 50°C for 30 min; 95°C for 1.5 min, 35 cycles of denaturation at 94°C for 45 sec, annealing at 58°C for 45 sec, extension at 72°C for 1 min; and a final extension at 72°C for 10 minutes. The ubiquitously expressed β-actin gene was amplified as an internal control. A reaction mixture devoid of template RNA was included in each PCR procedure as a control [17].

#### *Detection of SS18 rearrangement by fluorescence in situ hybridization*

We used the Vysis SS18 Break Apart FISH Probe Kit to determine the presence of translocations involving the SS18 gene in two SS cases. Fluorescence in situ hybridization (FISH) was performed according to the manufacturer's instructions. The slides were hybridized with the LSI SS18 (18q11.2) Dual Color, Break Apart Rearrangement Probe (Vysis, USA). Tumor tissue sections (4-µm) were dewaxed, rehydrated, incubated in 0.05% pepsin/0.1 M HCl solution at 37°C for 10 min, and washed twice in phosphate-buffered saline for 5 min. The sections were subjected to microwave pretreatment with 0.1 M sodium citrate buffer (pH 6.0), dehydrated in a graded series of ethanol solutions, incubated in acetone at -20°C for 10 min, fixed in Carnoy's solution at RT for 5 min and denatured in 70% formamide/2X standard sodium citrate at 73°C for 5 min. A 10-µl volume of probe mixture was also denatured at

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**Figure 1.** CT, gross and microscopic image of SS. A. Contrast-enhanced CT scan revealing a 4.0 cm soft tissue-like, low-density mass in the lower pole of the kidney, protruding from the renal contour, ill defined compared with the normal kidney and containing heterogeneous density. B. CTA demonstrated that branches of the renal artery supplied the blood of the mass. C. Macroscopically, a yellowish-grey, round 4.0×3.5×3.5 cm nodular tumor mass was identified in the upper polar region of the kidney. D. Tumor composed of short fascicles of spindle cells. E. Spindle tumor cells with indistinct cell borders, ovoid nuclei and indistinct nucleoli. F. The tumor cells infiltrated around non-neoplastic sclerosing glomeruli and entrapped renal tubules. G. Alternating hypo- and hyper-cellular areas with myxoid degeneration were observed. H. Foci with a hemangiopericytoma-like vascular pattern. I. Cysts lined with a cuboidal or flat epithelium. J. Poorly formed broad glandular-like structures that mimic the epithelial elements of adult Wilms' tumor. K. Glands components of the BSS. L. Tumors have foci of rhabdoid cells with eccentrically located nuclei, prominent nucleoli, and eosinophilic cytoplasm.

73°C for 5 min and applied to the denatured sample tissue sections. The sections were covered with a cover glass, sealed with rubber cement, and incubated at 37°C for 16 h in a humidified chamber. Post-hybridization washes were routinely performed, and the sections were counterstained with a DAPI-II antifade

solution (Vysis). The hybridization signals were visualized using a fluorescence microscope, and images were captured with a CCD camera. Only non-overlapping nuclei were included in the analysis. Two different investigators counted at least fifty nuclei exhibiting both green and orange signals. The percentages of green,

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**Table 1.** Clinical features and pathologic diagnoses of the patients

No.	Age (years)	Sex	Ethnicity	Site of the tumor	Treatment	Pathologic diagnosis	Overall survival time (months)	SS18 gene rearrangement
1	52	M	Han	right	Nephrectomy	BSS	N/A	+(RT-PCR)
2	25	F	Han	right	Nephrectomy one week after transcatheter renal artery embolization	BSS	27 (died of tumor)	+(RT-PCR)
3	27	M	Han	left	Radical nephrectomy	MSS	N/A	+(FISH)
4	28	M	Uyghur	left	Neoadjuvant chemotherapy; laparoscopic left radical nephrectomy	BSS	11 (died of tumor)	+(FISH)
5	21	M	Han	right	Radical nephrectomy	MSS	16 (died of tumor)	+(FISH)
6	31	F	Han	left	Radical nephrectomy	MSS	12 (died of tumor)	+(FISH)
7	30	F	Han	right	Radical nephrectomy	MSS	N/A	+(FISH)
8	28	F	Han	left	Partial nephrectomy	MSS	23 (survival without tumor)	+(FISH)
9	45	F	Han	left	Radical nephrectomy	BSS	N/A	+(FISH)
10	35	F	Han	left	Radical nephrectomy	MSS	37 (died of tumor)	+(FISH)
11	38	F	Han	right	Radical nephrectomy	MSS	3 (survival without tumor)	+(FISH)
12	32	M	Han	right	Radical nephrectomy	MSS	1 (survival without tumor)	+(FISH)

