Case Report

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Urology Section

Management of Metastatic Primary Renal Synovial Sarcoma with Inferior Vena Cava Thrombus: A Rare Case Report

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ABSTRACT

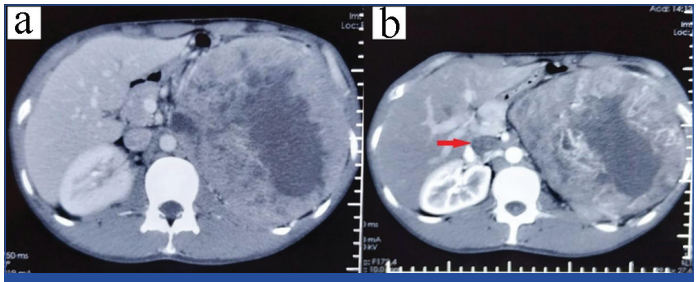
Synovial sarcomas constitute 5-10% of all soft-tissue sarcomas and tend to occur in the extremities, particularly the lower extremity. Renal sarcoma is a rare entity, constituting approximately 1-5% of all malignant renal tumours. Metastatic large renal synovial sarcoma with Inferior Vena Cava (IVC) thrombus is a rare surgical challenge that demands thorough preoperative evaluation and planning for its management. A 31-year-old young male presented with symptoms of left flank pain for 10 days, with a past history of similar complaints two years prior. On examination, a massively large renal mass was palpable. A Computed Tomography (CT) scan showed an IVC thrombus and metastatic deposits. The patient underwent a radical nephrectomy with ileal segment resection, followed by chemotherapy, which showed near-complete resolution of the metastatic deposits. Renal synovial sarcoma should be considered as a differential diagnosis in young patients presenting with a metastatic, exceptionally large renal mass complicated by an IVC thrombus, necessitating thorough preoperative evaluation for effective management.

Keywords: Kidney, Maligancy, Metastasis, Thrombosis

CASE REPORT

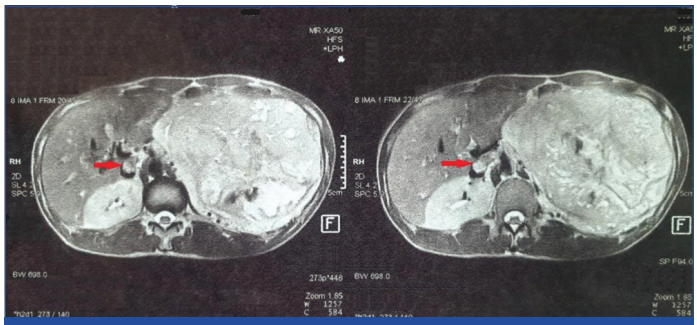
A 31-year-old male presented to the urology department of a tertiary care hospital with the chief complaint of left flank pain for the past 10 days. The pain was continuous and dull aching, radiating to the back and relieved by taking analgesics. The patient did report significant weight loss and anorexia, but there was no history of haematuria, bony pain, cough, or haemoptysis. There was a history of similar complaints two years prior, during which the patient underwent an abdominal ultrasound that revealed a small left renal mass, but the patient ignored the advised treatment.

On physical examination, a left hypochondrial mass extending to the epigastric and umbilical region was palpable, along with left flank fullness. The patient also had a left-sided grade 3 varicocele. His renal function tests were within normal limits; however, the patient had a low Body Mass Index (BMI) of 17.6 kg/m^2. Radiological investigations, including ultrasonography and Computed Tomography Intravenous Urography (CT IVU), suggested an exophytic predominantly solid mass lesion measuring 20×14 cm arising from the interpolar and lower pole of the left kidney, with central necrosis likely invading Gerota's fascia, causing a mass effect on the renal hilum with a supero-medial displacement of the left renal vessels. Additionally, a thrombus was observed in the left renal vein, extending into the IVC, reaching up to the infrahepatic IVC [Table/Fig-1a,b].



[Table/Fig-1]: a) CT axial cut section showing left large renal mass; b) Inferior Vena Cava (IVC) tumour thrombus (arrow).

Magnetic Resonance Imaging (MRI) with Magnetic Resonance Angiography (MRA) was performed to study the extent of the thrombus. The imaging showed the left renal vein displaced superiorly and medially with a filling defect extending into the IVC for a length of 4.5 cm and a thickness of 1.1 cm, reaching up to the mid part of the intrahepatic IVC [Table/Fig-2].

