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METASTATIC PRIMARY RENAL EWING'S SARCOMA WITH INFERIOR VENA CAVA THROMBUS

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CASE HISTORY

- 31-year-old, nil comorbid male
- Presented with
  - *Left flank pain since 10 days*
  - *continuous, dull and radiating to back*
  - *No nausea, vomiting or fever*
  - *No urinary complaints*
  - *H/O Significant weight loss and anorexia*
  - *No past surgical history*
  - *Patient had history of similar complaints 2 years ago, was diagnosed with a renal mass but ignored the advised treatment*

EXAMINATION

GENERAL EXAMINATION

- *Conscious, co-operative, well oriented to time, place, and person*
- *Moderately built*
- *Pulse: 82/min*
- *BP: 100/80 mmHg*
- *SpO2: 99% on room air*
- *Afebrile*

SYSTEMIC EXAMINATION

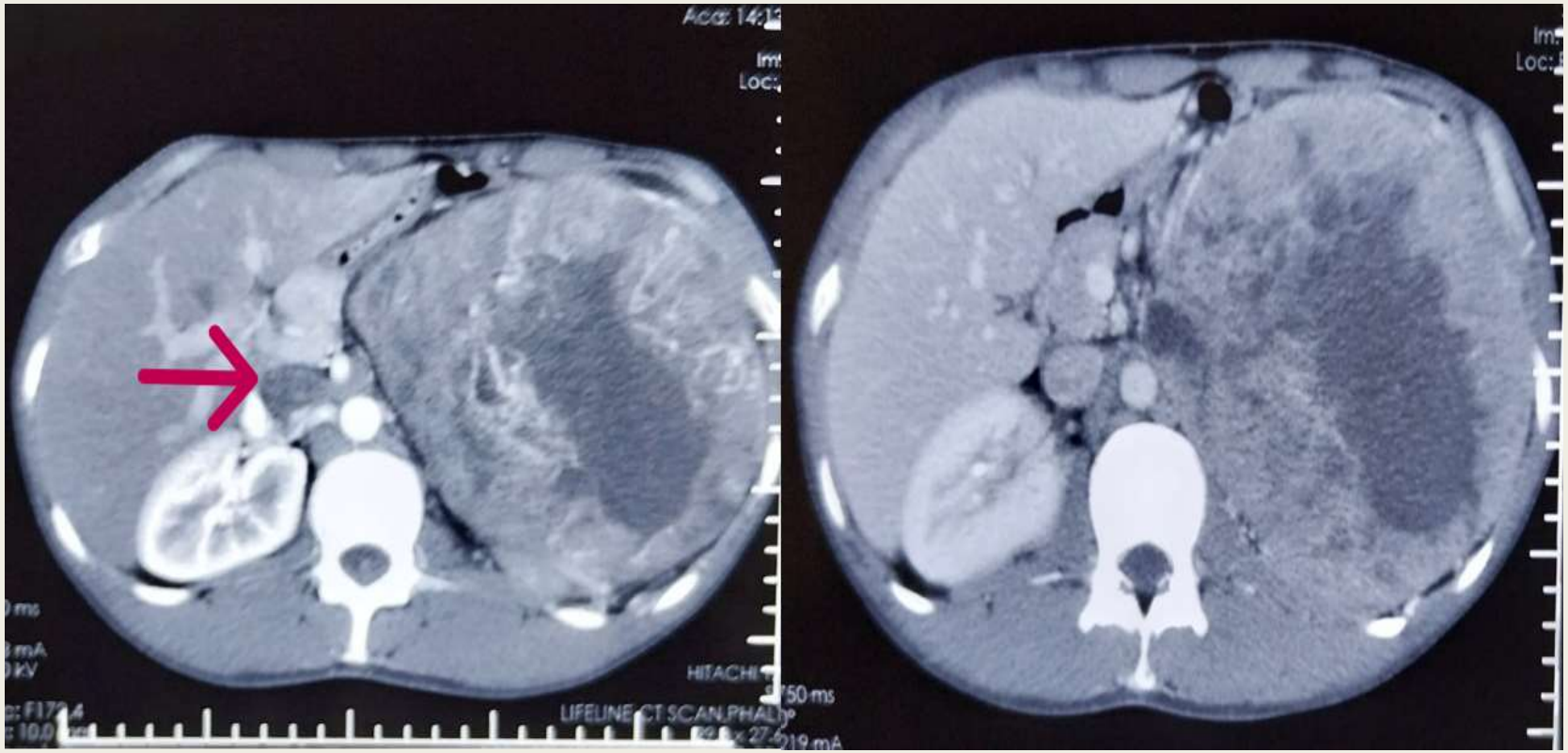
- *P/A: Palpable Left hypochondriac mass extending to umbilicus and Left iliac fossa with left flank fullness*
- *left grade 3 Varicocele*
- *CVS: S1, S2 present*
- *CNS: GCS 15/15*

