CASE REPORT

# Radical Nephrectomy of a Huge Renal Mass Adherent to the Liver, Diaphragm and Aorta: The Challenges in the Identification and Management of Synovial Sarcoma of the Kidney

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Synovial sarcoma (SS) is a mesenchymal tumor that more rarely affects the kidneys. As of 2020, only around 100 cases of renal SS have been reported. Herein described is a 28-year-old female with a huge mass occupying almost the entire abdomen. Her history, physical examination and imaging suggested a renal cell carcinoma with metastases to the diaphragm, liver and large vessels. A radical nephrectomy was successfully performed, removing an enormous 26-kilogram, 52cm x 37cm x 14cm right renal mass and providing immediate relief. Histopathology surprised the authors with findings of extra-gastrointestinal stromal tumor. 20 months post-surgery, there was tumor recurrence, and a metastasectomy was done. Immunostaining revealed renal synovial sarcoma. To date, the patient remains asymptomatic and disease free. The rarity of renal SS, as well as its ability to mimic more common tumors, makes clinicopathological diagnosis and management difficult. Moreover, the role of chemotherapy for SS remains unclear.

***Keywords***: synovial sarcoma of the kidney, renal tumor, primary renal synovial sarcoma

## Introduction

Renal Cell Carcinoma (RCC) make up the majority of malignant neoplasms of the kidneys, accounting for 95% of all renal cancers.[1] There are, however, other uncommon forms that may be mistaken for renal cell carcinoma in terms of presentation and progression. One of these is soft tissue sarcoma. These soft tissue sarcomas are rare mesenchymal tumors comprising less than 1% of all malignant tumors. Of these, synovial sarcoma (SS) represents only 5% and more commonly develops from the proximal limbs of young adult males (Table 1).[2,3] Primary renal SS is an even rarer entity. It makes up less than 1% of synovial sarcoma and less than 2% of all renal neoplasms. To date, there are only about 100 published cases of SS of the kidney. Presented here is a curious case of a patient with a painless but rapidly enlarging abdomen which clinically appeared to be RCC and histochemically as extra-gastrointestinal stromal tumor (eGIST). Immunohistochemical studies, however, revealed renal SS. Currently, there is no standard protocol on the diagnosis and management of renal SS, due to 1) its rare nature, and 2) its diagnosis only after immunohistochemical and genetic studies. As it is commonly mistaken for other urologic malignancies, specifically RCC, radical surgical excision remains as the standard of care. This ensures complete tumor removal thus decreasing the incidence for relapse and recurrence. This case may provide the medical field with more data and insights on the diagnosis and management of renal SS.

**Table 1**.Distribution of synovial sarcoma with its major subtype (2).

| Site | % | Predominant Type |
|---|---|---|
| Extremity | 45% | Malignant Fibrous Histiocytoma |
| Retroperitoneal/ Intra-abdominal | 27% | Leiomyosarcoma |
| Visceral | 13% | Leiomyosarcoma |
| Thoracic | 9% | Malignant Fibrous Histiocytoma |
| Head and Neck | 6% | Rhabdomyosarcoma |

## The Case

A 28-year-old female came in due to a 5-month long history of a palpable right upper quadrant mass, initially described as the size of a tennis ball, with associated weight loss. There was no abdominal pain, hematuria, jaundice or dyspnea. No consultation was done at the time. She is a non-smoker with no known associated co-morbidities. Family history revealed no history of cancer. The interim revealed rapidly progressive enlargement of the mass, with eventual exertional dyspnea and abdominal fullness. This prompted consultation. Patient was noted to be hyposthenic, with a height of 168cm and weight of 66kg. Her vital signs were in the normal range. Physical examination revealed a 41cm x 50cm firm, non-movable, abdominal mass, as shown in Figure 1. A grade 1 pitting edema of the right foot was also noted. An abdominal CT scan with contrast was done, revealing a 30cm x 19cm hypodense, heterogeneously enhancing right renal mass arising from the superior and middle poles (Figure 2). This exhibited a claw sign (Figure 2), a radiologic finding in renal masses distinctive of renal papillary necrosis. Furthermore, the right adrenal gland was not visualized, there was note of leftward shift and questionable infiltration of the inferior vena cava (Figure 2), leftward deviation of the left lobe of the liver, infiltration of the right lobe of the liver (Figure 2) and a suspicious extension of the mass through the right hemidiaphragm. At this point, a clinically stage IV RCC was considered. As this only has a 5-year survival rate of 0-10%, cytoreductive nephrectomy was advised.[1] The patient then was apprised for an exploratory laparotomy with either radical nephrectomy, right with possible metastasectomy from the liver, diaphragm and IVC, or simple biopsy if the mass was deemed unresectable. Intraoperatively, a midline incision was done, revealing a mass measuring 50cm x 40cm, as shown in Figure 3. The liver was displaced to the left subphrenic space and the spleen inferior to it. While the mass was noted to be well-circumscribed, it seems to have infiltrated the right lobe of the liver (Figure 3).

