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Case Report

A rare presentation of gastric gastro-intestinal stromal tumour managed by stomach preserving surgery: a case report

Arun S. Patil, Dnyaneshwar Raut, Aishwarya V. Swami*, Khadeija A. Hussain, Kanishk N. Patil

Department of General Surgery, Grant Government Medical College and Sir J. J. Group of Hospitals, Mumbai, Maharashtra, India

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***Correspondence:**
Dr. Aishwarya V Swami,
E-mail: swamiaishwarya14@gmail.com

ABSTRACT

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the alimentary canal that account for 0.1–3% of all gastrointestinal malignancies. Vast majority of these tumors have oncogenic gain-of-function mutations of the KIT receptor tyrosine kinase. The mainstay of treatment is complete surgical resection followed by adjuvant therapy with tyrosine kinase inhibitors (Imatinib). We present a case report of 47 years old male presenting with a large abdominal lump, later diagnosed as a Gastrointestinal stromal tumor and underwent stomach preserving sleeve gastrectomy.

Keywords: GIST, Abdominal lump, Imatinib, DOG1

INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the alimentary canal that account for 0.1–3% of all gastrointestinal malignancies. Lesions can occur anywhere along the gastrointestinal tract, with the majority arising in the stomach (60%) and small intestine (30%).[1] The median age of presentation is 60 years with equal incidence in males and females with no racial/ ethnic predilection. The majority of GIST is sporadic but hereditary GIST can occur in Von Recklinghausen's neurofibromatosis, Carney's triad, and Carney – Stratakis syndrome.[2] Sporadic mutations within the tyrosine kinase receptors of the interstitial cells of Cajal-pacemaker cells of the gastrointestinal tract, have been identified as the key molecular step in GIST carcinogenesis. Although many patients are asymptomatic, the most common associated symptoms include- abdominal pain, dyspepsia, gastric outlet obstruction, and anorexia. Rarely, GIST can perforate causing life-threatening hemoperitoneum. Most are ultimately diagnosed on cross-sectional imaging studies (i.e., computed tomography and/or magnetic resonance imaging in combination with upper endoscopy.[3]

CASE REPORT

A 47 years old male presented with an abdominal lump and vague pain in the abdomen on and off for 1 month. There were no other associated general or GI tract symptoms. Clinical examination revealed an averagely built male with pallor. Abdominal examination revealed mobile, firm to a hard, non-tender, mass of size approximately 16×14 cm occupying the epigastrium, left hypochondrium, and extending into left lumbar region. The rest of the abdominal examination was unremarkable. Routine blood investigations were normal except for low hemoglobin. Ultrasonography (USG) 2021 showed a well-defined heterogeneous solid cystic lesion of size 15×17×18 cm in the left hypochondrium and epigastric region. Mass was

causing displacement of the small bowel laterally and stomach superiorly. The stomach is not visualized separately.

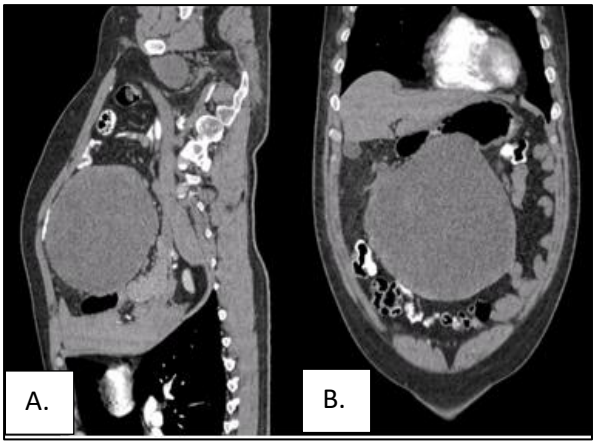

Figure 1: CECT scan showing exophytic mass arising from stomach displacing small bowel laterally (A) sagittal section (B) coronal section.