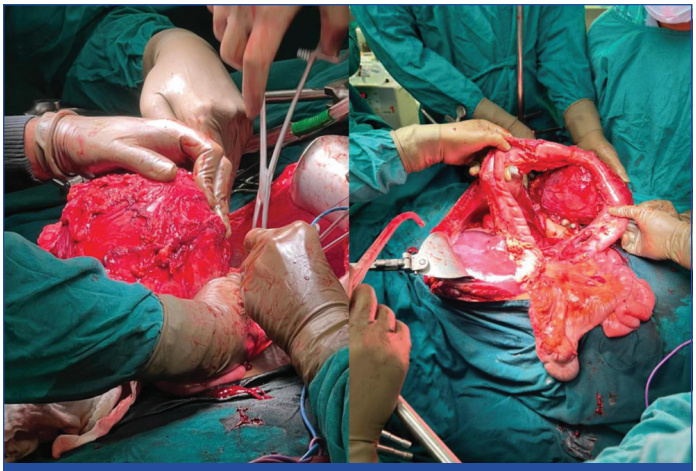


[Table/Fig-2]: MRI showing massive renal mass with tumour thrombus (arrow) extending into IVC and right renal vein.

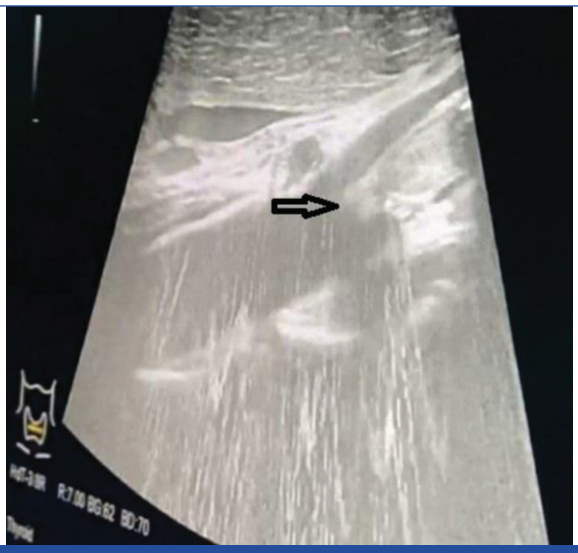
FDG-PET (18-Fluoro-deoxyglucose Positron Emission Tomography) revealed a few nodules in both lungs, FDG avid deposits in both the lobes of the liver, perisplenic deposits, and a few lytic skeletal lesions suggestive of metastatic renal cell carcinoma. After thorough counseling with the patient's relatives and obtaining a medical oncology opinion, the decision was made to proceed with upfront palliative surgery.

During laparotomy, a circumscribed left renal mass abutting the anterior abdominal wall was mobilised inferolaterally and superiorly after opening the line of Toldt's fascia. The mass was freed of its adhesions posteriorly up to the renal hilum after lifting it medially, and the left renal artery was dissected. The transverse and splenic flexure of the colon, along with its mesentery, approximately 10 cm in length, was found adhered to the anterior wall of the mass and was excised [Table/Fig-3].

The retrohepatic IVC was isolated after mobilising the right lobe of the liver using the Cattell-Braasch maneuver. An intraoperative ultrasound was performed to delineate the proximal extent of the thrombus and rule out IVC invasion [Table/Fig-4]. The thrombus was seen extending cranially up to the infrahepatic IVC. The infrarenal IVC, right renal vein, and suprahepatic IVC were clamped sequentially using vascular clamps after passing vascular loops, taking care not to dislodge the thrombus. Cavotomy was performed,

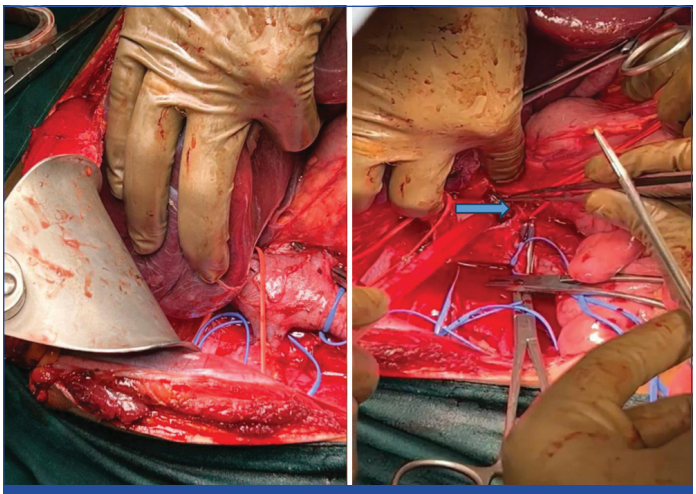


[Table/Fig-3]: Intraoperative tumour dissection with adherent colon and mesentery.



