Case Report

Multiple small bowel gastrointestinal stromal tumors presenting as acute abdomen: A rare case report

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ABSTRACT

Gastrointestinal stromal tumors (GISTs) are rare malignant mesenchymal neoplasms of the alimentary tract that account for a small proportion of gastrointestinal tumors and most commonly arise from the stomach and proximal small intestine. Multiple synchronous jejunal and ileal GISTs are exceedingly uncommon, and presentation with acute abdomen due to complications such as perforation and intussusception is particularly rare. A 55-year-old male presented with an acute onset of severe abdominal pain, bilious vomiting, and constipation. Clinical evaluation and imaging were suggestive of acute intestinal obstruction secondary to a mid-jejunal mass. Emergency laparotomy revealed multiple small bowel tumors involving the jejunum and ileum, including a 10 × 8 cm jejunal mass with anti-mesenteric perforation with densely adherent omentum and a 4 × 3 cm mid-ileal mass serving as a lead point for intussusception. Histopathological examination of the resected specimens showed a tumor arising from the muscularis propria composed of atypical round to spindle cells arranged in fascicles, bundles, and sheets, with areas of atypical mitoses, necrosis, and tumor emboli, features consistent with high-grade GIST. The postoperative period was complicated by refractory septic shock, and the patient unfortunately succumbed on postoperative day 2 despite intensive care. This case emphasizes the importance of considering GISTs in the differential diagnosis of small bowel masses presenting with acute intestinal obstruction and highlights the potential for aggressive clinical behavior and poor outcome in large, high-grade tumors complicated by perforation.

Key words: Acute abdomen, Gastrointestinal stromal tumor, Ileal gastrointestinal stromal tumors, Intestinal obstruction, Jejunal gastrointestinal stromal tumors, Multiple small bowel tumors

Gastrointestinal stromal tumors (GISTs) are rare mesenchymal neoplasms comprising <1% of all gastrointestinal tumors, with an annual incidence of 10–20 cases/million population [1,2]. They most commonly originate in the stomach (60–65%), followed by the small intestine (25–35%) [3], typically presenting with nonspecific abdominal pain, gastrointestinal bleeding, early satiety, or incidental detection on imaging [4]. Acute abdominal presentations due to obstruction, perforation, or bleeding occur less frequently [5,6]. Multiple synchronous small bowel GISTs involving both jejunum and ileum simultaneously represent an exceedingly rare subset, with only isolated case reports documented in medical literature [7,8]. These cases pose significant preoperative diagnostic challenges as imaging findings often mimic other causes of intestinal obstruction, intussusception, or perforation, with definitive diagnosis requiring histopathological confirmation and characteristic immunohistochemical markers (CD117/c-KIT, DOG1) [9]. The clinical relevance lies in recognizing this rare entity in acute surgical abdomen presentations to guide appropriate management [10,11].

This case represents the first reported instance from our region of quadruple synchronous small bowel GISTs presenting with simultaneous jejunal perforation and ileal intussusception, highlighting extreme diagnostic rarity and aggressive multifocal behavior in a resource-limited setting.

CASE REPORT

A 55-year-old male presented to the emergency department with abdominal pain, bilious vomiting, and constipation for 4 days, not relieved with medications.

On examination, he had persistent tachycardia. Per abdominal examination revealed a distended abdomen with diffuse tenderness, guarding, and sluggish bowel sounds. Per rectal examination showed an empty,

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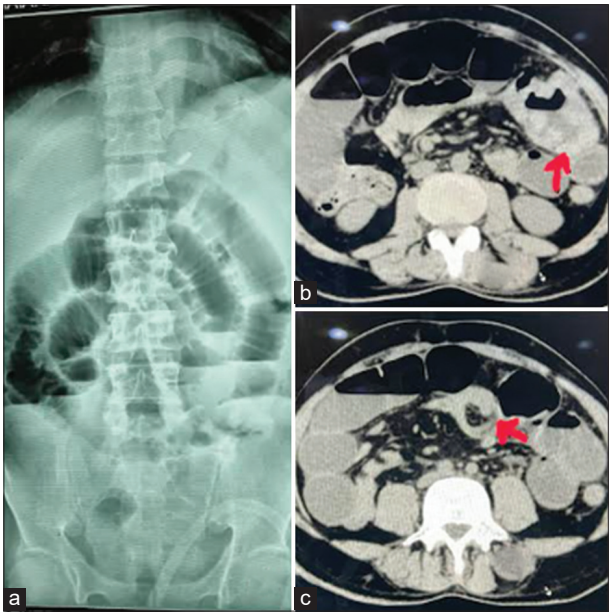

