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Gastrointestinal Stromal Tumour (GIST)

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Abstract:

- ***Introduction***

Gastrointestinal stromal tumors (GISTs) are the most frequent malignant mesenchymal lesions of the gastrointestinal tract believed to originating from the interstitial cells of Cajal. These tumors typically exhibits overexpression of the tyrosine kinase receptor, protein product of c-KIT gene (KIT) CD 117.they may appear in GI tract and most frequently found in the stomach(60%) .

- ***Objective:***

To analyze the clinical characteristics , diagnosis , treatment , approaches, recurrence, and survival rates of GISTS.

In this case, series a total number of 3 patients, admitted and operated in the surgical department of PDU Medical College and Hospital .The age range of the patients was 50 to 70years. The cases were evaluated for symptoms ,tumor location ,size,diagnostic findings , surgical approach and outcomes. Out of 3 cases, 2 occurred in the stomach, 1 in jejunum.

- ***Conclusion:***

GISTs are aggressive tumors, particularly when located in stomach . Diagnosis and treatment strategies depend on their size and location, surgical intervention remains the mainstay of treatment , with endoscopy playing a role in selected cases.

Keywords: *GISTS, Diagnosis, Differential Diagnosis, Management, Localization Histopathology.*

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I. INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are recognized as the most frequent malignant mesenchymal tumors of the gastrointestinal tract originating from the interstitial cells of Cajal. Often show overexpression of the tyrosine kinase receptor, c-KIT gene (KIT) proto-oncogene account for 1 to 2 % of all gastrointestinal malignancies.

GISTs are more frequently diagnosed in individuals between 50 and 70 years of age with a slightly higher incidence in males with sporadic presentation or as part of syndrome complex.Most common in stomach(60%),followed by small intestine(30%)and 5-10% any where in Gi tract.

Gastric lesions have wide range of clinical behavior,varying from small incidental findings to large lesion with aggressiveness.Symptoms are not specific and depend on size and location.

Small lesion are discovered incidentaly during endoscopy, abdominal surgery or radiological imaging .these lesions can cause non specific symptoms like dyspepsia, abdominal pain, nausea ,vomitting and in some cases gastrointestinal bleeding or may remain asymptomatic.

Mitotic activity and tumor size are important prognostic parameters of the risk of aggressive tumors.

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➢ *Objective :*

To describe the clinical presentation diagnostic findings , management, recurrence and survival of GISTs . Special attention is given to the impact of tumor size and location and the effectiveness of various therapeutic approaches on overall survival.

II. CASE SERIES

Information of A total of 3 cases of GISTs were collected by reviewing the medical recods of patients diagnosed with GISTs in our hospital .

Clinical, histopathological characteristics reported in biopsy were recorded as well as the surgical specimen of detected cases.

A. Case 1:

A 75 Year female patient with GIST in stomach undergone exploratory laparotomy with wide local excision from greater curvature of stomach.Her Past history included Asthama since birth. No any other significant family history. Patient complain of abdominal discomfort and fullness. No any other complains.

In investigation patient underwent for ultrasonography, CT scan of abdomen and pelvis which suggest approx. 9 cm in axial and 7 cm in craniocaudally size exophytic mass lesion arising from greater curvature of stomach. In histology tumor size more than 10 cm with mitotic count < 5/50hpf.

Patient underwent for exploratory laparotomy with wide local excision of GIST with adhesiolysis of adhesions from liver and spleen done.

B. Case 2:

A 70 yr female patient with diagnosed small bowel neuroendocrine tumor (GIST) underwent diagnostic laproscopy first and then exploratory laparotomy with excision of jejunal segment with mass and jejunojejunal anastomosis.patient complain of abdominal pain and nausea.

In investigation patient underwent for CT abdomen and pelvis which suggest approx. 2 cm size exophytic lesion in small bowel loops with loss of fat plane, in histopath 2cm size mass in jejunum suggest GIST .

Patient underwent for diagnostic laparoscopy converted to open exploratory laparotomy with excision of jejunal segment with mass with J-J anastomosis.

C. Case 3:

A 60 year male patient with GIST in somach underwent exploratory laparotomy with sleeve gastrectomy presented with complain of abdominal fullness and discomfort with upper GI bleed .patient was mildly obesed and no any other significant medical or past history.

In investigation patient undergone ultrasonography CT scan and upper GI scopy and mass diagnosed in body of stomach . in histopathology it suggest mix type GIST size 3 cm in size.

Patent inderwent open surgery by laparotomy and sleeve gastrectomy with resection of mass was done.

