

Case Report

Vanek's tumor presenting as ileocecal intussusception in an adolescent: a rare case report

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Received: 25 April 2025

Revised: 28 May 2025

Accepted: 07 June 2025

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ABSTRACT

A 17-year-old male presented with a 10-day history of abdominal pain and multiple episodes of vomiting. Clinical examination revealed tachycardia, pallor, and a mobile mass in the right iliac fossa. CT imaging showed ileo-ileal intussusception with anterior displacement of the cecum and wall thickening in the distal ileal loop. An emergency laparotomy revealed telescoping of the terminal ileum and cecum into the ascending colon, with free fluid in the pelvis. Histopathological examination confirmed Vanek's tumor as the lead point. This case underscores the importance of considering rare pathological entities in the differential diagnosis of abdominal symptoms in adolescents.

Keywords: Vanek's tumor, Inflammatory fibroid polyp, Small bowel intussusception, Ileal neoplasm, Intestinal obstruction

INTRODUCTION

Inflammatory fibroid polyp (IFP) is a rare, benign gastrointestinal lesion first described by Vanek in 1949 as a gastric submucosal granuloma with eosinophilic infiltration. While most commonly found in the stomach, IFPs may also arise rarely in the small bowel and, less frequently, in the colon.

Clinically, they can present with abdominal pain, bleeding, or obstruction, and may occasionally lead to intussusception. It typically occurs in middle-aged adults with a slight female predominance.¹ Occurrence in pediatric or adolescent male populations is exceedingly rare, with very few cases documented in the literature.² Here, we present a rare case of Vanek's tumor presenting as an ileocecal intussusception in a 17-year-old male, detailing the clinical presentation, diagnostic workup, surgical findings, and histopathological features, along with a brief literature review.

CASE REPORT

A 17-year-old male presented with a 10-day history of abdominal pain and multiple episodes of vomiting. On examination, he was tachycardic with fair hydration and pallor. Abdominal examination revealed a mobile mass measuring 6×4 cm in the right iliac fossa. Digital rectal examination showed an empty rectum with absent fecal staining. CT imaging of the abdomen and pelvis indicated ileo-ileal intussusception with anterior displacement of the cecum and wall thickening in the distal ileal loop, presenting a 'bowel within bowel' appearance in the right iliac fossa.

A diagnosis of ileocecal intussusception was made, and an emergency laparotomy was performed, revealing telescoping of the terminal ileum and cecum with the appendix- the intussusceptum, into the ascending colon- the intussusciens, causing significant bowel obstruction with free fluid noted in the pelvis. After manual

reduction, the intussusceptum along with the lead point was resected and an end ileostomy was fashioned.

The resected specimen was sent for histopathological examination which revealed a well-defined polypoid lesion, composed of a proliferation of spindle cell elements, small capillaries and inflammatory cells in the submucosa. The stroma was myxoid edematous, concentrically arranged around blood vessels with an “onion skin” pattern. The inflammatory cells were predominantly composed of eosinophils. IHC of spindle cells was positive for CD34, vimentin, and smooth muscle actin. All these findings were consistent with inflammatory fibroid polyp/ Vanek’s polyp. The patient subsequently, after 6 weeks, underwent reversal of the ostomy with ileocolic anastomosis and was discharged in stable condition with an uneventful postoperative course.

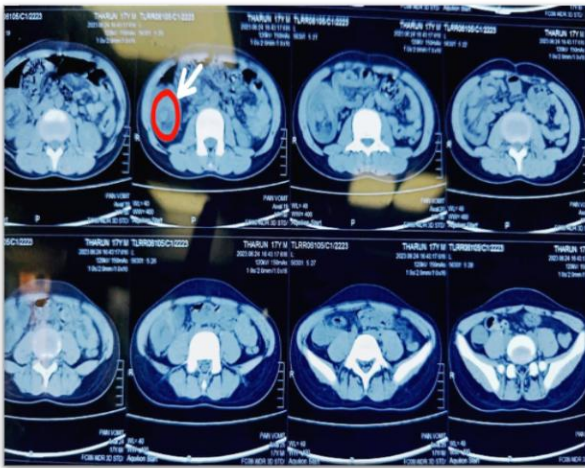


Figure 1: CT image of bowel within a bowel appearance.