Figure 1: (a) X-ray Erect Abdomen showing multiple dilated small bowel loops with multiple air-fluid levels suggestive of acute intestinal obstruction, (b) CT abdomen showing jejunal mass (Red arrow), (c) CT abdomen showing jejuno-ileal intussusception (Red arrow showing target sign)

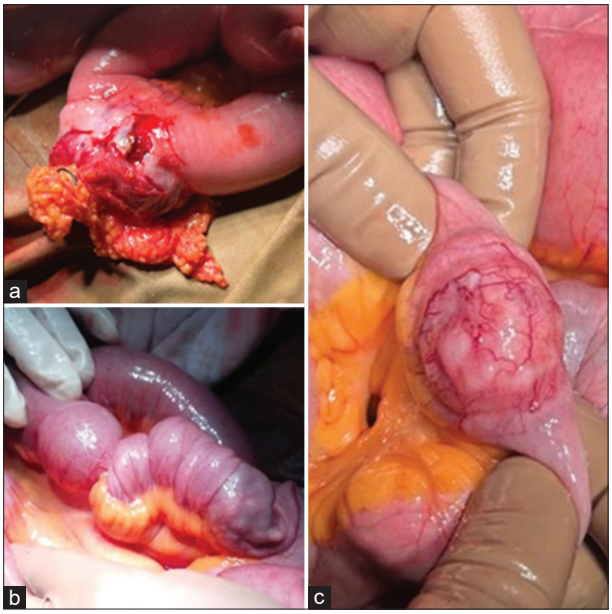

Figure 2: Intraoperative images. (a) 10 × 8 cm exophytic jejunal gastrointestinal stromal tumors (GIST) with antimesenteric perforation and omental adhesions, (b) 10–15 cm ileo-ileal intussusception secondary to 3 × 3 cm ileal lead-point mass, (c) Additional 3 × 3 cm ileal GIST

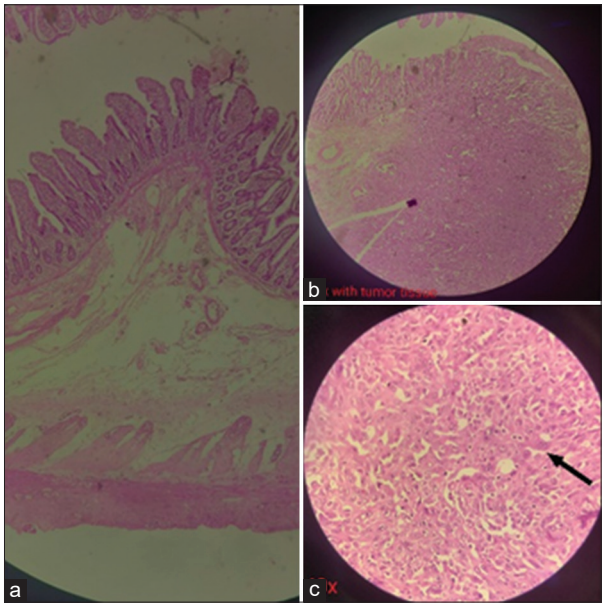

Figure 3: Histopathological images. (a) Normal small bowel mucosa showing crypts, villi, and intact muscularis propria (4×0), (b) Tumor arising from and completely replacing muscularis propria (×40), (c) Atypical round-to-spindle shaped tumor cells with nuclear polymorphism, high mitotic activity, and tumor emboli (black arrows, ×400)

Table 1: Laboratory investigations on admission

Parameter	Value	Reference range
Total leucocyte count	14,000/mm^3	4,000–11,000/mm^3
Hemoglobin	8.8 g/dL	13–17 g/dL
Serum creatinine	2.4 mg/dL	0.7–1.3 mg/dL
Serum albumin	2.4 g/dL	3.5–5.0 g/dL
Arterial blood gas	Metabolic acidosis with elevated lactate levels	

Table 2: Imaging Findings

Modality	Findings
X-ray abdomen	• Multiple dilated bowel loops with air fluid levels, suggestive of acute intestinal obstruction
CT abdomen	• Multiple grossly distended small bowel loops with air fluid levels with maximum bowel diameter 37 mm • Evidence of Narrowing at the level of mid-distal jejunum level suggestive Intraluminal/Mural Mass causing mechanical obstruction

collapsed rectum. The patient was resuscitated with intravenous fluids and analgesics, and a nasogastric tube and a Foley's urinary catheter were inserted.