Furthermore, it was noted to be densely adherent to the right hemidiaphragm with no apparent extension through the latter. Likewise, the IVC and aorta were spared from tumor extension. Inferiorly, the inferior pole of the already inferiorly displaced right kidney is seen. Figure 8 shows the middle and superior pole are no longer appreciated with the mass in its place. This large mass was mobilized from its adhesions and successfully removed, providing immediate relief (Figure 4).

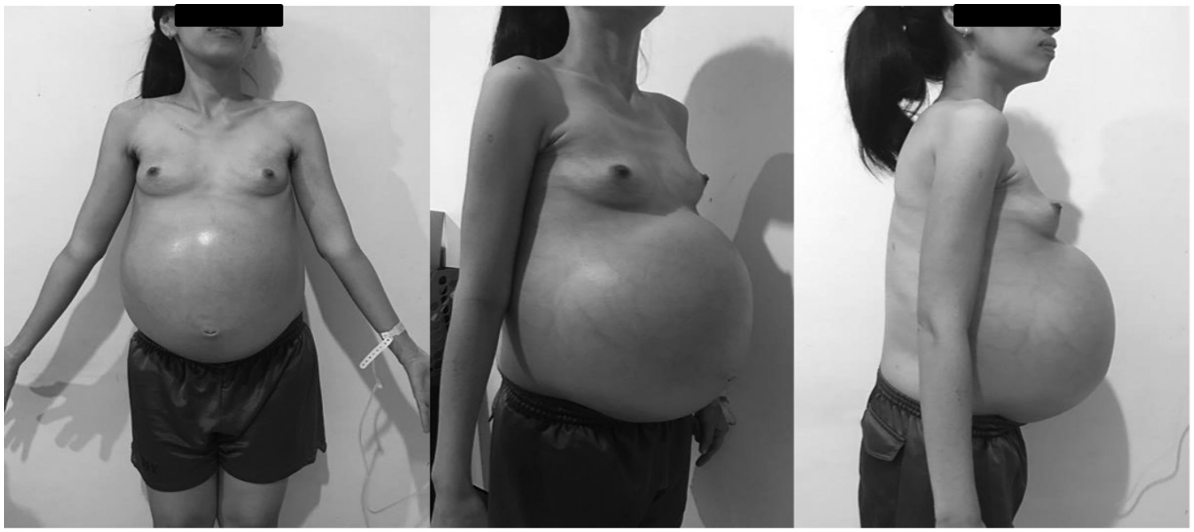

**Figure 1**. This 28-year-old female who presented with a 41cm x 50cm abdominal mass.

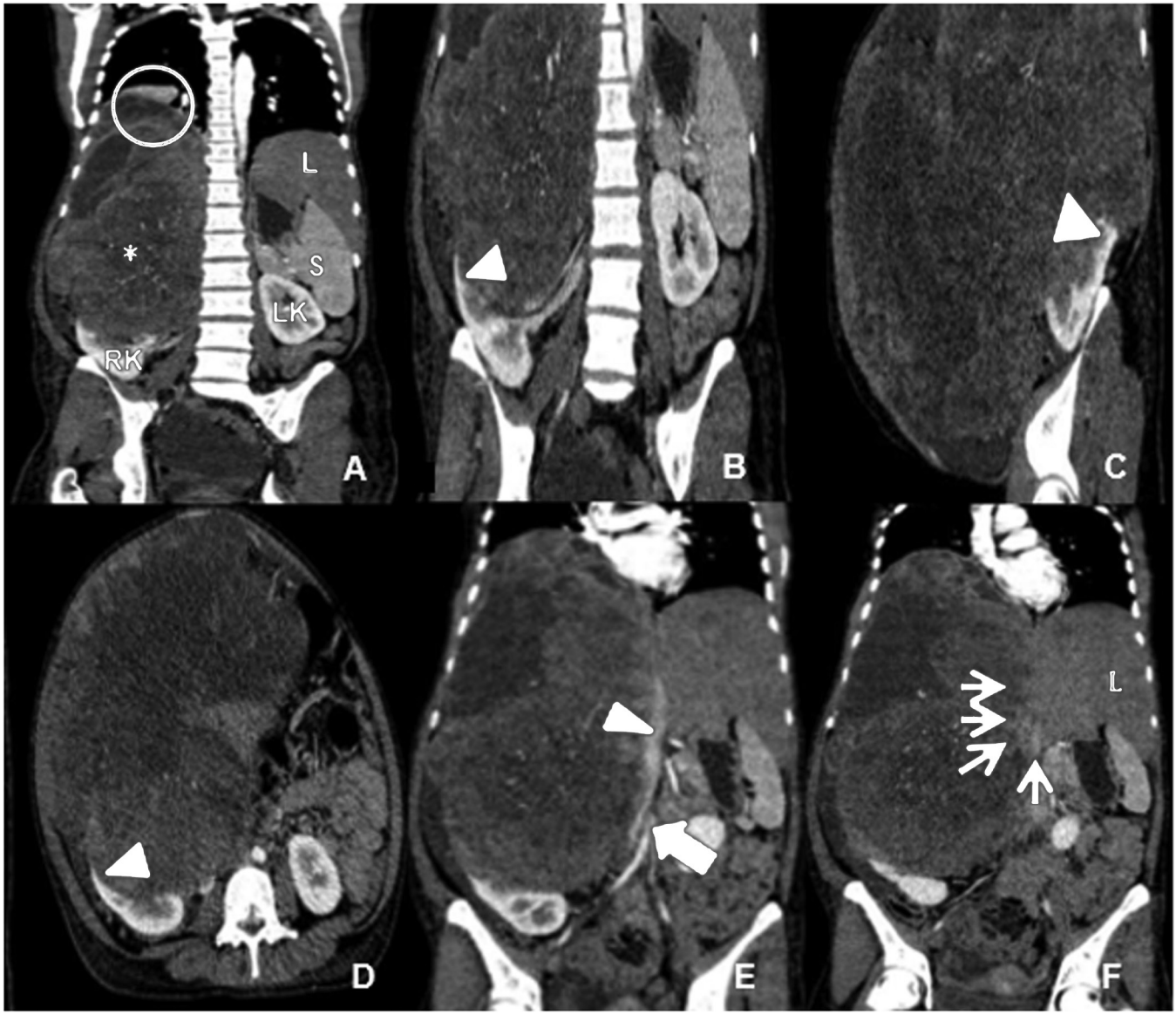


**Figure 2**. CT of the abdomen with contrast. A) CT scan with contrast revealed a mass 30cm x 19cm heterogeneously enhancing mass (*) arising from the superior and middle pole of the inferiorly displaced right kidney (RK). Note a suspicious infiltration into the right hemidiaphragm (encircled) with the liver shifted to the left hemiabdomen (L) thereby relocating the spleen (S) below it. The left kidney (LK) also appears to be shifted inferiorly, noted between L2 and L4. B) Radiographic findings suggesting that the mass originated form the kidney (arrow heads). C) Sagittal section showing the inferior vena cava (arrow head) and abdominal aorta (arrow) has been displaced to the left hemiabdomen. Alos note that the vena cava is compressed. D) Sagittal section shows no definite distinction betwee the mass and the right lobe of the liver (L).

Partial resection of the right lobe of the liver was done, with inadvertent injury to the right hemidiaphragm as well as the IVC. Primary repair was done and a chest tube was inserted. Intraoperative blood loss was at 2,500mL with episodes of hypotension noted during removal of the mass. Volume resuscitation with intravenous fluids, inotropes and blood transfusion was successful. Post operatively, the patient was placed in the SICU where she was monitored for 10 days. She was weaned off the ventilator and inotropes and her chest tube was removed.

Gross examination revealed a 52cm x 37cm x 14cm right kidney and weighed 26 kilograms. Cut section (Figure 5) shows a well-demarcated pale to whitish gray solid tumor with several nodular as well

