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Case Report

Inflammatory Fibroid Polyp of the Ileum: A Case Report of a Rare Tumor Causing Small Bowel Intussusception

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Abstract

An Inflammatory fibroid polyp (IFP), also known as Vanek's tumor, is a rare benign mesenchymal lesion of the gastrointestinal tract. IFP can be found throughout the gastrointestinal tract but most frequently in the gastric antrum or ileum. The clinical presentation is usually asymptomatic, yet the symptoms depend on the location of the polyp.

We report a case of a 30-year-old healthy male, who presented to the emergency department with symptoms of bowel obstruction. A computed tomography of the abdomen was done which demonstrated a small bowel obstruction due to an ileal intussusception.

A laparotomy was performed which revealed an ileo-ileal intussusception. After manual reduction, a pedunculated intraluminal mass was found at the terminal ileum and an enterectomy was performed with a small bowel anastomosis. The postoperative course was unremarkable and the patient was discharged home. Histopathology examination revealed the tumor to be a benign inflammatory fibroid polyp. This case report presents a rare benign tumor originating in the ileum, presenting as a small bowel intussusception that was discovered incidentally during an exploratory laparotomy.

Keywords: *Vanek's Tumor; Inflammatory Fibroid Polyp; Intussusception*

Abbreviations

IFP: Inflammatory Fibroid Polyp; CT: Computed Tomography; SBO: Small Bowel Obstruction; GIA: Gastrointestinal Anastomosis; GI: Gastrointestinal

Introduction

Intussusception is defined as an invagination of the proximal part of the bowel along with its mesentery into an adjacent segment. This can lead to impaired peristalsis, obstruction and possible vascular compromise [1]. It is a rare cause of intestinal obstruction in adults and accounts for approximately 1%-5% of all cases of bowel obstruction [2]. In adults presenting with intussusception, most cases are secondary to a pathology, such as benign tumors, primary malignancies, or metastatic tumors that serve as the lead point of intussusception [3]. Inflammatory fibroid polyps are mesenchymal tumors that arise in the submucosa and mucosa of the GI tract. It is usually localized in the gastric antrum, although it can occur throughout the gastrointestinal tract [4]. We present a rare case of an ileal IFP noted during a laparotomy of an adult presenting with small bowel obstruction (SBO).

Case Presentation

We present a 32-year-old male, with an unremarkable past medical and surgical history, who was referred to our emergency department due to a 3-day history of abdominal pain. His vital signs were within normal limits. During abdominal examination, his abdomen was distended and he also had localized tenderness and guarding on palpation of the right lower quadrant. His abdomen was also tympanic on percussion. In addition, the patient reported a history of constipation for the last year. Abdominal radiography revealed a stepladder sign with several small bowel loops of air fluid levels (Figure 1A). Abdominal computed tomography (CT) revealed dilated small bowel loops with a localized target sign, delineating a small bowel intussusception causing an obstruction (Figure 1B).

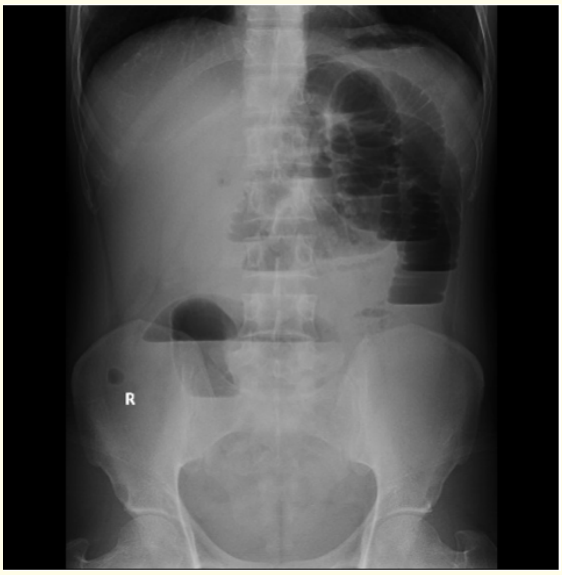

Figure 1A: *Abdominal x-ray showing multiple air fluid levels delineating a possible bowel obstruction.*

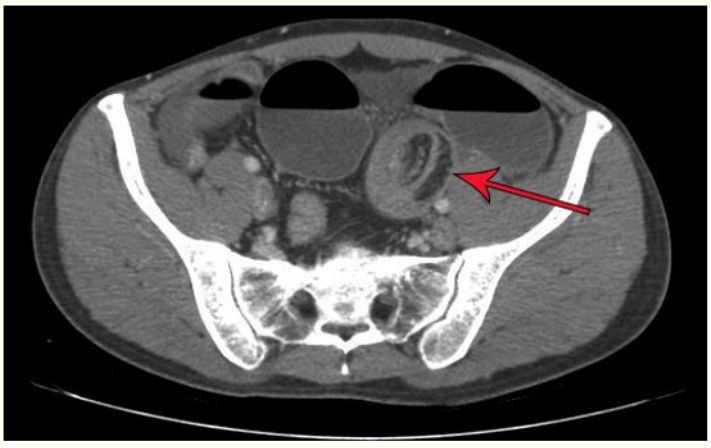

Figure 1B: *CT Scan (Axial view): Dilated small bowel loops with a localized target sign (Arrow) outlining a small bowel intussusception.*

Laboratory investigations are presented in the table below (Table 1).