INVESTIGATIONS

Radiological investigations-

Ultrasonography and Computed Tomography Intravenous Urography (CT IVU) were suggestive of an exophytic predominantly solid mass lesion (20×14 cms) from the interpolar and lower polar region 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 left renal vessels with a thrombus in left renal vein extending into the IVC, reaching up to the infrahepatic Inferior Vena Cava (IVC)

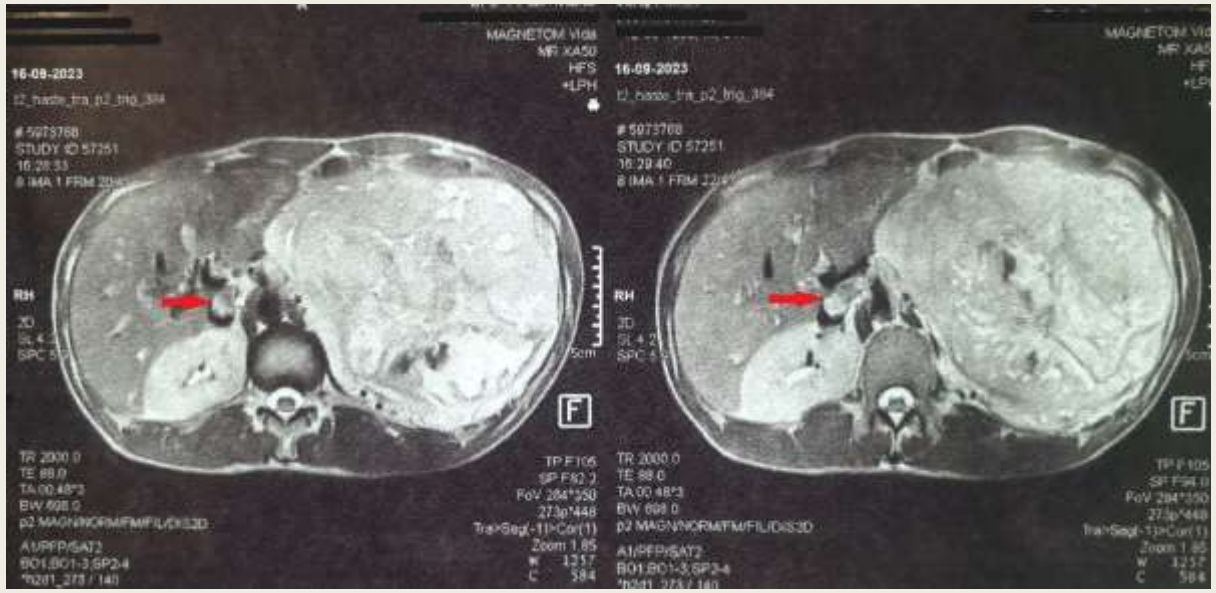
INVESTIGATIONS



Table/Fig 1- CT Axial Cut Section - Left Large Renal Mass With IVC Thrombus (arrow)

Magnetic Resonance Imaging (MRI)

The left renal vein was displaced superiorly and medially with a filling defect extending into the IVC, cranially for a length of 4.5 cms and 1.1 cm thickness reaching up to the mid part of infra-hepatic IVC



Table/Fig 2- MRI Showing Renal Mass with Tumour Thrombus (arrow) extending into IVC and Rt Renal Vein

Overall, a metastatic renal cell carcinoma was seen on FDG PET which revealed few nodules in both lungs with FDG avid deposits in both lobes of liver with peri-splenic deposits and few lytic skeletal lesions.

