

Ultrasonographic diagnosis of ileo-ileal intussusception secondary to Vanek's tumor

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Abstract. Intussusception is a common condition of bowel obstruction in pediatric patients. However, 5% of all cases occur in adults, mostly aged over fifty, with no difference based on sex, representing about 1% of all causes of bowel obstruction. Compared to pediatric population, it is triggered by a pathologic lead point in about 85% of cases, represented in 60% of cases by malignant and benign neoplasms. Among these neoplasms, an inflammatory fibroid polyp (IFP), a benign neoplastic submucosal lesion also known as Vanek's tumor, is considered a very uncommon cause of adult intussusception. Clinical presentation could differ by location and size of tumor, and may include abdominal pain, nausea, vomiting, diarrhea or constipation, bleeding, weight loss, palpable abdominal mass, bowel obstruction, and gastrointestinal bleeding. Considering its common and non-specific symptoms, radiologic imaging plays a key role in the diagnosis of an IFP, especially computed tomography (CT) scan, which represents the most sensitive modality to confirm intussusception. However, bowel sonography (BS) has become an accurate procedure in various pathological intestinal diseases, also including intussusception. In this paper, we report a rare case of ileo-ileal intussusception secondary to Vanek's tumor diagnosed by BS.

Key Words:

Vanek's tumor, Inflammatory fibroid polyp, Bowel sonography, Surgery.

Introduction

Intussusception – the telescoping of a proximal gastrointestinal segment into the lumen of the immediately distant portion – is considered a common condition of bowel obstruction in pediatric patients, with an overall incidence of 2-3 cases per million per year^{1,2}. However, 5% of all cases occur

in adults, mostly aged over fifty, with no difference based on sex, representing about 1% of all causes of bowel obstruction^{3,4}. Compared to pediatric population, it is triggered by a pathologic lead point in about 85% of cases, represented in 60% of cases by malignant and benign neoplasms, followed by postoperative adhesences, stricturing Crohn's disease, bowel infections, and Meckel's diverticulum⁵⁻⁷. Considering their localization, benign lesions are more often the cause of small bowel intussusception, while malignant lesions, particularly adenocarcinoma, usually trigger colonic intussusception⁸⁻¹⁰.

Of the benign and malignant neoplasms, an inflammatory fibroid polyp (IFP), a benign neoplastic submucosal lesion, also known as Vanek's tumor, is considered a very uncommon cause of adult intussusception¹¹. Described for the first time by Vanek in 1949 as a benign, solitary, submucosal granuloma of the stomach characterized by loose of connective tissues and vessels, with an eosinophilic inflammatory infiltrate, an IFP can also affect the small bowel (21.1%), colon (8.4%), esophagus (1.7%), appendix (0.7%), duodenum (0.5%), and gallbladder (0.2%)^{11,12}. An IFP occurs more frequently in women (1.3:1 F/M ratio), with a peak incidence during the fifth decade¹¹. Although the etiopathogenesis is still unclear, local infections, allergic reactions, autoimmune processes, and mutations in platelet-derived growth factor alpha (PDGFRA) have been linked to the development of an IFP¹³. Clinical presentation could differ by location and size of tumor, and may include abdominal pain, nausea, vomiting, diarrhea or constipation, weight loss, palpable abdominal mass, bowel obstruction, and gastrointestinal bleeding^{11,14}. Considering its common and non-specific symptoms, radiologic imaging plays a key role in the diagnosis of an IFP, es-

pecially computed tomography (CT) scan, which represents the most sensitive modality to confirm intussusception^{15,16}. However, bowel sonography (BS) has become an accurate procedure in various pathological intestinal diseases, also including intussusception. In effect, BS has shown a very high diagnostic accuracy in intestinal pathological condition, such as inflammatory bowel diseases^{17,18}, coeliac disease¹⁹, intestinal neoplasms²⁰ and bowel invaginations²¹. In the diagnostic setting of bowel intussusception, BS highlights the presence of a target sign on axial view with a “trident” image on longitudinal sections; sometimes a polypoid lesion or a stricture can be observed as the fulcrum of the invagination.

Endoscopy should be the treatment of choice for most IFPs, but surgery, performed as early as possible, is frequently recommended as the best treatment for intussusceptions secondary to an IFP¹³.