Regarding clinical presentation abdominal pain was most common symptoms, more in female than male the age range is 50-70 years, median age is 65 yr age , most common in stomach then small intestine and rare in colon and oesophagus. All tumor are low risk, spindel cell type histologically immunohistochemistry CD 117 and non mutated.

Most patient underwent open surgery followed by laproscopic surgery and no in tervention at all.in case of metastatic lesion patients may required medical management with neoadjuvnt therapy.

The patient's data (age and sex) and tumor characteristics (site, type, symtoms, histology, surgical management and follow up) were analyzed in these cases.

➢ *All the Cases are Elaborated in Table Shown Below*

Table 1 Details of All 5 Cases of GIST.

Variables	Case 1	Case 2	Case 3
Age (years)	75	70	60
Sex	Female	female	male
Clinical presentation	Mass	Mass	mass
Site	gastric	Jejunum	stomach
Size (cm^3)	9×7×	2×1.9×2	3.5×3×2
Histologic type	Spindle cell type	Mixed (spindle>epithelioid type)	Mixed (epithelioid>spindled)
Histologic grade	high grade	low grade	High grade
Mitotic rate (hpf)	>5/50	<5/50	>/5/50
Necrosis	Absent	absent	Present
IHC for CD117	Positive	Positive	Negative
Follow up	Doing well	Doing well	Lost to follow up

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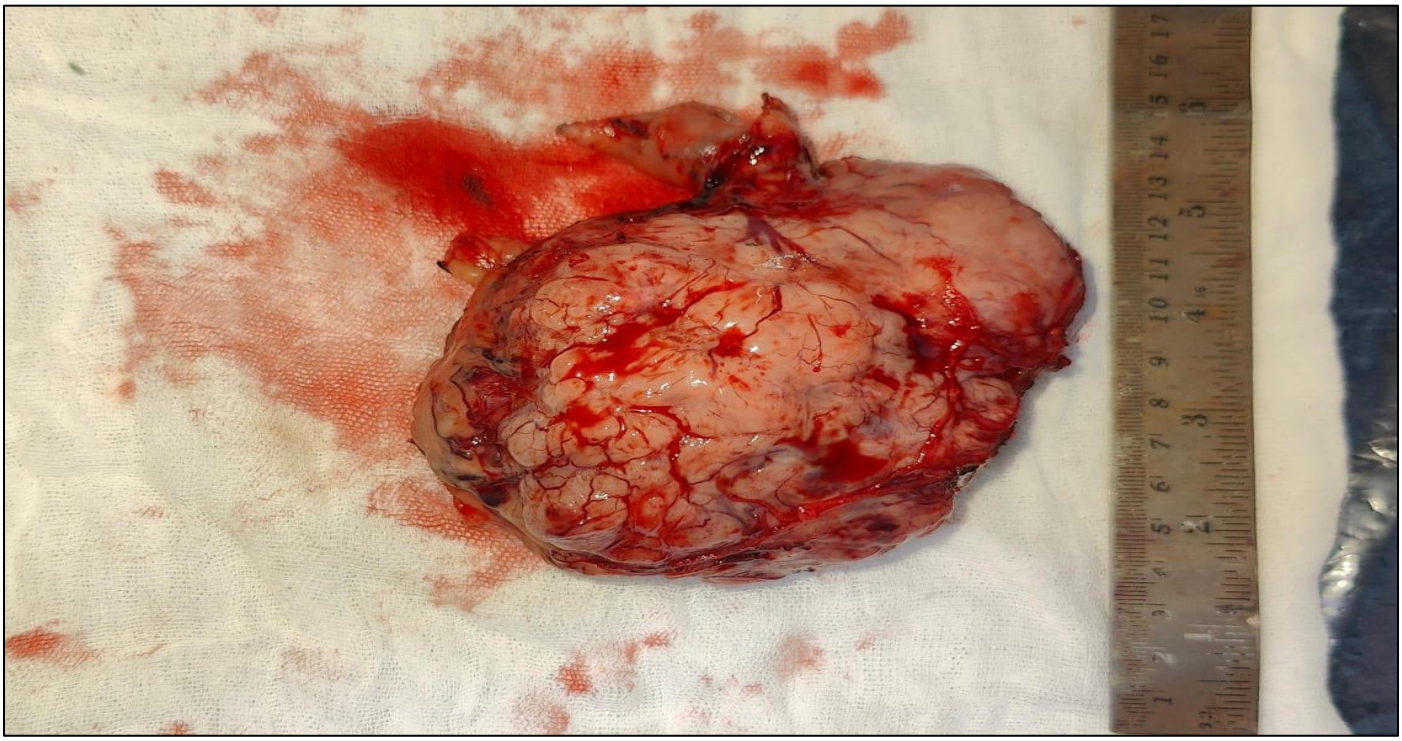