After thorough counselling and discussions with patient's relatives and a medical oncology opinion, a decision for upfront palliative surgery was taken.

MANAGEMENT

- On laparotomy, a circumscribed left renal mass abutting the anterior abdominal wall was mobilized infero-laterally and superiorly after opening the line of Toldt .
- The Mass was freed of its adhesions posteriorly upto the renal hilum after lifting it medially and left renal artery was dissected.
- The transverse and splenic flexure of colon along with its mesentery, approximately 10 cm in length was found adhered to the anterior wall of mass which was excised with adequate margins.

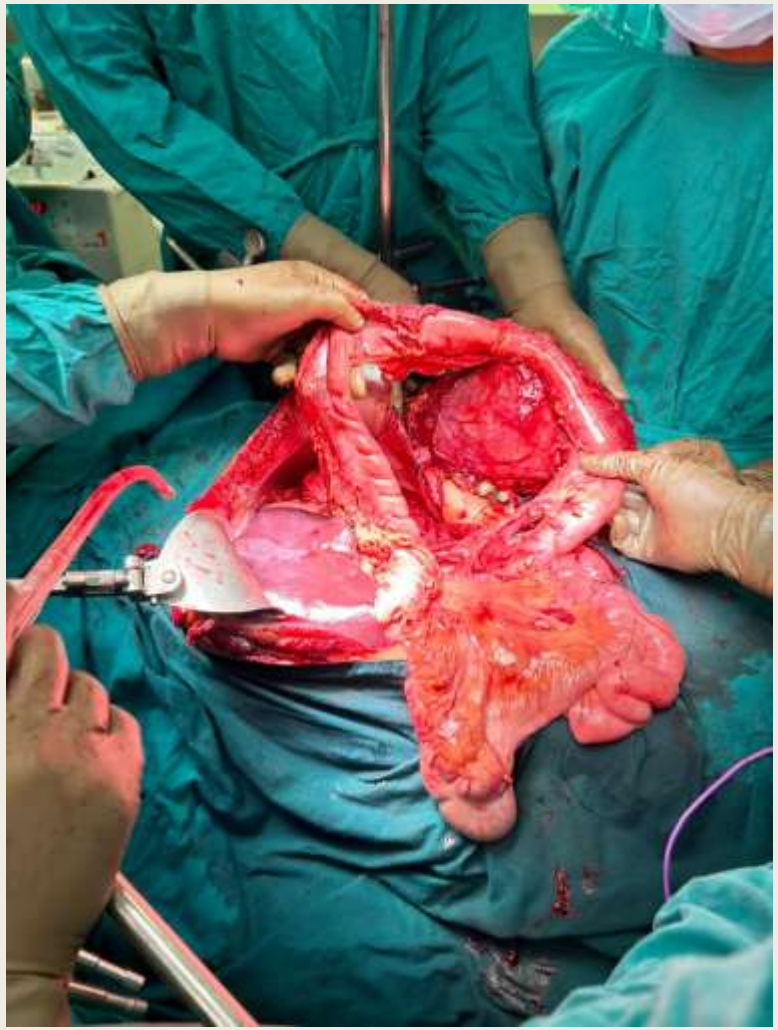


Fig 3- Intra-Op Tumour Dissection With Adherent Colon and Mesentery

- Retrohepatic IVC was isolated after mobilizing the liver. An Intra-operative Ultrasound was performed to delineate the proximal extent of the thrombus and rule out IVC invasion
- Thrombus was seen extending cranially upto the infra-hepatic IVC.
- The Infra-renal IVC, Right Renal vein, and Supra-hepatic IVC were clamped sequentially using vascular clamps after passing vascular loops, taking care not to dislodge the thrombus.
- A Cavotomy was performed, and thrombus was removed in-toto following which IVC repair was done using 6-0 prolene after flushing with heparinized saline. Radical nephrectomy with thrombectomy was performed followed by regional lymphadenectomy and ileal stoma formation