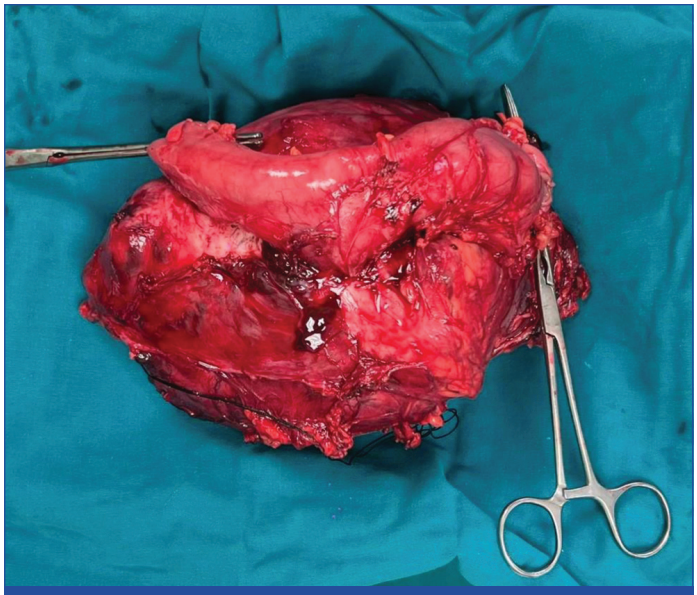
[Table/Fig-4]: Intraoperative 2D ECHO- delineating the proximal extent of IVC thrombus (arrow).

and the thrombus was removed in toto, following which IVC repair was done using 6-0 Prolene after flushing with heparinised saline. Radical nephrectomy with thrombectomy was performed, followed by regional lymphadenectomy and ileal stoma formation [Table/Fig-5,6]. The operative time was 180 minutes with an approximate blood loss of 500 mL.



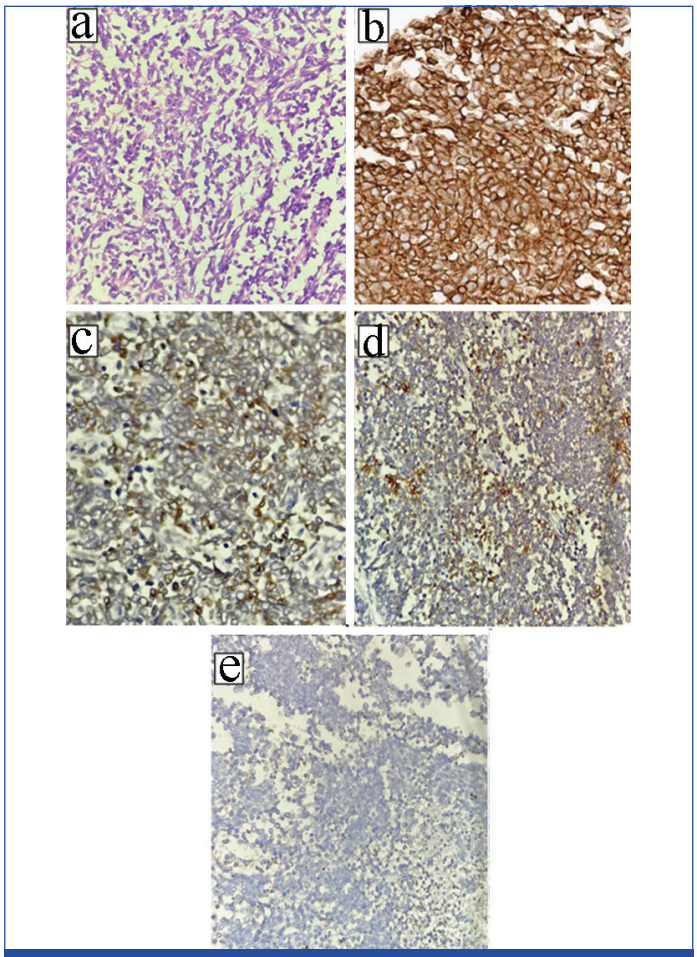
[Table/Fig-5]: Inferior Vena Cava (IVC) with its branches.
[Table/Fig-6]: Tumour thrombus extraction. (Images from left to right)

Grossly, a well-circumscribed tumour was noted in the mid and lower pole of the kidney, measuring 20×16.5×10.7 cm. The cut-section showed a greyish-white, friable mass with focal areas of haemorrhage and necrosis, weighing approximately 2.75 kg [Table/Fig-7]. The renal vein tumour thrombus measured 3×2×1.6 cm.