Laboratory investigations of the patient are shown in Table 1. Imaging findings are summarized in Table 2 and Figure 1. X-ray abdomen showed multiple dilated bowel loops with air fluid levels, suggestive of acute intestinal obstruction. Computed tomography abdomen showed multiple grossly distended small bowel loops with air fluid levels, with a maximum bowel diameter of 37 mm. Evidence of narrowing at the level of the mid-distal jejunum was suggestive of an intraluminal/mural mass causing mechanical obstruction. In view of the clinical and radiological features of acute intestinal obstruction (Figure 1a-c), the patient was taken up for emergency exploratory laparotomy through a midline incision.

Intraoperatively, a 10 × 8 cm exophytic jejunal mass was identified approximately 20 cm from the duodeno-jejunal junction, with dense omental adhesions and a 0.5 × 0.5 cm perforation at the proximal end of the mass (Figure 2a). A 10–15 cm ileo-ileal intussusception was noted, secondary to a 3 × 3 cm intraluminal/mural ileal mass located about 50 cm from the ileocecal junction (Figure 2b). Two additional mural masses, measuring

2 × 2 cm and 3 × 3 cm, were identified in the jejunum and ileum with patent lumina (Figure 2c). The remaining small and large bowel, stomach, and solid organs appeared normal.

Segmental resection of the involved jejunal segment with the large mass and the ileal segment containing the intussusception and associated mass was performed following oncological principles, with primary jejuno-jejunal and ileo-ileal anastomoses. All resected specimens were submitted for histopathological examination.

Histopathology revealed a tumor arising from the muscularis propria of the small bowel, composed of atypical round to spindle-shaped cells arranged in fascicles, bundles, and sheets, with areas of marked nuclear atypia, high mitotic index, necrosis, and tumor emboli, consistent with intestinal GIST (Figure 3a-c).

Immunohistochemical confirmation (CD117/c-KIT, DOG1 positivity) was not performed due to the lack of facilities at our center, but it is recommended for definitive diagnosis as per standard guidelines.

Despite resuscitation and postoperative intensive care, the patient developed refractory septic shock and, unfortunately, succumbed on postoperative day 2.

DISCUSSION

GISTs represent the most common mesenchymal tumors of the gastrointestinal tract, originating from interstitial cells of Cajal in the muscularis propria [3]. While gastric GISTs constitute 60–70% of cases, small bowel involvement occurs in 20–30%, with multifocal synchronous tumors being exceptionally rare and typically associated with familial syndromes like neurofibromatosis type 1 or Carney triad [12]. This case demonstrates an unusual presentation of multiple jejunal and ileal GISTs manifesting as an acute abdomen due to perforation and intussusception complications seen in less than 5% of small bowel GISTs [5,6].

The diagnostic challenge lies in the non-specific preoperative imaging findings mimicking mechanical obstruction from adhesions or Crohn's disease, underscoring the importance of maintaining high clinical suspicion for mesenchymal tumors in atypical obstruction patterns [4,9]. Similar cases include those reported by Kelly *et al*. [7] (multifocal small bowel intussusception) and Jemedafe *et al*. [1] (multifocal GISTs without perforation). Our case is unique for simultaneous perforation and intussusception from quadruple tumors [8,11].

Surgical resection remains the cornerstone of therapy for localized disease, though tumor rupture and multifocality portend poor prognosis [10,13]. The rapid postoperative demise from septic shock highlights the aggressive biology of high-risk GISTs (large size >10 cm, high mitotic index) and the critical need for multidisciplinary management incorporating tyrosine kinase inhibitors like imatinib for adjuvant therapy in high-risk cases [14].

The limitation of this case lies in the unavailability of immunohistochemical confirmation (CD117/c-KIT, DOG1) due to the lack of facilities at our center, though recommended for definitive GIST diagnosis and risk stratification per ESMO guidelines [9]. Histopathology alone (spindle cells, muscularis origin, and high mitotic index) strongly supports GIST diagnosis. Referral to tertiary centers for IHC/molecular testing is advised for similar cases.

CONCLUSION

This rare case of multiple small bowel GISTs presenting with acute intestinal obstruction, perforation, and intussusception illustrates the diagnostic and therapeutic challenges of these aggressive mesenchymal neoplasms. The coexistence of a perforated 10 × 8 cm jejunal mass with omental adhesions and an ileal intussusception secondary to a lead-point tumor underscores the potential for life-threatening complications despite oncologically sound resection. Postoperative mortality from refractory septic shock emphasizes the dismal prognosis associated with tumor rupture, multifocality, and high-grade histopathological features. Clinicians must consider GISTs in the differential diagnosis of small bowel obstruction with mass lesions, particularly when preoperative imaging reveals transition points suggestive of mural pathology. Early surgical intervention following oncological principles remains essential, though outcomes in advanced presentations remain guarded.

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