Investigation		Normal Range	Unit	Investigation		Normal Range	Unit
White blood cells	10.8 H	4.23-9.07	10^9/L	Eosinophils#	0.01 L	0.04-0.54	10^9/L
Red blood cells	5.46	4.63-6.08	10^{12}/L	Basophils#	0.01	0-0.08	10^9/L
Hemoglobin	146	137-175	g/L	Creatinine	108 H	60-106	umol/L
Hematocrit	0.441	0.4-0.51	L/L	Na (Sodium)	136	135-146	mmol/L
Mean Corpuscular Volume	80.8	79-92.2	fL	K (Potassium)	4.70	3.5-5.3	mmol/L
Mean Corpuscular Hemoglobin	26.7	25.7-32.2	pg	CL (Chloride)	90 L	95-107	mmol/L
Mean Corpuscular Hemoglobin Concentration	331	323-365	g/L	eGFR (CKD - EPI)	77	More than 90	ml/min/1.73m^2
Red Blood Cell Distribution Width	12.4	11.6-14.4	%	Prothrombin Time	17.80	13.1 - 16.7	Seconds
Platelet count	298	140-400	10^9/L	International Normalized Ratio	1.330		
Neutrophils#	9.38 H	1.56-6.13	10^9/L	Activated partial thromboplastin time (APTT)	36.4	25.4 - 36.9	Seconds
Lymphocytes#	0.79 L	1.32-3.57	10^9/L	APTT Ratio	1.30		(Ratio)
Monocytes#	0.57	0.35-1	10^9/L				

Table 1

An urgent exploratory laparotomy was performed, which revealed a segmental small bowel intussusception at the terminal ileum. Gentle manual traction was performed to reduce the intussusceptum from the intussusceptor. Once reduced, the bowel segment did not look ischemic, but on palpation, an intraluminal mass was found in the distal ileum, located approximately 50 cm from the ileocecal valve (Figure 2A and 2B). Further bowel exploration did not reveal any other masses or abnormalities. An enterectomy was performed encompassing the intraluminal tumor and a stapled side-to-side anastomosis was created.

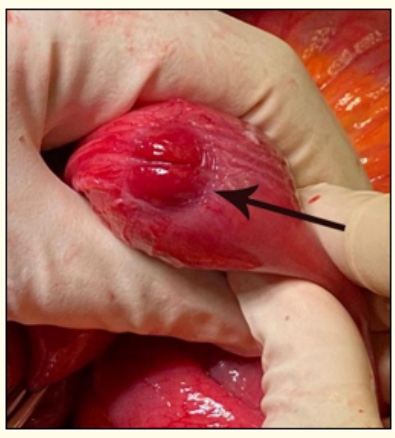

Figure 2A: *External view of the small bowel segment containing the lesion, illustrating an intact ileum with a firm mass located within the bowel wall as pointed by the arrow.*

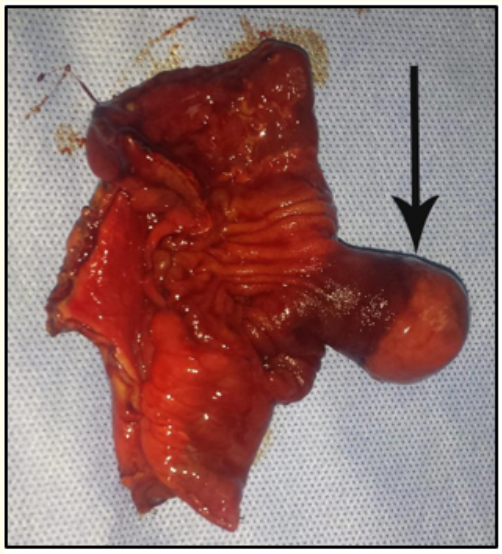

Figure 2B: *Gross examination of the enterectomy specimen showing an intraluminal pedunculated polypoid lesion originating from the small bowel wall (Arrow). The rest of the specimen appears to be grossly normal.*

The patient was discharged on the 4th postoperative day following an uneventful recovery.

The histopathology specimen revealed a firm greyish brown mass arising from the submucosa, measuring 3.5 X 2.5 X 2cm with a smooth outer surface. Macroscopically, the sections revealed a polypoid lesion with focal ulceration overlying the mucosa, formed of spindle and stellate cells along with inflammatory cells rich in eosinophils. On immunohistochemical analysis, the lesion was focally positive for CD34, Cyclin D1, Bcl2 and strongly positive for Vimentin. It was negative for CD117, Dog 1, S100, PanCK, Desmin, CD1a, EMA, SMA, ALK1, Beta-Catenin, NSE, CD99 and Calponin. The characteristics of the specimen thus revealed the tumor to be consistent with an inflammatory fibroid polyp of the small bowel.