BSS, biphasic type SS; MSS, monophasic type SS.

orange, and separated signals were calculated. Tissue sections in which more than 20% of the nuclei exhibited separated signals were considered positive. A case of SS known to harbor the SS18-SSX fusion gene was used as positive control. A rhabdomyosarcoma case was included as a negative control.

### Results

#### Clinical features

Clinically, the patients had either no symptoms or non-specific manifestations, such as abdominal pain or hematuria. In two cases, the masses were discovered during a routine physical examination. One case's imaging data were obtained. Abdominal B ultrasound examination revealed an abnormal heterogeneous hypoechoic signal in the kidney. Computed tomography (CT) scans of the abdomen and pelvis showed a heterogeneous solid or cystic-solid enhancing mass in the kidney with potential necrosis or hemorrhage (**Figure 1A**). CT angiography (CTA) demonstrated that branches of the renal artery supplied the blood of the mass (**Figure 1B**). The clinical features and pathological diagnoses are presented in **Table 1**. The age of the 12 patients ranged from 21 to 52 years (mean, 36 y). The tumors were identified in 7 females and 5 males. Eleven patients were the Han Ethnic group, 1 was the Uyghur. Six cases involved the right kidney, and 6 involved the left kidney. The patients were treated with radical nephrectomy in 10 cases, nephrectomy in 1 case and partial nephrecto-

my in 1 case. One case underwent preoperative transcatheter renal artery embolization, and another underwent neoadjuvant chemotherapy.

