

Primary Renal Synovial Sarcoma



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Abstract

Synovial sarcomas are tumors that often occur in the proximity of large joints of young adults and adolescents. Primary synovial sarcoma of the kidney is very rare and difficult to diagnose. To date, fewer than 50 cases of primary renal synovial sarcoma (SS) have been reported in literature published in English. There are no established guidelines regarding the management of this tumor given the limited number of cases reported. Although primary surgical resection is the treatment of choice for primary renal synovial sarcoma, the prognosis is poor with surgery alone. Here, we describe a case of a 25-year-old female with primary renal synovial sarcoma that was treated successfully by radical nephrectomy followed by adjuvant chemotherapy. Physicians should be aware of the possibility of malignancy in renal masses and raise the suspicion of synovial sarcoma, especially when patients with renal masses are young adults.

Keywords: synovial sarcoma, renal

Synovial sarcoma occurs primarily in the soft tissues, mostly in para-articular regions of the extremities in adolescents and young adults. They account for about 5% to 10% of adult soft-tissue sarcomas.¹ These tumors have been described in other unusual locations, including thoracic and abdominal wall,² head and neck region, including pharynx and larynx,³ retroperitoneum,⁴ and bone⁵ as well as visceral organs, such as lung⁶, pleura⁷, or prostate.⁸ Primary renal SS is a rare tumor first described by Argani et al. in 2000.⁹

Primary synovial sarcoma of the kidney is a rare entity: to date fewer than 50 cases have been reported. This tumor presents a diagnostic dilemma because it is quite difficult to differentiate it from other renal neoplasms, such as metastatic sarcoma, renal cell carcinoma with sarcomatoid differentiation, which may have similar histological features.

Case Report

A 25-year-old Pacific Islander female presented with 2 weeks' history of left flank pain and gross hematuria. CT whole abdomen revealed 11.3x10.3x9.5 cm (AP x transverse x vertical) lobulated exophytic heterogeneous necrotic mass from the anterior part of left kidney, without internal calcification or fat. The mass markedly compresses the left renal pelvis. Left renal artery and vein are patent (Figure 1). Patient subsequently underwent left nephrectomy.

Grossly, there is one solid hemorrhagic globular tumor protruding from the renal hilum, measuring 9.0x7.0x7.0 cm. The sectioned midvertical surface of the kidney and tumor reveal extensive hemorrhagic necrosis and sclerotic areas with no cystic space of tumor tissue. The interface of the tumor and the adjacent kidney is well demarcated. The capsular surface of the tumor is covered by a thin band of blood clots and fibrous material on top of perinephric fat.

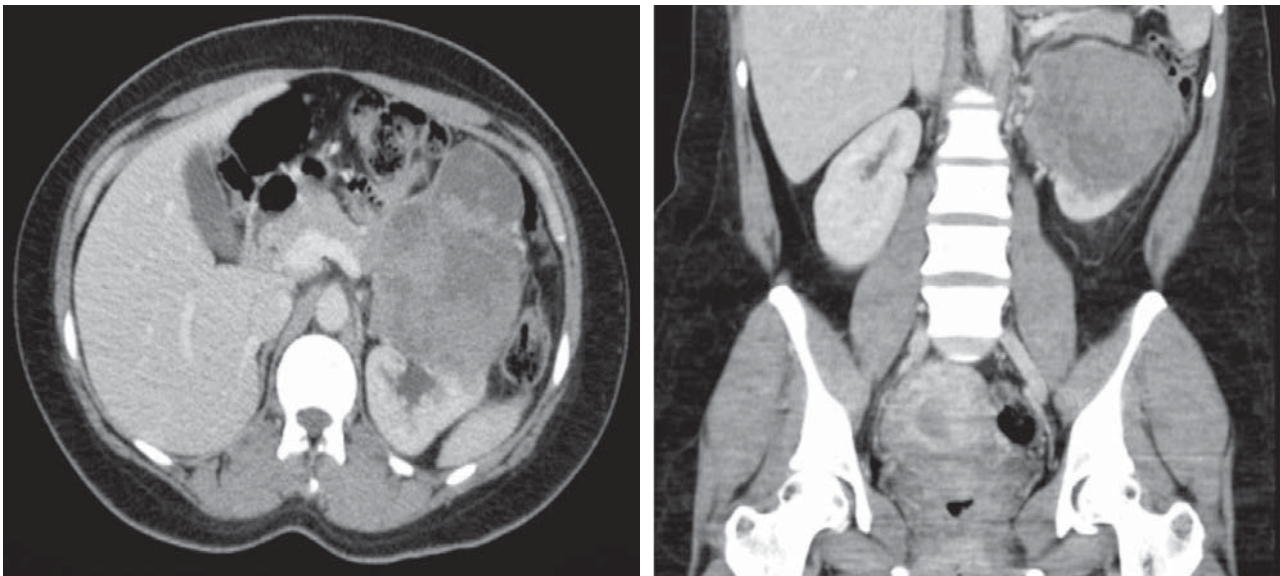


Figure 1: CT abdomen revealed 11.3x10.3x9.5 cm lobulated exophytic heterogeneous necrotic mass with contrast enhancement from anterior part and upper pole of left kidney.



