A Rare Case of Adult Ileocolic Intussusception Due to a Vaneks Tumour

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Abstract

Intussusception occurs when a more proximal portion of the bowel invaginates into the more distal bowel. Intussusception in adult is a rare condition, its presentation is acute with clinically vague signs. Incidence of vaneks tumour presenting as intussusception is 8.6%. Vaneks tumour is a least common benign small bowel neoplasm. We present to you a case of a 32 year old female presenting with an acute abdomen, which was later diagnosed to be an ileocolic intussusception. The diagnosis was made based on Computed Tomographic images of the abdomen showing bowel within a bowel appearance. A classical right Hemicolectomy with ileotransverse anastamosis was successfully performed. Post-operative histopathological picture confirmed lead point causing the intussusception as inflammatory fibroid polyp or Vaneks tumour.

Intussusception in adults is a rare condition, its presentation is acute with clinically vague signs, making initial diagnosis difficult and tricky. Once diagnosis is confirmed intervention should be prompt and appropriate as it is lifesaving. We share this case report for the rarity of this condition and to reinforce the knowledge of this atypical presentation of intussusception.

Introduction

Intussusception occurs when a more proximal portion of the bowel (intussusceptum) invaginates into the more distal bowel (intussuscipiens). Intussusception is common in children. Only 5%-16% of intussusception occurs in adults. Most of adult intussusception has a lead point. Vaneks tumour is one such lead point. Vaneks tumour, or inflammatory fibroid polyp, is one of the least common benign small bowel tumours. Its Peak incidence occurs in sixth and seventh decades of life, with a slight male preponderance. It’s incidence in GIT is 0.3-0.5%. Incidence of Vaneks tumour in terminal ileum is 18-20%. A incidence of Vaneks tumour presenting as intussusception is 8.6%. We herein present a rare case of a 32 year old female with ileocolic intussusceptions due to IFP.

Case Report

A 32 year old female who was apparently normal two weeks back, presented with a three day history of colicky upper abdominal pain, predominantly of the right hypochondrium. History of multiple episodes of loose stools in the past 2 weeks. History of (2-3) episodes of bilious vomiting since the past 1 week. History of weight loss present. No history of fever, loss of appetite. No history of blood in stools or melena. Patient does not have any co-morbid disease. No history of previous surgery. P2L2 normal delivery, sterilised. On clinical examination, pallor present. Patient was afebrile with stable vital signs. On abdominal examination, abdomen was found to be soft with mild distension. Right hypochondrial tenderness was present. No guarding/rigidity. Bowel sounds on auscultation were sluggish. Our clinical diagnosis included cholecystitis. Routine blood investigations were within normal limits. On ultrasound, few dilated bowel loops were seen, otherwise normal. Abdomen x-ray erect and chest x-ray were normal.

On CECT abdomen, right hypochondrium and right lumbar region showed diluted ileum, caecum and proximal ascending colon seen invaginating into the distal ascending colon, hepatic flexure and seen up to the proximal transverse colon. Mesenteric fat and mesenteric vessels also seen invaginating along with the above mentioned bowel loops. All these features were representative of an ileocolic intussusception at ileo-caecal junction “Target sign- positive”atien was taken for laparotomy and proceed. Intra-operatively, an ileocolic intussusception was identified as a huge mass in the ascending colon. Appendix was in right iliac fossa, identified at the point of ileum invaginating into the caecum. 10cms of ileum has intussuscepted into right colon & presented as a huge mass in the ascending colon. Intussusceptiens (large bowel) and intussusceptum (small bowel) (Figure 1). A brief trial of reduction was done, but was unsuccessful. Viability
of the bowel was good. A classical right hemicolectomy & end
to side ileo-transverse anastomosis with hand sewn anastomosis
& mesentery rent was closed. On cutting open the specimen, a
11cm long intussusceptum and the returning segment formed by
ileum both along with the intussusceptors component formed by
the ascending colon appeared as a bulky mass, with a huge 5x4
cm ulcerative proliferative polyp found in the apex as the lead point
(Figure 2). Post op period was uneventful.

![Inflamatory fibroid polyp](image1.png)

**Figure 1:** Showing a segment of terminal ileum measuring 7.5cm,
partially cut open large intestine (along taenia coli) measuring
22cm, appendix measuring 9.5cm. Intussusceptum measuring
21cm.

