EDORIUM Journals

J Case Rep Images Pathol 2018;4:100020Z11AG2018.
www.edoriumjournals.com/case-reports/jcrp

CASE REPORT PEER REVIEWED | OPEN ACCESS

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

Ayaz Ghani, Saad Baqai, Nayan Mainkar, Hani El-Fanek

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.

Ayaz Ghani[1], Saad Baqai[2], Nayan Mainkar[3], Hani El-Fanek[2]

Affiliations: [1]Chief Resident, Department of Pathology, Danbury Hospital, Western Connecticut Health Network, 24 Hospital Ave, Danbury, CT, USA; [2]Attending Pathologist, Department of Pathology, Danbury Hospital, Western Connecticut Health Network and University at Buffalo, 24 Hospital Ave, Danbury, CT, USA; [3]Medical Student, Department of Pathology, Danbury Hospital, Western Connecticut Health Network and Ross University School of Medicine, 24 Hospital Ave, Danbury, CT, USA.

Corresponding Author: Ayaz Ghani, MD, Danbury Hospital, Western Connecticut Health Network, 24 Hospital Ave, Danbury, CT 06810, USA; Email: ayaz133@yahoo.com

Received: 23 May 2018
Accepted: 22 June 2018
Published: 20 July 2018

Keywords: Bowel obstruction, Gastrointestinal tract, Inflammatory fibroid polyp, Intussusception, Vanek's tumor

How to cite this article

Ghani A, Baqai S, Mainkar N, El-Fanek H. Small bowel intussusception secondary to giant inflammatory fibroid polyp of the ileum: A case report and review of the literature. J Case Rep Images Pathol 2018;4:100020Z11AG2018.

Article ID: 100020Z11AG2018

****

doi:10.5348/100020Z11AG2018CR

INTRODUCTION

The inflammatory fibroid polyp (IFP) is a benign mesenchymal growth arising in the submucosa of the gastrointestinal tract that was first described by Vanekin 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(10^3/µl), anemia with hemoglobin of 8.7 g/dl, and platelet count of 671(10^3/µl). CT scan of abdomen and pelvis revealed an unusual, tubular, elongated, intraluminal and partially extraluminalileal 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 lesioncomposed of bland spindled cells embedded in a loose fibromyxoid stroma. The stromais 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

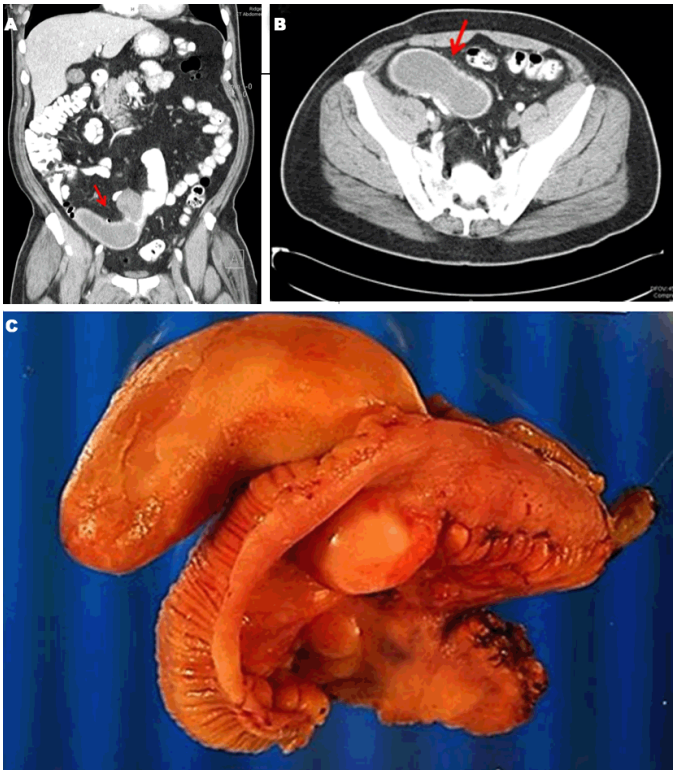

Figure 1(A–C): Coronal and axial CT scan of abdomen and pelvis revealing a tubular, elongated ileal mass (red arrows) causing partial obstruction and intussusception (A, B). Gross image with a large solid tan-pink elongated luminal and extraluminal tumor mass protruding into the small bowel lumen on one side and pushing the serosal surface on the other side (C).

