RIGHT ILIAC FOSSA MASS – Ileal leiomyomoscarcoma

UNIT VI

Under the guidance of

UNIT VI Faculties .

Introduction

- **Leiomyoscarcoma is a benign / potentially malignant tumour arising from the smooth muscle cells.**
- Commonly seen in uterus, retroperitoneal or dermis of the extremities.
- Primary intestinal leiomyosarcomas are extremely rare and only a 26 cases are reported following robust IHC techniques in medical literature .

- The best treatment modality so far is surgical resection.
- In this case report we present a extremely rare case of the patient with gastrointestinal LMS with clinical presentation, blood test report , imaging, histological and immunohistochemical finding.

Case presentation

A 45 year old male came with complaints of pain in lower abdomen since 1 month which was :

-insidious in onset ,gradually progressive ,

-radiating towards umbilicus and back .

-Mild to moderate in intensity , dull aching in nature .

-Aggravates on walking and had no specific relieving factor .

-Gives H/O passing hard stools {on and off} .

- No H/O fever
- No H/O abdominal fullness , dyspepsia.
- No H/O vomiting , nausea , constipation or loose stools.
- No H/O of similar complaints in the past .
- No H/O bleeding per-rectum .

On examination

PER ABDOMEN :

Inspection:

-The abdomen is fatty .

-Umbilicus central and inverted.

-No visible lump .

-Abdomen move equally with respiration .

-No dilated veins or scar present.

- All the hernial orifices appears normal .

Palpation:

- The abdomen is soft and not distended .
- Tenderness (+) in right iliac fossa.
- A lump of size 6x6 cm (+) in right iliac fossa.
- With well defined margins ,
- lower border could be palpated
- Smooth surface,
- Restricted mobility ,
- Firm in consistency .

Percussion :

Liver dullness + 6^{th} -12^{th} intercoastel space .

Tympanic note present all over abdomen .

Auscultation:

bowel sounds present in all the quadrants

PER-RECTAL EXAMINATION:

No skin tags +

No e/o fissure or fistula

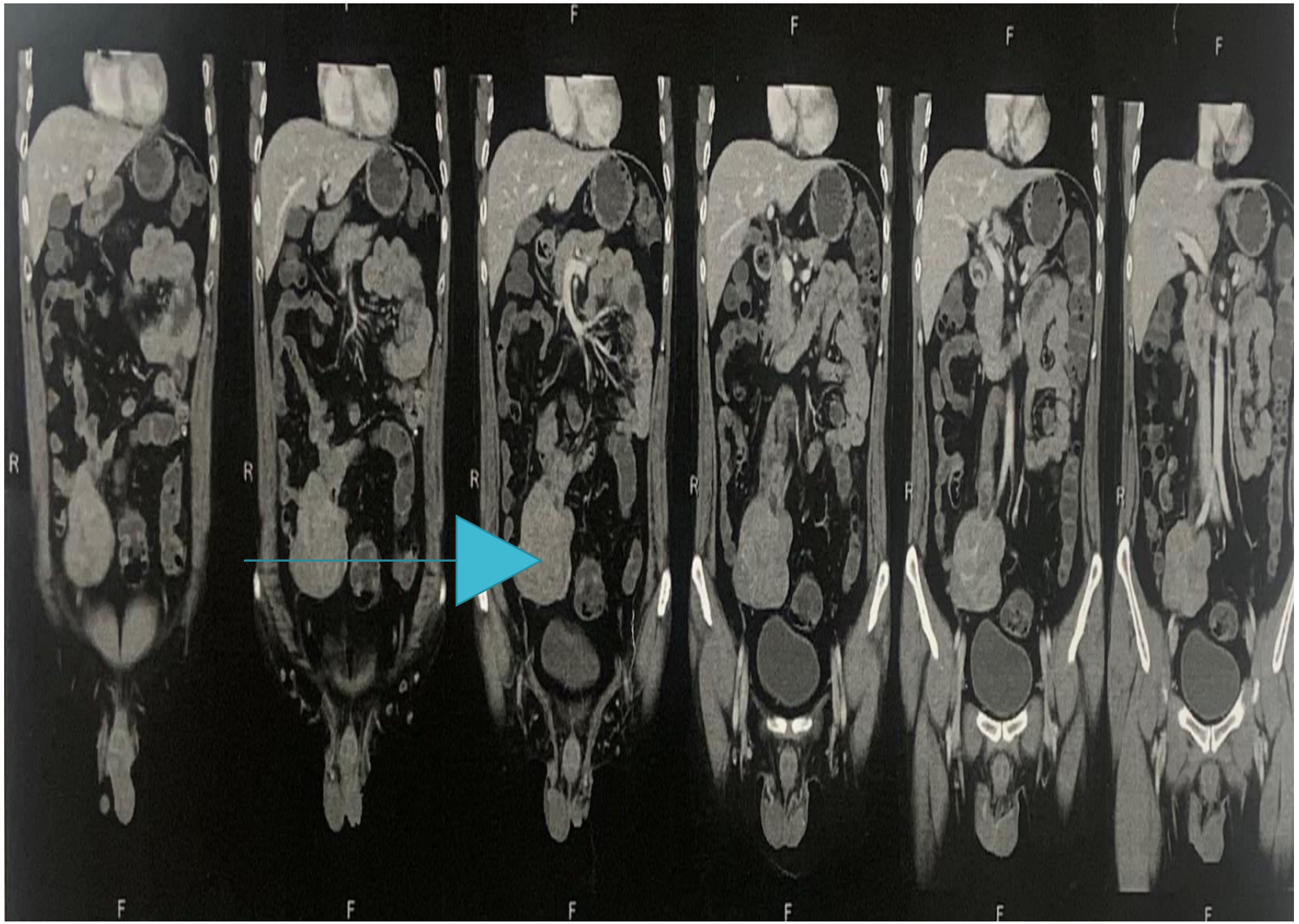
No active bleeding or discharge

Soft stools +

Investigation

- All the hematological and the blood biochemical investigation were within normal limits .
- Abdominal ultrasound revealed a large heterogeneous hypoechoic oval mass in the right iliac fossa of size 9*9*6 cm merging with serous layer of the ileal loop , however does not seem to cause any obstruction .

- **Dynamic contrast enhanced computed tomography** revealed a large well defined lobulated heterogeneously enhanced solid mass of size 8.4*5.4*8.1cm is noted in the right iliac fossa in the relation to mesenteric border of distal jejunal /proximal ileal loop anterior to adjoining right psoas and right common iliac vessels .
- The mass shows contrast – opacified vasculature . No calcification noted .
- It is seen extending supero-inferiorly from level of L5 to S1 Vertebrae .
- Posteriorly the mass is abutting the right psoas muscle with obliterating of intervening fat planes.
- Other fat planes between the mass and surrounding structures appear normal .



CT films showing the mass in the right iliac fossa .

- **USG guided biopsy** was done :

  -showed e/o spindle cell tumor .

  s/o smooth muscle tumor V/S gastrointestinal stromal tumor.

HRCT : no obvious abnormalities noted.