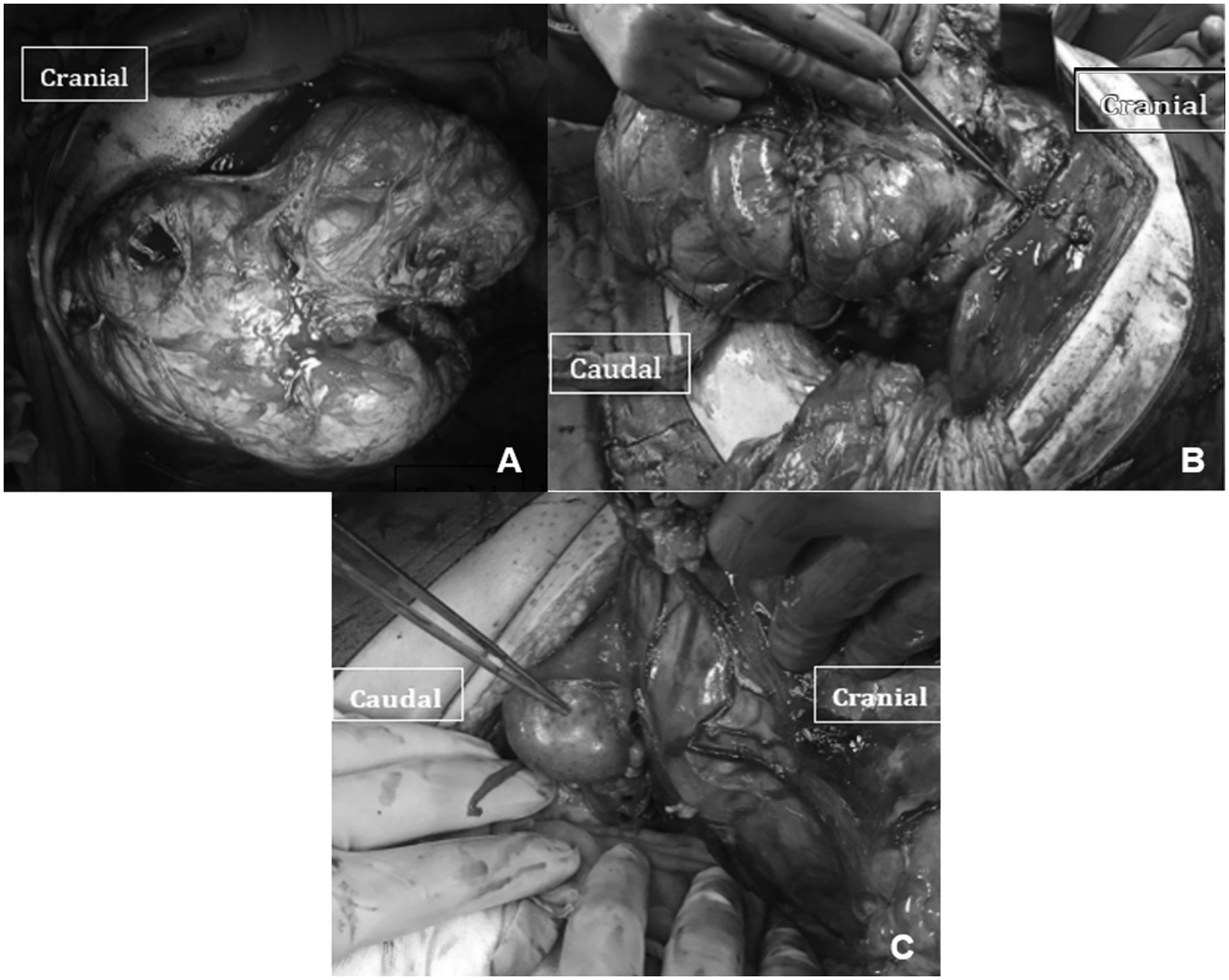


**Figure 3**. Gross findings. A) A huge 50cm x 40cm mass occupying the entire abdomen. While dissection proved to be difficult at the level of the right hemidiaphragm, visually, there was no apparent infiltration through it. B) The superior aspect of the mass was especially adherent to the liver with note of infiltration at the lateral and posterior aspect of the right lobe of the liver. C) An inferiorly displaced normal looking pole of the right kidney is seen with the mass arising from the middle and superior pole of the same kidney.