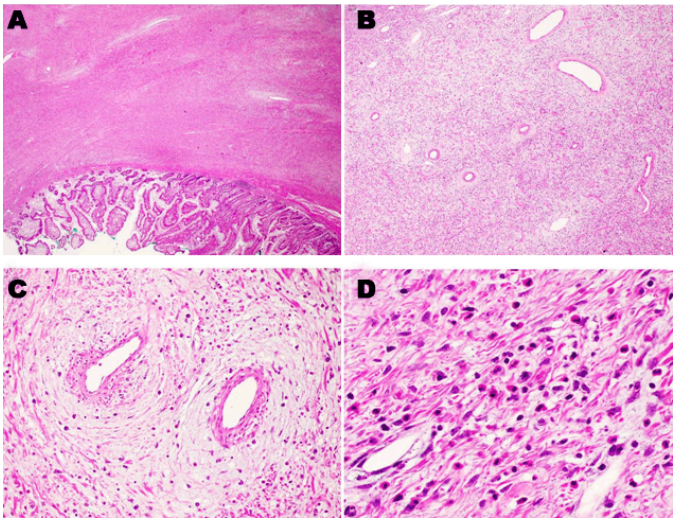

Figure 2(A–D): Histopathologic findings: The submucosal tumor is composed of hyper and hypocellular areas with proliferation of bland spindle and stellate-shaped mesenchymal cells embedded in a loose fibromyxoid stroma (A, B). The stroma demonstrates prominent vascularity with aggregation of mesenchymal cells around the vessels in an onion skinning pattern (C). Diffuse infiltration of eosinophils and few scattered plasma cells (D).

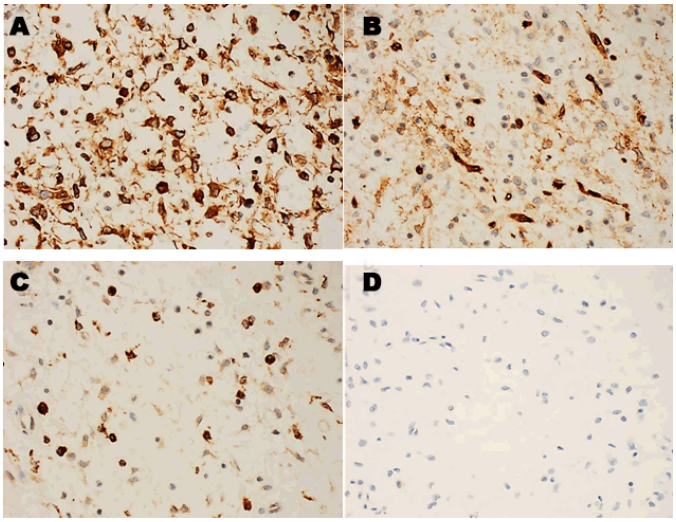

Figure 3(A–D): Immunohistochemical studies reveal tumor cells positive for vimentin (A), CD34 (B), CD68 (C) and negative for c-Kit (D).

throughout the GI tract, more commonly in the stomach, but larger tumors are often in the intestines [2]. Typically, patients are asymptomatic and the IFP is discovered incidentally during endoscopy or laparoscopy [6]. If the lesion is symptomatic, it typically presents during the 6th or 7th decade, but there have been reports of young adults with chronic manifestations such as weight loss and intermittent constipation [7]. Clinical presentations of intestinal lesions range from insidious bleeding resulting in anemia to acute intestinal obstruction or perforation [2]. Our patient presented at the age of 54 with partial obstruction, which was a cause for urgent treatment.

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 IFP has been confused with lipoma on radiography [8, 13].

On gross appearance, the resected tumor is usually polypoid in appearance with a tan, yellow, or gray surface and the mucosa is usually ulcerated [14]. This gross description is consistent with our patient; however, the overlying mucosa was intact, despite the lesion being so large. IFP lesions leading to mechanical intraluminal obstruction without intussusception have been reported [3]. This lesion can pose some difficulty for pathologists due to its histopathological similarities to many other spindle cell neoplasms of the GI tract including gastrointestinal stromal tumor (GIST). GIST stains positive for c-kit and DOG-1, whereas IFP will be negative for c-kit, DOG-1 and positive for CD34 and vimentin [7, 15]. IFP's are submucosal lesions that have prominent vasculature and spindle cells. The characteristic finding of "onion skinning" is helpful in diagnosis as well as abundance of eosinophils.

