

Jejunojejunal Intussusception in an Adult Due to a Gastrointestinal Stromal Tumour

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ABSTRACT

Intussusception in an adult is a rare entity responsible for less than 1% of all intestinal obstructions. It is far more common in children, where the etiology is more often idiopathic. In adults most cases have a demonstrable lesion as the lead point which invariably needs surgical treatment. A gastrointestinal stromal tumour is a rare tumour of mesenchymal origin responsible for only 0.2-1% of all gastrointestinal neoplasms. There have been only a

few reports of such tumours causing intussusception in adults. The case being reported is of a 37 years old male who presented to the hospital with an abdominal lump and anaemia. On investigating it was discovered that he had jejunojejunal intussusception due to a gastrointestinal stromal tumour. The high index of suspicion, appropriate use of pre-operative imaging studies and regular post-operative follow-up contributed to a successful outcome.

Keywords: Laprotomy, Gastrointestinal Stromal Tumor, Intestinal obstruction, Intussusception

CASE REPORT

A 37 years old male presented to the hospital with complaints of pain in abdomen, constipation and multiple episodes of vomiting over the previous 3 days. On inquiry he revealed intermittent pain in abdomen over the past few weeks. On examination he had a distended abdomen with a palpable lump in the left lumbar region of the abdomen. The patient was pale and his rectal exam revealed blackish stool. Investigations revealed anaemia (Hemoglobin- 5.5g/dl) but a plain erect abdominal radiograph did not reveal multiple air fluid levels. Resuscitation was carried out including a packed red cell transfusion. An abdominal CT scan revealed a bowel in bowel appearance or the target sign involving the proximal jejunum [Table/Fig-1]. A mass lesion arising from the bowel

wall was seen within the lumen of the invaginated bowel loop. A decision to perform an exploratory laprotomy was made keeping the above findings in mind. At laprotomy a proximal jejunojejunal intussusception was discovered [Table/Fig-2]. A limited resection and anastomosis was performed. The lead point was found to be a tumour arising from the jejunal wall that had ulcerated the mucosa [Table/Fig-3]. Histopathological examination revealed the tumour to be a gastrointestinal stromal tumour (c-kit positive) 5.5x3.4x2.2cm dimensions with resection margins free from tumour. The tumour had a low mitotic rate (<5/50 hpf), it had ulcerated the mucosa and stained positive for SMA focally but not for DOG 1 and CD 34. The patient made an uneventful recovery in the post-operative period. The patient was stratified as having a moderate risk for



[Table/Fig-1]: CT scan showing characteristic "target sign". **[Table/Fig-2]:** Operative photograph showing jejunojejunal intussusception. **[Table/Fig-3]:** Intra-luminal GIST having ulcerated the mucosa.

disease progression and a follow-up plan was designed. He is currently doing well following up with regular clinic visits at the hospital.

DISCUSSION

Intussusception in adults is a rare clinical entity. It is a common cause of intestinal obstruction in children. The exact mechanism that initiates the invagination is not known. However, the hypothesis is that when there is a lesion in the bowel wall that may interrupt normal peristalsis it may serve as a leading point to invagination [1]. It is more common in locations that mark the junction between relatively fixed and freely mobile segments of the intestine [2]. In adults an overwhelming majority of patients (~ 90%) who present with intussusceptions have a demonstrable lesion that serves as the lead point. Malignant lesions serve as lead points for as many as 30% when small bowel is involved and 66% when large bowel [3].

In adults there is a spectrum of clinical presentation from acute, subacute to chronic [4]. Adults mostly present with chronic symptoms. Abdominal pain is the most common complaint but the patient may also demonstrate signs of intestinal obstruction. Abdominal lumps are present in only about 24-42% as reported in various case series [4]. The finding of an abdominal lump as in this patient is a significant finding and if appearance of lump coincides with symptoms and disappearance of lump coincides with relief, intussusception or volvulus should be suspected. The presence of vague non-specific symptoms usually means that the patient is subjected to a battery of tests. Very often the first imaging ordered is a plain abdominal radiograph which may reveal a picture of intestinal obstruction. If done with barium it may reveal a "stacked coin appearance" in upper GI series or "cup shaped defect" in enema series. The ultrasound of the abdomen may reveal the characteristic "target sign" or the "pseudo-kidney sign". Colonoscopy has also been used to diagnose ileo-colonic intussusception at times [5]. However, in recent times abdominal CT (with the "target sign" or bowel in bowel appearance) has shown to be the most accurate investigation (58-100% in various series) to detect and delineate intussusception [4].

There is consensus that once detected the treatment for intussusception is always surgical in adults. There is a debate regarding the extent of resection and whether reduction should be attempted before resection. Primary reduction is in cases with suspicion of malignancy is avoided due to fear of intraluminal seeding, perforation causing dissemination and venous embolization from areas of ulcerated mucosa. The extent of resection should usually enable a R0 resection and may include a lymphadenectomy if malignancy is suspected, as histopathological diagnosis is rarely available in the pre-

operative setting. There is a role of laparoscopic resection as shown by few of the recent case reports.

The lesions that may cause an intussusception in adults may be benign neoplasms, inflammatory lesions, Meckel's diverticuli, appendix, and adhesions—or malignant lesions. Gastrointestinal Stromal Tumour (GIST) is a rare mesenchymal tumour (representing <1% of all GI malignancies) of the gastrointestinal system. There are very few reports of a GIST leading to intussusceptions in adults. GISTs usually occur in middle aged to older population and occurrence under the age of 40 (as in this case) is rare. They may arise anywhere in the GI tract from the esophagus to the anus. The common sites are stomach (40–60%) and jejunum/ileum (25–30%) [6]. While duodenum (5%), colorectal (5–15%), and esophagus ($\leq 1\%$) are less common sites. They are usually asymptomatic and may be discovered incidentally at laprotomy or on imaging. They may give rise to vague abdominal pain or mass lesion in advanced stages of the disease. They are highly vascular tumours and have known to bleed into intestinal lumen if they ulcerate the mucosa. Some patients as the one in this case may have a history suggestive of melena and anaemia at presentation. It rarely may lead to intestinal obstruction.

The goal of therapy is complete surgical excision with negative margins whenever possible. Risk stratification is done for these patients after auditing the surgical specimen. The factors considered include location of the GIST, mitotic rate, tumour size and resection margins [7]. Surgical resection is adequate for a low-risk GIST while adjuvant Imatinib therapy is given to moderate and high risk GISTs [8]. Imatinib, a tyrosine kinase inhibitor is useful as it is active against c-kit positive GISTs. In advanced cases, metastases or when it may not be possible to achieve surgical clearance Imatinib may be used in a neoadjuvant setting.

CONCLUSION

Intussusception occurs rarely in adults. It almost always warrants surgery as it is due to a demonstrable lesion more than 90% of the times. The diagnosis is difficult to make many times pre-operatively due to non-specific symptoms. The liberal use of imaging modalities like CT scan contributes to the surgeon's ability to diagnose it before surgery. The surgeon should not only know how to deal with intussusception in adults who present with intestinal obstruction but also know how to deal with the lesion at its lead point (GIST in this case) that may require specific treatment in the post-operative setting.

CONSENT

Written informed consent was obtained from the patient for publication of this Case report and accompanying images.

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