Fig 1 Showing Specimen of Tumor

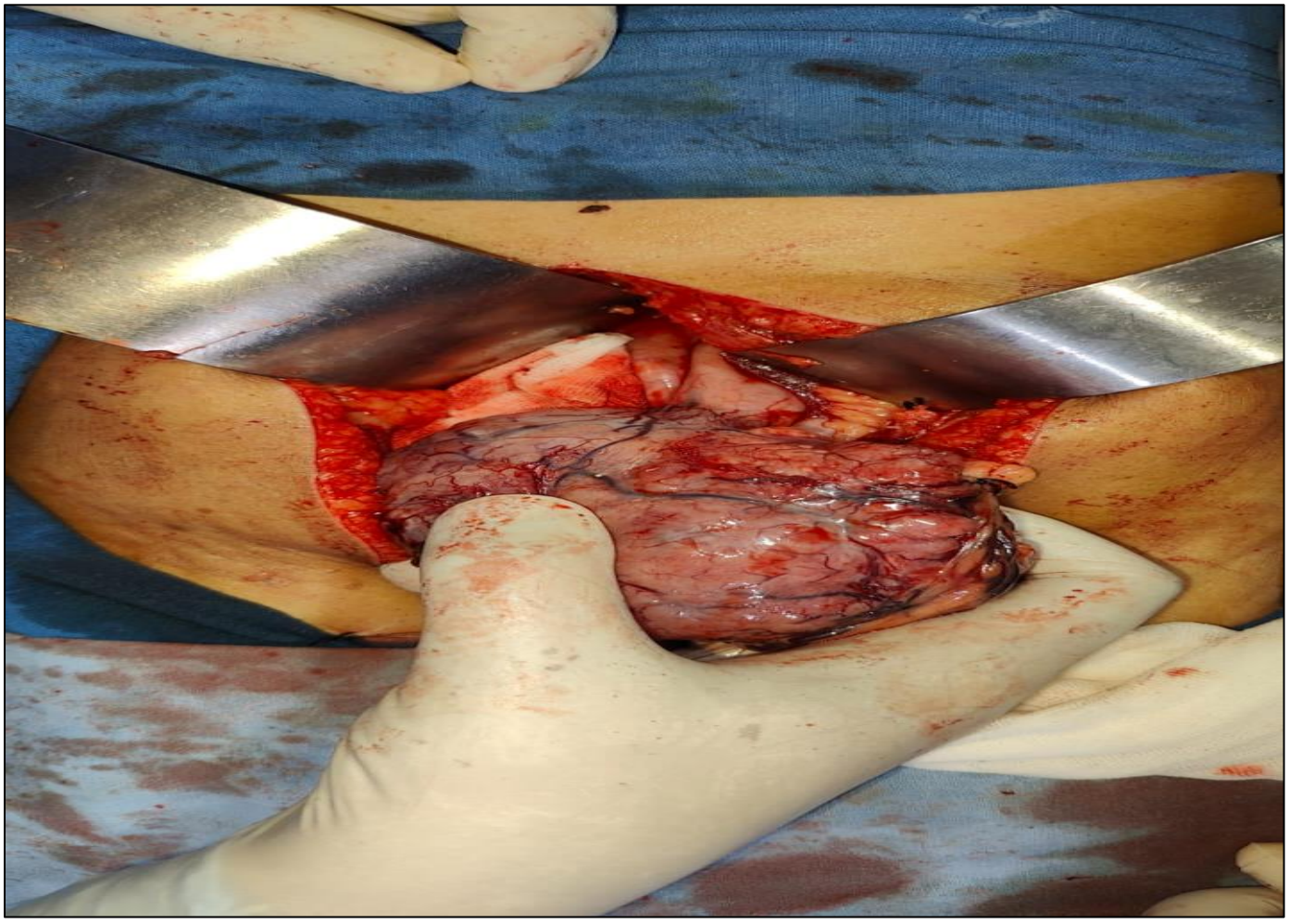

Fig 2 GIST at Greater Curvature of Stomach