Treatment of the intussusception is either primary reduction or resection of the involved bowel. The majority of small intestine lead point lesions are benign, whereas in the colon they are primarily malignant. Therefore, it is recommended that colonoscopy and reduction of any small intestinal intussusception be the first modality of treatment if possible in order to avoid unnecessary surgery [4, 8, 10]. As a rule, colonic lesions in patients over the age of 60 should not be reduced due to high likelihood of malignancy [9]. In patients who are prone to having many polyps such as Peutz-Jager syndrome, endoscopic and laparoscopic reduction with polypectomy should be preferred in an effort to preserve bowel length [9]. However, recurrence is more likely if the 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 PDDGFR-A 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

1. Vanek J. Gastric submucosal granuloma with eosinophilic infiltration. Am J Pathol 1949 May;25(3):397–411.
2. Acero D, Garijo G, Hombrados M, et al. Gastrointestinal inflammatory fibroid polyps. Clinical characteristics and follow-up in a series of 26 patients. [Article in Spanish]. Gastroenterol Hepatol 2005 Apr;28(4):215–20.
3. Hiremath S, Nanjappa N, Kamath S. Inflammatory fibroid polyp (IFP) of the terminal ileum presenting as acute intestinal obstruction without intussusception. BMJ Case Rep 2015 Sep 7;2015.
4. Wang N, Cui XY, Liu Y, et al. Adult intussusception: A retrospective review of 41 cases. World J Gastroenterol 2009 Jul 14;15(26):3303–8.
5. Costamagna D, Erra S, Zullo A, Servente G, Durando R. Small bowel intussusception secondary to inflammatory fibroid polyp of the ileum: Report of a case. Chir Ital 2008 Mar–Apr;60(2):323–7.
6. Bhutia CT, Das D, Bhutia P. Inflammatory fibroid polyp of the ileum presenting with acute intestinal obstruction in an adult patient: A case report. J Clin Diagn Res 2016 Jan;10(1):EJ01–2.
7. Rais M, Chahdi H, Elfahssi M, Albouzidi A, Oukabli M. An unusual cause of intestinal obstruction in a young adult patient: Inflammatory fibroid polyp. Case Rep Surg 2017;2017:3675848.
8. Akbulut S. Intussusception due to inflammatory fibroid polyp: A case report and comprehensive literature review. World J Gastroenterol 2012 Oct 28;18(40):5745–52.
9. Eisen LK, Cunningham JD, Aufses AH Jr. Intussusception in adults: Institutional review. J Am Coll Surg 1999 Apr;188(4):390–5.
10. Yakan S, Caliskan C, Makay O, Denecli AG, Korkut MA. Intussusception in adults: Clinical characteristics, diagnosis and operative strategies. World J Gastroenterol 2009 Apr 28;15(16):1985–9.
11. Takeuchi K, Tsuzuki Y, Ando T, et al. The diagnosis and treatment of adult intussusception. J Clin Gastroenterol 2003 Jan;36(1):18–21.
12. Tan KY, Tan SM, Tan AG, Chen CY, Chng HC, Hoe MN. Adult intussusception: Experience in Singapore. ANZ J Surg 2003 Dec;73(12):1044–7.
13. Joyce KM, Waters PS, Waldron RM, et al. Recurrent adult jejuno-jejunal intussusception due to inflammatory fibroid polyp - Vanek's tumour: A case report. Diagn Pathol 2014 Jun 27;9:127.
14. Shimer GR, Helwig EB. Inflammatory fibroid polyps of the intestine. Am J Clin Pathol 1984 Jun;81(6):708–14.
15. Kolodziejczyk P, Yao T, Tsuneyoshi M. Inflammatory fibroid polyp of the stomach. A special reference to an immunohistochemical profile of 42 cases. Am J Surg Pathol 1993 Nov;17(11):1159–68.
16. Kang SH, Kim SW, Moon HS, et al. Inflammatory fibroid polyp in the jejunum causing small bowel intussusception. Ann Coloproctol 2015 Jun;31(3):106–9.
17. Liu D, Wang J, Chen M, et al. Inflammatory fibroid polyp of the gastrointestinal tract: A clinicopathologic features of 37 cases. [Article in Chinese]. Zhonghua Bing Li Xue Za Zhi 2016 Jun 8;45(6):381–6.
18. Huss S, Wardelmann E, Goltz D, et al. Activating PDGFRA mutations in inflammatory fibroid polyps occur in exons 12, 14 and 18 and are associated with tumour localization. Histopathology 2012 Jul;61(1):59–68.
19. Rehman S, Gamie Z, Wilson TR, Coup A, Kaur G. Inflammatory fibroid polyp (Vanek's tumour), an unusual large polyp of the jejunum: A case report. Cases J 2009 May 18;2:7152.

****

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
Nayan Mainkar – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published
Hani El-Fanek – 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

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None.

Consent Statement

Since no direct patient identifier was used, so 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.