Sub acute intestinal obstruction - A Vanek's tumor induced ileo-ileal intussusception


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Abstract : Vanek's tumor is a rare benign inflammatory fibroid polyp like localized lesions originating in the submucosa of the gastrointestinal tract. Intussusceptions due to inflammatory fibroid polyps are uncommon; moreover, ileo-ileal intussusception presenting as subacute small bowel obstruction has rarely been reported. We report a 49-year-old man who came with complaints of indigestion, flatulence, altered bowel habits with abdominal distension for the past 1½ year that aggravated since a week. Abdominal examination revealed distension, signs of intestinal obstruction. The patient underwent laparoscopic assisted resection of intussuscepted ileal segment and four-layer side to side anastomosis. Histopathological analysis showed an ulcerative lesion with variable cellularity, formed by spindle cells with small number of mitosis and an abundant inflammatory infiltrate comprising mainly eosinophils. Immunohistochemistry confirmed the diagnosis of ileal Vanek's tumor. Although inflammatory fibroid polyps are seen very rarely in adults, they are among the probable diagnoses that should be considered in obstructive tumors of the small bowel causing Intussusceptions.

Keywords : bowel, Ileo-ileal intussuception, inflammatory polyps

I. Introduction

Adult Intussusception is a rare entity for which the treatment almost always is surgical and most often the diagnosis is made only during surgery in around 50% of the cases. Adult Intussusception remains a rare cause of persistent or intermittent chronic abdominal pain in contrast to its paediatric counterpart which is a well described and frequently reported entity[1].Vanek's tumours are solitary inflammatory polyloid pseudotumours also called eosinophilic granuloma form one of the least common benign small bowel pathology causing Intussusceptions. They fall under the classification of submucosal connective tissue tumours. They typically present in the 5th to 7th decade of life and can be found throughout the gastrointestinal (GI) tract but most commonly in the gastric antrum (70%) or ileum (20%), but rarely in the duodenum and jejunum [2]. The first six cases of the disease were described in 1949 by Vanek as a submucosal granuloma of the stomach with eosinophilic infiltration. Helwig and Ranier in 1953[3] introduced the term Inflammatory Fibroid Polyp (IFP) to define the disease. IFP affects both sexes and all age groups, with a peak of incidence in the fifth and sixth decades [4, 5]. At the time of diagnosis most IFPs, as in the present patient, have a diameter of 3 to 4 cm, however, there is also a report of a case with an IFP 12.5 cm in size [6]. The lesions have always been recorded as solitary polyps.

A review of the literature on small bowel Intussusceptions and gastro-intestinal IFP is made, based on a well illustrated case of ileo-ileal Intussusception secondary to a Vanek’s tumour.

II. Case History :

A 56-year-old male was admitted with a 45-day history of post-alimentary intermittent and generalized abdominal colics. Abdominal examination revealed intense and diffuse abdominal pain with distension and clanging intestinal sounds. Digital rectal examination did not show the presence of feces, mucus or blood. Laboratory investigation showed leukocytosis (12,100/mm3), Hb 14.9 g/dl, Htc 48%, creatinine 1.2 mg/dl, urea 36 mg/dl, sodium 141 mmol/l and potassium 3.4 mmol/l. A plain abdominal radiograph and ultrasonography did not show any features of obstruction. CT scan showed ileo-ileal small bowel Intussusception (Figure 1). A provisional diagnosis of sub acute intestinal obstruction due to intestinal Intussusception was made. Based on this observation, a laparoscopic exploration was performed. During this procedure the diagnosis of small bowel Intussusception was confirmed and an extracorporeal partial small bowel resection with laterolateral mechanic anastomosis was carried out. Macroscopic examination of the specimen revealed a dumbbell-shaped, circumscribed fleshy white mass measuring 6.5 cm in maximum dimension (Figure 2). Histopathology revealed variable cellularity, with spindle cells having bland nuclei and clear cytoplasm. There was an abundant
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inflammatory infiltrate comprising plasma cells, lymphocytes and eosinophils (Figure 3). The morphological features were typical of IFP.

Figure 1; Abdominal CT-scan. The arrow shows ileo-ileal small bowel Intussusceptions.

Figure 2: Surgical specimen showing a dumbbell shaped white mass.

Figure 3. Histopathological examination of specimen revealed variable cellularity, and spindle cells having bland nuclei, and clear cytoplasm. There was an abundant inflammatory infiltrate comprising plasma cells, lymphocytes, and eosinophils.
Non-specific abdominal pain is the commonest cause of acute surgical admissions for abdominal pain. The diagnosis may not be established in 40-65% of the patients. The investigations are often directed towards excluding ailments like acute appendicitis, intestinal obstruction and the like. At times this may turn chronic resulting were the usual target of investigation is malignancy, inflammatory bowel disease, acid peptic disease and hepato-biliary causes and also gynaecological. Intussusception lie somewhere between and is most often a surgical diagnosis. Intussusception is defined as the prolapse of one segment of the intestines into a distal segment. Intussusception is classified according to its gastrointestinal location: enteric, ileoceleal, and colonic. However, it is difficult to distinguish the different subtypes clinically. Ileoleal intussusceptions usually are secondary to an organic pathology of which Vanek's tumour is one of the rarest causes.

The pathogenesis of Intussusception results from the development of venous and lymphatic congestion and ultimately intestinal oedema if not treated immediately, the most common presenting sign and symptom of an adult patient with Intussusception is colicky abdominal pain (71% to 90% of patients). Nausea and vomiting, bleeding, diarrhoea, constipation, or a change in bowel habits is other non-specific findings at the time of presentation. Palpation of an abdominal mass during clinical examination is only reported in less than 50% of patients. Clinically, ileoileal Intussusception presents either as acute pathology with signs and symptoms of abdominal obstruction or as a more chronic pattern with recurrent colicky abdominal pain. It can be explained by transient Intussusception and self-reduction of the affected bowel and the duration of symptoms is longer in benign and enteric lesions usually. Only 5% of all Intussusceptions occur in adults. In 63% cases of adult small bowel Intussusceptions a benign underlying lesion can be found. Inflammatory Fibroid Polyp (IFP) is an uncommon, benign, sub-mucosal lesion which is usually found incidentally in asymptomatic patients. However, patients can also present with abdominal pain, weight loss, bleeding, dyspeptic symptoms and obstruction, with the pattern of symptoms dependent on the site and size of the lesion.

In the main, IFPs rarely reach more than 6 cm, and their size is presumably related to the likelihood of symptoms. Schildhaus et al., 2008 described 25 cases of small bowel IFPs, the largest of which measured 7 cm and none were found in the jejunum. Recently a paper has described an IFP in the ileum of 15 cm. Other recent papers have described ileal IFPs of smaller size presenting with symptoms of obstruction. There have been reports of retroperitoneal IFPs measuring up to 20 cm and of colon IFPs measuring up to 14 cm. About 15 cases of jejunal IFP have been described in the current literature and these rarely exceed 3-4 cm. Many IFPs are identified incidentally during endoscopy or laparotomy. When symptomatic, the clinical presentation relates to the site of the tumour. Surgical excision is the mainstay of treatment and the tumours are not thought to recur following complete resection.

In the current case report, the patient presented with chronic pain abdomen which later turned acute due to the vanek's tumour induced intussusception which resolved immediately after surgery. To the best of our knowledge, this is the only reported case in which the tumour has presented as ileoileal intussusception. The tumour in this case was not palpable on abdominal examination. This case highlights the importance of performing a thorough examination, even when patients present with unusual symptoms that are suspicious of functional pain.

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