

## Case Report

# A rare case of gastrointestinal stromal tumors presenting with jejunum-jejunal intussusception and anemia in elderly

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### ABSTRACT

Jejunum-jejunal intussusception is a rare occurrence in the context of a gastrointestinal stromal tumor (GIST). We present the case of an elderly female with recurrent episodes of small bowel obstruction and anemia due to blood loss caused by jejunal GIST with jejunum-jejunal intussusception.

**Key words:** GIST, Jejunum, Anemia, Intussusceptions

## INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are rare, representing 0.1-3% of gastrointestinal cancers with an estimated incidence of 14.5 per million.<sup>1</sup> Gastrointestinal stromal tumor (GIST), although rare, is the most common mesenchymal tumor of the intestinal tract and is thought to arise from pluripotential mesenchymal stem cells programmed to differentiate into the interstitial cell of Cajal. These act as pacemaker cells in the intestines.<sup>2</sup>

About 50-70% of GISTs have gastric origins whilst 20-30% has their origins in the small intestine.<sup>3</sup> Clinical features vary based on the location of tumor. GISTs may have intraluminal and extraluminal elements. They can be asymptomatic or with symptoms which can be vague initially. A majority of tumors are found incidentally on investigations like endoscopy or on computed tomography (CT) scanning.<sup>4</sup>

In spite of their clinical diversity, GISTs share common genetic alterations. Activating mutations in KIT or platelet-derived growth factor receptor alpha (PDGFRA)

have been identified in up to 80% and 10% of GISTs, respectively, and these mutually exclusive gain-of-function mutations play a fundamental role in GIST development by constitutively activating tyrosine kinase receptors.<sup>5,6</sup> Treatment of GIST involves surgery to remove the lesion and the use of Imatininib mesylate (ST1571; Gleevec, Novartis, East Hanover, NJ) a selective tyrosine kinase inhibitor to target local and metastatic disease.

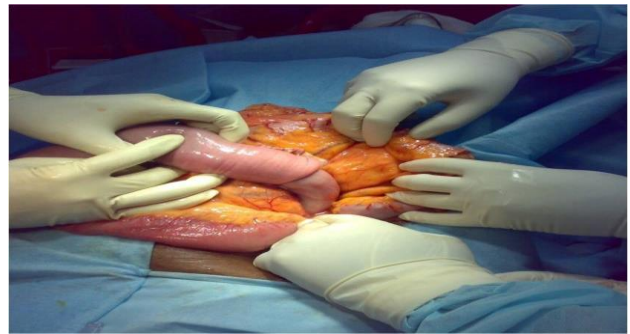
## CASE REPORT

An 80 year old woman was referred to gastroenterology department with history of recurrent episodes of sub-acute small bowel obstruction since last 3 months. On evaluation she had complaints of central colicky abdominal pain, abdominal distension and bilious vomiting of five days duration. She also had history of malena initially for two days following the onset of pain. She complained of easy fatigability, dyspnoea on exertion and palpitation of recent onset. She had history of hospitalization for similar symptomatic episodes one month and three months back. She had undergone upper GI endoscopy and colonoscopy three months which were

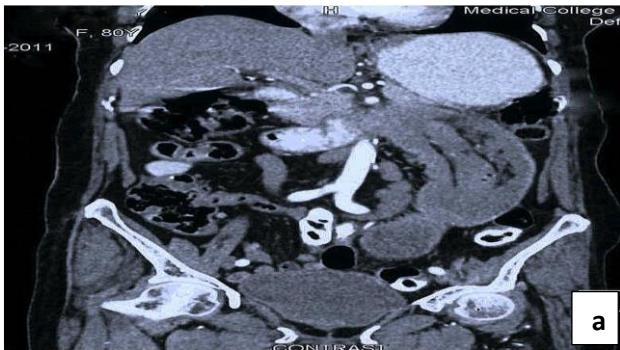
normal and had received helicobacter pylori eradication therapy. She is hypertensive on amlodipine 5 mg for last 15 years. She has no history of any abdominal surgeries. No family history of GI malignancy. On clinical examination she was pale. Vitals were stable. Per abdominal examination revealed an umbilical region mass of size 7 x 7 cms size, non-tender, firm, smooth surface, with slight transverse mobility. Examinations of other systems were unremarkable except for grade II hypertensive retinopathy. Routine investigations showed that she had iron deficiency anemia as evidenced by Hb of 6.5 gm%, serum ferritin was 12 µg/L, MCV was 61.8 fL, MCH was 17.4 pg, peripheral smear showed hypochromic microcytic anemia. Ultrasonography revealed a bowel related mass with pseudokidney sign in left upper quadrant. CECT abdomen (Figure 1a) showed bowel mass of length 18 cm craniocaudally. Superiorly sausage shaped and inferiorly with target appearance (Figure 1b). Radiological features were of jejunio-jejunal intussusception. Involved bowel loops showed diffuse wall thickening with sub mucosal edema (1.2 cm). An ill-defined soft tissue density at the apex of intussusception could be made out. No proximal dilatation or gangrene of bowel loops was seen. She underwent elective laparotomy. Intra operatively jejunio-jejunal intussusceptions (Figure 2) was present. There was an intramural mass arising from the wall of jejunum making the lead point of intussusception. There were no signs of bowel gangrene. There was no free fluid and inter bowel adhesions. Surgical team resected the jejunal segment bearing the mass and an end to end jejunio-jejunal anastomosis was done. Postoperative period was uneventful and she recovered well. She was referred to regional oncology centre for targeted therapy with imatinib. Pathologic examination of resected jejunal segment cut open through antimesenteric border identified polypoid looking 3x3 cm growth. Surface was lobulated, fibrous in appearance with necrotic areas and was originating from the muscular layer. Histology showed gastrointestinal stromal tumor of spindle cell type with mitotic index of 8/50 HPF (Figure 3a and 3b). Immunohistochemistry was positive for CD 117.