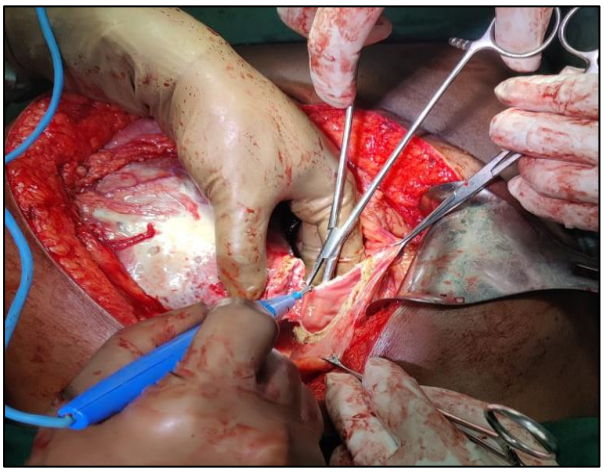

Figure 2: Intraoperative imaging showing gastrotomy with 1 cm margin resection.

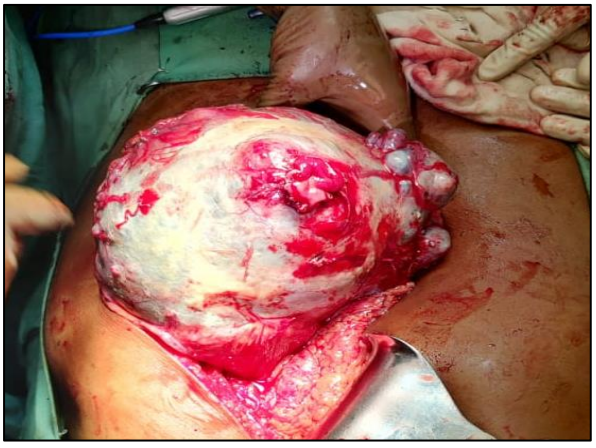

Figure 3: On table tumor removed completely without rupture.

Contrast-Enhanced Computed Tomography (CECT) 2021(Figure 1) showed A large well defined exophytic heterogeneously lesion with a thin rim of peripheral enhancement and central non enhancing component measuring 13.6×17.5×17 cm is noted arising along the greater curvature of the distal part of the body of the stomach for a length of 6.4 cm. It was protruding into the lumen with loss of fat planes with the stomach wall. Overlying gastric mucosa shows mild thickening. The rest of the part of the stomach wall appears uninvolved. Anteriorly the lesion is seen abutting the transverse colon with maintained fat planes. Other organs were unremarkable.

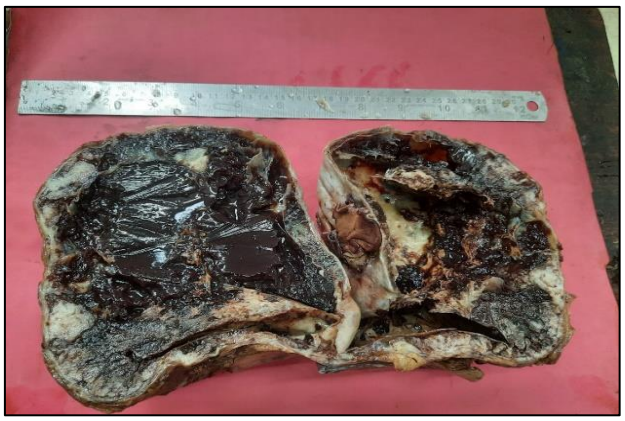

Figure 4: Histopathological specimen (gross) showing central necrosis.

