

Small Bowel GIST: Clinical Presentation as Intussusception and Obscure Bleeding: Case Reports

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ABSTRACT

Intussusception and obscured GI bleeding are rare presentations of small intestinal GIST. Here, we report two cases one 14 year old male and another 58 year old female who had jejunal GIST found on exploratory laparotomy. In this article, we discuss patients' clinical findings, investigations and management. It is hoped that this report would increase awareness of such conditions.

Keywords: GIST (Gastrointestinal Stromal Tumor), Obscured Bleeding, Intussusception.

INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are rare clinical entities, representing less than 0.2% of all gastrointestinal tumors and only 0.04% of small intestinal malignant neoplasms. GISTs may occur anywhere along gastrointestinal tract, but most commonly arise in the stomach (40–60%) and jejunum/ileum (25–30%).^{1, 2} The clinical presentation of GISTs is highly variable depending on their size, location and presence of mucosal ulceration. Abdominal pain and gastrointestinal bleeding (when mucosal ulceration is present) are the most common symptoms or signs although persistent significant bleeding is unusual. Intussusception and subsequent obstruction is a very uncommon presentation of these lesions because of their tendency to grow in an extraluminal fashion.^{3,4} Very few cases of small bowel intussusceptions from stromal tumor in adults have been described in medical literature.¹⁻⁵ Intussusception is rather infrequent in adults, accounting for 0.1% of all surgical admissions and 1–5% of mechanical bowel obstructions. In contrast to childhood, where intussusception is idiopathic in 90% of cases, in adults a definable pathologic lesion is established in over 90% of cases, with neoplasms considered to be the cause in 65% of them. Preoperative diagnosis is often difficult because the symptoms are nonspecific.¹⁻³ Intussusceptions is correctly diagnosed preoperatively in only one-third of cases.^{1,3} The relative rarity of GISTs combined with nonspecific presentation results in delayed diagnosis. Frequently, this is only possible after surgery and histological examination.^{2,3,5} We report two unusual cases of small bowel GIST, one of which presented with a long-standing obscure gastrointestinal bleeding, established by double balloon enteroscopy examination, the later presented with acute intestinal obstruction as diagnosed clinically and straight X ray abdomen erect posture followed by ultrasonography which suggested it to be intussusception.

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CASE DESCRIPTION

Case 1

A 14 year old boy was previously hospitalized for gastrointestinal bleeding (melena) requiring frequent blood transfusion. Standard gastrointestinal endoscopic study (upper endoscopy, colonoscopy) revealed no gastrointestinal lesions. The patient underwent double balloon enteroscopy, which identified a submucosal growth in the proximal-jejunum of 4 cm size with ulceration but no active bleeding or oozing. The lesion was partially occluding the lumen and stained with India Ink (Fig. 1). Biopsies were performed. Computed tomographic scan showed in the proximal jejunum a hypervascular mass with 1.8cm X 2.2 cm suggestive of GIST (Fig. 2). The patient underwent laparotomy that identified a jejunal tumor (Fig. 3). The lesion was resected (Fig. 4A and B) and histological examination revealed a GIST tumor (R0).

The recovery was uneventful and the patient was discharged on the 7th postoperative day. There was no clinical or radiologic evidence of recurrence at follow-up (Stage I- T2, N0, M0).

Case 2

A 58-year-old woman was admitted with acute pain abdomen, clinically found to be acute intestinal obstruction. Straight X ray abdomen erect posture shows multiple air fluid level (Fig. 5). Ultrasonography was done in the emergency room shows target sign (Fig.6). The patient underwent laparotomy that identified a jejunal tumor (Fig. 7) causing jejunoileal intussusception (Fig.8). The lesion was resected (Fig. 9A and B) and histological examination revealed a GIST tumor (R0). The recovery was uneventful and the patient was discharged on the 9th postoperative day.

CASE 1



Fig 1: Lesion stained with India Ink



Fig 2: CT Scan showing Hypervascular mass



Fig 3: Jejunal Tumour



Fig 4 A: Resected mass



Fig 4 B: Resected mass

CASE 2



Fig 5: Straight X ray abdomen showing multiple air fluid level



Fig 6: USG showing Target Sign



Fig 7: Jejunal Tumour (found during surgery)



Fig 8: Jejuno-ileal Intussusception



Fig 9 A: Resected Mass



Fig 9 B: Resected mass (cut open)

DISCUSSION

In the present study, we have reported two cases of jejunal GIST with uncommon presentation.

GISTs are the most common sarcomatous tumors of gastrointestinal tract. They are known to be a distinct tumor arising from interstitial cells of Cajal, an intestinal pacemaker cell. Approximately, 40-60% of GISTs arise in the stomach, with 25-30% arising from small intestine.^{1,2}

Velasco et al., reported that the jejunal GIST accounts for approximately 10% of all the cases.⁶ The clinical presentation of GIST is erratic. Furthermore, only 70% of patients are symptomatic, where as 20% are asymptomatic and 10% are detected at autopsy.⁷

In the review of Miettinen and Lasota, the most common presentation is reported as gastrointestinal bleeding.⁷ Jejunoileal intussusception of jejunal GIST is rare.

A similar kind of case has been reported recently by Silvia Giestas et al., 2016.⁸ The tumors smaller than 2 cm in size are generally asymptomatic and larger tumors maybe present with upper abdominal pain, palpable intra-abdominal mass, vomiting, weight loss, perforation, or rupture. On exploration, both patients were found to have small growth that can be only appreciated after palpation intraoperatively.

GISTs have an unpredictable behavior, and a long-term follow-up is essential for all patients, independent of their benign or

malignant characteristic. According to the National Comprehensive Cancer Network guideline, contrast CT of the abdomen and pelvis is recommended every 3-6 months for 3-5 years and then yearly. Positron emission tomography is a very useful tool for postoperative follow-up of patients receiving imatinib.

CONCLUSION

Though rare, GIST in the jejunum may present as acute abdomen resulting from intussusception of tumor or may present with long term obscured GI bleeding. Oncologic surgery principles including clear resection margins and resection without spillage or rupture should be tried in such cases. Based on histopathology and IHC adjuvant chemotherapy with imatinib and close follow-up should be advocated.

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