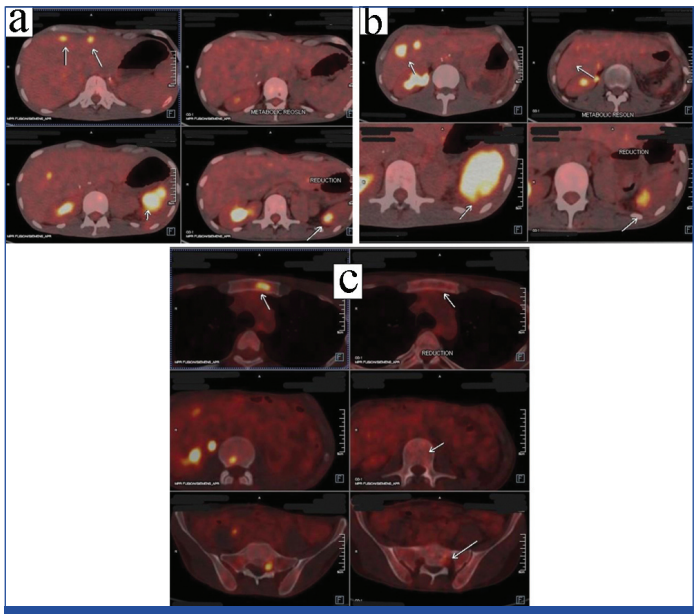
[Table/Fig-7]: Gross tumour specimen.

Microscopy revealed a highly cellular tumour with uniform morphology comprising fascicles of dysplastic short spindle cells with hyperchromatic nuclei and coarse chromatin, with scanty stroma and extensive lymphovascular emboli [Table/Fig-8a]. The tumour was found infiltrating the renal pelvis, perirenal fat, and Gerota's fascia. Sections from the renal artery and ureteric cut margin were free from tumour. Immunohistochemistry revealed positivity for BCL2, synaptophysin, CD99, and CD56, while being negative for desmin and CD34, along with histological characteristics favouring synovial sarcoma [Table/Fig-8b-e].



[Table/Fig-8]: a) Histological evaluation by high power magnification field of the biopsy specimen (H&E, 400x) showing uniform fascicles of monomorphic dysplastic short spindle cells with hyperchromatic nuclei; b) Immunohistochemically the tumour cells (400x) showing CD99 strong positivity; c) IHC the tumour cells (100x) showing BCL-2 positivity; d) IHC the tumour cells (100x) showing CD56 positive; e) IHC the tumour cells (100x) showing CD34 negative.

The patient was discharged on the 8th postoperative day. Multidisciplinary evaluation was initiated with doxorubicin and ifosfamide-based chemotherapy. Four months after surgery, the patient had undergone three cycles of chemotherapy, which showed a reduction in size and metabolic resolution of FDG avid deposits in both lobes of the liver, with residual ametabolic small lesions [Table/Fig-9a]. There was also a reduction in size and metabolic activity in the perisplenic and skeletal lesions observed [Table/Fig-9b,c]. No FDG avid deposit was seen at the tumour bed. Six months after surgery, the patient was currently under an intensive follow-up protocol with stoma care and adjuvant cycles of chemotherapy.



[Table/Fig-9]: FDG PET showing preoperative: a) Liver metastatic deposits and resolution after three cycles of chemotherapy, six months postsurgery; b) Perisplenic deposit and reduction after three cycles of chemotherapy, six months postsurgery; c) Skeletal deposits and reduction after three cycles of chemotherapy, six months postsurgery.

DISCUSSION

Approximately, 6-10% of soft-tissue sarcomas are synovial sarcomas, a rare type of sarcoma that is more commonly found in the extremities but can also originate from the kidney, pleura, lung, mediastinum, or ovary [1,2]. The first report of synovial sarcoma of the kidney was by Argani P, and males were more frequently affected compared to females [3]. Primary renal synovial sarcoma is a relatively rare condition, with around 205 documented cases [4,5]. However, cases of renal synovial sarcoma with an IVC thrombus are extremely uncommon, with only about six reported instances [6,7]. There is very limited literature on managing metastatic renal synovial sarcoma with an IVC thrombus. Modi G et al., demonstrated a partial response with neoadjuvant chemotherapy for an unresectable tumour due to mass encasing the aorta [8].