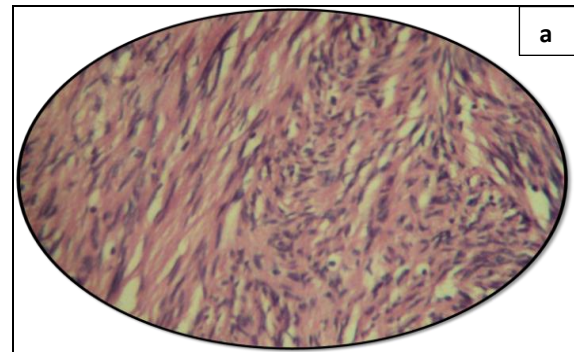
**Figure 1b: CECT abdomen coronal view showing Target sign.**



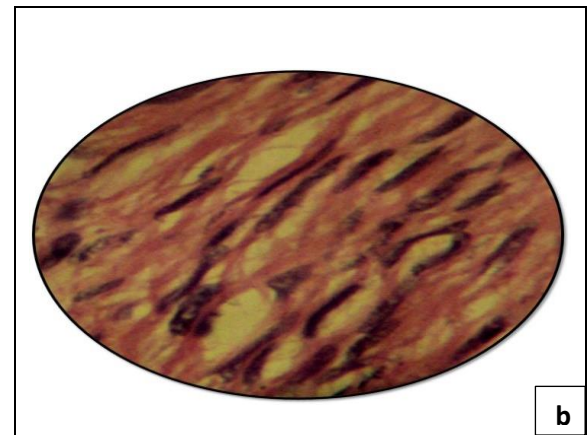
**Figure 2: Jejunio-jejunal intussusception at laparotomy.**



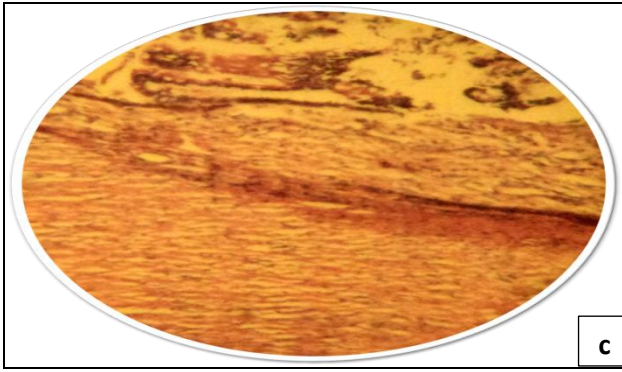
**Figure 1a: CECT abdomen reconstructed coronal view showing jejunio-jejunal intussusceptions.**



**Figure 3a: Low power microscopic appearance.**



**Figure 3b: High power microscopic appearance.**



**Figure 3c: C-kit immunostaining microscopic appearance.**

## DISCUSSION

GIST presenting as a focus for jejunojejunal intussusception is a very rare presentation. Clinical presentation of GISTs can be varied and quite often diagnosed incidentally. A study showed that ninety percent of GISTs were detected clinically due to symptoms 69% or were incidental findings at surgery 21% the remaining 10% of GISTs were found at autopsy.<sup>1</sup> Symptoms in GISTs depend on their location and both mass effect and intraluminal bleeding can occur. Gastrointestinal tract GIST can induce significant hemorrhage causing haematemesis, melena or occult bleeding with anemia. They have been noted to cause dysphagia in the esophagus, biliary obstruction around the ampulla of Vater, hypoglycemia, abdominal pain due to torsion of an exophytic tumor, presentation as content in a hernial sac, intraperitoneal bleed and mimicking acute appendicitis.<sup>7-10</sup> In the literature there is description of jejunal intussusception due to jejunal polyp, adenomyoma, heterotopic gastric mucosa and of Jejuno gastric/jejunal duodenal intussusception or retrograde intussusception as a severe complication of gastric surgery.<sup>11</sup>

This case shows how GIST can rarely present as recurrent sub-acute intestinal obstruction due to jejunojejunal intussusception and as cause of anemia due to blood loss in elderly.

## CONCLUSION

GISTs are rare tumors with varied clinical presentations. The clinician needs to be aware of rare presentations of these tumors. A multidisciplinary approach is essential for diagnosis and successful management of these patients.

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