Management

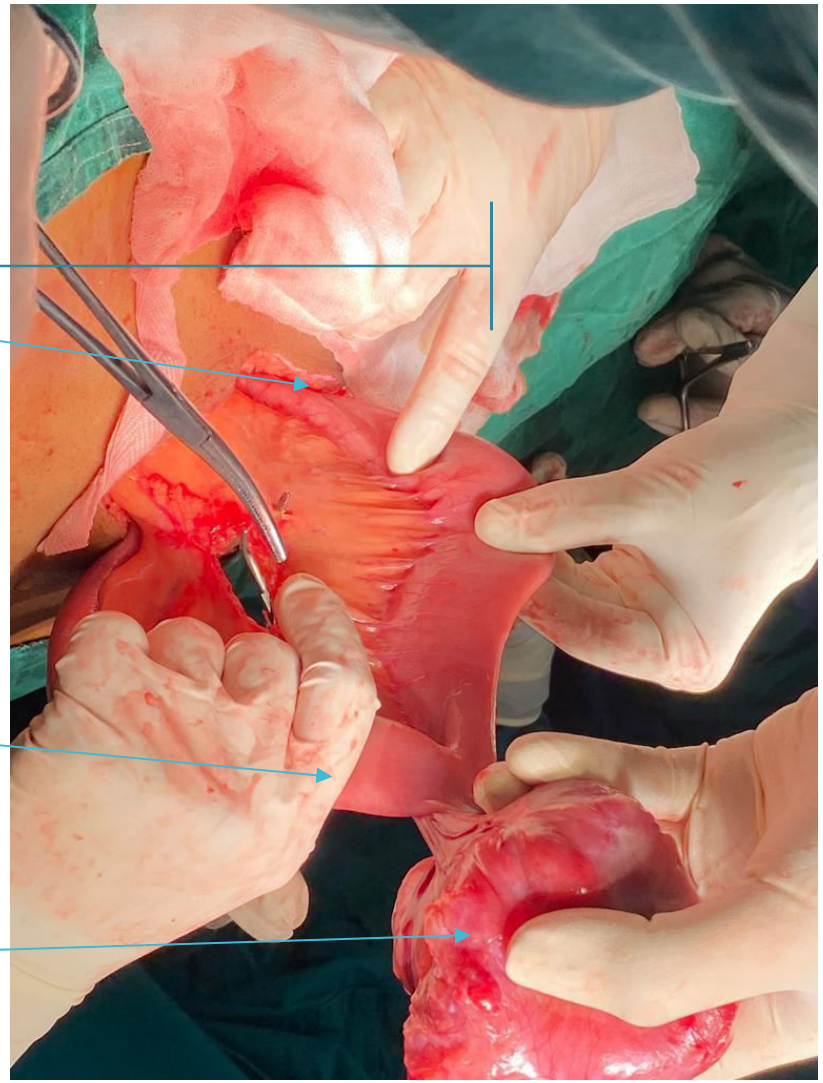
- **Exploratory laparotomy** was done with excision of the proximal ileal mass with ileoileal anastomosis .
- **Findings** : a tumor of mass 7x6x5cm at proximal ileal loop over the anti mesenteric border 20 cm from the ileoceacal valve . Ileal patency and circumference was maintained.
- The tumor was encapsulated , smooth surface with regular margin.

