

Spontaneous perforation of GIST of the Small Intestine: A Rare Cause of Perforation Peritonitis

Amandeep Saharan¹, Satish Dalal², Mahavir Singh³, Chisel Bhatia⁴, Rajeev Sen⁵

ABSTRACT

Gastrointestinal stromal tumors (GIST) are relatively rare and often present with vague symptoms. Their first clinical manifestation as acute abdomen due to perforation is extremely rare. We report a rare case of spontaneous perforation of exophytic gastrointestinal stromal tumor (GIST) causing perforation of jejunum. A 61 year, male patient presented with symptoms of acute abdomen. After evaluation, a laparotomy was performed, where perforation of a tumor in the jejunum was found. The perforated part along with the tumor was resected and the histopathological examination showed that the tumor was GIST. In emergency setting the patients with perforation peritonitis, a diagnosis of spontaneous perforation of GIST though rare should be kept in mind specially in elderly patients.

Key words: Gastrointestinal stromal tumors (GISTs), Perforation peritonitis, Small intestine.

**Amandeep Saharan¹,
Satish Dalal², Mahavir
Singh³, Chisel Bhatia⁴,
Rajeev Sen⁵**

^{1,2,3,4}Department of Surgery, Post Graduate Institute of Medical, Rohtak, Haryana, INDIA.

⁵Department of pathology, PGIMS, Rohtak, Haryana, INDIA.

Correspondence

Dr. Amandeep Saharan

House No. 185, Sector 1, Rohtak, Haryana 124001 INDIA.

Phone: +918198911209

Email: amandeephane@gmail.com

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INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are rare, representing only 0.2% of all GI malignancies.^[1] The possibility of presence of malignancy is 20-30%.^[2] GISTs are difficult to diagnose and are often at advanced stage at the time of definitive treatment. They are characterized by indolent clinical symptoms including vague abdominal pain, weight loss, occult GI bleeding and obstruction, which is caused by the growing tumor. GISTs can occur anywhere in the gastrointestinal tract. They are submucosal lesions, which most frequently grow endophytically in parallel with the lumen of the affected structure. GISTs have a wide range of malignancy degree and it is preferable that they be considered and treated as potentially malignant lesions.^[1] Small tumors that are completely resected and are of intermediate malignancy seldom create postoperative complications, while larger tumors may recur after resection, or present with intraperitoneal infiltration or hepatic metastasis. Prognosis in perforated GISTs is poor until there is complete surgical removal of the tumor. We present the diagnosis and management of a patient, who presented in emergency with acute abdomen resulting from a spontaneous perforation of gastrointestinal stromal tumor (GIST).

CASE REPORT

A 61-years old, male presented in emergency department with complaints of a pain abdomen, vomiting and abdominal distension for two days with poor chest condition. Patient was referred from a private hospital with diagnosis of perforation peritonitis with severe anemia. His haemoglobin was docu-

mented as 5.8 g/dl. There was medical history of passing dark colored stool for past six months. He had no prior abdominal operations. Abdominal examination revealed a tense, tender and distended abdomen with rigidity. Routine laboratory investigation in our institute revealed haemoglobin of 6 g/dl and a raised total leukocyte count i.e. 13500/mm.^[3] Abdominal X-ray (erect) showed gas under both domes of diaphragm. A diagnosis of perforation peritonitis was made, patient was stabilised with IV fluids, antibiotics and two unit blood were transfused before the patient was taken up for emergency operation. Intraoperatively, around 1000 ml of bilious fluid was present in abdominal cavity and there was a tumor mass of 6×4.5cm present in jejunum about five feet from dedunojejunal junction at antimesenteric wall with perforation of size 2×1 cm over the growth (Figure 1a and b). The part of jejunum containing the tumor was excised with about five cm margins from both side of tumor and an end-to-end jejunojejunal anastomosis was done. On microscopic examination, the diagnosis of the GIST was confirmed (Figure 2). Mitotic activity was >5 per 10 hpf. On immunohistochemistry, the tumor was strongly positive for CD117 (Figure 3) and positive for vimentin (Figure 4) while negative for CD34, S-100 and SMA. The patient was shifted to Respiratory ICU considering the poor chest condition. He was stable from surgical point but his chest condition did not improve despite aggressive treatment and he ultimately expired on 4th post-operative day

DISCUSSION

Gastrointestinal stromal tumors are rare neoplasms of the gastrointestinal tract ranking a distant third af-

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Figure 1(a): Perforation in small-intestine exophytic gastrointestinal stromal tumor.



Figure 1(b): Perforation in small-intestine exophytic gastrointestinal stromal tumor.

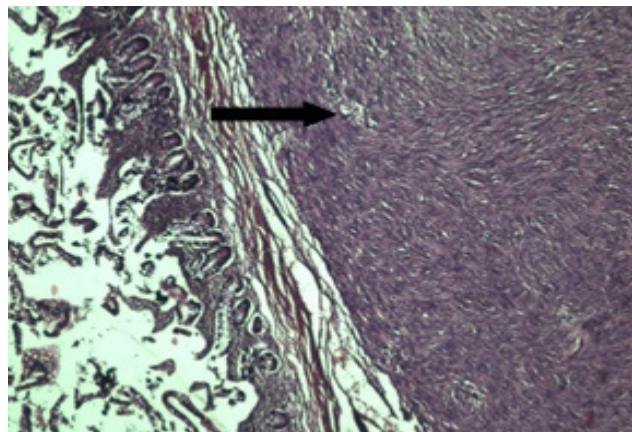


Figure 2: Showing tumor cell on H & E stain (black arrow).