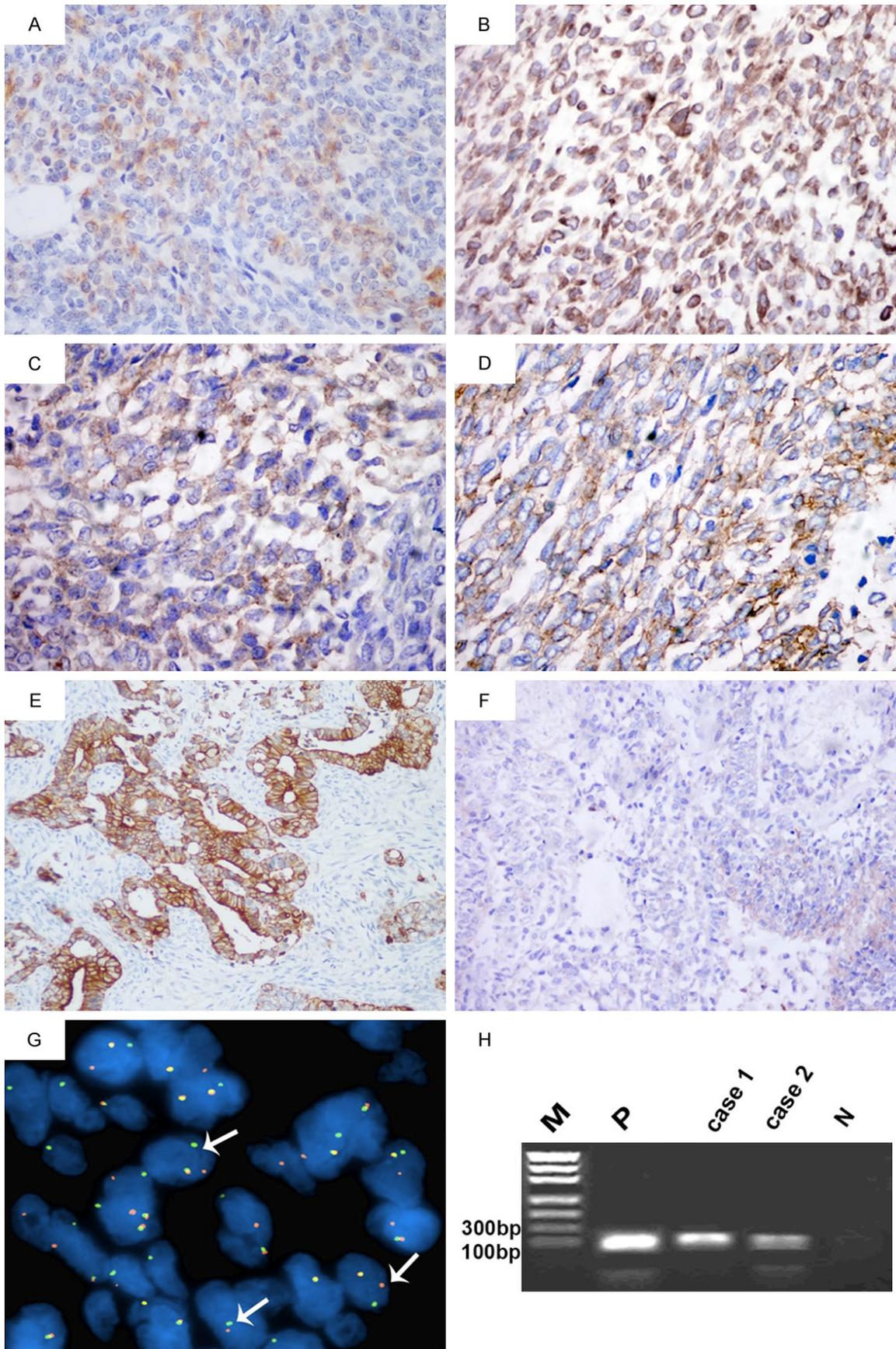
Clinical outcome data were available for 8 of 12 patients (66.7%) (Range 1 to 37 mo; mean, 16.3 mo). Follow-up revealed that five patients (62.5%) died of the disease within 11 to 37 months of diagnosis (mean, 20.6 mo). Three patients (37.5%) were alive without evidence of disease 1 to 23 months after diagnosis (mean, 9 mo). Another three patients were alive without tumor at 6, 7, and 12 months after diagnosis and were then lost to follow-up.

#### Pathological findings

Grossly, the tumors were gray-white, soft to rubbery solid masses (**Figure 1C**), many with partially necrotic and hemorrhagic areas. Cysts were apparent in one case. The tumor size ranged from 3.1 to 17.0 cm (mean, 7.2 cm).

Microscopic examination revealed that 8 cases were MSS and 4 cases were BSS. MSS were composed of short fascicles, sheets of spindle cells (**Figure 1D**) with indistinct cells borders, ovoid nuclei and indistinct nucleoli (**Figure 1E**) infiltrating around non-neoplastic, entrapped sclerosing glomeruli and renal tubules (**Figure 1F**). Scattered mitotic figures could be seen. Focal alternating hypo- and hyper-cellular areas with myxoid degeneration were observed in 2 cases (16.7%) (**Figure 1G**). Foci with a hemangiopericytoma-like vascular pattern (**Figure 1H**)

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**Figure 2.** Immunostains and molecular detection of SS. (A) The spindle cells were weakly focally positive for EMA and (B) diffusely positive for Vimentin, (C) BCL2, and (D) CD99. (E) The glandular components of the BSS were positive for AE1/AE3 and (F) weakly focally positive for EMA. IHC,  $\times 400$ . (G) Fluorescence in situ hybridization (FISH) for the detection of SYT rearrangement; positive signal: separation of the orange and green signals (arrow); co-localized orange and green signals are observed in intact cells. (H) The tumor was positive for the SYT-SSX fusion gene (98 bp) as detected by RT-PCR. M: Marker, P: Positive control, N: Negative control.

and cysts lined by a cuboidal or flat epithelium (**Figure 1I**) were present in some areas of 1 case (8.3%, 8.3%), respectively. A poorly formed broad glandular-like pattern was apparent in 1 case (8.3%) (**Figure 1J**), increasing the potential for misdiagnosis as an adult Wilms' tumor. Plump epithelial cells forming glands, which were negative for PAX8, excluded entrapped renal tubules and suggested BSS (**Figure 1K**). One case had foci of rhabdoid cells with eccentrically located nuclei, prominent nucleoli, and eosinophilic cytoplasm (8.3%) (**Figure 1L**).