![Transverse colon](image2.png)

**Figure 2:** Intussusceptum showing a pedunculated polyp
measuring 4x3x2cm. Stalk measuring 1cm, with a region of
ulceration and slough.

Histopathological examination of the specimen-

A - Proximal resected margin- section showed wall and
mucosa of small intestine which appears viable.

B - Distal resected margin- showed colonic wall and mucosa
which appears viable with no specific pathology.

C - Tip of appendix- shows wall of appendix with mucosal
lymphoid hyperplasia.

D, E - From polypoidal lesion- shows a polypoidal lesion
covered with colonic and ileal mucosa with focal ulceration
covered with neutrophilic exudates.

The submucosa is expanded and shows myxomatous stroma
with benign spindle shaped cells (Figure 3), prominent thick-
walled vessels and many proliferating capillaries. There was
scattered inflammatory cell infiltrate composed was plasma cells,
lymphocytes and eosinophils. No evidence of dysplasia/malignancy
seen. IHC markers were positive for CD30, negative for SMA and
S100. Features were suggestive of INFLAMMATORY FIBROID
POLYP (IFP).

![Histopathological examination](image3.png)

**Figure 3:** (H&E, 10x) The polyp composed of benign spindle
shaped cells, thick walled blood vessels, proliferating capillaries,
inflammatory cell infiltrate of plasma cells, eosinophils and few
lymphocytes.

**Discussion**

In 1674, Barbette first described intussusception. Later
in1789, Hunter described about intussusception. First surgeon
to operate on a child diagnosed with intussusception was Sir
Jonathan Hutchinson in 1871(4). In adults intussusception is
very rare & its incidence being 2-3 per 10,00,000 per year (1).
Its defined as invagination of proximal part of small intestine
along with its mesentery into the adjacent segment of bowel
& leads to various complications like obstruction, impaired
peristalsis & gangrene (4). Any lesion in bowel wall/ irritant
in the lumen is the leading point for intussusception & initiates
invagination (4). Ingested food and subsequent peristaltic activity
of the bowel produces an area of constriction above the stimulus
and relaxation below, thus telescoping the lead point through
the distal bowel lumen (4). Freely moving part of bowel &
retroperitoneally or adhesively fixed parts are the most common
locations for intussusception (2). Intussusceptions have been
classified according to location into three major categories, i.e,
enteroenteric, ileocolic or ileocecal, and colocolic (4). In ileocolic
intussusception, the ileum invaginates through the ileocecal valve
Adult intussusception occurs more frequently in small bowel (50%-88%) than in the large bowel (12%-50%) (4). In adults, the aetiology, presentation & management is very different from that of children (1). Idiopathic or secondary viral illness are the most common causes in children. In adults, various causes are present. In 90% cases, a lead point is found to cause intussusception (1). Most lead points in gastrointestinal tract involve primary or metastatic malignancy, lipomas, leiomyomas, adenomas, neurofibromas, postoperative adhesions, meckels diverticulum, foreign bodies, vascular anomalies, lymphoid hyperplasia, trauma, celiac disease, cytomegalovirus colitis, lymphoid hyperplasia secondary to lupus, henoch-schonlein purpura, wiskott-aldrich syndrome, appendiceal stump, or inflammatory fibroid polyps (IFP) (4). Benign lesions account for almost 25% cases of intussusception in adults (1). The commonest benign lesion is a lipoma in the colon (1).