Figure 2: Solid hemorrhagic globular tumor protruding from renal hilum with extensive hemorrhagic necrosis

Renal pelvis is compressed by tumor margin. Structures in the renal pelvis and renal sinus including the artery, veins and the pelvocalyceal wall show no gross tumor invasion and no lymph node. There are subcapsular scars of renal parenchyma along the tumor margin (Figure 2). Histologic section reveals a highly cellular tumor composing of monomorphic, plump short spindle cells growing in a solid sheet with indistinct cell borders. Cytoplasm is scant and ill defined. Nuclei are ovoid to fusiform with coarse chromatin and variable-sized nucleoli. Mitosis is frequent. There are extensive areas of hemorrhage and necrosis. The cystic space is not clearly seen. Invasion of renal parenchyma, renal capsule, renal pelvis or renal vein is not identified. Renal parenchyma shows segmental chronic pyelonephritis. Immunohistochemistry revealed positive CD99 and BCL-2; negative synaptophysin, chromogranin, WT-1 (Figure 3).

The patient underwent 4 courses of MAI (mesna (M), doxorubicin (A) and ifosfamide (I)) chemotherapy. Doxorubicin 60 mg/m² day 1, ifosfamide 2500 mg/m² day 1-3, and mesna 2500 mg/m² day 1-4. Pegfilgrastim was given on day 5 as a primary neutropenia prophylaxis. The patient has tolerated chemotherapy very well and there is no evidence of disease recurrence at the 5-month follow up.

Discussion

Sarcoma is classified according to histologic type: leiomyosarcoma, rhabdomyosarcoma, liposarcoma, fibrosarcoma, malignant fibrous histiocytoma, hemangiopericytoma, and osteosarcoma.¹⁰⁻¹² Sarcoma originating from the kidney is very rare. The most common primary renal sarcoma is leiomyosarcoma, which accounts for 40–60%