INTRA-OPERATIVE - ULTRASOUND

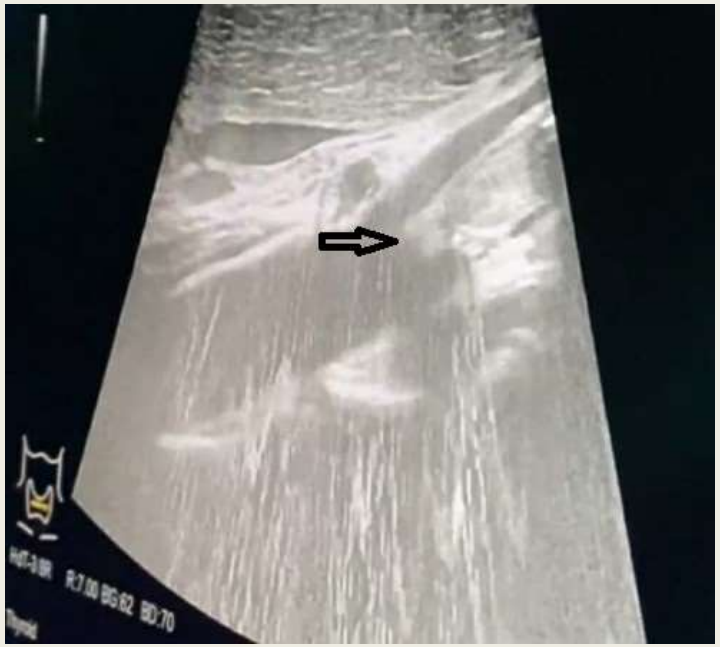


Fig 4- Intra-op USG– Delineating the Proximal extent of IVC

THROMBUS (arrow)