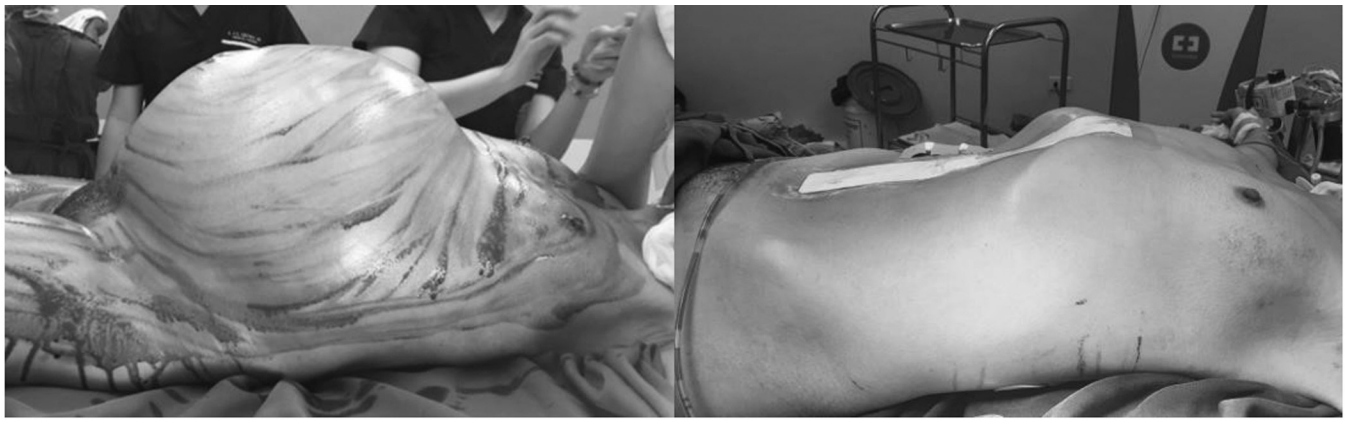

**Figure 4**. Pre- and post-operative photo of the patient's abdomen

as cystic areas occupying almost the entire kidney. Hematoxylin and eosin staining revealed multiple sarcomatous elongated spindle cells with mitotic figures (Figure 6). Surprisingly, these findings were suggestive of eGIST.

The patient made a full recovery and was eventually discharged stable and improved on the 21st post-operative day. While she agreed to follow up, the patient refused further investigation. The interim was unremarkable until 20 months postsurgery when repeat imaging revealed a 24cm tumor recurrence in the right renal fossa (Figure 7). A metastasectomy was done.

Intraoperatively, a midline incision was done, revealing a retroperitoneal mass measuring 35cm x 17cm. The mass was noted to have infiltrated through the right hemidiaphragm. Still, the IVC and aorta appeared to be spared from tumor extension. Posteriorly, it was densely adherent to the psoas muscle and retroperitoneal bed. Mobilization from its adhesions, as well as a right diaphragmatic full thickness resection with pleurectomy and mesh

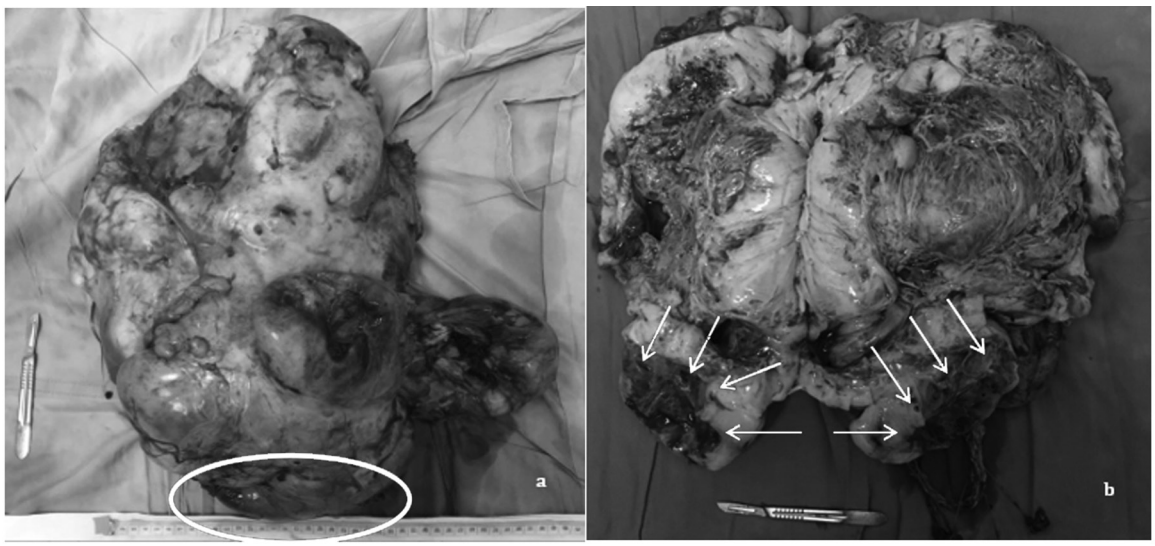


**Figure 5**. The gross specimen. A) An enormous 52cm x 37cm x 14cm mass was removed. Note the normal looking inferior pole of the right kidney (encircled) while the middle and superior poles have evolved into part of the mass. B) On cut-section, the mass is observed to affect all three poles of the right kidney, leaving few normal-looking parenchyma.