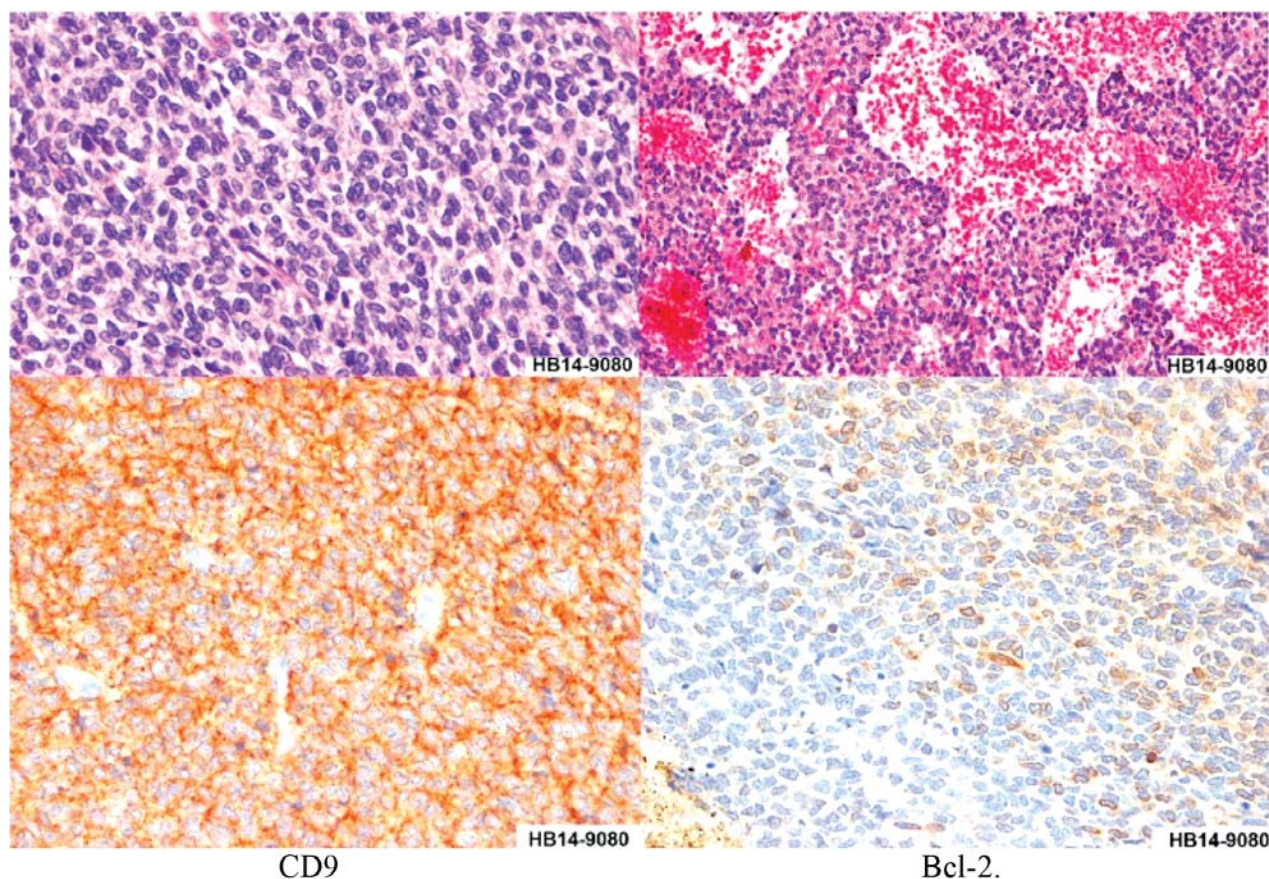


Figure 3: Monomorphic, plump short spindle cells growing in a solid sheet with indistinct cell borders. Mitosis is frequent. Tumors are positive for CD 99 and Bcl-2.

of the all reported cases, while primary synovial sarcoma in the kidney is extremely rare.^{10,11} Less than 50 cases have been described in the literature to date.

The limited number of primary renal synovial sarcoma cases reported has shown a gender ratio close to one, a mean age at diagnosis of 37 years (ranging between 13 and 67).¹³ There is no clinical or imaging characteristic that can indicate the diagnosis. The diagnosis is difficult due to the rarity of the tumor and its similar presentations when compared to other renal tumors. Most of these neoplasms were interpreted radiologically as renal cell carcinoma. Grossly, they were generally well-defined, irregular large masses ranging from 5 to 20 cm. Extension into inferior vena cava was reported in three cases. Grossly identifiable smooth-walled cysts were identified in eight cases.^{9,14-16} Reverse transcriptase-polymerase chain reaction (RT-PCR) analysis of synovial sarcoma shows a unique chromosomal translocation $t(X;18)(p11.2;q11.2)$ that results in the fusion of SYT gene on chromosome 18 with SSX family gene on chromosome X and helps to confirm the diagnosis.¹⁶

The final pathological diagnosis of this case was based on morphological and immunohistochemistry features, which depicted a pattern typical of synovial sarcoma, due to the presence of epithelial and mesenchymal markers.

We have requested a chromosomal translocation study but because we were not able to extract quality DNA from the tumor tissue, therefore, we were not able to carry out the chromosomal study to confirm our diagnosis.

Synovial sarcoma is considered to be an aggressive form of sarcoma and often has systemic spread. Although primary surgical resection is the treatment of choice for synovial sarcoma, the prognosis is poor with this treatment alone. It is considered to be more sensitive to anthracycline-based chemotherapy with approximately 53% response rate.¹⁷

However, the prognosis of primary renal synovial sarcoma is unclear due to the limited number of reported cases. From previously published data, renal synovial sarcomas are believed to have aggressive clinical courses and poor outcomes.^{18,19} There are no established guidelines regarding the management of this tumor given the limited number of cases reported. Primary surgical treatment is considered to be the treatment of choice; prognosis is poor with this treatment alone. The value of chemotherapy is yet to be proven but synovial sarcoma may be sensitive to high doses of Ifosamide-based regimens. Park et al.²⁰ reported complete remission of metastatic lung lesion using Ifosamide and doxorubicin. Due to this uncertainty

about the benefits, there is a higher possibility for adjuvant chemotherapy to be used in younger patients and larger tumors, in which a net benefit of treatment could be achieved as in our reported case.

Conclusion

Primary renal synovial sarcoma is rare and commonly affects young adults. Physicians should be aware of the possibility of synovial sarcoma in renal masses especially when patients with renal masses are young adults. Additional adjuvant ifosfamide based chemotherapy with surgical resection might be beneficial specifically when the patient is young and harbors a large tumor.

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