### *Immunohistochemical findings*

Immunohistochemical staining revealed that MSS were scattered, individual cells positive for AE1/AE3 (3/8) and focally positive for EMA (5/8) (**Figure 2A**). In all 12 cases, the spindle cell components were diffusely positive for Vimentin (100.0%, 12/12) (**Figure 2B**), BCL2 (83.3%, 10/12) (**Figure 2C**), and CD99 (33.3%, 4/12) (**Figure 2D**). CD10, CD34, SMA, and Desmin were negative in all cases. Between 5% and 25% of tumor cells were positive for Ki-67. The epithelial components of 4 BSS were positive for AE1/AE3 (4/4) (**Figure 2E**), EMA (4/4) (**Figure 2F**), and negative for PAX8.

### *Expression of SS18-SSX transcripts and SS18 gene rearrangement*

Ten cases of SS were positive for rearrangement of the SS18 gene as evaluated by FISH using a break-apart probe. More than 20% of the nuclei had separated signals in the tumor samples (**Figure 2G**). Separation of signals is indicative of the t(X;18) translocation. Two cases of SS were positive for SS18-SSX fusion transcripts (98 bp) as detected by One-Step RT-PCR using a consensus primer for SSX1 and SSX2 (**Figure 2H**). We could not evaluate the SS18 gene rearrangement by FISH in these two consultative cases because tissues were unavailable.

### **Discussion**

Primary SS of the kidney was first reported in 2000 by Argani P et al [8]. Only case reports or

small series studies have been published. Lacovelli et al summarized 21 case reports and 9 small series from 2000 to 2011 and published a meta-analysis of 64 cases of renal SS (including the series of Argani and colleagues) in 2012 [19]. The findings of the present series comprising 12 genetically confirmed renal SS, including 4 BSS and 8 MSS with follow up information extend those of Argani and Schoolmeester et al.

Renal SS occur most commonly in adolescents and young adults, but the age at presentation can range from 17 to 61 years. The age of the 12 patients in our series ranged from 21 to 52 years. The mean age of patients with renal SS in our series is 36 years, which is very similar to that reported in the Argani series (37 years) and somewhat younger than that in the Schoolmeester series (46 years). Clinically, patients commonly present with flank pain and/or hematuria or are asymptomatic. Neither clinical features nor imaging modality are diagnostic. The outcome for renal SS is poor, both in the present and prior series. Approximately 57% of patients die from the tumor within 3 years. Because of the low number of cases, the data on the outcome in our series are helpful to clearly define poor prognosis of renal SS.

Histologically, in contrast to Argani et al and Schoolmeester et al, who reported 15 and 16 cases of renal MSS, respectively, our series included 4 cases of BSS, reflecting differences in our patient populations.

Renal tumors have special characteristics compared with tumors in other organs. The spectrum of renal tumors differs in children and adults. Embryonal tumors, such as Wilms' tumor, mesoblastic nephroma, rhabdoid tumor, and clear cell sarcoma mainly occur in children. Renal cell carcinoma (approximately 90%) and rarely mesenchymal tumor are more common in adults. However, some embryonal tumors can occur occasionally in the kidneys of young adults. Because of the wide range of age of onset (range from 15-61 years old), the rarity of

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**Table 2.** Differential diagnosis of synovial sarcoma

	AE1/3	EMA	SMA	ER/PR	BCL2	CD10	CD34	CD99	S-100	WT-1	Molecular
Sarcomatoid renal cell carcinoma		+				+					
Malignant epithelial and stromal sarcoma	+	+		+							
Wilm's Tumor	+	+								+	
MPNST									+		
Ewing's sarcoma/PNET								+			EWS/FLI1
Hemangiopericytoma							+				
Solitary fibrous tumor					+		+				NAB2-STAT6
Synovial Sarcoma	+	+						+	+/-		SS18-SSX

the tumor, and the lack of specific immunomarkers, renal SS are easily misdiagnosed for morphologically similar child and adult renal tumors.