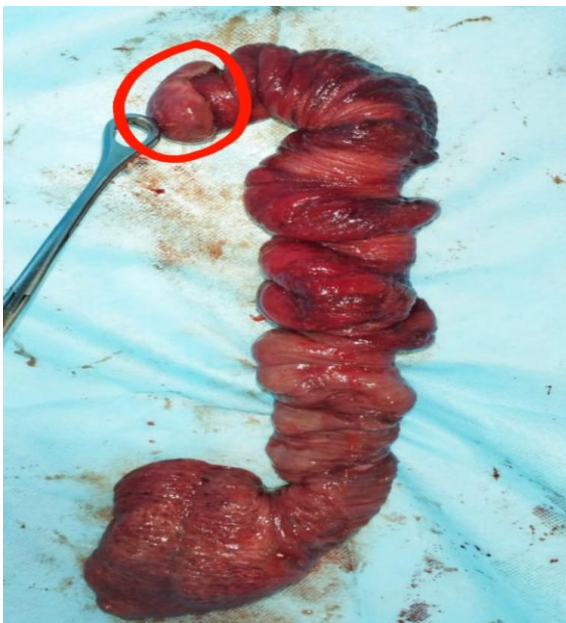


Figure 2: Resected specimen with inflammatory fibroid polyp.

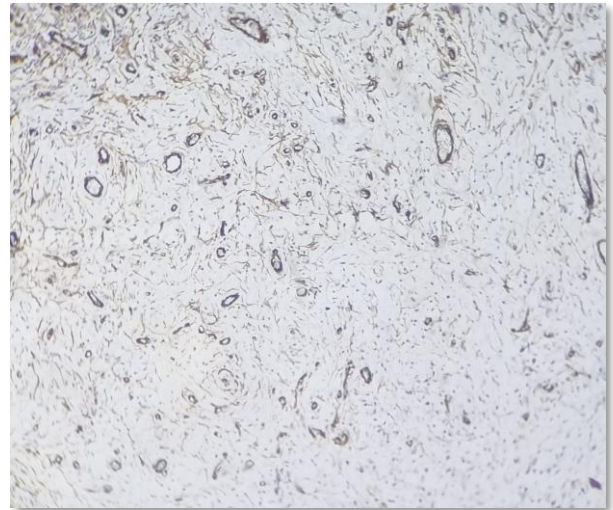


Figure 3: Staining positive for CD34.

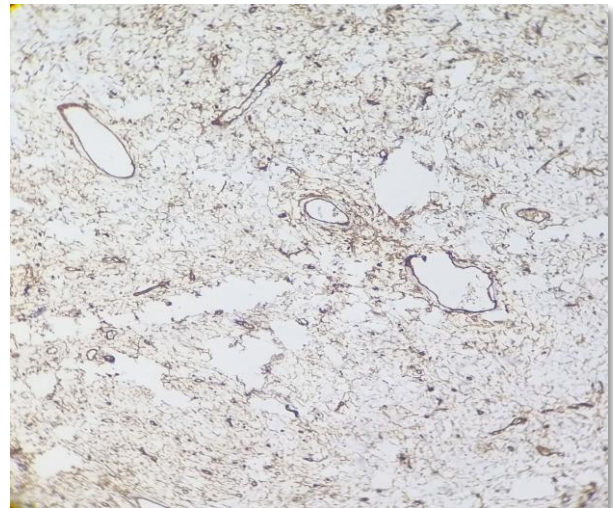


Figure 4: Staining positive for smooth muscle actin.

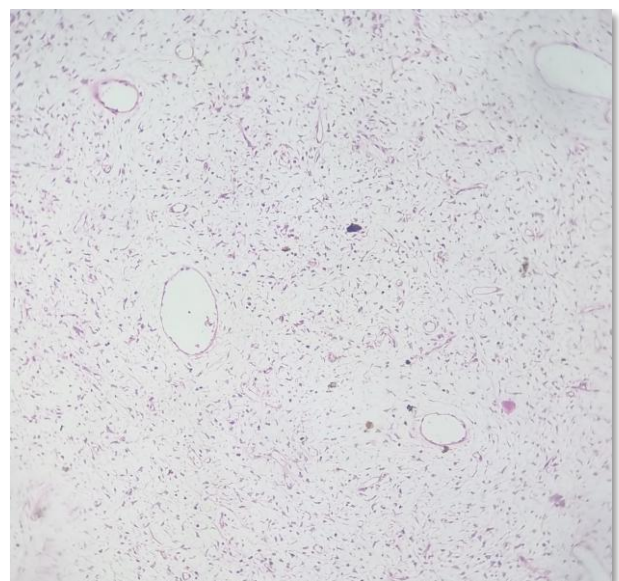


Figure 5: Amplification of granulated tissue with a proliferation of spindle cell elements.