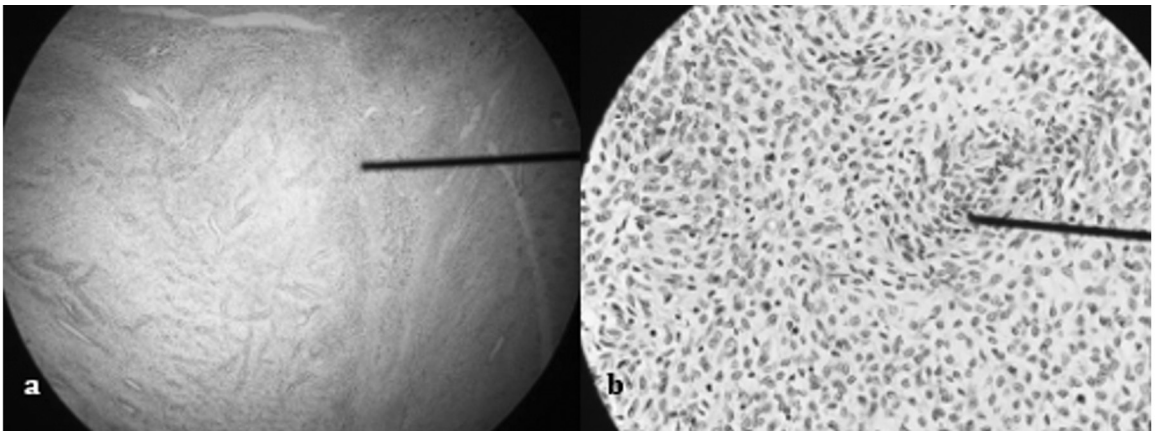


**Figure 6**. Microscopic evaluation of the specimen. A) multiple sarcomatous spindly elongated cells in a richly cellular network, and B) dark blue mitotic figures.

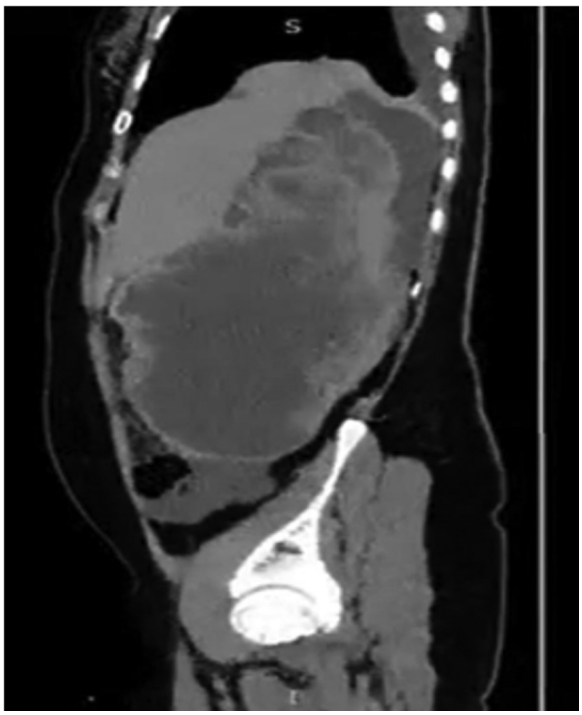

**Figure 7**. Repeat CT scan imaging revealed a 24cm x 13cm x 15cm mass displacing the liver anteriorly.

repair was done and this large mass was successfully removed.

A chest tube was inserted on the right. Intraoperative blood loss was at 1000mL. Volume resuscitation with intravenous fluids, inotropes and blood transfusion was successful. Postoperatively, the patient was placed in the SICU where she was monitored for 7 days. There she was weaned off the ventilator and inotropes and her chest tube, removed.

Gross examination revealed a 35cm x 17cm x 15cm retroperitoneal mass and weighed 12 kilograms. Cut section (Figure 8) shows a pale to whitish gray solid tumor with several nodular as well as cystic areas. Differential diagnosis at this time included Sarcomatoid RCC, Hemangiopericytoma, Leiomyosarcoma, eGIST and primary renal SS. Immunohistochemical staining was done, revealing monophasic synovial sarcoma of the kidney (Table 2).[4] Thirty-two months post-radical nephrectomy and 12 months post-metastasectomy, the patient was stable with neither evidence of metastasis nor recurrence. She was set to undergo repeat laboratories and imaging studies on her continued follow-up.

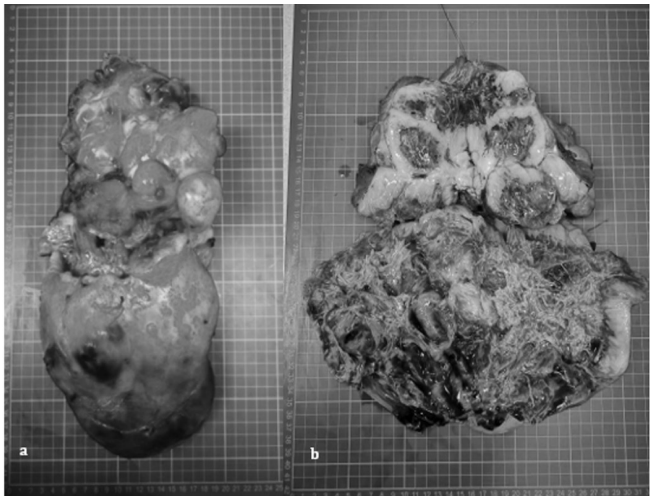


**Figure 8**. Gross and cut section of the recurrent mass. A) Gross and B) cut-section shows a 35 cm well-delineated, nodular, predominantly solid mass with cystic areas as well as tumor necrosis.

