

Primary Renal Synovial Sarcoma

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Abstract

Primary renal synovial sarcoma is a rare malignancy that may present similarly to other renal neoplasms. The diagnosis of synovial sarcoma is performed through the identification of a *SYT-SSX* gene fusion. Here, we present a case of a primary renal synovial sarcoma in a patient who presented with renal mass initially thought to be renal cell carcinoma until further pathological characterization. After undergoing radical open nephrectomy, the patient developed pulmonary and psoas metastases and was treated with systemic therapy.

Keywords: Synovial sarcoma, kidney neoplasm, *SYT-SSX*

Introduction

A synovial sarcoma is a malignant mesenchymal tumor that notably presents adjacent to large joints. A primary synovial sarcoma arising from the kidney is rare with limited reports in the literature. As such, it is unlikely to be on the differential diagnosis list of most urologists (1). More likely, the diagnosis is made upon renal mass biopsy or after surgical extirpation.

Characterization of the synovial sarcoma is performed histologically and at the molecular level through the presence of a *SYT-SSX* gene fusion of t(X;18)(p11.2;q11.2) (2). Because of the rarity of this malignancy, reports outlining the most effective management are scarce (3). Furthermore, due to the rarity of primary renal synovial sarcomas, descriptive case presentations can provide valuable insight into the clinical presentation and management of this malignancy to improve medical and surgical management (1).

Here, we present a case of a primary renal synovial sarcoma in a 35-year-old female patient who presented with a renal mass initially thought to be renal cell carcinoma, but was determined to be a primary synovial sarcoma upon pathological investigation. Informed consent by the patient to publish the details of this case was obtained.

Case Report

A 35-year-old female patient presented to her family physician with sudden and severe right-sided flank and abdominal pain. She underwent computed tomography scans revealing a 5.2 cm heterogeneous enhancing right-sided renal mass with associated retroperitoneal hematoma (Figure 1). A prominent retroperitoneal lymph node adjacent to the right ureter was present and appeared stable in size (8 mm) compared to previous imaging. The left kidney was unremarkable. Bloodwork noted an acute decrease in serum hemoglobin, which was treated with 1 U of packed erythrocytes. The patient denied constitutional symptoms such as weight loss or fever and denied any history of hematuria.

The patient's medical history was unremarkable apart from morbid obesity. A right renal biopsy was performed and the specimen was sent to pathology that detected a *SYT-SSX* translocation through a multiplex real-time polymerase chain reaction assay. Pathological investigation determined the presence of a primary monophasic synovial sarcoma with a mitotic rate of 21 per 100 high power field and a pathologic staging (pTNM) of pT1b. Immunohistochemical analysis demonstrated strong positivity for BCL-2 and moderate positivity for CD99 (Figure 2). Liver lesions discovered upon imaging were biopsied

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and characterized as benign and not metastases associated with the renal synovial sarcoma. No other sites of disease were identified, and after multidisciplinary discussions with medical oncology, radical open nephrectomy and a paracaval lymph node dissection were recommended.

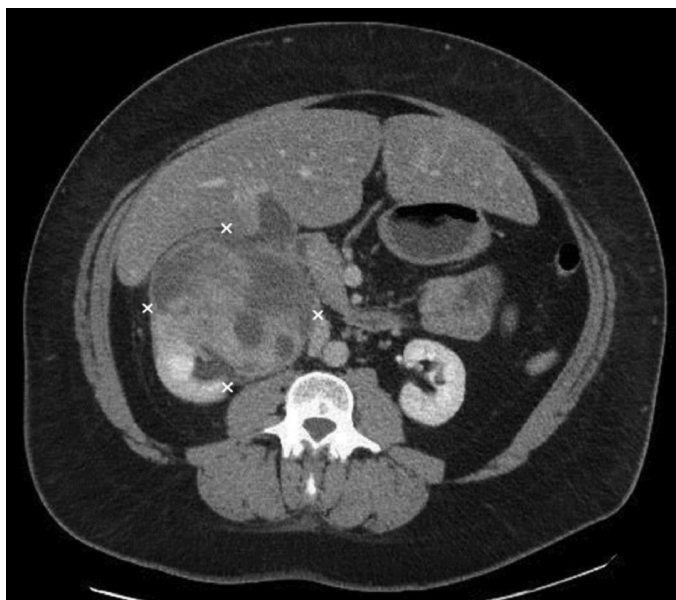


Figure 1. CT scan showing a right renal synovial sarcoma located within the collecting system of the upper, middle, and lower pole. A right perinephric hemorrhage with mass effect on the kidney and right ureter was also noted