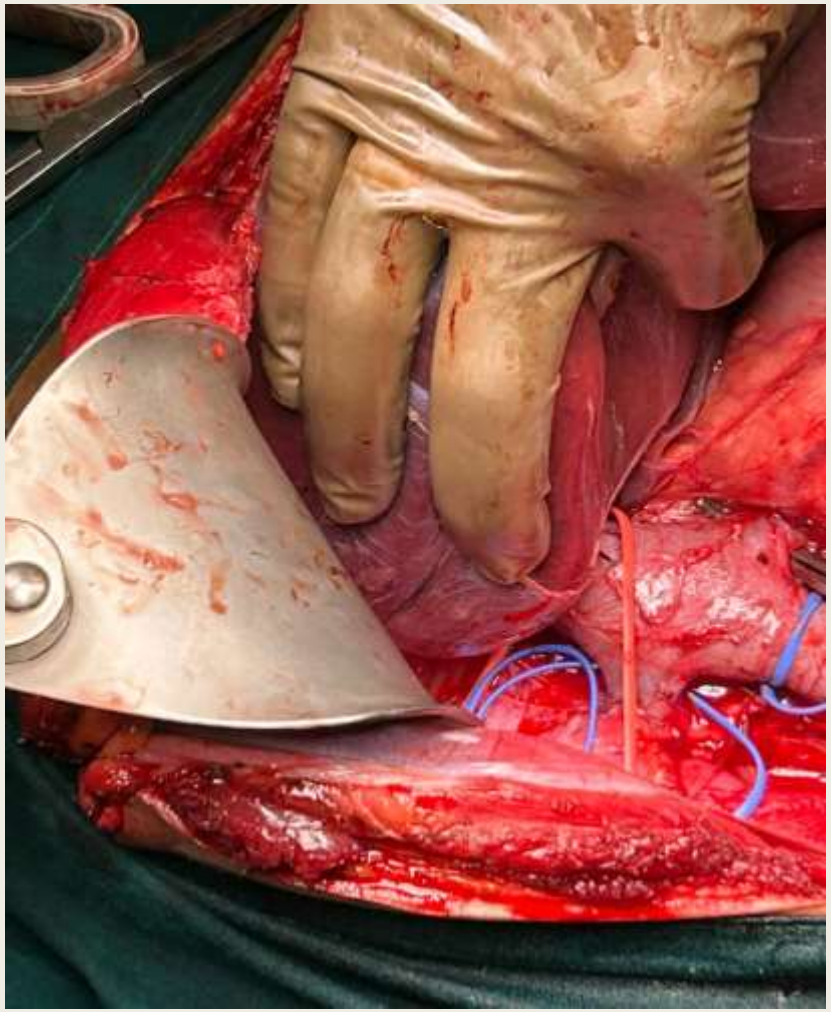


Fig 5- Inferior Vena Cava with Its Branches