**Table 2**. Immunochemical staining results.[4,17]

| Stain | Tumor | SS | eGIST | Sarcomatoid RCC | Heman-giopericytoma | Leiomyosarcoma |
|---|---|---|---|---|---|---|
| Vimentin | + | + | + | + | + | + |
| Cytokeratin | - | - | - | + | - | + |
| EMA | + | + | +/- | + | - | - |
| CD-99 | + | + | + | - | - | +/- |
| BCL-2 | + | + | + | - | +/- | +/- |
| TLE-1 | + | + | - | - | +/- | - |
| Desmin | - | - | +/- | - | - | + |
| Myogenin | - | +/- | + | - | - | - |
| S-100 | - | - | - | - | - | +/- |
| CD-34 | - | - | + | - | + | +/- |
| RCCA | - | - | - | + | - | - |
| CD-117 | - | - | + | - | - | - |

## Discussion

SS are relatively uncommon, making up only 5% of all soft tissue sarcomas. These are usually seen in the lower extremities of young males. Other sites include the retroperitoneum, thorax, head and neck and viscera (Figure 9). SS primary to the kidney is an even rarer condition, accounting only <1% of all SS and <2% of renal cancers.[5,6,7] Since its discovery by Argani in 2000.[8], only about 100 cases have been published. To the authors' knowledge, this is the first case reported in this institution, the first in Mindanao and only one other case noted in the Philippines.[9] Of all published cases, the average age at diagnosis was 40 years with a similar incidence between genders.[10,11] This is in stark contrast to RCC's onset at a later age (55-75 years) with a male predominance.[1] The most frequent presenting symptom is flank pain, followed by hematuria and a palpable flank mass.

In this case, a rapidly enlarging abdominal mass was the chief complaint. On imaging studies, a renal SS is often seen as large, well-delineated heterogeneously enhancing, soft tissue lesion that may extend into the renal pelvis. These tumors present as predominantly cystic with solid components.[12,13] These findings, however, prove to be non-specific as other tumors may present similarly. This is also the reason why this was managed as a case of RCC, the most common renal tumor. Renal SS are predominantly large tumors, with a mean size of 8cm (519cm). Grossly, they appear as well-delineated, tan and rubbery tumors with admixed multilocular cysts. Necrosis and hemorrhage are predominant.[14] At 52 cm wide and weighing a massive 26 kilograms, this case is the largest reported case to date.

Histologically, renal SS arises from mesenchymal cells and consists of either spindle cells only or epithelial cells only(monophasic), both spindle and epithelial cells (biphasic) or may be of poorly-differentiated cells.[15] The presence of elongated spindle cells with a mitotic rate of >5/50 hpf was what prompted the pathologist to identify the present case as a high risk eGIST. While there have been reports of extra-gastrointestinal stromal tumors (eGIST), none have been reported to arise from the kidney.[16] Furthermore, the clinical profile of

