CASE REPORT

Inflammatory fibroid polyp (IFP) of the terminal ileum presenting as acute intestinal obstruction without intussusception

Srikantaiah Hiremath,¹ Nikhil Nanjappa,¹ Sulatha Kamath²

¹Department of General Surgery, M S Ramaiah Medical College & Hospitals, Bangalore, Karnataka, India
²Department of Pathology, M S Ramaiah Medical College & Hospitals, Bangalore, Karnataka, India

Correspondence to

Dr. Nikhil Nanjappa,
 drnikhilnanjappa@gmail.com

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SUMMARY

Inflammatory fibroid polyps (IFPs) are rare entities. They commonly occur in the stomach, and a fraction of them are present in the small intestines. The exact aetiopathogenesis for IFPs remains unknown. Most small bowel IFPs are asymptomatic and usually go undetected until they produce symptoms. IFPs are responsible for roughly 2% of all small bowel obstructions. They act as a lead point for intussusception, by telescoping into the distal bowel loops. Nearly 85 such cases have been reported in the literature. However, if the IFP is sufficiently large and pedunculated, it could cause mechanical intraluminal obstruction without intussusception. We report one such case in a middle-aged man who had an impending perforation of the terminal ileum caused by an IFP. Resection and anastomosis of the offending segment of the small intestine remains the standard of care. IFPs lack malignant potential and recurrences are rare.

BACKGROUND

The list of aetiologies for small bowel obstruction is extensive. There are certain causes that are specific for certain age groups. Usually, in the paediatric age group, the most common causes are benign lesions. However, in middle aged or elderly patients, it is attributed either to adhesions or malignant lesions. Inflammatory fibroid polyps (IFPs) are a rare cause for small intestinal obstruction. When they do obstruct, it is usually because the polyp acts as a lead point for intussusception. We report an extremely rare case of a terminal ileum IFP presenting as acute intestinal obstruction without intussusception.

CASE PRESENTATION

A 46-year-old man from rural India presented to the emergency room after 4 days of painful abdominal distension. He also had fever and obstipation since the onset of the pain. The pain was present diffusely as a dull ache. The patient had a fracture of the neck of the left femur a year prior and surgical fixation was performed. He did not have any medical comorbidity. He did not have any history of haematochezia or malaena. He had not undergone any abdominal surgery in the past.

On examination, he was febrile and his pulse rate was slightly elevated. The abdomen was distended and diffusely tender on palpation. There was no obvious external hernia. No bowel sounds were heard. On digital rectal examination, the rectum

was empty and there was no blood tinge on the examining finger.

INVESTIGATIONS

A routine blood panel and other radiological investigations were carried out. The patient's total leucocyte count was elevated, suggesting an inflammatory process. An erect X-ray of the abdomen was taken, showing multiple dilated small bowel loops and air fluid levels (figure 1), suggestive of intestinal obstruction. A contrast-enhanced CT (CECT) of the abdomen with both intravenous and water soluble oral contrast was carried out. The CECT showed dilation of the small bowel loops and multiple air fluid levels with the transition point at the distal ileum. There was a long segmental narrowing of the distal ileum up to the ileocaecal junction, about 8.5 cm in length. The caecum was pulled up and small bowel faeces sign was present (figure 2). This was in line with our clinical diagnosis of acute intestinal obstruction. The CECT did not shed light on the possible cause for the obstruction.

The histopathological examination of the resected small bowel revealed the following features. On grossing the specimen, a 4×3.5×2.8 cm polyp was found. Microscopic examination of the polyp showed small intestinal mucosa with unremarkable glands. The submucosa and muscular layer showed a lesion composed of aggregates of small thin-walled blood vessels surrounded by

Figure 1 Erect X-ray of the abdomen showing dilated small bowel loops and air fluid levels.

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Figure 2 Contrast CT showing dilation of the small bowel loops and multiple air fluid levels with the transition point at the distal ileum. Long segment narrowing of the distal ileum is seen up to the ileocaecal junction for about 8.5 cm.

spindle cells with cytoplasmic processes. Dense mixed inflammatory cell infiltrates composed of lymphocytes and eosinophils were seen. A scant focus of fibrinoid necrosis along with serosal haemorrhage and ulceration was seen (figures 3 and 4). The two lymph nodes found along with the resected specimen appeared to be reactive. Given the above microscopic picture, the pathologist reported it as an IFP of the terminal ileum. On immunohistochemistry, CD34 was positive, which was consistent with IFP. However, CD117 was negative and the possibility of gastrointestinal stromal tumours (GIST) was ruled out (figures 5 and 6).