Discussion

Inflammatory fibroid polyps are rare benign mesenchymal tumors arising from the mucosa and submucosa of the gastrointestinal tract. Multiple factors have been identified as initiators of the inflammatory process that would eventually lead to the development of IFP. These include trauma, allergy, genetic factors, as well as bacterial, physical, chemical and metabolic stimuli.

The exact mechanism leading to intussusception is still unknown. It is believed that lesions in the bowel wall or any irritant within the lumen that may alter the normal peristaltic bowel movement can result in the invagination process leading to the intussusception.

Intussusceptions have been classified according to their locations into four categories: entero-enteric, colocolic, ileocolic, or ileocecal [5].

Intussusception in adults occurs secondary to a pathological condition involving a lead point. The etiologies causing intussusception in the small bowel and the colon are diverse.

In the small intestine, benign processes are predominant in up to 90% of cases. These include adhesions, hamartomas, Peutz-Jeghers syndrome, lipomas, leiomyomas, neurofibromas, adenomas, IFP, Meckel's diverticulum, lymphoid hyperplasia and tuberculosis.

IFP is considered a benign gastrointestinal tract lesion. It develops in the submucosa and is composed of mononuclear spindle-shaped cells and prominent eosinophils. Vanek first discovered inflammatory fibroid polyps in the stomach in 1949. He described the lesion as a submucosal granuloma with eosinophilic infiltration [6]. There are different names for IFP in literature including eosinophilic granuloma, hemangiopericytoma, neurofibroma, and polypoid fibroma. Gastrointestinal stromal tumor (GIST) may have the same pathological features as an IFP and immunohistochemistry can be used to differentiate them. Although both are positive for CD34 and vimentin, CD117 is specific for GIST. Recent histopathology studies from Schildhaus., *et al.* [12] have shown that inflammatory fibroid polyps express a platelet-derived growth factor receptor alpha (PDGFRA) in more than 90% of cases, and the majority harbor activating PDGFRA mutations. Therefore, inflammatory fibroid polyps represent true benign mesenchymal tumors of the gastrointestinal tract, and although many years thought to be reactive, they are now regarded to be true PDGFRA-driven benign neoplasms [7].

The most common sites of occurrence are the gastric antrum (60 - 70%), small bowel (18 - 20%), colon and rectum (4 - 7%), and far less common (1%) in the esophagus, duodenum, gallbladder, and appendix (< 1%) [8-11]. They are usually asymptomatic and are discovered incidentally during endoscopic procedures or surgical exploration. The clinical symptoms depend on the location and size of the tumor. Abdominal pain is the main symptom in patients with lesions in the stomach. Intussusception and obstruction tends to be the most frequent initial symptoms when the polyp is located in the small intestine. Other GI symptoms include vomiting, diarrhea, and bloody stool, where larger polyps tend to erode and ulcerate. Tenesmus and change in bowel habits are also seen, although their frequencies are low [12].

Imaging studies such as CT scans usually diagnose intussusception because the polyp's clinical manifestation and history is nonspecific and present as small bowel obstruction.

In our case, ileo-ileal intussusception was apparent on CT scan. However, CT did not reveal any apparent lesions that might have served as a leading point in this case because of the overlapping ileo-ileal wall and edematous and dilated small bowel loops. The finding of the intussusception secondary to the polyp was discovered during laparotomy. The primary treatment for adult intussusception is surgical resection.

According to the current literature, IFP is a benign localized tumor with no risk of recurrence after complete excision and no malignant potential [13].

Due to the unknown origin of the lesion and rarity of the disease, there are no specific guidelines regarding follow up. Nevertheless, an article about clinical surveillance of the affected children suggested that follow up with endoscopy should be done only for the cases of endoscopically removed polyps [14]. In cases such as ours where the tumor was found incidentally, an upper gastrointestinal endoscopy and colonoscopy follow up may be warranted to make sure that there are no other co-existent polyps present in the areas examined by endoscopy. Due to the low incidence of these tumors, data gathering and analysis through an international registry may aid in further understanding the etiology and prognosis of these tumors in order to set guidelines for optimal patient management.

Conclusion

In conclusion, we report a rare case of an ileal IFP presenting with small bowel intussusception that was detected during laparotomy. Although rare and benign, this tumor tends to only be diagnosed once surgical or endoscopic resection has been performed. While management with resection seems to be sufficient due to the benign nature of the tumor, further investigations on the pathogenesis, pathophysiology, and associative features of inflammatory fibroid polyps are necessary to aid in optimizing detection, treatment and management of these lesions.

Conflict of Interest

There are no conflicts of interest to declare by all the authors.

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