A midline laparotomy approach was used with a full retroperitoneal exposure, including mobilization of the root of the mesentery and temporary evisceration of the bowel. A significant mass effect on the vena cava with dense adhesion was noted. At the level of the renal hilum, an excision of the renal vein os with primary closure of the inferior vena cava was performed. A paracaval lymph node dissection was then performed.

Tumor dimensions were 13.5 cm x 12.5 cm x 8.5 cm, showing rapid growth in the 3 months between initial imaging and surgery. Tumor involvement of the collecting duct region with extension into the perinephric fat and the renal vein was found. Surgical margins were negative for tumour involvement. The paracaval lymph nodes were negative for metastasis.

In hospital, the patient recovered uneventfully and was discharged home on post-operative day 5. Unfortunately, 6 months after surgery on the first surveillance imaging, multifocal pulmonary metastases developed and the patient was started on doxorubicin single-agent systemic therapy. After 5 cycles of doxorubicin treatment with an unsatisfactory response, the patient began single-agent ifosfamide treatment. Fifteen months after surgery, a right psoas metastasis developed and the patient was started on gemcitabine and docetaxel. Twenty-four months after surgery the patients remain alive and with stable disease.

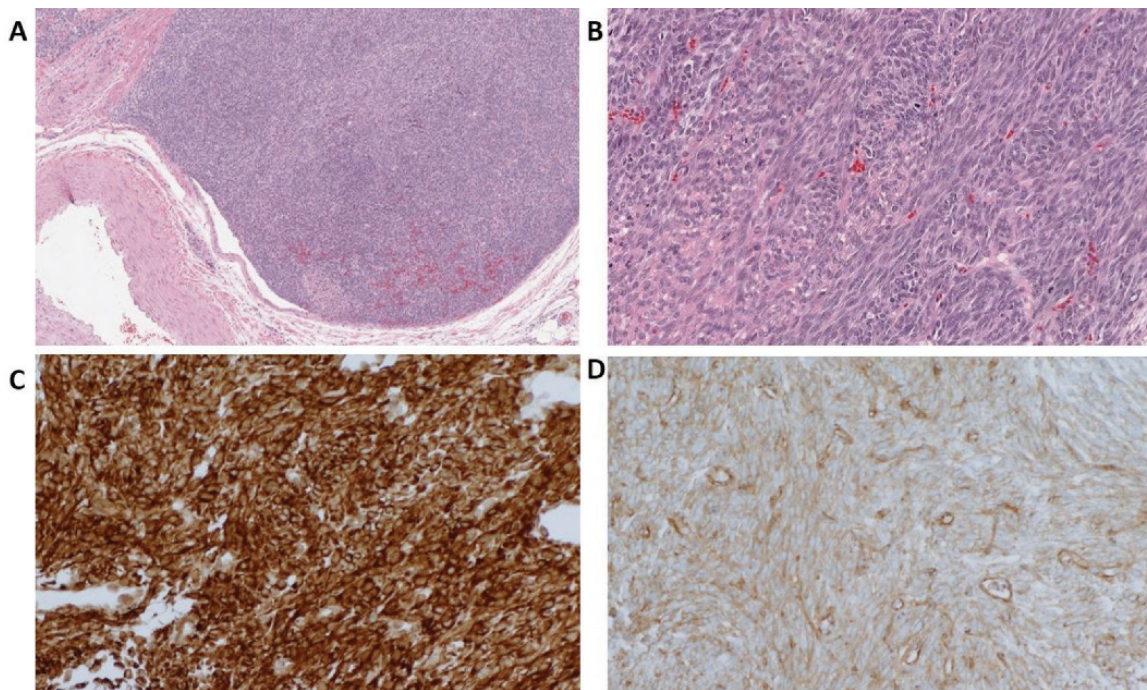


Figure 2. (A) Renal synovial sarcoma (4x HE Stain) showing interlacing fascicles of spindle neoplastic cells with high mitotic index and tumor invasion into a renal vein branch. (B) Dot-like keratin expression without distinct epithelial component, histologic (FNCLCC) grade of 3 (Differentiation score: 3, Mitosis score: 3, Necrosis score: 1) (20x HE Stain). (C) Strong BCL-2 expression (membranous and cytoplasmic pattern) detected in the tumor cells (100x magnification). (D) Moderate level of CD99 expression (membranous pattern) detected (100x magnification)

Discussion

Primary renal synovial sarcoma is a rare malignancy that presents similarly to a renal cell carcinoma and is associated with high rates of metastasis (3). The median age for diagnosis is 36.5 years with a male-to-female ratio of 1:1 (1,3). As this median age is approximately half that of renal cell carcinoma (61 years), there is an increased relative likelihood of renal synovial sarcoma being the cause of malignancy in younger patient presentations (4). The median overall survival in patients who underwent radical nephrectomy is 48 months (3).

The initial clinical presentation in cases of primary renal synovial sarcoma has been described to include abdominal or flank pain (67%) and hematuria (38%) (3,5). Here, our patient presented initially with abdominal and flank pain, which progressed to life-threatening hemorrhage.