DISCUSSION

In 1920, Konjetzny was the first to report what is now recognized as an inflammatory fibroid polyp (IFP), initially describing it as a ‘polypoid fibroma’. Vanek named it “gastric submucosal granuloma with eosinophilic infiltration” and described six cases with an incidence of 0.5%. IFP lesion that arises from the GI tract submucosa are classified as submucosal tumors of connective tissue.³ Although the exact cause remains unclear, certain cases have been linked to mutations in the PDGFRA gene. Theories include a localized eosinophilic inflammatory response, chronic irritation, or trauma.

The antrum is the most frequently affected site, accounting for approximately 70% of cases, followed by the ileum, which represents around 20%. Ileal involvement is particularly rare but clinically significant, as it can serve as a lead point for intussusception. Less commonly, inflammatory fibroid polyps are found in the colon, jejunum, duodenum, and esophagus, in that order of frequency. They rarely exceed 6 cm in size however, an exceptional case measuring 12.5 cm has been documented. These lesions are most often identified in middle-aged to elderly individuals. While they are more frequently seen in infants, their occurrence in adolescents is uncommon.⁴ IFPs are frequently detected incidentally during evaluations, as many patients remain asymptomatic. Clinically, Vanek’s tumor can present with non-specific gastrointestinal symptoms or be discovered incidentally during imaging or endoscopy. Depending on the size and location of the lesion, patients may present with dyspepsia, vomiting, in addition to gastrointestinal bleeding, and iron-deficiency anemia.

Complications like intussusception, which require immediate surgical intervention, are rare, with an incidence of 8.6%, and are usually diagnosed intraoperatively. Imaging modalities such as ultrasound, CT, and MRI aid in preoperative diagnosis.⁵ Ultrasound has the highest reported sensitivity (up to 100%), while MRI demonstrates superior accuracy in identifying obstructive causes compared to CT. While it is an uncommon cause of intussusception, especially in adolescents, it should be considered in differential diagnoses when imaging suggests a lead point.

Definitive treatment involves surgical resection, but surgical management remains debated. While some advocate for reduction before resection, especially in benign cases, others recommend primary resection without reduction to avoid the risk of spreading malignancy. While IFPs are considered non-recurrent and non-malignant, with only a single case of recurrence reported.⁶ A selective approach based on patient age, lesion location, and suspicion of malignancy is often preferred. Resection minimizes the risk of tumor spread, whereas prior reduction may help preserve bowel length and prevent short bowel syndrome. Timely management

is essential to avoid serious complications, including ischemia, tissue necrosis, and perforation.⁷ In the face of diagnostic ambiguity, histopathology stands as the gold standard, unveiling the rare culprit behind this adolescent intussusception. On histological examination, IFPs are characterized by a proliferation of fibroblastic cells accompanied by a prominent eosinophilic infiltrate. Immunohistochemistry typically reveals positivity for vimentin and CD34, suggesting a fibrovascular origin, while markers like cytokeratin, SMA, Desmin, S100, and CD117 are usually negative, ruling out smooth muscle, neural, or epithelial differentiation.⁸

Though often dismissed as benign curiosities, inflammatory fibroid polyps can unmask themselves dramatically, reminding clinicians to keep even the rarest culprits in mind when common symptoms conceal uncommon diagnoses.

CONCLUSION

This case highlights the importance of considering rare causes like Vanek’s tumor in adolescents presenting with intussusception. Prompt diagnosis and surgical management are vital for favorable outcomes. Our case, being a rare entity with a rarer presentation of the disease, was managed accordingly.

ACKNOWLEDGEMENTS

The authors would like to thank the surgical team and radiology department for their contributions to this case.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Kingston VA, Sudamani S. Vanek's tumor presenting as ileocecal intussusception in an adolescent: a rare case report. *Int Surg J* 2025;12:1185-8.