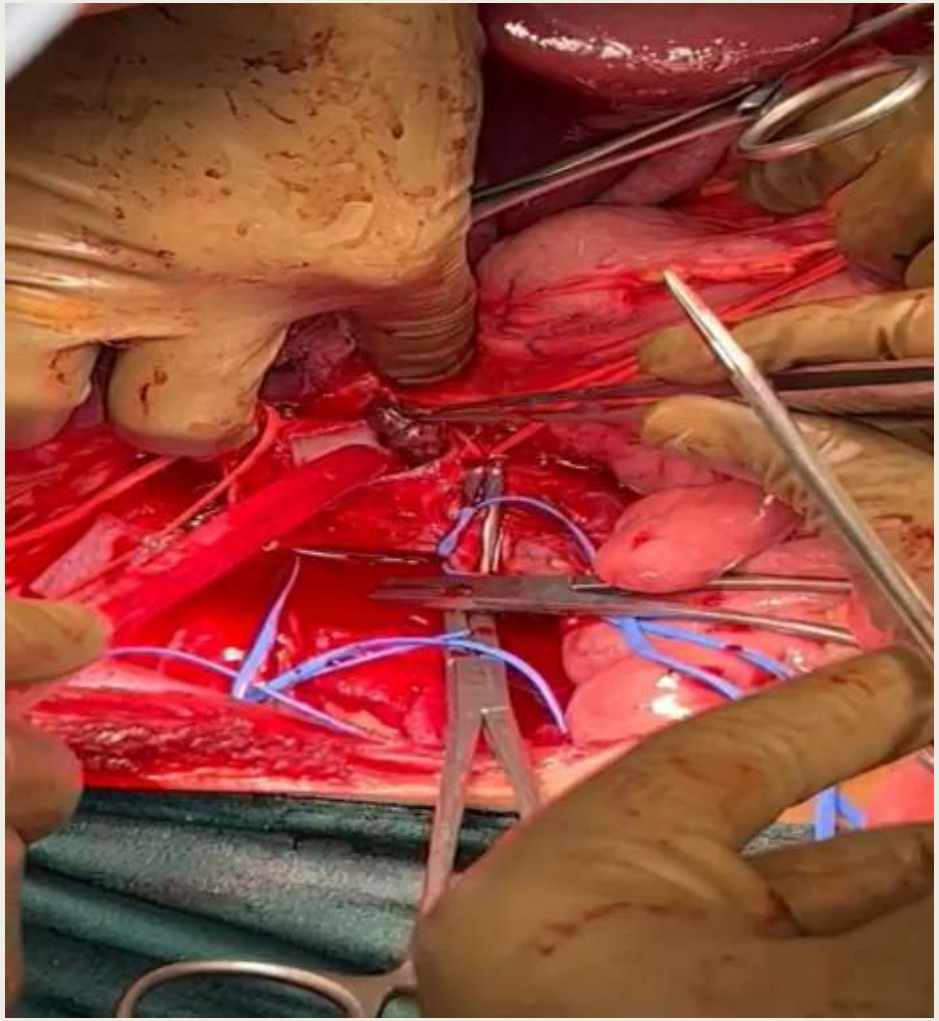
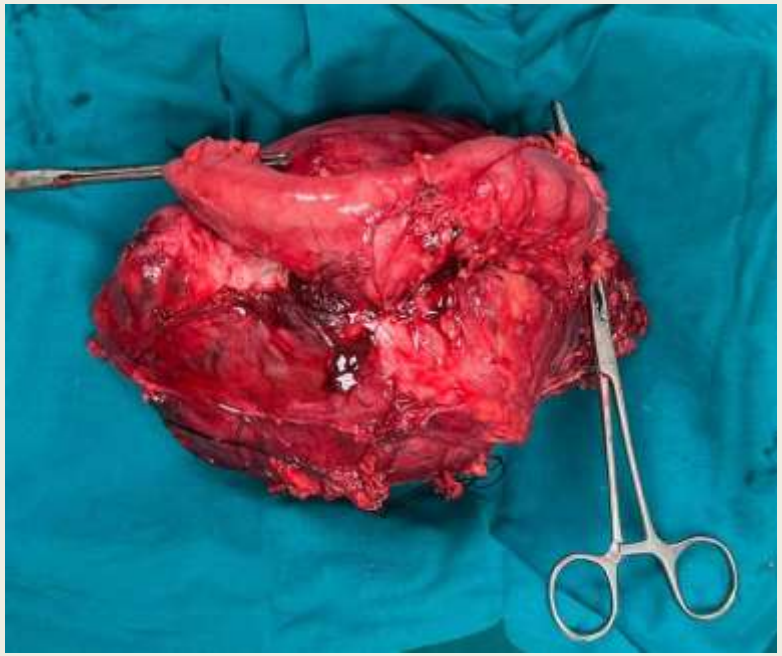