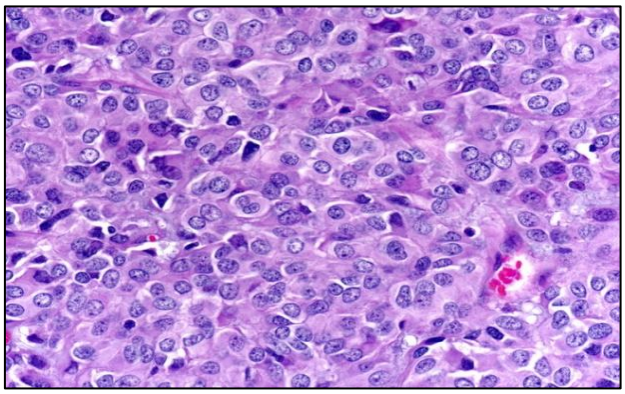

Figure 5: Microscopic image showing epithelioid cells of GIST.

Upper gastrointestinal endoscopy (2021) shows a smooth round indentation in the posteroinferior wall of the stomach, likely arising from the greater curvature, with no features of ulceration. No biopsy was done and a decision was taken to go for surgery.

Exploratory laparotomy revealed a 16×14×10 cm tumor arising from the greater curvature of the stomach, exophytically, with a sessile base. There was no infiltration of the mass into the surrounding structures, nor any evidence of metastases or lymphadenopathy. The tumor was excised with a sleeve of the greater curvature of the

stomach with a 1 cm margin and repaired in 2 layers (Figure 2). The tumor is seen attached closely to the transverse mesocolon which was separated as close to the tumor and distally from the root of the mesentery. Tumour excised as a whole (Figure 3). On re-inspecting, the bowel, a 10cm segment of the transverse mesocolon appeared dusky with a compromised blood supply. Resection of the dusky bowel segment and side to side anastomosis of the prox and distal loops of the colon was done. Histopathology (Figure 4) of the tumor revealed 16×14×10 cm-sized neoplasm arising from the gastric muscularis propria. Gastric mucosa and submucosa are free. Tumour is composed of epithelioid cells (Figure 5). Mitosis was less than 5/50 HPF. These findings favored the diagnosis of a gastrointestinal stromal tumor of intermediate risk of recurrence. The resected margin was reported clear of the tumor. On immunohistochemistry, the tumor is showing positive for DOG1/CD34 and negative for CD117(c-kit)/S100 protein.

DISCUSSION

GISTs are the mesenchymal tumor that arises from the interstitial cells of Cajal. Representing only 0.2% of all gastrointestinal (GI) tumors. In one study where a systematic review was done on patients undergoing Bariatric surgery and quoted that GISTs were found 2% incidentally.[4] It is the most common sarcoma of the GI tract which has to be differentiated from leiomyoma and leiomyosarcoma. The median age of presentation is 65 years. Equal in male and female (1:1). 95% of them are sporadic, while 5% are associated with congenital syndromes. Incidence was difficult to assess previously. But with the discovery of KIT (CD117) (95%) and CD34 expression (70%) in GIST, incidence is estimated to be around 7-15 cases / 10,00,000. The vast majority of these tumors have oncogenic gain-of-function mutations of the KIT receptor tyrosine kinase.[5] Most cases that lack a KIT mutation will have a PDGFRA mutation. Site of presentation are stomach (56%), small bowel (32%), colon and rectum (6%), esophagus (0.7%), other sites (5.5%).

Clinical presentation is variable and reflects the variability of appearance, location, and biological behavior. Small tumors are more likely to be asymptomatic, regardless of location. The tumor tends to grow intramural, bulging intraluminal or extramural if large enough, while larger tumors are known to ulcerate and cause GI hemorrhage. It can present with non-specific gastrointestinal symptoms (example- abdominal pain, nausea, vomiting). Aggressive tumors may present with metastases or symptoms relating to local disease.

CECT of the abdomen shows the mass of heterogeneous origin with central areas of necrosis with peripheral enhancement without any lymph node enlargement. Sometimes in the case of malignant disease focal lesions can be seen. On upper GI endoscopy smooth appearing, the round, submucosal tumor can be identified. Diagnosis can be done by Endoscopic Ultrasound (EUS) guided biopsy, which has a diagnostic accuracy of sensitivity 82% and specificity of 100%.

