Small bowel intussusception secondary to giant inflammatory fibroid polyp of the ileum: A case report and review of the literature

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ABSTRACT

Inflammatory fibroid polyps (IFP) are benign mesenchymal tumors described first by Vanek, who reported a series of cases occurring in the gastric submucosa. Since then, there have been varying presentations of these benign tumors, many of them have shown to infiltrate into surrounding tissue and can cause significant symptoms including intestinal obstruction and intussusception. This can result in peritonitis and bowel necrosis, and can change the management of a typical bowel obstruction. Adult intussusceptions are rare, as this phenomenon occurs most often in infants and children. IFP can occur anywhere in the gastrointestinal tract but most commonly in stomach and small intestine. The largest IFP arising from the gastrointestinal tract described in English literature has been 15 cm at total length. Our case is unique in that it is the largest IFP reported to date occurring in the GI tract.

INTRODUCTION

The inflammatory fibroid polyp (IFP) is a benign mesenchymal growth arising in the submucosa of the gastrointestinal tract that was first described by Vanek in 1949. It was reported to be made of collagenous tissue with fibroblasts, lymphocytes, neutrophils and eosinophils [1]. Today it is defined as a poorly circumscribed collection of proliferating cells with an inflammatory infiltrate arranged around capillaries. It is also known to have varying degrees of infiltration into local tissues [2]. Patients with this type of tumor can present in many different ways depending on location, size and infiltration into local tissues. IFP is a rare cause of intestinal obstruction, representing about 2% of cases. Rarely, a large IFP can act as a lead point for intussusception and allow for telescoping of bowel segments [3]. Only 71 cases have been reported in literature. Adult intussusceptions represent 5% of all intussusceptions, as most cases occur in the pediatric population [4]. To the best of our knowledge, the largest gastrointestinal IFP that has been reported in literature...
has been 15 cm described by Costamagna et al. [5]. We report a case of a giant IFP measuring 20.5 cm in length and causing intussusception and obstruction.

**CASE REPORT**

**Clinical**

A 56-year-old male patient with a history of multiple polyps in the colon presented with a history of 5-7 days of intermittent colicky mid-abdominal pain that had been progressively worsening over the last 48 hours. The patient did endorse foul smelling, dark diarrhea, but denied nausea or vomiting. On physical examination, abdomen was soft, slightly distended, tympanic to percussion and tender in left lower quadrant and suprapubic areas. Bowel sounds were hyperactive. Significant laboratory data included leukocytosis of 13.1(10³/µl), anemia with hemoglobin of 8.7 g/dl, and platelet count of 671(10³/µl). CT scan of abdomen and pelvis revealed an unusual, tubular, elongated, intraluminal and partially extraluminal ileal mass causing small bowel obstruction and short segment intussusception Figure 1 (A and B). There were also several enteric nodes measuring up to 7 cm by imaging. Together these findings were suspicious of neoplasm and patient was taken to the operating room for an exploratory laparotomy and small bowel resection.

**Pathology**

Opening the small bowel revealed a large solid tan-pink elongated submucosal mass with intact overlying mucosa measuring 20.5 cm in greatest dimension (Figure 1 C). Histologic sections showed a non-encapsulated fibro-histiocytic lesion composed of bland spindled cells embedded in a loose fibromyxoid stroma. The stroma is rich with eosinophils and contain multiple thin walled blood vessels with characteristic “onion skin” arrangement of spindled cells around vessels Figure 2 (A–D). No mitoses or necrosis is identified. Immunohistochemical studies reveal tumor cells strongly positive for CD 34, vimentin and CD 68 and negative for CD 117 (c-Kit) Figure 3 (A–D). Other immunostains like actin, desmin and S-100 were also performed and were negative. These histologic findings were consistent with a benign, non-neoplastic tumor-like mass, also known as Giant Inflammatory Fibroid Polyp-Tumor Like (Vanek’s Tumor).

**DISCUSSION**

Inflammatory fibroid polyp (IFP) was first identified by Vanek, noted to arise in the gastric submucosa. He reported several cases of these tumors causing presentations ranging from mild recurrent pain to stenosis [1]. Since then, IFP’s have been identified...
Intussusception in adults is relatively rare and represents 10% of all intussusceptions. It is more common in the pediatric population and is often idiopathic. Conversely, in adults, 90% of intussusceptions are due to a pathological etiology that acts as a lead point for development of secondary intussusception [8, 9]. Most common symptoms were abdominal pain, nausea and vomiting, and a palpable mass only occurred in 24–42% of patients [10]. Yakan et al. found that adult intussusceptions were 85% in the small intestine and 15% in the colon [10]. Previous standards have encouraged the use of ultrasonography in the diagnosis of intussusception, which should show a characteristic pseudo kidney sign or target sign [7]. More recently, the CT scan has become widely used and accepted as the best diagnostic test for intussusception. In some cases, 90.5% of intussusception is diagnosed with CT scan and it has an accuracy of 58–100% [4, 11, 12]. In a CT scan, a characteristic target or sausage appearance has a high diagnostic yield and this modality can also provide information on metastasis or local invasion [4, 10, 11, 13]. Lipomas are the most common benign cause of adult intussusception and there have been cases where intussusception is only reduced as in the case that Joyce et al reports. The lead lesion was thought to be a lipoma and was treated laparoscopically without resection. This resulted in a recurrence of intussusception with complete obstruction [13]. Our patient had a very large lead lesion; therefore, surgical resection was preferred due to the possibility of future complications of an unresected lesion. Conservative treatment such has nasogastric tube decompression and bowel rest has also shown to be less effective than surgical intervention [16].

Although the etiology of IFP’s is still debated, the PDGFRA mutation has been implicated in causing these lesions. A mutation in exon 12 is associated with small intestinal lesions and a mutation in exon 18 is associated with gastric IFP [17, 18]. Small intestinal IFP’s typically develop to be larger than gastric lesions, but the largest gastrointestinal IFP reported has been 15 cm [5]. There have been reports of retroperitoneal IFP measuring up to 20 cm; however none that were part of the GI tract [19]. To the best of our knowledge, this case of an inflammatory fibroid polyp measuring 20.5 cm in greatest dimension is the largest IFP reported to date.
CONCLUSION

IFP is a rare cause of intestinal obstruction, representing about 2% of cases. There have been varying presentations of these benign tumors, many of them have shown to infiltrate into surrounding tissue and can cause significant symptoms including intestinal obstruction and intussusception. To the best of our knowledge, this case of an inflammatory fibroid polyp measuring 20.5 cm in greatest dimension is the largest IFP reported to date.

REFERENCES


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Author Contributions

Ayaz Ghani – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Saad Baqai – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published
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Hani El-Fanek – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

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Consent Statement

Since no direct patient identifier was used, no written or verbal informed consent was obtained from the patient for publication of this case report.

Conflict of Interest

Authors declare no conflict of interest.

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