CASE REPORT

Vanek’s tumor causing ileoileal intussusception in a middle-aged man

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Abstract

Intestinal obstruction is a common surgical emergency requiring urgent intervention. Small bowel obstruction secondary to intussusception is rarely encountered especially when inflammatory fibroid polyp (IFP) is the lead point. A 41-year-old gentleman with intestinal intussusception secondary to IFP presented to us with a classic symptom of intestinal obstruction. Computed tomography revealed a target or sausage-shaped soft tissue mass with a layering effect, which was confirmed by intraoperative findings. Histopathology was consistent with IFP and supported by immunoreactivity of CD34 and negative immunostaining for CD117. He recovered without any surgical complication or recurrence. Even intussusception can be managed via non-surgical technique in children; surgery is the mainstay of treatment in adults.

INTRODUCTION

Intestinal obstruction is a common surgical emergency, which is always encountered by the surgical fraternity. It can be caused by a malignant tumor, hernia, adhesion, volvulus and rarely an intussusception. In principle, intussusception happens as a result of a telescoping of a proximal bowel either small or large intestinal into the distal bowel lumen. The aetiologies of intussusception in the small bowel and colon are distinct. In small bowel pathology, the lead points are mainly due to benign entities such as hamartomas, lipomas, adenomas, Peutz–Jeghers syndrome, Meckel’s diverticulum, lymphoid hyperplasia, tuberculosis and rarely inflammatory fibroid polyp (IFP) or also known as Vanek’s tumor [1]. Ileum is the most common site of small bowel intussusception encountered.

On the other hand, the intussusception of the large bowel is more likely to have a malignant etiology. Colon adenocarcinoma or malignant polyp is the most important cause of malignant large bowel intussusception [1]. The classification of intussusception follows the involved segment namely ileoileal, ileocolic, colocolic and colorectal intussusception. The surgical intervention differs between organs involved as well as the histological diagnosis. Herein, we describe a middle-aged gentleman with intestinal intussusception secondary to IFP.

CASE REPORT

A 41-year-old man presented with abdominal distension and intermittent discomfort for 1-month duration. He also had an
Figure 1: CT scan showing a target or sausage-shaped soft tissue mass with a layering effect (arrow) involving the ileum suggesting of an ileoileal intussusception.

absolute constipation for 3 days duration in addition to nausea, faeculent vomiting, loss of weight and appetite. He denied having per rectal bleeding, blood or mucus-stained feces or tenesmus. Upon assessment, he was dehydrated with coated tongue. He was tachycardic with pulse of 105 beats per minute with low volume. The blood pressure, however, was within normal range. The abdomen was distended with high-pitched bowel sound. Biochemical investigations revealed acute kidney injury with urea of 15 (normal: 2.5–6.7 mmol/L) and creatinine of 115 (normal: 50–98 umol/L). Otherwise, full blood count and coagulation profiles were within normal readings. Abdominal radiograph was consistent with small bowel obstruction, showing centrally located valvulae conniventes. Computed tomography (CT) of the abdomen (Fig. 1) showed the intussusception of distal ileum.

The patient was managed in the view of classic intestinal obstruction. He was fluid resuscitated in the view of risk of third space loss, bowel decompression with a nasogastric tube and urine output chart by using continuous bladder drainage chart. He later was subjected to an exploratory laparotomy in the view of such findings. Intraoperative findings revealed a hard ileal mass, which was telescoping into the distal bowel lumen suggestive of intussusception (Fig. 2A). It was located 120 cm from the ileocaecal valve. A single mesenteric lymph node was seen with minimal ascites. We proceeded with small bowel resection and primary anastomosis in addition to the excision of the mesenteric lymphadenopathy.

Macroscopically, there is a bulging/polypoidal mass measuring 40 × 40 × 36 mm causing bowel obstruction (Fig. 2B). Other parts of bowel are normal. Microscopically, from hematoxylin and eosin (H&E) stain, the small bowel tissue shows a poorly circumscribed fibroinflammatory mass centered in the submucosa (Fig. 3A). The mass is composed of a mixture of haphazardly arranged spindle and mixed inflammatory cells, particularly eosinophils, in a background of prominent capillaries and fibromyxoid, granulation like tissue sarcoma (Fig. 3B). The spindle stromal cells display vesicular nuclei with small conspicuous nucleoli and moderate eosinophilic cytoplasm. In areas, these cells typically whorl around vessels, resulting in an onion skinning appearance. There is no cytological atypia and mitotic figures noted. The spindle cells are immunoreactive for CD34 and negative for CD117 (Fig. 3C and D). The mesenteric lymph node shows preserved lymphoid architecture with T-lymphocytes rich zone by mature lymphocytes, histiocytes, plasma cells and variable sizes of B-follicles with reactive germinal centers. No multinucleated giant cells or granuloma is seen.