DIFFERENTIAL DIAGNOSIS

Adhesions as a cause for acute intestinal obstruction were unlikely as the patient had not undergone surgery in the past and had no history of tuberculosis. However, since the caecum appeared to be pulled up, ileocaecal tuberculosis had to be considered. The patient had no obvious external hernias that could have caused obstruction. Crohn's disease was not considered, as it is very rare in India. Acute appendicitis was a possible differential diagnosis but imaging ruled that out. Since the patient was middle aged, we suspected that the obstruction could have been due to a malignant cause. Small bowel malignancy was the most likely cause followed by ileocaecal tuberculosis.

Figure 3 Submucosal lesion comprised of fibrous tissue and inflammatory cells (H&E staining, ×100).

Figure 4 Fibroblastic tissue with eosinophil-rich mixed inflammatory cell infiltrate (H&E staining, ×200).

TREATMENT

The patient was posted for an emergency laparotomy after his hydration and electrolyte status was corrected. On exploration, we found an intraluminal polypoidal lesion in the terminal ileum, 13 cm proximal to the ileocaecal junction, with impeding serosal disruption (figure 7). The loops of small bowel proximal to the obstruction were dilated and the distal loops were collapsed. Resection and manual single layer extra mucosal end-to-end anastomosis of the terminal ileum was performed. A 28 Fr abdominal drain was placed and secured.

The resected specimen was examined by the authors and it showed a large, firm pedunculated polyp completely obstructing the lumen of the ileum (figure 8).

OUTCOME AND FOLLOW-UP

The patient had an uneventful postoperative recovery and was started on oral feeds once his bowel motility was established. The abdominal drain was removed subsequently and he was discharged.

He was followed up for 6 months and there were no other problems.

DISCUSSION

Obstruction of the small or large intestine occurs due to mechanical or functional causes. The probable aetiologies make for

Figure 5 Perivascular CD34 positivity (Immunohistochemical staining, ×200).

Figure 6 Negative staining for CD117 (Immunohistochemical staining, ×200).

an extensive list. Mechanical cause for obstruction of the small intestine could be classified into three subcategories: those caused by extraluminal compression, sources in the intestinal wall and intraluminal obstruction. The cause of small bowel obstruction has changed significantly during the past century.¹ In the early 20th century, hernias were responsible for over 50% of mechanical intestinal obstructions. Intra-abdominal adhesions due to previous abdominal surgery are by far the most common cause of small bowel obstruction today in developed as well as developing countries. Malignant tumours are responsible for about 20% of small bowel obstructions. The majority of these are caused by metastasis to the small bowel from other parts of the gastrointestinal tract. Primary small bowel malignancy can cause obstruction but is extremely rare. Hernias are responsible for roughly 10%, followed by Crohn's disease and intra-abdominal abscesses. In developing countries, ileocaecal tuberculosis still remains an important cause for small bowel obstruction. Miscellaneous causes are responsible for roughly 2–3%, but must be part of the differential diagnosis. They most often include intussusception, which in the adult is usually secondary to a pathological lead point such as a polyp

Figure 7 Terminal ileum impeding serosal disruption secondary to obstruction caused by large firm polyp. Dilated ileal loops proximal to the obstruction.

Figure 8 Cut section of the resected terminal ileum showing a large firm polyp almost completely occluding the lumen, allowing only a thin blade handle to pass through the lumen.

or tumour.² Nearly three quarters of all adult intussusceptions are caused by a malignant or benign lesion appearing at the head of the invagination. In the large bowel, it is more likely to be a malignant lesion. Malignant lesions in the small intestine are a rare entity. Various benign lead points of an intussusception are lipomas, leiomyomas, neurofibromas, adenomas and IFPs.³ Inflammatory IFPs in adults presenting as acute intestinal obstruction is a rarity, and only 85 cases have been reported in the literature.⁴ However, we did not find any reference to an IFP causing small bowel obstruction without intussusception. The authors felt the need to report this case as it suggests that IFPs in the small intestines may cause acute obstruction without intussusception.

IFP is defined as a gastrointestinal tract tumour characterised by spindle and stellate cells, set in an inflammatory, myxoid stroma.⁵ It is also referred to as Vanek tumour. The nomenclature of IFP remained inconsistent in the past. It was referred to as an inflammatory pseudotumour, eosinophilic granuloma and granuloma with eosinophils, among other names. Helwig and co-authors referred to it as an IFP, which is its current term.⁶

Nearly three quarters of all IFPs are gastric, and small intestinal IFPs account for only 18%.⁷ The exact aetiology of IFPs is still unknown, but trauma, allergy, genetics, as well as bacterial, physical, chemical and metabolic stimuli, have been suggested as initiators of the inflammatory process.⁴