In this paper, we report a case of ileo-ileal intussusception secondary to Vanek's tumor diagnosed by BS.

Case presentation

In October 2020, a 61-year-old female (height 1.63 m, weight 100 kg), with a 2-month history of recurring and self-limiting right-sided abdominal pain associated with abdominal distention and obstipation, was admitted to our Gastroenterology Unit. The patient signed the informed consent. She reported hypertension, type-2 diabetes and asthma in her anamnesis. She underwent an appendectomy about 50 years ago. She had no familiar history of gastrointestinal cancer. Physical examination revealed a palpable abdominal mass in her right iliac fossa. Laboratory findings showed mild leukocytosis (WBC $10.76 \times 10^9/L$) and increased levels of inflammatory biomarkers (CRP 26.6 mg/L). Ultrasonography, performed by a gastroenterologist experienced in BS, showed a well-defined “target-type” lesion (measuring 20 x 15 mm) derived from an ileo-ileal intussusception on a polypoid fulcrum of about 3 cm with a stable dilatation of the upstream intestinal loop (3.5 cm) (Figure 1a). The invagination appeared to be distant 10-15 cm from the ileo-cecal valve. To surely detect the polypoid lesion, and to firstly attempt an endoscopic resection, ileo-colonoscopy was performed. The endoscopic procedure revealed an ileal pedunculated polyp (measuring 3.5 cm) sited 10 cm beyond the ileo-cecal valve, not treatable by endoscopic polypectomy because of the

rigid and fixed visceral angle (Figure 1b). So, the patient underwent a laparoscopic surgical resection of the ileum with a side-to-side stapled anastomosis (Figures 1c-d). Histopathological analysis revealed an IFP characterized by the presence of spindle and epithelioid cells, with immunohistochemistry negative for anaplastic lymphoma kinase (ALK), in the context of an abundant lymphoplasmacellular infiltrate.

After 6 months of follow-up, the patient was symptom-free with BS showing a normal picture.

Discussion

An IFP is a rare intraluminal polypoid, benign mesenchymal lesion characterized by proliferation of highly vascular fibrous tissue and infiltration by different inflammatory cells, which can affect any part of the gastrointestinal tract, mostly the stomach^{11,12}. Different names have been suggested to describe Vanek's tumor, such as eosinophilic granuloma, granuloblastoma or gastric fibroma with eosinophilic infiltration, hemangiopericytoma, and inflammatory fibroid tumor. However, in 1953, Helwig and Ranier²² concluded that these different conditions represented the same pathological entity, named an IFP, which thus became the preferred term for this lesion.

The clinical presentation of an IFP could differ from more frequent asymptomatic cases, diagnosed incidentally during endoscopy or laparotomy, to acute abdomen, caused by intussusception²³.

Differential diagnosis between an IFP and other neoplasms of the gastrointestinal tract include spindle cell lesions, such as inflammatory fibrosarcoma, spindle cell carcinoids, and gastrointestinal stromal tumors¹⁴. Macroscopically, an IFP appears typically solitary and well circumscribed, arising by the submucosa, pedunculated or sessile lesion, with erosion or ulceration of the overlying mucosa, and with a diameter of 2-5 cm²⁴. Microscopically, an IFP – the gastric variant – is composed of a loose mixture of spindle-shaped and stellate to epithelioid stromal cells positive for CD34 and vimentin and negative for CD117 on immunohistochemistry, with an inflammatory infiltrate predominantly composed of lymphocytes and eosinophils but also of plasma cells and histiocytes, and small blood vessels in an edematous/myxoid background²⁵. In contrast, the intestinal variant is paucicellular and rich in collagen with epithelioid stromal cells negative for CD34²⁴.