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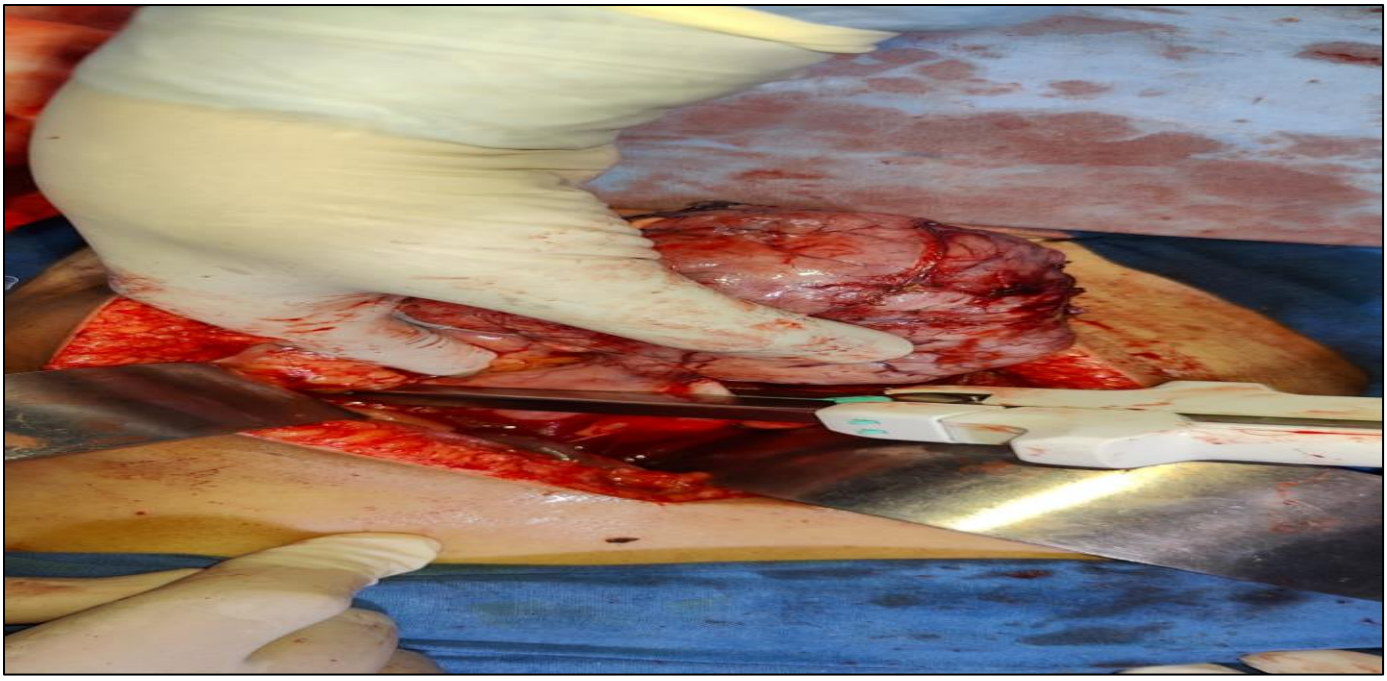