There are no standard criteria for assessing aggressive behavior and predicting the clinical prognosis of GISTs, although the NIH and AFIP criteria are widely recommended. It is commonly accepted that all GISTs are considered to have malignant potential. Through multivariate analysis, higher mitotic rate and tumor metastasis or local invasion before treatment were revealed to be associated with poor survival in GIST patients, and non-gastric disease has a high recurrence rate.[6]

Table 1: NIH criteria for risk assessment.[7]

	Tumor size (cm)	Mitotic count (per 50 HPF)
Very low risk	<2	<5
Low risk	2-5	<5
Intermediate risk	<5	6-10
	5-10	<5
	>5	>5
High risk	>10	Any
	Any	>10

The mainstay of treatment is complete surgical resection. The curative intent in the treatment is operative excision with a clear margin, R0 without causing tumor rupture. As GIST does not infiltrate the gastric wall, a wide normal mucosa margin is not needed, although some recommend a 1cm margin. However, the microscopic negative margin is sufficient for R0 resection.[8]

In a study of 48 patients, 33 underwent laparoscopic GIST resection and 15 underwent open resection. The laparoscopic group had a higher incidence of R1 resection but none developed recurrence or metastasis during follow-up.[9]

In a study, 50 patients to undergo significantly less extensive surgical procedures and resulted in a stomach preservation rate of 96% and R0 resection in 94%.[10] The Laparoscopic surgery is recommended for GISTs that are less than 5 cm and located in the stomach and small bowel. laparoscopic transgastric resection (LTGR) of a gastrointestinal stromal tumor (GIST) near the esophagogastric junction (EGJ) can be performed safely and effectively in a patient with morbid obesity.[11] All GISTs ≥2 cm in size should be resected when possible because none of them can be considered benign. Wedge or full-thickness partial gastrectomy is an effective strategy for the tumor that is located along the lesser or greater curvature of the stomach.[8] Posteriorly based gastric GIST often require transgastric resection through an anterior longitudinal gastrotomy; the tumor is everted and its pedicle divided with a linear stapling device.[12]

As in the present case-patient had only large GIST from the stomach with no metastasis or lymphadenopathy, hence excision of the tumor along with a sleeve of stomach and hand sewen closure in 2 layers was done. The Histopathology report confirmed the mass as a Gastrointestinal stromal tumor with the size of 16×14×10 cm with a mitotic count <5/50 HPF which falls in the intermediate-risk category according to the National Institute of Health (NIH) criteria.

Imatinib is a TKI that works by binding to the ATP binding sites on CD117 and PDGFRA, blocking signal transduction. GISTs that are CD117 and PDGFRA positive are thought to benefit from this therapy. One study developed a nomogram to predict recurrence-free survival (RFS) based on tumor size (cm), location (stomach, small intestine, colon/rectum, or other), and mitotic index (<5 or ≥5 mitoses per 50 high-power fields). The nomogram accurately predicts RFS after resection of localized primary GIST and could be used to select patients for adjuvant imatinib therapy.[13]

Imatinib can also be used for downstaging in unresectable tumors or preventing recurrence after surgery has been performed. It should be given for 12 months and can be extended to 36 months. One study compared with 12 months of adjuvant imatinib, 36 months of imatinib improved survival of GIST patients with a high risk of GIST recurrence.[14] In the present case-patient was started on Imatinib (dose- 400 mg daily) after consultation with an oncologist.

CONCLUSION

Gastrointestinal stromal tumors (GISTs) are a group of biologically distinct tumor which is different from other smooth muscle and neural tumors of the gastrointestinal tract (GIT). They are diagnosed whenever a large mass submucosal in location is seen to arise from the stomach. Management of such large GIST should be wide surgical resection (stomach preserving) followed by tyrosine kinase inhibitors (Imatinib) depending on Immunohistochemistry.

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