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GIST JEJUNUM Presented as HOLLOW VISCUS PERFORATION –A rare Aetiology

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ABSTRACT

A 55yr old diabetic, hypertensive male with prior history of coronary stenting presented to the emergency department with complaints of abdominal pain and distension of duration 4 days. The abdomen was grossly distended with diffuse tenderness and guarding The Patient was evaluated resuscitated, investigated, and found to be Hollow viscous perforation with peritonitis. The Patient was subjected to emergency laparotomy. The surprise finding in the Operating room was mass arising from jejunum with perforation. The Resected specimen histopathology revealed GIST jejunum which was an unusual cause of acute abdomen.

KEYWORDS: Gastrointestinal stromal tumor, GIST, Jejunum, Perforation.

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INTRODUCTION

Gastrointestinal stromal tumors are the most common sarcomatous tumors of the GI tract. Malignant GISTs constitute about 20% of malignant neoplasms of the small bowel. These tumors are more common in jejunum and ileum, diagnosed in the fifth and sixth decades of life. GISTs vary considerably in their presentation and clinical course, ranging from small benign tumors to massive lesions with necrosis, hemorrhage, and wide metastasis, but rarely do they present with bowel perforation. In this journal, we would like to discuss the points to be aware of while managing such a scenario.

CASE REPORT

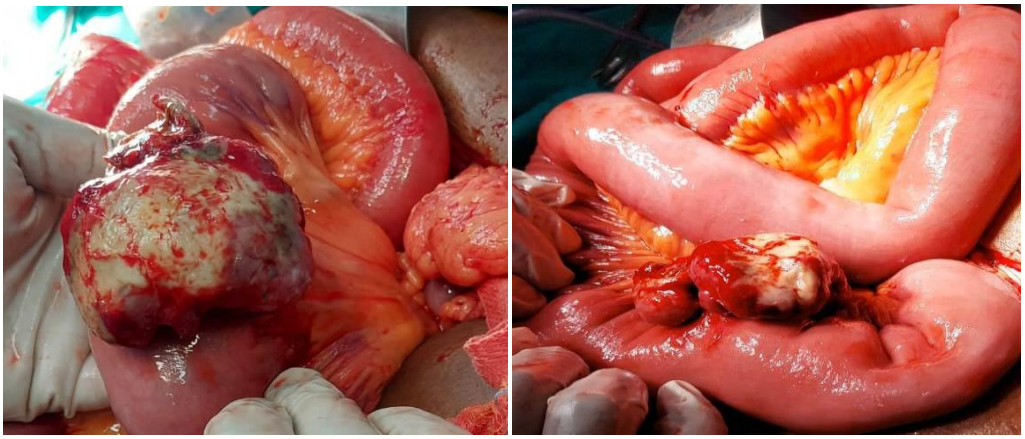
INITIAL PRESENTATION

A 55-year-old male, diabetic, hypertensive, with Coronary artery disease with a history of PTCA presented to the emergency department with abdominal pain and distension of 4 days duration. The pain was initially mild in intensity in the upper abdomen but later it was severe and diffuse. The patient had a history of constipation for 1 day. On general examination, there was pallor, temperature of-100-degree Fahrenheit, a pulse rate of 120/minute, and blood pressure of 90/60 mm hg. There was abdominal distension with diffuse abdominal tenderness and guarding. The Liver dullness was obliterated with an absence of the bowel sound. The clinical diagnosis was made as Hollow viscous perforation with peritonitis.

The patient was resuscitated with crystalloids, intravenous antibiotics were initiated. Nasogastric aspiration and catheterization were done for input-output monitoring. Following initial stabilization, an X-ray abdomen revealed pneumoperitoneum. Abdominal ultrasound revealed free fluid in the abdomen with normal solid viscera. Blood investigations revealed neutrophilia with a leukocyte count of /L, and raised renal parameter. The retroviral status was negative. The patient was planned for Emergency Laparotomy.

SURGICAL COURSE

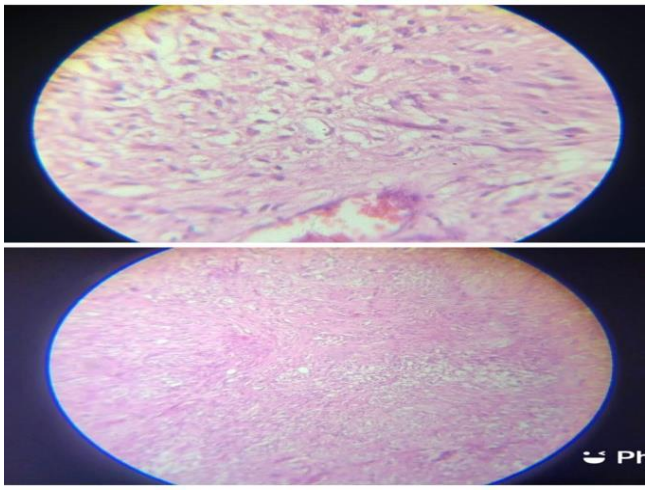
The patient underwent an emergency midline laparotomy. There were sero-purulent collections of 750 ml in the right paracolic gutter and pelvis. There was a mass of size approximately 7*6cm arising from the proximal jejunum from the serosal surface in the mesenteric border (Figure 1).