Fig 3 Excision of GIST Using Endostapler

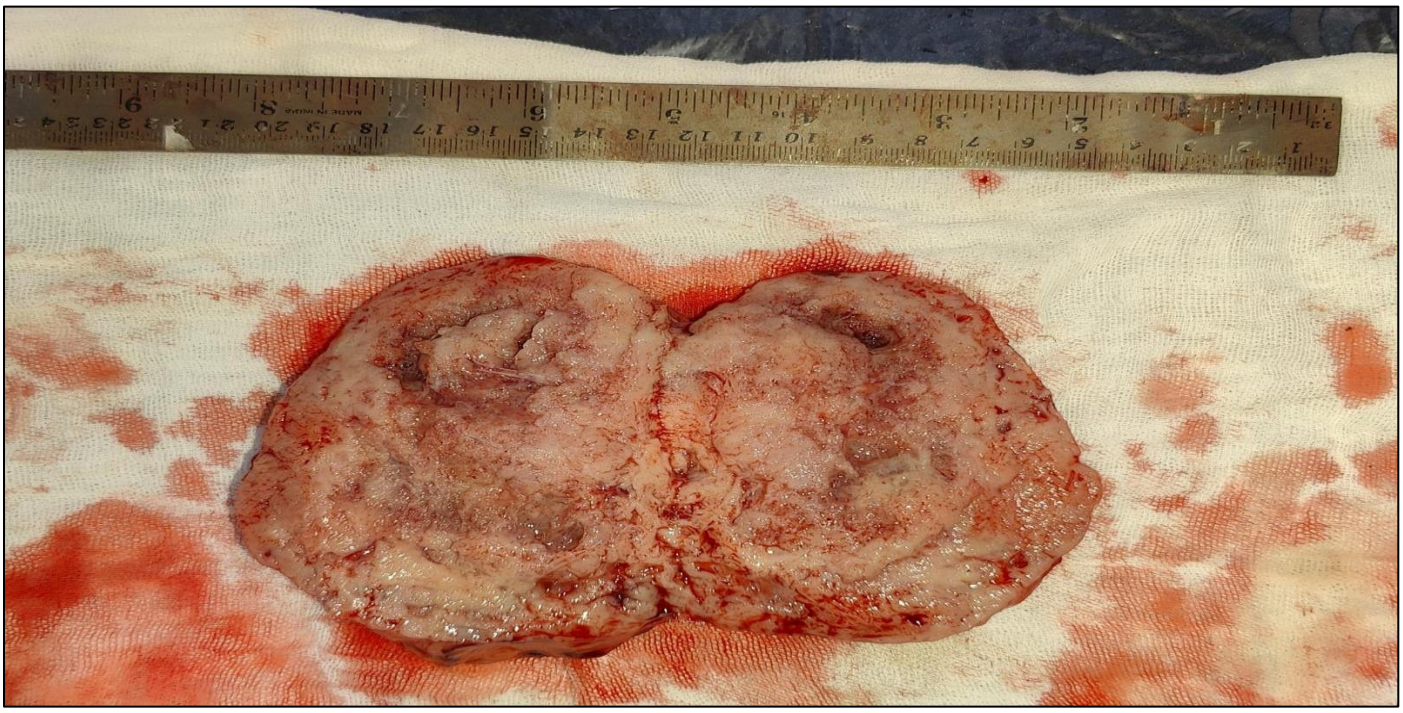

Fig 4 Cut Specimen of GIST

III. DISCUSSION

GISTs commonly occur in the age group of 50-70 years with rare cases in the paediatric age group, slightly more common in male. GISTs most commonly seen in the stomach (60-65%), followed by jejunum and ileum (25-30%), duodenum (5%), colorectum (4%), and esophagus or appendix (1%).

Patients can present with abdominal pain, bleeding, perforation, obstruction or sometimes may be asymptomatic. Symptoms are highly dependant on size (largest one 20 cm and smallest one 2 cm) identified incidentally on GI endoscopy, radiological procedure or autopsy. Majority of GISTs are sporadic, but have also seen in association with neurofibromatosis type 1 (NF1), Carney triad and Carney Stratakis syndrome. CD 117 and C-KIT is the most specific immunohistochemical marker.

GISTs are well defined tumour masses and on cut section tan-white with areas of haemorrhage, necrosis and degeneration.

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➢ *GISTS have three Subtype :*

- Spindle cell type (70%)
- Epihelioid cell type (20%)
- Mixed type (10%)

Immunohistochemistry for CD 117 (KIT) shows positivity in 90% of GISTs with DOG 1 showing positivity for 1/3rd of CD 117 negative cases showing that it is a promising new marker with even greater sensitivity and specificity for GIST. CD 34 shows positivity in 50-90% of cases.

The differential diagnoses for spindle cell GISTs are leiomyoma, leiomyosarcoma, intra-abdominal desmoid fibromatosis, schwannoma, inflammatory myofibroblastic tumor and solitary fibrous tumor. The differential diagnoses for epithelioid GIST include neuroendocrine neoplasms.

Diagnostic intervention may include Computed tomography (CT), upper GI scopy and PET scan. Endoscopic ultrasound with FNAC is useful in upper GI tract GIST. Gastric lesion have more favarable prognosis compared to other location.

Management of GISTs include surgical removal with negative margin and use of tyrosine kinase inhibitors (imatinib)as adjuvant therapy. Depending on the location and size of lesion surgical intervention is indicated.endoscopic resection , open or laproscopic surgical resection , resection with combined procedure and transgastric surgery may be performed.

IV. CONCLUSION

Gastrointestinal stromal tumors (GISTs)are the most prevalent mesenchymal tumors of the gastrointestinal tract. The most common location in the stomach is body. Abdominal pain , abdominal lump , bleeding are common symptoms. . Understanding the specific genetic mutations associated with GIST is important for guiding the treatment and prognosis of patients .depending on the size and location surgical intervention is required. The only limitation of the study is that it is a retrospective case series. Additionally molecular profiling of GISTS was not possible due to limitation in resources and infrastructure.

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