

## CASE REPORT

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# An Unusual Presentation of a Rare Tumor: Primary Renal Synovial Sarcoma

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**ABSTRACT** Primary renal synovial sarcomas are very rare and highly aggressive tumors. They are seen at an average age of 35 years. Common symptoms include side pain, abdominal pain, and hematuria. Imaging modalities have no specific findings indicating their presence, and the diagnosis is made based on immunohistochemical methods, which may need to be confirmed by cytogenetic analysis. Radical nephrectomy is the best treatment option currently available, with a combination of anthracyclines and ifosfamide being administered as adjuvant chemotherapy. In this case report, we present a patient with a primary renal synovial sarcoma presenting with spontaneous massive retroperitoneal bleeding due to tumor rupture. To the best of our knowledge, this is only the second case of such clinical presentation reported in the literature.

**Keywords:** Kidney neoplasms; synovial sarcoma; hemorrhage

Synovial sarcomas (SS) account for 5-10% of all soft tissue sarcomas. They usually occur between the third and fifth decades of life. In 65% of cases, they are located in the lower extremities, with the most common location being around the knee. Unusual areas of formation include the head, neck, heart, lungs, and kidneys.<sup>1,2</sup>

Primary renal SS are very rare and highly aggressive tumors with the common symptoms of side pain, abdominal pain, and hematuria. Low back pain, back pain, palpable mass, fever, and hypertension can also be seen, albeit rarely.<sup>3</sup> In the literature, only one case with renal rupture and spontaneous retroperitoneal hemorrhage was previously reported.<sup>4</sup> To our knowledge, ours is the second case with a similar clinical presentation.

In this case report, we aimed to draw attention to a condition that is often overlooked in retroperi-

toneal hemorrhage and evaluate renal SS in the light of literature.

## CASE REPORT

A 37-year-old male patient presented to the emergency department with left side pain, hematuria, fever and fatigue. There was no history of trauma. The results of his laboratory test were as follows: white blood cell count  $21.7 \cdot 10^3/\mu\text{L}$ , hemoglobin  $11.27 \text{ g/dL}$ , hematocrit 34.11, platelets  $267.7 \cdot 10^3/\mu\text{L}$ , urea  $57 \text{ mg/dL}$ , and C-creatinine:  $2.9 \text{ mg/day dL}$ . Due to the elevated creatinine levels, non-contrast abdomino-pelvic computed tomography was performed, which revealed a hematoma filling the retroperitoneum around the left kidney (Figure 1). The patient was admitted to hospital with the diagnosis of spontaneous massive retroperitoneal bleeding. Hemodynamic monitoring was provided. During the follow-up, he-

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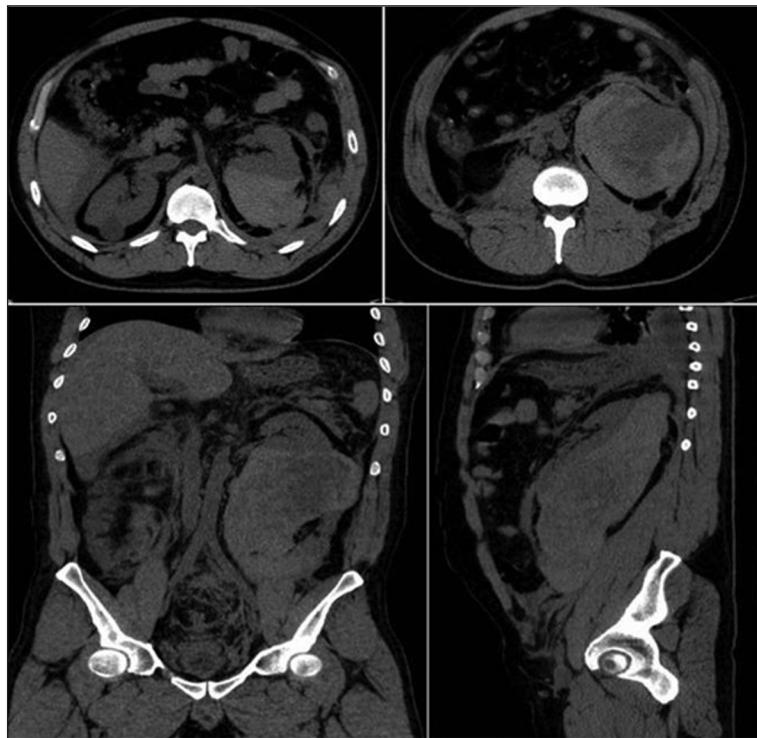