The postoperative recovery was uneventful. He was discharged after 3 days following enhanced recovery after surgery protocol. Upon follow-up at 6 weeks, he was well without any surgical complications or recurrence.

DISCUSSION

IFP is among the least common benign gastrointestinal (GI) tumors, which was first described by Vanek in 1949 [1]. It accounts for <3% of all gastric polyps [1]. It can be found throughout the GI tract, mostly in the gastric antrum or ileum, but rarely in the duodenum and jejunum. On average, the diameter of IFP found is 3–4 cm, but the largest case of IFP is 18 cm in size, which was reported before [2]. IFP affects both sexes, but male gender shows predominance among all cases. The causative factors are unexplained, but some aetiologies have suggested chemical, physical or metabolic triggers. Even, genetics also play a role whereby a mutation of platelet-derived growth factor receptor alpha is involved [3].

IFP mostly is asymptomatic, but it can manifest as abdominal pain, vomiting, altered bowel habits, GI bleeding and loss of
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Figure 3: (A) IFP (arrow) arising from the submucosal layer (H&E stain, original magnification x4). (B) Histopathological examination showing variable cellularity, and spindle cells with bland nuclei and clear cytoplasm. There is an abundant inflammatory infiltrate comprising plasma cells, lymphocytes and eosinophils (H&E stain, original magnification x40). (C) Positive immunostaining for CD34 (original magnification x20). (D) Negative immunostaining for CD117 (original magnification x20).

weight. Compared with children, a triad of intermittent colicky abdominal pain, currant jelly stool and palpable right lower quadrant sausage-shaped mass is not present in our case [4]. Hence, the clinical judgment is limited among adults, requiring an assistance from a complementary imaging. Imaging modalities are undoubtedly trivial in managing acute abdomen. Causes such as obstruction, perforation and abscess can be visualized well with the assistance of the imaging. Abdominal radiography and ultrasonography are important, but CT is currently the confirmatory modality to diagnose intussusception with sensitivity between 86–100% especially on multislice CT [5]. The features that can be visualized include a target or sausage-shaped soft tissue mass with a layering effect [4].

Histopathological assessment is crucial because the clinical presentations are equivocal. Among the differential diagnoses in intussusception include inflammatory myofibroblastic tumor (IMT), schwannoma and gastrointestinal stromal tumor (GIST). IMT is characterized by myofibroblastic spindle cells with inflammatory infiltrates of plasma cells and intermingled mast cells [6]. It produces a diffuse cytoplasmic staining for vimentin, desmin and actin but not on myogenin, myoglobin, S100, CD117 and epithelial membrane antigen [7]. GI schwannoma is an abnormal growth that originates from Schwann cells, which histologically show the presence of Antoni A and Antoni B areas of spindle-shaped cells with formation of Verocay bodies [8]. Since they are neurogenic tumors, they are usually positive for S100 protein and vimentin, whereas negative for desmin, keratin, smooth muscle actin and CD34 [9]. GIST is another important tumor arising from a mesenchymal or interstitial cell of Kajal, develops from the submucosal layer, which is identical to IFP. Both IFP and GIST show immunopositivity for CD34 and vimentin, but IFP does not express CD117 as compared with GIST [1]. Immunostains for c-KIT, DOG-1, S100 and EMA are consistently negative.

The treatment of IFP-induced intussusception depends on the tumor size and its clinical presentation. Small IFP can be removed endoscopically via endoscopic submucosal resection. Exploratory laparoscopy or laparotomy is best performed in a larger sized IFP. Since it is benign in nature, resection is not following the oncologic manner. However, in large bowel intussusception with majority of the cases are due to malignant polyp, surgery following the oncologic resection is required following the vascular supply. For example, in cases of malignant polyp in the cecum or ascending colon, resection involves right hemicolectomy; meanwhile, left-sided tumor requires left hemicolectomy. In addition, colorectal intussusception mostly ends up with anterior resection. The surgery should be performed as soon as possible to prevent ischemia, necrosis and subsequent perforation of the invaginated bowel segment.

In conclusion, IFP can manifest as small bowel intussusception. The classical triad of intermittent colicky abdominal pain, red currant jelly stool and palpable right lower quadrant sausage-shaped mass is absent in adults with intussusception. Non-oncologic bowel resection is warranted in a non-viable bowel in addition to a primary anastomosis.

CONFLICT OF INTEREST STATEMENT
None declared.

FUNDING
There is no funding.
ETHICAL APPROVAL
No approval was required.

PATIENT CONSENT
Written informed consent was obtained from the patient for publication of this article and the figures related to it.

GUARANTOR
Firdaus Hayati.

ACKNOWLEDGEMENTS
We would like to thank the Director General of Health Malaysia for his permission to publish this article as case report.

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