Some of the cases in our cases group were consultation cases from the other hospitals. The initial diagnosis included Wilm's tumor, subtype undetermined; spindle renal tumor, possibly malignancy. The differential diagnosis includes sarcomatoid renal cell carcinoma (sarcomatoid RCC), Mixed epithelial and stromal tumor (MEST) [20, 21]. Here, we describe the key criteria for differential diagnosis of primary SS from its mimics, and list the useful diagnostic immuno-markers and molecular features in **Table 2**. Sarcomatoid RCC is characterized by sheets of whorled or interlacing bundles of pleomorphic spindle cells in a storiform pattern and may focal positive for epithelial markers, which mimics synovial sarcoma. Both the sarcomatoid RCC and SS may present hemangiopericytoma like pattern. In contrast to synovial sarcoma, sarcomatoid RCC show classic RCC components with adequate sampling, which are mostly clear cell RCC, less common are chromophobe RCC, papillary RCC, unclassified RCC and collecting (Bellini) duct carcinoma, and negative for SS18-SSX. Epithelial lined tubular or cystically dilated structures admixed with spindle mesenchymal components in MEST can mimic BSS, especially when the tumor is cellular, which with increased stromal cellularity, and high mitotic index. Most of the MESTs are benign renal tumor. Bland epithelium, ovarian-type stromal components, and estrogen-receptor and progesterone-receptor positivity of the stromal are characteristics of MEST [22].

Additional, SS of the kidney need to differentiate from Adult Wilms' tumors, malignant peripheral nerve sheath tumor (MPNST), Ewing sar-

coma/PNET, and solitary fibrous tumor (SFT). Wilms' tumor is characterized by more pleomorphic and complex components, immunopositivity for WT1, and absence of SS18-SSX translocation. S100 antigen is not helpful in differentiating SS and MPNST because 30% of SS express S100. Morphologically, MPNST has more pleomorphic cells with tapering nuclei, large gaping vascular spaces, perivascular plump tumor cells, geographic necrosis with tumor palisading at the edges (resembles glioblastoma multiforme), and frequent mitotic figures. MPNST may feature bizarre cells. Immunohistochemically, MPNST is negative for EMA, CK7, CK19, CD99, and the SS18-SSX fusion gene, while the SS is positive for EMA (91%), CK7 (57%), CK19 (57%), and CD99 (30%) [23]. Ewing sarcoma/PNET may have focal neuroendocrine features, including Homer-Wright rosettes. RT-PCR and FISH analyses have indicated that 90% contain an EWS/FLI1 fusion product due to t(11;12)(q24/q22;q12). Multiple arrangement patterns and diffuse strong CD34 positivity indicate the diagnosis of the SFT, which has been proved harbor the NAB2-STAT6 gene fusions recently. Occasionally, angiomyolipoma of the kidney, which is lack of the adipose and vascular components and mainly composed of myoid spindle cells, may confuse with MSS. Both of the tumors may variable positive for calponin. Bland and without atypical spindle cell, positive for HMB45, support the diagnosis of variable angiomyolipoma.

Our study has some limitations. Some studies reported that type of SS18-SSX translocation were independent survival factors. Patients with SS18-SSX1 fusion gene had worst prognosis [24]. Because some of the cases were consultation cases from other hospital, the materials were not available for further study. We did not detect the specific SS18-SSX1 or SS18-SSX2 fusion respectively for every case. In

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addition, the longest follow up time in our serious was 37 months, which was less than five years. We will do the further follow up to clarify the five years or ten years survival rates of the renal SS in our series.

In conclusion, we have described 12 cases of primary SS of the kidney. This rare tumor is easily confused with other spindle cell tumors or biphasic differentiated tumors of the kidney. Primary SS should be included in differential diagnosis when evaluating spindle cell tumors and biphasic differentiated tumors of the kidney, especially for the young patients. Adequate sampling, immunohistochemical staining, and Identifying characteristic genetic changes are essential for accurate diagnosis.

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### Disclosure of conflict of interest

None.

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