The current gold standard for renal synovial sarcoma is the demonstration of the translocation (X; 18) (p11.2, q11.2) using Reverse Transcriptase Polymerase Chain Reaction (RT-PCR), involving fusion of the SYT gene on chromosome 18 to either the SSX1 or the SSX2 gene on chromosome Xp11 found in 90% of patients [9]. The mean age of patients with primary renal synovial sarcoma was 38.0 years, which is half the median age for renal cell carcinoma diagnosis, with a slight male predominance (1.82:1) [10]. A typical misinterpretation of renal cell carcinoma occurs when patients with primary renal synovial sarcoma present with haematuria (46.3%), alone or in conjunction with other symptoms like discomfort (43%), palpable mass (37%), or both, making it indistinguishable from other renal malignancies based on clinical and radiological backgrounds. Chemotherapy based on ifosfamide and doxorubicin is preferred for sarcomas, with acceptable results of the possibility of complete remission, similar to the present case [11,12].

The present study demonstrates the role of aggressive upfront surgical therapy for patients with resectable large renal synovial sarcoma with IVC thrombus followed by adjuvant therapy, in the presence of metastatic disease, thereby emphasising the importance of cytoreductive nephrectomy. In this study, there was a tumour-free renal bed with near-complete resolution and reduction of metastatic deposits after three cycles of chemotherapy, six months postsurgery, contributing to disease-free survival. [Table/Fig-10] presents previously published cases of renal synovial sarcoma and IVC thrombus with metastasis [5-8,12-14].

Due to the tumour's rarity, with a poor median survival postsurgery (48 months), definitive management guidelines are lacking [15]. By general agreement, the best course of treatment is surgical resection either with a curative or palliative intent. Radiation therapy is preferred for localised disease, and adjuvant chemotherapy is recommended for cases that have metastasised [11,15].

Authors name and year of publication	Place	Age (years)/ gender	Findings	Management	Follow-up
Guimarães T et al., [13] (2023)	Lisbon, Portugal	69/M	8 cm left renal mass with thrombus in left renal vein	Robotic-assisted left radical nephrectomy	Developed metastasis, nine months after surgery and died
Lohani R et al., [5] (2022)	Lucknow, India	22/F	Right 14×19.1 cm renal mass lesion with no IVC thrombus/metastasis	Right radical nephrectomy with no adjuvant therapy	Patient disease free at six months of follow-up
Dutt UK et al., [6] (2018)	Pondicherry, India	21/M	Right kidney lower polar mass of 6.5 cm×6.4 cm with IVC thrombus extending upto suprahepatic IVC and inferiorly up to bilateral and femoral veins	Open right radical nephrectomy with IVC and bilateral femoral vein thrombectomy. Adjuvant chemotherapy with Adriamycin and Ifosfamide	–
El Chediak A et al., [12] (2018)	Beirut, Lebanon	26/M	Right 6 cm renal mass followed by recurrence six months after surgery at tumour bed with IVC invasion	Right radical nephrectomy with Ifosfamide+Doxorubicin based chemotherapy for recurrence	Complete pathological response, an year after surgery
Chandrasekaran D et al., [7] (2016)	Chennai, India	44/M	9×9 cm mass lesion involving upper pole (left kidney) with left renal vein thrombosis	Left radical nephrectomy with no adjuvant therapy	Disease free survival of 24 months
Modi G et al., [8] (2014)	Gujarat, India	41/M	5×5 cm left renal mass with IVC thrombus and mass encasing aorta	Neo-adjuvant chemotherapy with adriamycin+ifosfamide	Partial response after three cycles of chemotherapy
Dassi V et al., [14] (2009)	Mumbai, India	20/F	14.3×9.4 cm mass in left kidney with thrombus in left renal vein and adjacent IVC	Left radical nephrectomy	–
Present case, 2024	Pune, India	31/M	20×14 cm left renal mass with thrombus extending up too infrahepatic IVC involving left hepatic flexure of colon with metastatic lesions	Left radical nephrectomy with caval thrombectomy and hepatic flexure colon resection followed by adj. chemotherapy with ifosfamide+doxorubicin	Significant reduction in metastatic deposits after three cycles of chemotherapy at six months postsurgery

[Table/Fig-10]: Similar cases from English literature [5-8,12-14].

CONCLUSION(S)

Primary sarcoma of the kidney presents a formidable challenge in clinical management due to its aggressive nature, potential for metastasis, poor outcomes, and median survival postsurgery. Renal synovial sarcoma should be considered as a differential diagnosis in young patients presenting with a large renal mass. The most important prognostic indicators are tumour size, histologic grade, and the preferred course of treatment is aggressive resection, which can be a surgical challenge. Furthermore, adjuvant chemotherapy improves the disease-free survival rates of the patients. However, the need for further defined guidelines with preoperative identification and planning, along with patient education, is imperative for successful outcomes.

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