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Dr. Dharmendra K Shah
Head of Unit, Department of
Surgery, Medical College
Baroda, SSG Hospital, MS
University, Gujarat, India

Dr. Shivani Chaudhary
Senior Resident, Department
of Surgery, Medical College
Baroda, SSG Hospital, MS
University, Gujarat, India

Dr. Nriya Trivedi
Intern, Department of
Surgery, Medical College
Baroda, SSG Hospital, MS
University, Gujarat, India

Corresponding Author:
Dr. Nriya Trivedi
Intern, Department of
Surgery, Medical College
Baroda, SSG Hospital, MS
University, Gujarat, India

Ruptured haemorrhagic gastrointestinal stromal tumour causing spontaneous hemoperitoneum: A case report with Review of Literature

Dr. Dharmendra K Shah, Dr. Shivani Chaudhary and Dr. Nriya Trivedi

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Abstract

Gastrointestinal stromal tumour (GIST) is a common mesenchymal tumour. Often GIST is associated with intraluminal gastrointestinal bleeding but rarely undergoes rupture causing spontaneous hemoperitoneum. This case report talks about spontaneous hemoperitoneum being the initial presentation of a GIST and emphasizes that a high index of suspicion and prompt diagnosis could prevent its deadly complications. In our case, radiological investigations and later, excision revealed it to be of a diameter of 16cm; though interestingly, producing only mild abdominal discomfort in the patient till it underwent a sudden rupture.

Keywords: Hemorrhagic GIST, Spontaneous Hemoperitoneum

Introduction

Spontaneous hemoperitoneum is rare and a life-threatening entity with many possible differential diagnoses. GIST is an infrequent cause responsible for producing this condition [1, 2]. Other rare causes of spontaneous hemoperitoneum are ruptured cystic artery pseudoaneurysm, ruptured splenic artery aneurysm, and spontaneous variceal rupture due to portal hypertension [2]. GIST are mesenchymal smooth muscle tumours that may arise in any part of the GI tract. They account for <1% of all GI tract cancers [3]. The most common site of GIST is the stomach (60%), followed by jejunum- ileum (30%) and duodenum (5%), colo-rectum, and oesophagus. It can also occur in the retroperitoneum, mesentery, and omentum (i.e., extra gastrointestinal GIST) [4, 5].

GISTs may present as a part of the Carney triad (other lesions including paragangliomas and Para-chondromas) [6]. In a large retrospective series of gastric GISTs [7], was noted that these tumours occurred in patients 40 years and above, a finding confirmed by Indian studies [8, 9]. These tumours present with a median diameter of 6 cm [8].

Case Presentation

A 45-year-old lady presented to us with complaints of abdominal pain, vomiting, and for two days. The pain was diffuse & not accompanied by anorexia, diarrhoea, jaundice, vomiting, or fever. On examination, the patient had tachycardia and tachypnoea with hypotension. On per abdominal examination mild generalized tenderness present with an approximately 6x6 cm sized single, firm lump palpable in epigastrium with smooth surface, lump doesn't move with respiration and dull on percussion, which is separate from liver dullness. After stabilizing, laboratory tests revealed low haemoglobin (3.80 gm/dl). The contrast-enhanced computed tomography (CECT) of the abdomen and pelvis with a CT angiogram of the abdominal aorta revealed a large heterogeneously enhancing hypodense lesion with non-enhancing hypodense areas. (Figure 1) A neoplastic mass with haemorrhagic collection (in the region of gastro hepatic ligament and extending into the lesser sac) possibility of ruptured haemorrhagic GIST with hemoperitoneum. The patient was optimized by transfusing packed red blood cells (PCV) and posted for definitive surgery.

The patient underwent a laparotomy with a rooftop skin incision and drained approximately 500 millilitres of hemoperitoneum. A 16 cm x10 cm x6 cm ruptured tumour mass along with total gastrectomy (Figure 2, 3) followed by Roux-en-Y end to side esophago-jejunostomy and side-to-side jejuno-jejunostomy was done.

Histopathology analysis (Figure 4) confirmed the tumour type to be of low-grade GIST (grade 1) with mitotic rate $< 5/5\text{mm}^2$ and with tumour-free resection margins, Pathological grading as per AJCC (American joint

committee of cancer 8th edition) was pT4pNxMx. Adjuvant Imatinib therapy (400mg daily) started. The postoperative course was uneventful. We discharged the patient after suture removal on the 14th day postoperatively.