Fig 6- Tumour Thrombus Extraction

HISTO-PATHOLOGY

- Grossly, a well-circumscribed tumour was noted measuring 20 x 16.5 x 10.7 cm
- cut section showing a greyish white, friable mass with focal areas of hemorrhage and necrosis and weighed approx. 2.75 kgs
- Renal vein tumour thrombus measured 3 x 2 x 1.6 cm.



- Microscopy revealed a highly cellular tumour with uniform morphology comprising of fascicles of short spindle cells with hyperchromatic nuclei and coarse chromatin with scanty stroma and extensive lymphovascular emboli.
- Tumour was found infiltrating renal pelvis, peri-renal fat and Gerota's fascia.
- Sections from renal artery and ureteric cut margin was free from tumour.
- Immunohistochemistry revealed BCL2 – positive, synaptophysin positive, CD 99 and CD 56 positive, while negative for desmin and CD34.

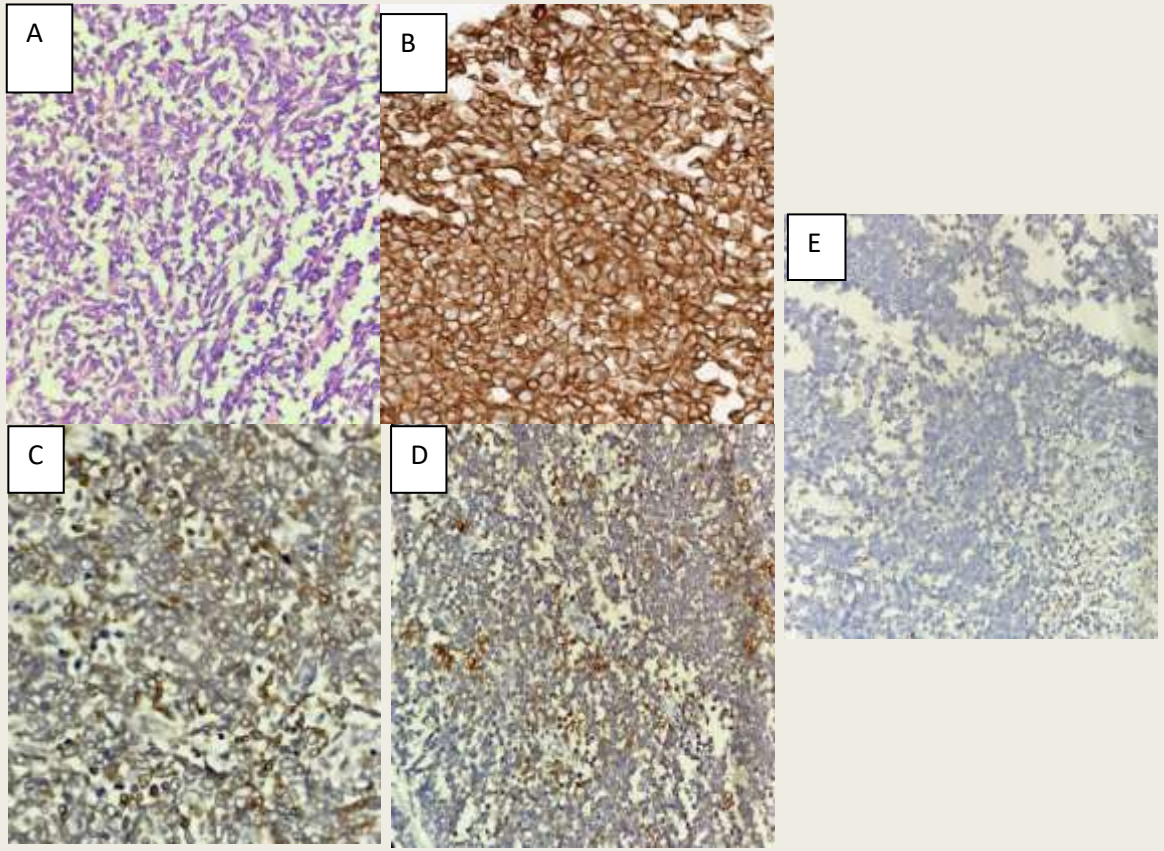


Fig 8(A)- Histological evaluation by high power magnification field of the biopsy specimen (400x) showing uniform fascicles of monomorphic short spindle cells with hyperchromatic nuclei; 8(B)- Immunohistochemically the tumour cells (400x) showing CD99 strong positivity; 8(C) – IHC the tumour cells (100x) showing BCL-2 positivity; 8(D) – IHC the tumour cells (100x) showing CD56 positive; 8(E) – IHC the tumour cells (100x) showing CD34 Negative

- The patient was discharged on postoperative day 8
- After multidisciplinary evaluation, patient was started on vincristine-based chemotherapy. Four months after surgery patient had undergone 3 cycles of chemotherapy which showed reduction in size and metabolic resolution of FDG avid deposits in both lobes of liver with residual ametabolic small lesions.
- There was also reduction in size and metabolic activity in perisplenic and skeletal lesions.
- No FDG avid deposit was seen at tumour bed.
- Six months after surgery, patient is currently under an intensive follow up protocol with stoma care and adjuvant cycles of chemotherapy

