Case Report

Gastrointestinal stromal tumor presenting as ileo-ileal intussusception: a rare case report

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ABSTRACT

Gastrointestinal stromal tumors (GISTs) are mesenchymal tumors that can arise in any part of gastrointestinal tract. It is commonly seen in the fifth or sixth decade of life with slight male preponderance. Intussusception and subsequent obstruction is a very uncommon presentation of these lesions because of their tendency to grow in an extraluminal fashion. In the literature, very few cases of small bowel intussusceptions from a stromal tumor in adults have been described. We report a rare case of GIST presenting as acute intestinal obstruction due to intussusception in an elderly male.

Keywords: GISTs, Intussusceptions, Neoplasm

INTRODUCTION

Gastrointestinal stromal tumors (GISTs) arise from the interstitial cells of Cajal in the mesenchymal tissue and constitute less than 3% of all gastrointestinal malignant neoplasms and represent only 20% of small-bowel malignant neoplasms.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.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. GISTs presenting as intussusception is very rare especially since GISTs grow in an extraluminal fashion. Here, we report a rare case of GIST presenting as acute intestinal obstruction due to intussusception in an elderly male.

CASE REPORT

A 62 year old male presented to the surgical emergency with chief complaints of abdominal pain and distension for past 2 days with history of non-passage of stools and flatus and multiple episodes of bilious vomiting. He did not have any previous similar episodes. He had no history of any other comorbidities. On examination, he was having tachycardia, hypotensive and his abdomen was distended, tense with diffuse guarding and rigidity and exaggerated bowel sounds. Per rectal examination revealed roomy, ballooned out and empty rectum with no mass palpable in the surroundings. All blood investigations were within normal limits. X-ray abdomen revealed dilated small bowel loops with multiple air fluid levels. A provisional diagnosis of acute intestinal obstruction was made and patient was taken up for emergency laparotomy. Intra operatively, ileo-ileal intussusception was present 2 feet proximal to ileo-caecal junction (Figure 1). Rest of the bowel and solid organs were normal with no free fluid in the abdomen. 1 feet of
bowel containing the intussusception was resected and anastomosed.

(GIST) with IHC marker positive for CD 117 (Figure 4 and 5). There was no recurrence till 1 year of follow up.

On exploring the resected bowel segment, a 2x2 cm intraluminal growth was seen which acted as the lead point for the intussusception (Figure 2 and 3).

The bowel segment with the tumor mass was sent for histopathological examination. The patient had an uneventful postoperative period. The histopathological report was suggestive of Gastrointestinal stromal tumor (GIST) with IHC marker positive for CD 117 (Figure 4 and 5). There was no recurrence till 1 year of follow up.

DISCUSSION

GISTs are mesenchymal tumors arising in any part of gastrointestinal tract but 50% will be found in the stomach. In small bowel, it is more common in the jejunum than the ileum. It is commonly seen in the fifth or sixth decade of life with slight male preponderance.1,2
These tumors are universally associated with mutation in the tyrosine kinase c-kit oncogene. GISTs are most commonly incidentally found either in CT scan or UGI endoscopy. It may present as bleeding in case of mucosal rupture, rarely as obstruction or free perforation due to haemorrhagic necrosis. GISTs arise from the muscularis propria and generally grow extramurally. Differentiation between benign and malignant GISTs is difficult even on histological examination.

Large size (usually more than 5 cm) and high levels of c-kit (CD 117) are associated with malignant potential. GISTs metastasize to liver, lungs and peritoneum. Lymph nodal metastases is very rare. The initial diagnostic test is an abdominal ultrasound.

Small GISTs (less than 2 cm) found incidentally in surgical specimens don't require further treatment. Imatinib mesylate is a tyrosine kinase inhibitor which is the first line drug in treatment of malignant GISTs. It is used as adjuvant therapy in high risk diseases.

Intussusception is the telescoping of one portion of intestine into the other; invariably it is the proximal into the distal. It is most common in children where it is usually idiopathic. Adult intussusception has a definable pathologic lesion in over 90% of cases, with neoplasms considered to be the cause in 65% of them. Any intraluminal lesion, especially polyps, which irritates and alters normal peristaltic activity, is able to trigger an intraluminal invagination finally causing an intussusception. Subsequent peristaltic bowel activity produces an area of sequence constriction and relaxation, thus telescoping the leading point through the distal bowel lumen. The malignancy is more likely to be located in the colon rather than in the small bowel. Less common etiologies of intussusception in adults include postoperative factors (adhesions, suture lines, etc.), polyps, Meckel’s disease, sprue, cecal duplication and intramural hematoma. Intussusception consists of three parts; 1) the entering or inner tube (intussusceptum), 2) the returning or middle tube, 3) the sheath or outer tube (intussuscepiens).

The presentation of adult intussusception is usually subacute or chronic. Only up to 20% of all cases present with complete bowel obstruction and acute onset. In the literature, very few cases of small bowel intussusceptions from a stromal tumor in adults have been described. The initial diagnostic test is an abdominal ultrasound which shows ‘target sign’ in transverse view or ‘pseudo-kidney sign’ in longitudinal view. Hydrostatic reduction by enema using contrast material is the therapeutic procedure of choice in children and surgery in indicated in recurrent cases. In adults, surgery is the mainstay of treatment.

CONCLUSION

This case highlights the diagnostic difficulty and unusual presentation of GIST. The relative rarity of GISTs combined with nonspecific presentation results in delayed diagnosis. Frequently, this is only possible after surgery and histological examination.

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REFERENCES