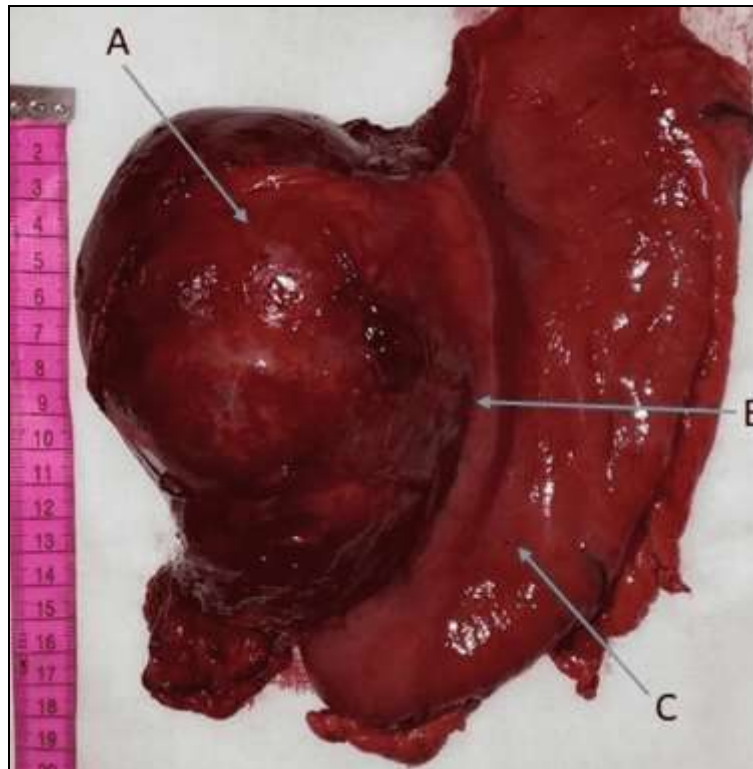


Fig 1: Post-operative specimen showing A: anterior surface of GIST tumour with intact capsule, B: lesser curvature of stomach, C: stomach



Fig 2: Post operative specimen showing A: stomach, B: posterior surface of ruptured GIST

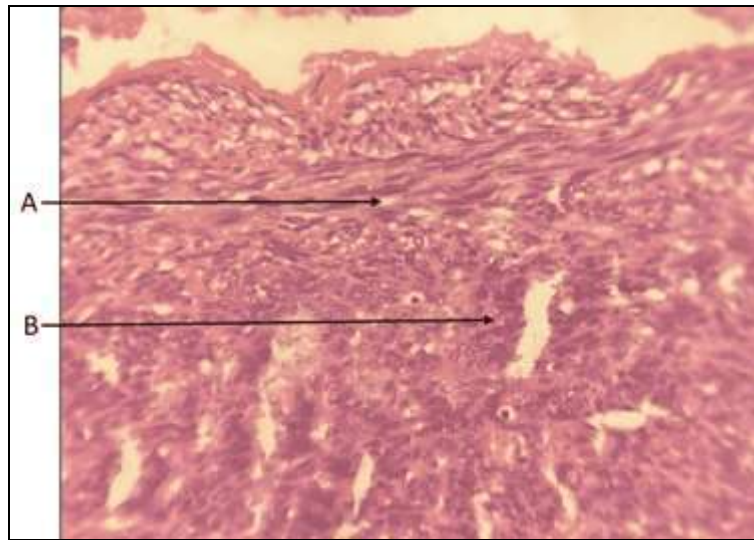


Fig 3: A: Spindle cells in fascicles B: oval and spindle shaped tumour cells with bland nuclei

Discussion

Large GIST may ulcerate or present with GI bleed ^[1]. The presence of only vague, mild abdominal symptoms in a patient harbouring approximately a 16cm GIST (as in our patient) is unusual as a population-based study showed that the tumour size is 8.9 cm in patients with clinical symptoms, which is about 70% of GISTs studied, 2.7 cm in patients without symptoms, 20%, and 3.4 cm in patients with GISTs detected at autopsy, 10% ^[10] Such a large and more or less asymptomatic GIST producing spontaneous hemoperitoneum as its initial presentation is not a common occurrence.

The most common aetiologies for sudden intraperitoneal haemorrhage are gynaecologic, splenic, hepatic, and vascular. Studies have reported ovarian cyst and post-procedure haemorrhage as the two most common causes of hemoperitoneum in women ^[6]. Gastrointestinal stromal tumours (GISTs) are commonly associated with intraluminal bleeding but rarely with spontaneous hemoperitoneum ^[1, 2].

Most GISTs remain silent or asymptomatic. Symptoms produced depend on location and size -the plethora of symptoms being- anorexia, dyspepsia, upper GI bleed, and abdominal pain. Lymph node metastasis is uncommon. Spread to the liver or lungs in advanced and aggressive cases are reported ^[11]. Tumour rupture is a risk factor predictive of recurrence ^[12] even after macroscopic and complete resection of gastrointestinal stromal tumours. Tumour rupture indicates a defined interval or even lifelong adjuvant therapy with imatinib ^[13].

Early events in the development of GIST are activating mutations in the KIT or the PDGFRA gene. These encode for mutated tyrosine receptor kinases. The mutations occur in most GIST tumours. A small minority of GISTs may have germline mutations in SDH, suggesting a potential role of SDH in pathogenesis. A mutation in the DOG1 gene was identified in GIST pathogenesis recently ^[14]. Identifying the occurrence of these mutations in these receptors is of prognostic importance as these are potential chemotherapeutic targets. Tyrosine Kinase Inhibitors like Imatinib mesylate, SU111248 - inhibit Tyrosine Kinase & block PDGFRA. Drugs like Sunitinib and Dasatinib are used in Imatinib refractory cases.

Conclusions

A spontaneously occurring hemoperitoneum is an uncommon sequela/complication of a gastrointestinal stromal tumour. This occurrence significantly raises concerns and increases chances of the reappearance of this tumour and significantly reduced expected life expectancy. More such case reports with a longer duration of follow-up are needed to comment on the prognosis and survival of ruptured haemorrhagic GIST.

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