In our case, the lead point was found to be a Vanek's tumour. Vanek's tumour/ Inflammatory fibroid polyp (IFP) are rare clinically benign mesenchymal tumours originating in submucosa of GIT. Incidence being unknown (3). It’s incidence in GIT is 0.3-0.5%. Incidence of Vanek’s tumour in terminal ileum is 18-20%. A Vanek’s tumour presenting as intussusception is 8.6% (5). In 1949, Vanek first described this as “gastric submucosal granuloma(s) with eosinophilic infiltration”. These lesions were found throughout the GIT (3). A few identified events of IFP’s are reactive inflammatory process with trauma, allergic reaction, and bacterial, physical, chemical or metabolic stimuli (3). Recently it was identified that reports of familial occurrence & recognition of activating platelet-derived growth factor receptor alpha (PDGFRα) mutations in these tumours suggest that IFPs represent true neoplasms (3). GIST & IFP share a common oncogenetic pathway since they have similar PDGFRα gene mutations (3,12) Immunohistochemically, IFPs are negative for CD117 and variably positive for CD34 (3). In contrast, GISTs have characteristically positive CD117 and CD34 immunostaining (3). The most common site is the gastric antrum (60-70%), followed by small bowel (18-20%), colorectum (4-7%), and far less commonly (1%) in oesophagus, duodenum, gallbladder, and appendix (3,6,8) The polyps are typically solitary, but rare metachronous lesions have been reported in familial cases (3). Most IFPs grow intraluminally and are smaller than 4cm (3). In this case the polyp measured 4x3x2cm in dimensions.

Clinical manifestations depend largely on tumour location and size. Often IFPs are asymptomatic and are identified incidentally either during endoscopic or surgical procedures (3). When present in small intestine they are more likely to present with chronic colicky abdominal pain, small bowel obstruction, intussusception, and weight loss (6,9) GI bleeding is a rare presenting symptom, and if present, it may indicate significant ulceration or ischemia (3).

The clinical presentation, most often in adults, intussusception presents as chronic condition but with non specific symptoms suggestive of intestinal obstruction (1.2,15) Abdominal pain, nausea, vomiting, diarrhoea and bleeding per rectum are the common symptoms (11,13,14) Acute intestinal obstruction is a very rare presentation of intussusception (1). Classical triad of abdominal pain, sausage shaped palpable mass in per abdomen examination & passage of red current jelly stools is very rarely seen in adults (1). Only in 24-42% of patients, palpable abdomen mass is felt. Intussusception is suggestive when a shifting abdominal mass or mass that’s only palpable when symptoms arises (2).

Various radiological methods are used to describe intussusception: x-ray abdomen erect, ultrasound abdomen, barium studies, angiography & radionucleotide studies (2). Plain abdominal x-ray may shows signs of intestinal obstruction if its present (2). On ultrasonography, a classical “target/ doughnut sign” on transverse view and the “pseudokidney sign” in longitudinal view is identified. The major disadvantage being, gas filled bowel loops (2). Abdominal CT is the most useful technique in diagnosing intussusception with accuracy 58%-100%. Signs of target/ sausage, mesenteric fat & vessels are identified. Metastasis & lymphadenopathy can also be viewed.

In the adult population, once intussusceptions is diagnosed, prompt surgical intervention is warranted to avoid complications of ischemia, necrosis, and perforation (3). Traditionally, surgical resection is the treatment of choice for symptomatic IFPs. Resection is curative, and only one case of polyp recurrence is found in the literature (3). The appropriate management of adult intussusceptions remains controversial, with the debate focusing mostly on the issue of primary en bloc resection vs initial reduction followed by more limited resection (4). Reduction by surgery before resection may theoretically permit more limited resection; however, the risk of potential intraluminal seeding or venous tumour dissemination during the manipulation of a malignant lesion should also be taken into consideration (4). The incidence of malignancy as the cause of small intestinal intussusceptions ranges from 1% to 47%, and the majority of lesions are metastatic (4). Therefore, recent reports have recommended initial reduction of externally viable small bowel prior to resection (4).

Conclusion

Intussusception in adults is a rare occurrence and when a adult patient presents with a slightly chronic symptoms of abdomen pain with diarrhoea and a tender spot in a quadrant of abdomen, surgeon should think about intussusception as a differential diagnosis. CT scan is the diagnostic imaging of choice. And surgery should not be delayed in order to prevent bowel ischemia and gangrene. Chances of malignant lesion as a lead point should be thought about and a proper resection of bowel with anastomosis will be an ideal form of surgical treatment.
The debate focuses mostly on the issue of primary en-bloc resection vs initial reduction followed by more limited resection. Reduction by surgery before resection may theoretically permit more limited resection; however, the risk of potential intraluminal seeding or venous tumor dissemination during the manipulation of a malignant lesion should also be taken into consideration. In our patient, due to failed attempt at reduction, primary en-bloc resection was carried out.

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