IFPs in the small intestines are usually asymptomatic, and are typically found incidentally. Gastric IFPs may present with abdominal pain and may be diagnosed on upper gastrointestinal endoscopy. Small bowel IFPs are unlikely to produce symptoms unless they lead to obstruction either with intussusception or without.^{8–10} Vomiting, diarrhoea, haematochezia and tenesmus do occur, but are rare.¹¹ Most IFPs usually measure between 2 and 5 cm in diameter. However, giant IFPs up to 20 cm in diameter have also been reported.^{10 12} GIST may have the same pathological features as IFPs. Immunohistochemistry is useful to differentiate them. Although both are positive for CD34 and vimentin, CD117 is specific for GIST.¹³

CECT, enteroscopy and capsule endoscopy are the various diagnostic modalities available to diagnose IFPs of the small intestines. But all these diagnostic entities may miss smaller IFPs. Diagnostic laparoscopy may be useful. Resection and anastomosis of the small bowel remains the gold standard of treatment of small bowel IFPs. Stomas are reserved for a few cases that may present late after perforation, with severe peritoneal contamination that may be hostile for anastomotic healing. They are well localised tumours with no risk for local recurrence after complete excision and lack malignant potential.

Patient's perspective

I came in with severe pain in my stomach for about 4 days. I did not pay much heed to it until it increased significantly. I was referred to this tertiary care centre for further care. The doctors explained to me what the possible causes were after running a few tests and advised surgery. I readily agreed as I was in excruciating pain. After I recovered from general anaesthesia and was oriented, my doctors told me they found a benign tumour, which they removed successfully from my intestine. The drain tube did cause me some inconvenience but I put up with it as it was for my betterment. I was started on soft foods 2 days after my surgery and I was discharged after the stitches were removed. I am comfortable now and I thank my doctors for the prompt treatment that saved my life. My family is very grateful too for the safe and affordable treatment.

Learning points

- ▶ Inflammatory fibroid polyps (IFPs) are an infrequent cause for acute small bowel obstruction.
- ▶ IFPs usually cause small bowel obstruction by acting as lead points for intussusception.
- ▶ However, IFPs can also cause small intestinal obstruction without intussusception, if they are large enough to cause intraluminal obstruction.
- ▶ If the obstruction is not relieved surgically, there is a grave risk of perforation of the proximal small intestine.
- ▶ A combination of laparoscopic resection and anastomosis is the standard of care, and recurrences are rare.

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Contributors SH was the chief surgeon who operated on the patient, contributed to the research and helped write the manuscript. NN was the first assistant surgeon, and was involved in the research and writing of the manuscript. SK was the pathologist who reported the HPE findings and provided insights on the pathological aspects.

Competing interests None declared.

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REFERENCES

- 1 Hayanga AJ, Bass-Wilkins K, Bulkley GB. Current management of small-bowel obstruction. *Adv Surg* 2005;39:1–33.
- 2 Twonsend CM, Beauchamp RD, Evers MB, et al. Sabiston textbook of surgery. 18th edn, volume II. Philadelphia: Saunders Elsevier, 2008:1290–1.
- 3 Karamercan A, Kurukahvecioglu O, Yilmaz TU, et al. Adult ileal intussusception: an unusual emergency condition. *Adv Ther* 2006;23:163–8.
- 4 Akbulut S. Intussusception due to inflammatory fibroid polyp: a case report and comprehensive literature review. *World J Gastroenterol* 2012;18:5745–52.
- 5 Bosman FT, Carneiro F, Hruban RH, et al. eds. *WHO Classification of Tumors of the Digestive System*. IARC, Lyon, 2010.
- 6 Malik KA, Pande GK, Aftab Z, et al. Inflammatory fibroid polyp of the ileum causing intussusception. *Saudi Med J* 2005;26:995–8.
- 7 Johnstone JM, Morson BC. Inflammatory fibroid polyp of the gastrointestinal tract. *Histopathology* 1978;2:349–61.
- 8 Coulter B, Maldague P, Broze B, et al. Ileal inflammatory fibroid polyp causing ileocolic intussusception. *JBR-BTR* 2008;91:149–52.
- 9 Dawson PM, Shousha S, Burn JL. Inflammatory fibroid polyp of the small intestine presenting as intussusception. *Br J Clin Pract* 1990;44:495–7.
- 10 Deschamps L, Bretagnol F, Couvelard A, et al. Inflammatory fibroid polyp in Crohn's disease revealed by ileoileal intussusception: case report and review of the literature. *Inflamm Bowel Dis* 2008;14:1317–20.
- 11 El Hajj II, Sharara AI. Jejunojejunal intussusception caused by an inflammatory fibroid polyp. Case report and review of the literature. *J Med Liban* 2007;55:108–11.
- 12 de la Plaza R, Picardo AL, Cuberes R, et al. Inflammatory fibroid polyps of the large intestine. *Dig Dis Sci* 1999;44:1810–16.
- 13 Cawich SO, Gibson TN, Mitchell DI, et al. Adult intussusception from an inflammatory fibroid polyp: a case report and review of the literature. *Internet J Pathol* 2008;7:1.

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