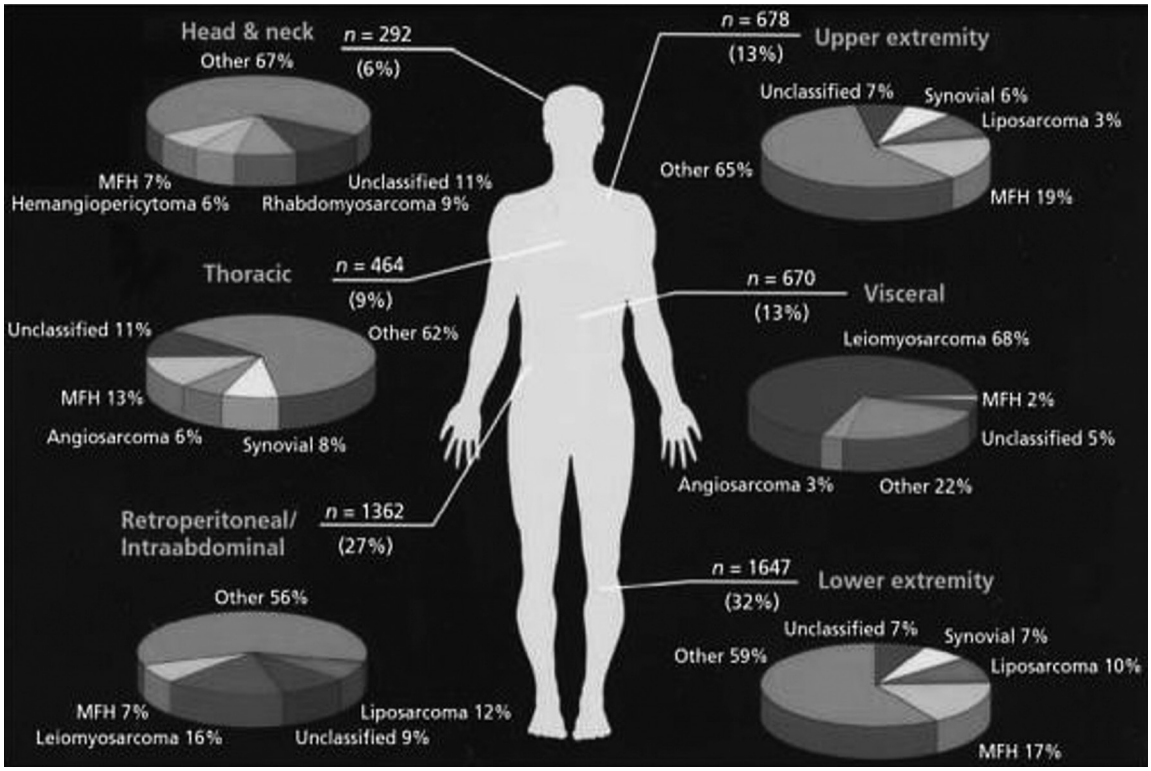


**Figure 9**. Anatomic distribution with histologic subtypes of patients seen at Texas MD Anderson Cancer Center (June 1996- June 2005)

eGIST is non-specific as well, hence the decision to push through with immunohistochemical studies. As monophasic renal SS share a myriad of clinicohistopathologic features as other tumors, immunostaining can aid in the differential diagnosis (Table 2). Renal SS stains positively for TLE-1, CD99, vimentin, EMA, Bcl-2 and does not stain for cytokeratin, S100, CD34. eGIST stains for CD34, CD117, making its diagnosis unlikely. Hemangiopericytoma in contrast to SS, has no mitotic activity, and is CD34 positive. Sarcomatoid RCC stains positively for EMA, vimentin and RCCA, but not TLE-1. Leiomyosarcoma is positive for desmin and keratin, may be positive S100 and CD34, and does not stain for CD117.[17,18,19] Confirmation of primary renal SS is done by cytogenetic analysis through reverse transcriptase polymerase chain reaction (RT-PCR). Renal SS exhibits a translocation between the SYT gene on chromosome 18 and either SSX1 or SSX2 on chromosome Xp11.[20] However, a negative RT-PCR does not totally discount a diagnosis of renal SS. In a review by Argani, only 4 out of 15 cases displayed the SYT-SSX fusion transcripts. Overall, about 90% of renal SS showed the t(X;18)(p11.2,q11.2) transcription.[8] For the present case, albeit still with some unanswered questions, there is strong evidence to support a diagnosis of primary renal SS.

Due to its rarity, there are no current guidelines in the management of SS. Complete surgical resection of the tumor seems to be the mainstay management. In this case, pre- and intra-operative assessment leaned more towards a renal cell carcinoma, and was managed with radical nephrectomy, the same procedure had this been pre-identified as a soft tissue sarcoma. Nevertheless, recurrence rates are high, and prognosis is poor. Even with negative surgical margins, up to 50% of patients developed distant metastasis, usually to the lungs and liver. 5-year survival rate was estimated to be at 25%.[21]

High dose ifosfamide-based adjuvant chemotherapy has been proven to be chemosensitive in some soft tissue tumors and shows promise in preventing recurrence and improving relapse-free survival in renal SS. Several studies have even confirmed complete remission with doxorubicin and ifosfamide. However, these studies have yet to be established into existing protocol and require more data and research.[22]

## Conclusion

Primary renal synovial sarcoma is an extremely rare tumor that can mimic other more common tumors, making diagnosis extremely difficult. To date, there is no specific protocol in the management of renal SS. As such, radical nephrectomy appears to be the cornerstone of management. Chemotherapy in the form of ifosfamide has shown promising results, but no definite protocol has been established. With the increasing literature on SS, there might be hope in better understanding these patients prior to surgery.

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