FIGURE 1: Non-contrast computed tomography images revealing a diffuse hematoma in the left retroperitoneal region.

moglobin was reduced to 6.9 g/dL and hematocrit to 20.52, and his hemodynamics were not stable. Since angioembolization could not be performed in our hospital, surgical exploration was planned and blood transfusion was undertaken. The retroperitoneum was reached by a flank incision and observed to have been completely filled with the hematoma. The kidney was released from the surrounding tissues, and nephrectomy was performed (Figure 2). When the nephrectomy material and hematoma were examined, tissue loss due to tumor rupture in the lateral of the middle pole of the kidney and tumor fragments within the hematoma were observed. During the follow-up, the hemodynamics stabilized, there was no additional problem, and the patient was discharged without any complication. The pathological diagnosis of renal SS was classified as the monophasic type. Necrosis was positive, cell type was spindle, mitosis was Grade 6, and Ki-67 proliferation index was 30%. Immunohistochemical (IHC) staining showed diffuse staining for CD56, TLE1, and bcl-2, focal weak positive staining for CD99, and diffuse positive staining for epithelial membrane antigen (EMA), vimentin and synapto-

phsin (Figure 3). The tumor had infiltrated the renal cortex but there was no perirenal fat tissue invasion. Clinical staging was performed, and no metastasis was detected. The patient was referred to an external center to receive adjuvant chemotherapy with a combination of ifosfamide and adriamycin. No local recurrence or distant metastasis was seen at the 36<sup>th</sup>-month follow-up. Written informed consent was obtained from the patient for publication of this case report.

## DISCUSSION

Primary renal SS are very rare tumors. Approximately 60 cases of primary renal SS have been reported in the literature since the first case published in 2000 by Argani et al.<sup>5</sup> These tumors are seen at a similar same rate in both sexes, and they usually occur at around 35 years of age. They usually present with common symptoms, such as side pain, abdominal pain, and hematuria, while rare symptoms include low back pain, back pain, palpable mass, fever, and hypertension.<sup>3</sup> Our case presented with massive retroperitoneal bleeding. If our hospital was equipped to perform angioembolization, this procedure would

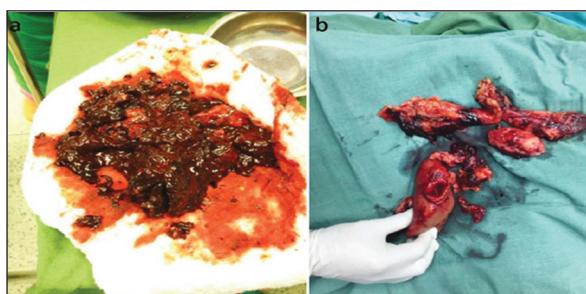


FIGURE 2: a) Hematoma; b) Nephrectomy material.

probably have been our first intervention. However, a successful angioembolization could have led to overlooking the diagnosis of renal SS.

SS can be confused with the Wilms tumor, Ewing sarcoma or sarcomatoid renal cell carcinoma, which have similar histological features in diagnosis.<sup>6</sup> Histologically, it can be classified as renal synovial sarcoma, biphasic synovial sarcoma, monophasic spindle synovial sarcoma, and monophasic epithelial synovial sarcoma. In our case, the pathological diagnosis of renal SS was classified as monophasic type. Necrosis was positive, cell type

was spindle, mitosis was Grade 6, and Ki-67 proliferation index was 30%. A definitive diagnosis is made by adjuvant immunohistochemistry, which is confirmed by molecular genetic analysis. The IHC analysis of primary renal SS shows diffuse staining for bcl-2, CD56, vimentin, and CD99 and focal positive staining for EMA.<sup>7</sup> Similarly, in our case, the IHC analysis showed diffuse weak positive staining for CD56, TLE1, and bcl-2, focal weak positive staining for CD99, and diffuse positive staining for EMA, vimentin, and synaptophysin. In most cases, molecular techniques, fluorescent in situ hybridization or polymerase chain reaction (PCR), or both can be used to confirm the diagnosis. Routine detection of t(X;18) translocation by PCR may provide useful additional assistance in the diagnosis of synovial sarcoma. This diagnosis can be confirmed by the detection of the t(X;18) (p11, q11) translocation using the reverse transcriptase PCR involving the fusion of SYT.<sup>8</sup> Genetic analysis could not be performed as we did not have the facility in our hospital.