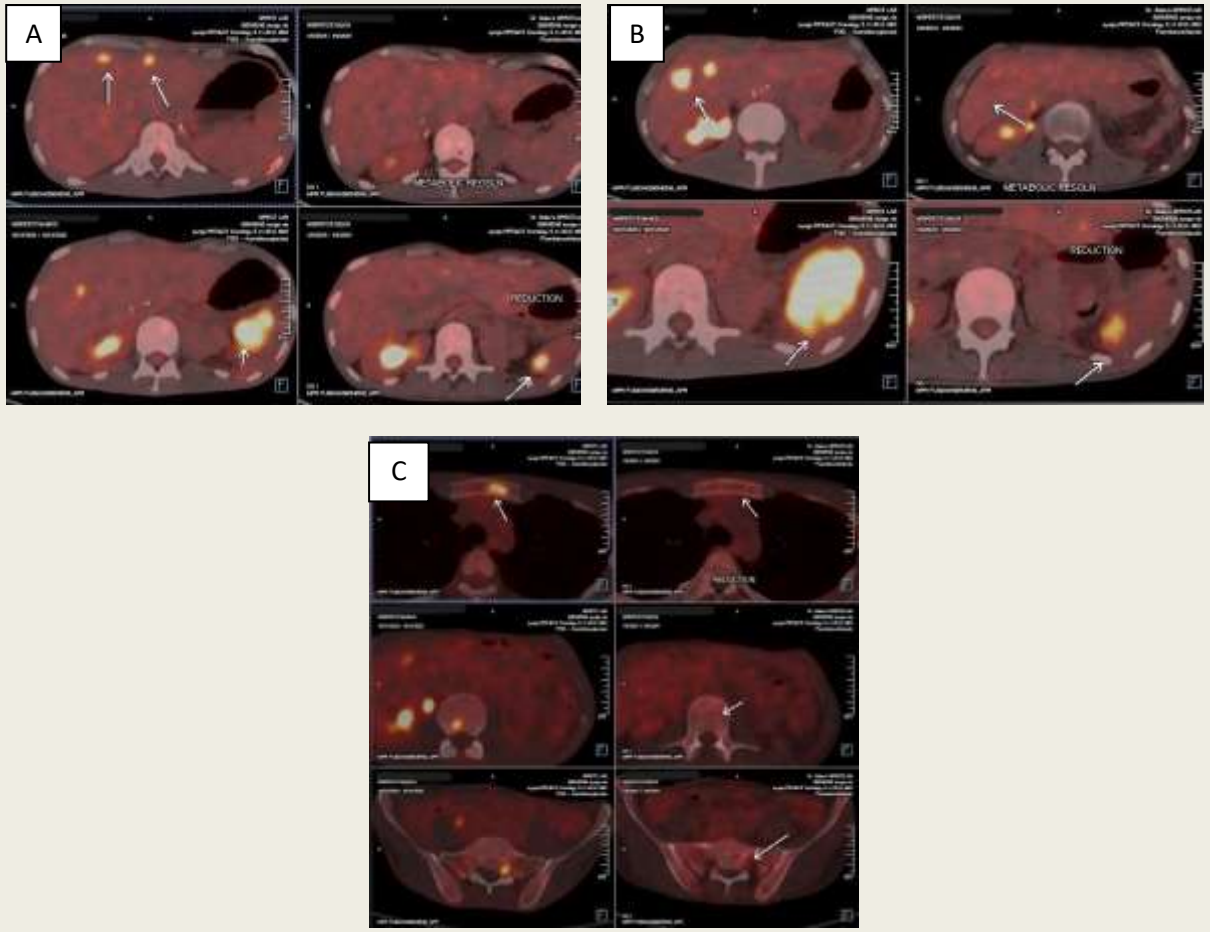


Fig 9 - FDG PET showing pre-op (A) Liver Metastatic Deposits and Resolution after 3 cycles of Chemotherapy, 6 months post-surgery.
(B) Peri-splenic Deposit and Reduction after 3 cycles of Chemotherapy, 6 months post-surgery.
(C) Skeletal Deposits and Reduction after 3 cycles of Chemotherapy, 6 months post- surgery

DISCUSSION

Primary Sarcoma of the kidney presents a formidable challenge in clinical management due to its aggressive nature, potential for metastasis with poor outcome and median survival post-surgery. Renal sarcoma should be considered as differential diagnosis in young patients presenting with large renal mass. The most important prognostic indicators are tumour size with 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 pre-operative identification and planning along with patient education is imperative for successful outcome.

THANK YOU