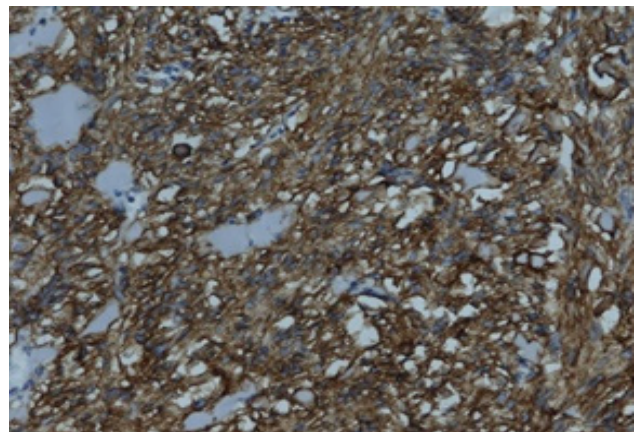


Figure 3: Immunohistochemistry positive for CD117.

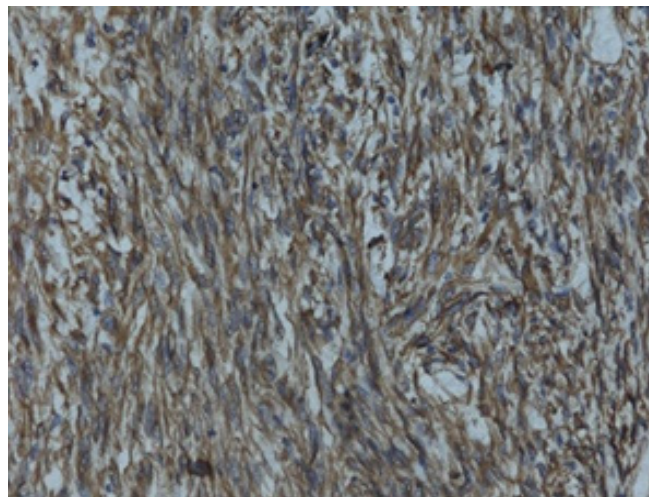


Figure 4: On Immunohistochemistry tumor is also positive for Vimentin.

ter adenocarcinomas and lymphomas.^[4] Clinical presentation of patients varies according to site, size and aggressiveness of the tumor. Symptoms manifest when the tumor is large (>5cm) or situated in a critical anatomical location (gastric outlet obstruction).^[4] Symptoms may include abdominal pain, mass, nausea, vomiting, anorexia and weight loss. Patients may also present with gastrointestinal bleeding (due to pressure necrosis and ulceration of the overlying mucosa) or with obstructive symptoms. GISTs can occur anywhere in the gastrointestinal tract. They are sub-mucosal lesions that often grow intraluminally. Sometimes they have an extraluminal exophytic component. The stomach (60%) is the commonest site followed by small intestine (30%), duodenum (5%), colon/rectum (5%) and esophagus (<1%).^[5] GIST is observed predominantly in adults at a median of around 60 years. The incidence is equal in men and women. GIST arises from interstitial cells of CAJAL, pacemaker cells of the gastrointestinal tract responsible for initiation of peristalsis.^[6] GISTs can be categorized as low or high-risk tumors by taking into account the possibility of metastasis at recurrence. However, the main prognostic factor is the mitotic count. A prognostic classification was defined by Fletcher *et al* and is widely accepted and used today.^[7]

Immunohistochemical examination of GISTs is always positive for KIT

Table I: GISTs classification by Fletcher *et al*⁷

Risk of malignancy	Size of tumor (largest dimension in cm)	Mitotic count(/50HPF)
Very low	<2	<5
Low	2-5	<5
Intermediate	<5	6-10
	5-10	<5
High	>5	>5
	>10	Any counts
	Any size	>10

protein (CD117 antigen), while the positivity regarding other markers varies (Table II).^[1-3]

Table II : Proportion of GISTs positivity for various immuno-histochemical markers

Marker	Positivity
KIT	100%
CD34	70%
SMA	20-30%
S100	10%
Desmin	<5%

The treatment of choice for GISTs is the complete surgical excision of the tumor, where ever possible, including the tissues that are infiltrated. Patients after complete surgical resection have 48-65% five-year survival. Partial resection should only be performed in cases of large tumors, for palliative purposes or complications such as compression of other or-

gans, hemorrhage, or pain.^[1] When the tumor presents with symptoms or signs such as perforation, multifocal location or metastatic lesions prognosis is generally very poor. Perforation of the tumor lowers the five-year survival to 24%, probably due to peritoneal dissemination.^[8] These patients have a similar prognosis as patients with incomplete tumor resection, with shorter disease-free survival. GIST response to conventional chemotherapy is very poor (<10%), while radiotherapy is only used in cases of intraperitoneal hemorrhage, when the precise location of the tumor is known, or for analgesic purposes. Imatinib, was found to act as a powerful selective inhibitor of tyrosine kinases (c-ABL, bcr-ABL), of PDGFR receptor (platelet derived growth factor receptor) and of c-kit receptor shows good result in metastatic or non resectable tumor or as adjuvant therapy following complete resection.¹

CONCLUSIONS

In emergency setting the patients presenting with diffuse peritonitis and severe anemia, a diagnosis of spontaneous perforation of GIST though rare should always be kept in mind specially in elderly patients. A high degree of suspicion is necessary in view of the high morbidity rates resulting from a delayed diagnosis of the disease.

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CONFLICT OF INTEREST

No conflict of interest are declared.

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