As the clinical and radiological presentation of synovial sarcoma is indistinguishable from other renal malignancies, the gold standard for diagnosis is through pathologic examination demonstrating a *SYT* gene translocation (1). Metastatic disease upon diagnosis is rare (8%), although there is an increased incidence of metastasis post-nephrectomy (36%) with the median time of metastasis development occurring 33 months post-operation (3). Additionally, the most common sites of metastasis post-nephrectomy include the lung (42%), abdominal lymph nodes (29%), and liver (24%), highlighting the importance of follow-up investigations in patients with this malignancy (1,3).

For the treatment of primary synovial sarcoma without evident metastatic disease, radical nephrectomy is the first-line approach to attempt to achieve local control and reduce metastasis and recurrence risk. For metastatic cases, surgical resection and chemotherapy (ifosfamide and doxorubicin) has shown success, with meta-analyses highlighting the efficacy of doxorubicin in reducing overall recurrence, promoting remission and reducing tumour volume (6). Although studies have shown success in tumour-volume reduction, there is controversy regarding the impact of overall survival in patients undergoing adjuvant chemotherapy due to the sparsity of randomized controlled trials (7). Recently, immunotherapy has shown promise for the treatment of synovial sarcoma (8). For metastatic or surgically unresectable locally advanced sarcoma, pembrolizumab, an anti-PD-1 monoclonal antibody, has demonstrated a 10% objective response rate in a phase II trial (9). For patients with advanced synovial sarcoma who have progressed on other approved therapies, targeted therapies such as pazopanib have demonstrated non-inferior progression-free survival and similar overall survival compared with doxorubicin (10).

Conclusion

This case describes a rare primary renal synovial sarcoma in a 35-year-old female presenting with retroperitoneal hemorrhage that was initially thought to be renal cell carcinoma. These rare malignancies can pose surgical challenges and have a high potential for metastatic progression. Prompt extirpation of the localized disease provides the best chance for cure.

Ethics

Informed Consent: Informed consent by the patient to publish the details of this case was obtained.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: R.A.R., B.C., C.W., K.D.H., J.M., R.J.M., Concept: W.M., R.A.R., B.C., K.D.H., Design: W.M., K.D.H., Data Collection or Processing: W.M., C.W., K.D.H., Analysis or Interpretation: W.M., R.A.R., B.C., C.W., K.D.H., Literature Search: W.M., Writing: W.M., C.W., K.D.H.

Conflict of Interest: No conflict of interest was declared by the authors.

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