On examining the growth and observing the mesenteric border of the jejunum, a 1*1cm perforation was found (Figure 2).

As there is no major vessel involvement and minimal contamination with adequate proximal jejunum available, the plan was taken for resection of the specimen leaving a 5cm margin on either side.

Resection of the involved bowel segment containing growth and jejuno-jejunal end-to-end suture anastomosis was done. Histopathological examination revealed a malignant gastrointestinal stromal tumor of the jejunum of mixed type, with histological grade-G2 which is of high grade with free surgical margins (Figure 3).



pTNM stage - T3N0Mx. The postop patient was referred to medical oncology for further management.

DISCUSSION

Gastrointestinal stromal tumors (GISTs) are the more common sarcomatous tumors of the GI tract derived from the interstitial cells of Cajal, an intestinal pacemaker cell. Although GIST can arise in any portion of the GI tract, from the esophagus to the rectum, the small bowel is the second most common site of involvement (30%-40%), after the stomach (40%-60%)[1].

Primary GIST arising in the GI tract presents symptomatically in 69% of cases and is diagnosed incidentally at the surgery in 21% of cases, with the remaining 10% found incidentally at autopsy[2].

The Risk factors for GIST include a History of neurofibromatosis 1(Von Recklinghausen disease), male, age of 50 or more, and family history of GIST.

The diagnosis of GIST of small bowel may be delayed for several reasons, including its relatively low incidence, and non-specific and variable symptoms. The wide spectrum of radiological appearances, intestinal thickening, and the presence of overlapping loops of the intestine make imaging studies difficult. All of the above lead to delayed or misdiagnosis of GIST of the small bowel.

CLINICAL FEATURES

Symptoms of GISTs at presentation may be related to mass effects. Mucosal ulceration can cause blood loss, and approximately half of individuals with GIST, present with anemia or related symptoms. GISTs may also be discovered as an incidental finding during radiologic imaging, endoscopy, or abdominal surgery performed for other reasons.

The clinical symptoms associated with small bowel GIST are usually nonspecific and varied and are usually associated with tumor size and anatomical site. Jejunal GISTs, which comprise 10% of all GISTs [3], are usually symptomatic and patients suffer from abdominal pain and early satiety. They

may also have symptoms secondary to obstruction or hemorrhage. Perforation with acute diffuse peritonitis is rare. [4,5].

Other relatively rare presentations of small bowel GIST include anemia, intraperitoneal hemorrhage, and altered bowel motility. [6]. Between 12% and 18% of patients with small bowel, GIST has been reported to be asymptomatic, with the tumors detected incidentally [7].

The preoperative diagnosis of primary GIST of the small bowel is difficult to make due to the relative accessibility of the small bowel to conventional endoscopic examination. Also, it can be difficult to determine the nature of mass solely on imaging of the abdomen. An abdominal ultrasound scan is often the initial imaging test employed in the investigation of a patient with abdominal pain or mass, but ultrasound is not an accurate method for the detection of tumors of the small bowel. CT imaging helps in diagnosing small bowel GIST and can be used for detection, localization, staging, surgical planning, and the evaluation of response to therapy.

Although computer tomography is a viable imaging modality for patients suspected of having intraabdominal GIST, the magnetic resonance imaging modality provides a more accurate preoperative picture [8].

GISTs can be quite large, as much as 30cm in diameter. They usually form a solitary, well-circumscribed, fleshy mass covered by ulcerated or intact mucosa, but can also project outward toward the serosa.

The cut surface shows a whorled appearance. Metastasis may take the form of multiple serosal nodules throughout the peritoneal cavity or as one or more nodules in the liver, spread outside of the abdomen is uncommon.

GIST composed of thin elongated cells are classified as spindle cell type, whereas tumors dominated by epithelial appearing cells are termed epithelioid type; mixtures of the two patterns also occur.

Tissue histology, supported by immunohistochemistry, is required for the definitive diagnosis of GIST. GIST tumors have variable markers, including C-KIT (CD 117), DOG -1, CD 34,
SMA,S100, and Desmin, of which, are almost always positive for C-KIT and DOG -1 [9,10]. Approximately 75% to 80% of all GISTs have oncogenes, the gain of function mutations in the receptor tyrosine kinase KIT.

DOG 1, a recently defined monoclonal antibody against a chloride channel protein expressed by GIST, is positively expressed in 95% of GISTs.

MANAGEMENT

Surgery is the standard treatment for non-metastatic GIST. The main objective of surgical treatments is to achieve negative surgical resection margins (R0) and to resect the tumor without causing tumor rupture.

A complete en bloc resection is recommended, whenever feasible, in cases where contiguous organs are involved [11]. A laparoscopic approach in patients with large tumors is strongly not recommended. Radiologic criteria for unresectability include infiltration of the celiac trunk, superior mesenteric artery, or portal vein. Imatinib is first-line standard therapy for unresectable, metastatic, or recurrent GIST, and the standard dosage is 400mg/day [12].

PROGNOSIS

Prognostic factors include the anatomic location of the primary tumor, age at presentation, histomorphology, molecular genetics, and immunohistochemistry, of which tumor size is the most important [13]. It is difficult to diagnose a jejunal GIST preoperatively due to the nonspecific and variable clinical symptoms, and it is also difficult to distinguish the tumor based solely on images. The definitive diagnosis of GIST is revealed by histopathological examination of the specimen.

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