Radical nephrectomy is recommended to relieve symptoms and control local disease. Local recurrence

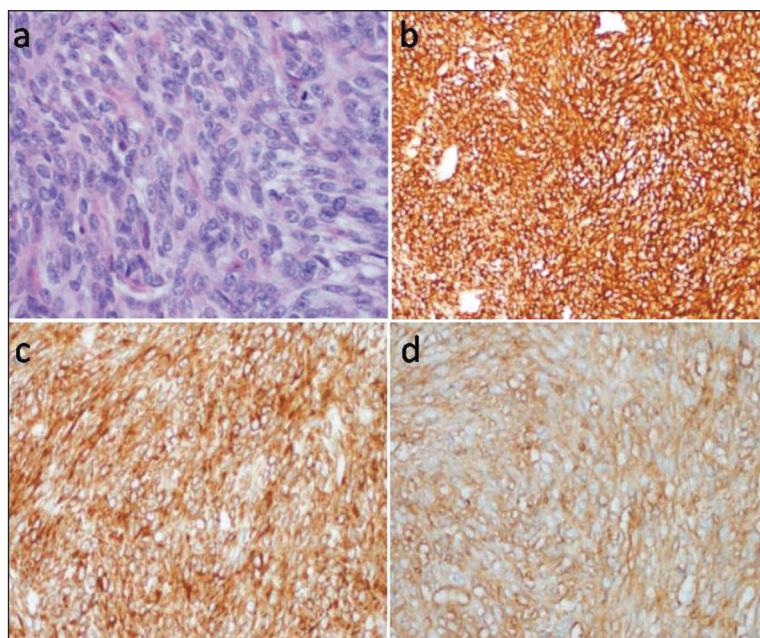


FIGURE 3: a) Immunohistochemical analysis revealing spindle/oval infiltration mostly consisting of uniformly chromatinated cells with a large nucleus and indistinct cytoplasm. Mitosis is seen in the top right corner (H&E, x400); b) Immunohistochemical vimentin positivity (Vimentin, x200); c) Immunohistochemical Bcl2 positivity (Bcl2, x400); d) Immunohistochemical CD99 positivity (CD99, x400).

or metastatic abdominal lymph nodes may occur in approximately one third of renal SS cases.<sup>7</sup> The role of adjuvant chemotherapy is controversial, but there are studies reporting a positive response to the combination of anthracyclines (e.g., doxorubicin) and ifosfamide in patients with metastatic disease.<sup>8</sup> Our case received four sessions of combined ifosfamide and adriamycin therapy at an external center. There is no standard treatment protocol for synovial sarcomas. Since there is no study showing the efficacy of radiotherapy in the treatment of primary renal synovial sarcoma, radiotherapy was not applied to our case.

The median overall survival in renal SS is only about 48 months. In our case, no local recurrence or distant metastasis was detected at the 24<sup>th</sup> month of follow-up.

In conclusion, renal SS are very rare and aggressive tumor that may present with massive retroperitoneal bleeding. Routine detection of t(X;18) translocation by PCR may provide useful additional assistance in the diagnosis of synovial sarcoma. Due to the rarity of the disease, no specific guidelines have been established for the treatment of renal SS. Radiotherapy is thought to be ineffective. The combination of radical nephrectomy and adjuvant

chemotherapy seems to be the best treatment option currently available.

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#### **Conflict of Interest**

*No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.*

#### **Authorship Contributions**

**Idea/Concept:** Ali Çift, Mehmet Özgür Yücel; **Design:** Ali Çift, Mehmet Özgür Yücel; **Control/Supervision:** Ali Çift, Mehmet Özgür Yücel, Can Benlioğlu, Bedreddin Kalyenci, Emine Kilinç; **Data Collection and/or Processing:** Bedreddin Kalyenci, Emine Kilinç; **Analysis and/or Interpretation:** Ali Çift, Mehmet Özgür Yücel, Can Benlioğlu, Bedreddin Kalyenci, Emine Kilinç; **Literature Review:** Ali Çift, Mehmet Özgür Yücel; **Writing the Article:** Ali Çift, Mehmet Özgür Yücel; **Critical Review:** Ali Çift, Mehmet Özgür Yücel, Can Benlioğlu, Bedreddin Kalyenci, Emine Kilinç; **References and Fundings:** Ali Çift, Mehmet Özgür Yücel.

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