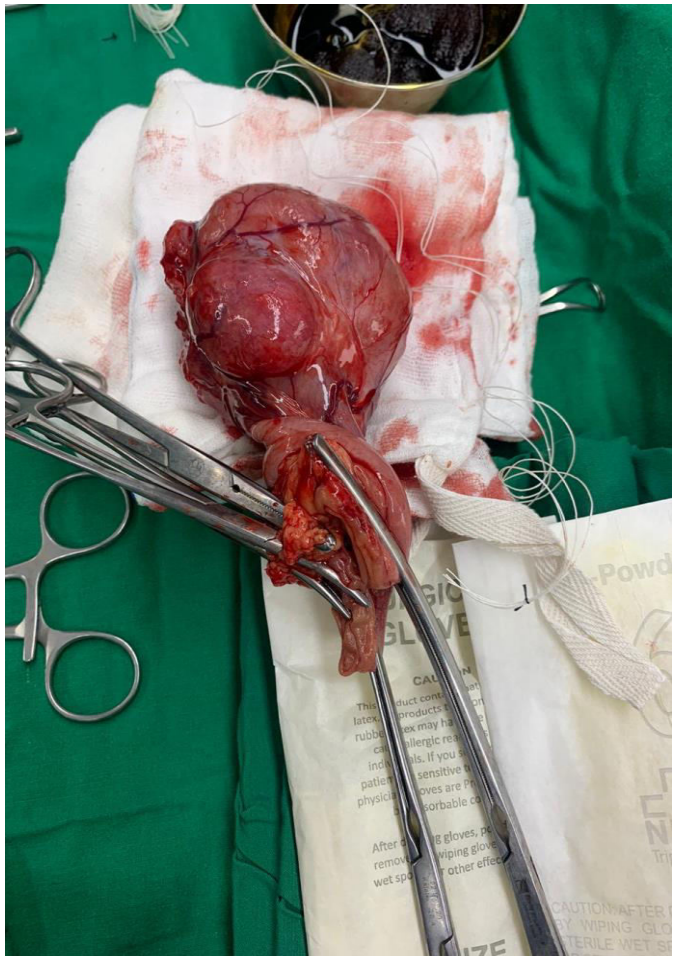
Intra op photos

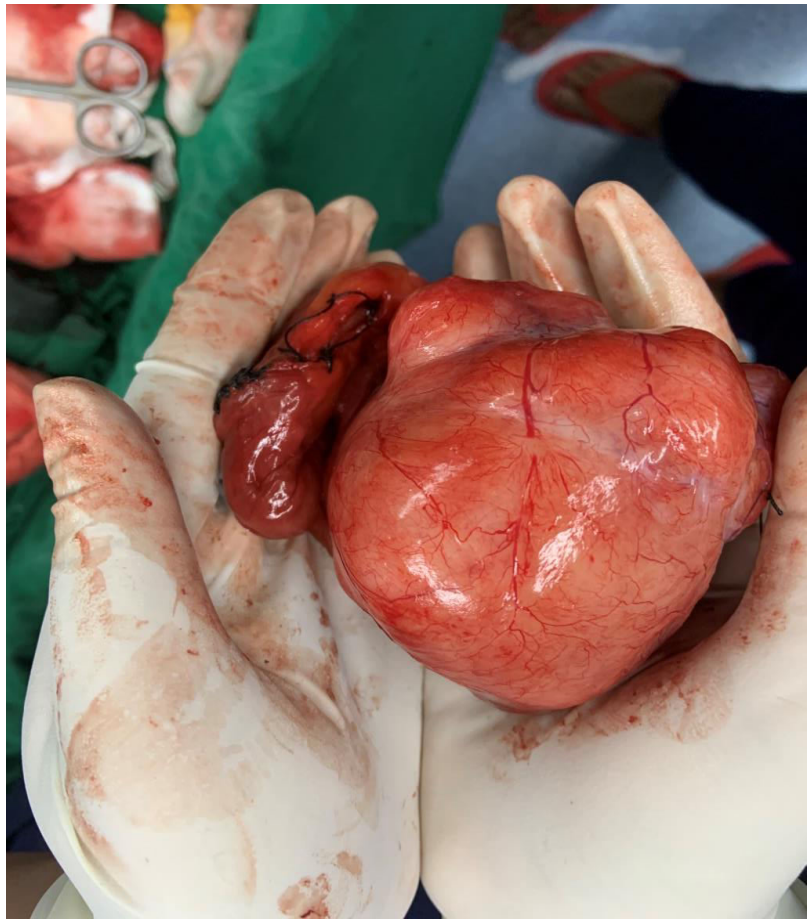
Proximal ileum

Distal ileum

Mass present in the ileal loop on the anti mesentric border







Resected specimen of size 7x6x5cm approximately with the ileoileal loop.

Histopathalogical examination .

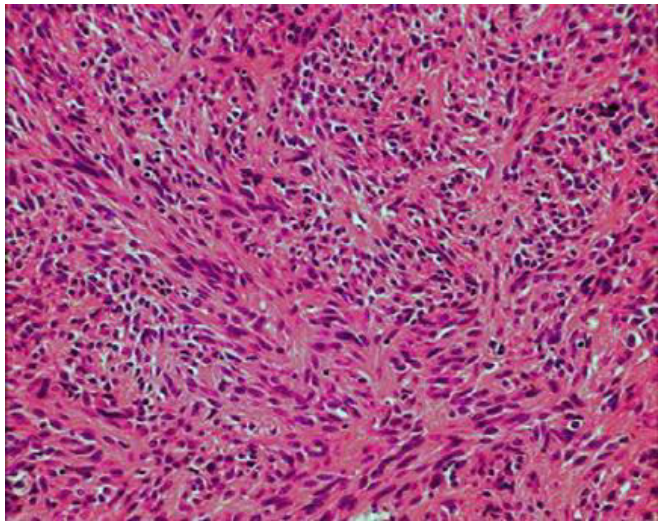
The specimen was sent for the histopathological examination:

- On gross examination: a mass of 200gms measuring 7.5x6.0x4.5cm which was encapsulated , greyish white . Cut surface showed whorled pattern.
- On microscopic examination:s/o leiomyosarcoma with mitotic rate: 1-2/10HPF Pathological stage classification –pT1 .
- IHC studies showed CD 117 : Negative in tumor cells

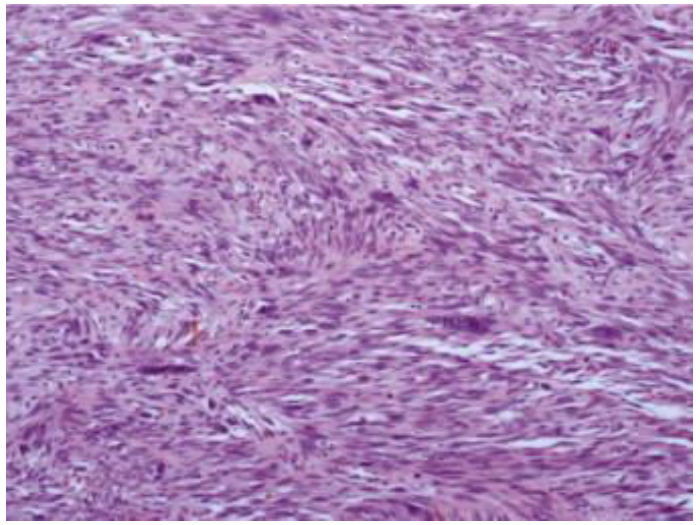
CD 34 : negative in tumor cells

SMA : positive in tumor cells

DESMIN : positive in tumor cells



H&E stain demonstrated spindle cells .



The tumour is arranged in fascicles of spindle cells with “Cigar shaped”, atypical nuclei (H&E stain).

Discussion

- The incidence of small bowel tumours is 22.7 cases per million, with sarcomas accounting for only 1.2% of these.
- Leiomyosarcomas of the small bowel are therefore extremely rare entities, to the extent that the World Health Organisation can provide no meaningful data on demographic or clinical features as a result. Only 26 documented cases of small bowel leiomyosarcomas were identified .
- The diagnosis of small bowel tumours can prove difficult, as the presentation can be delayed and traditional management of ominous symptoms such as weight loss, constipation and rectal bleeding consists of OGD and colonoscopy in the first instance.

- Furthermore, Small bowel tumours are easily missed by both modalities, and further imaging is therefore needed . Currently, magnetic resonance enterography (MRE), CT colonography (CTC) and wireless capsule endoscopy (WCE) have all been shown to be effective in reaching the diagnosis.
- Despite advances in imaging, the differentiation between benign and malignant tumours remains extremely difficult pre-operatively. The aetiology of many cases does not become apparent until after definitive resection, the case presented here being a prime example.

IHC MARKERS	GIST	LEIOMYOSARCOM
CD34	+VE	-VE
CD117	+VE	-VE
SMA	-VE	+VE
DESMIN	-VE	+VE

- Histologically, leiomyosarcomas are differentiated from GISTs by the lack of CD117 (c-KIT), DOG1 and CD34 as well as presence of smooth muscle actin (SMA) and desmin.
- The treatment of all small bowel tumours remains centred around radical surgical resection.
- Tumour size and histological staging are independent prognostic factors for 5 year disease-specific survival (DSS) in patients with small bowel sarcomas; these appear to be more favourable than small bowel adenocarcinomas.
- In summary, leiomyosarcoma in the small bowel is a rare finding; the management relies on accurate diagnosis and a predominantly surgical approach.

Conclusion

- Small bowel leiomyosarcomas are rare with only 26 cases reported so far .
- Despite the advances in imaging the differentiation between benign and malignant tumours is extremely difficult preoperatively in a case of leiomyoscarcoma ,with IHC staining it was diagnosed as the leiomyosarcoma .
- Treatment of such tumours are surgical resection and prognosis depend on the tumor size and histological staging .

THANK YOU .