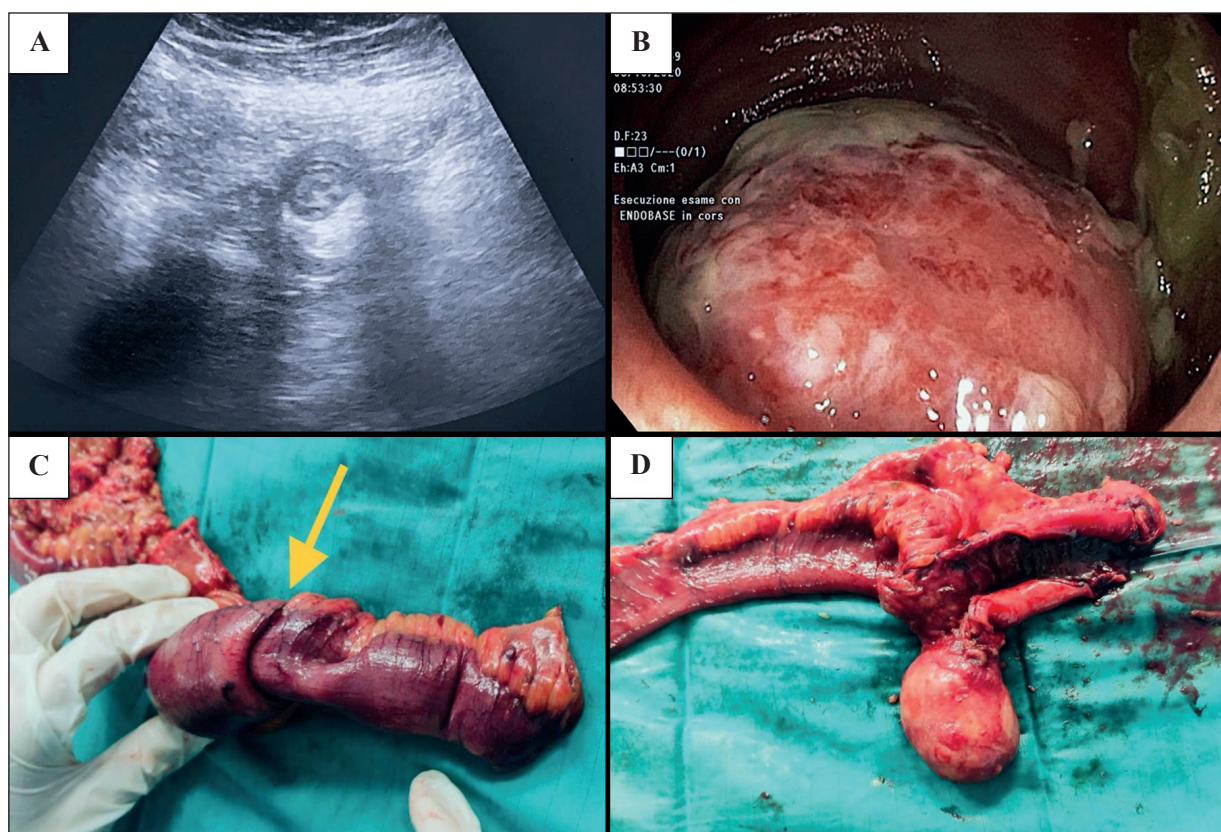


Figure 1. A, Vanek tumor on sonography; B, Vanek tumor on ileoscopy; C, Ileo-ileal intussusception (*arrow*) during surgery; D, Vanek tumor (open surgical finding).

CT scan is now considered the most sensitive modality for diagnosing intussusception^{15,16}. BS has also proven to be a useful technique for detecting the presence of intussusception, showing the classical “target sign” and “pseudo-kidney sign” in transverse and longitudinal view, respectively²⁶.

A very few cases of Vanek’s tumor complicated by intestinal invagination have been described in literature. Interestingly, only recently, a similar condition was described by Johan and colleagues²⁷. However, in this case described by Malaysians Authors the diagnosis of bowel intussusception due a polypoid lesion was mainly performed by CT scans²⁷.

The present case report, which is written by gastroenterologists, radiologists and other clinicians experienced in BS, highlights, once again, the high diagnostic accuracy of ultrasonography, confirming it to be a safe, cost-limited and well-accepted procedure also in difficult and a rare diagnostic setting as an ileal-ileal intussusception due to a Vanek’s tumor.

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Conflict of Interests

The authors declare that they have no conflict of interest.

Authors’ contributions

AR and RdS wrote the manuscript; AR performed ultrasonography and ileo-colonoscopy; GL and FT performed ileocolic resection; LDB and SR followed-up the patient; GN critically revised the manuscript for important intellectual content. All authors approved the final version of the manuscript.

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