La Pediatria Medica e Chirurgica

La Pediatria Medica e Chirurgica 2026; 48:366
doi:10.4081/pmc.2026.366

Primary intestinal lymphangiectasia presenting with intussusception in a child

Kavita Gaur, Mukul Choudhary, Smita Singh, Kiran Agarwal, Abu Suraih, Subhasis Roy Choudhary

Department of Pathology, Lady Hardinge Medical College, Shaheed Bhagat Singh Marg, New Delhi, India

Abstract

Intestinal lymphangiectasia is a protein losing intestinal disease caused by congenital malformation or obstruction of intestinal lymphatics. Based on etiology, it is classified as primary or secondary. Primary Intestinal Lymphangiectasia (PIL) is a rare disorder of intestinal lymphatic dysfunction and may present with a wide spectrum of clinical manifestations but it very infrequently presents as intussusception in children. To the best of our knowledge this is the second only reported case in children, both cases being described from the Indian subcontinent. A 12-year-old male presented complaining of abdominal pain for one week. Examination revealed a palpable mass in the right upper quadrant and abdominal tenderness. A provisional diagnosis of acute intestinal obstruction was made and the child underwent an emergency laparotomy which revealed a segment of distal ileum displaying intussusception. On cut opening, no growth was seen. Histopathology of the telescoped segment showed many ectatic and dilated lymphatic channels filled with lymph. PIL should be considered in the differential diagnosis in children presenting with intestinal obstruction.

Key words: intestine, lymphatic vessels, pediatric.

Correspondence to: Mukul Choudhary, Department of Pathology, Lady Hardinge Medical college, Shaheed Bhagat Singh Marg, 110001 New Delhi, India. Tel.: +91.9250296962. E-mail: mukulchoudhary2209@gmail.com

Introduction

Lymphangiectasia represents a rare entity characterized by the dilatation of lymphatic channels, commonly afflicting areas such as the axilla, neck and the head. Other reported sites include the lungs and intestines including the mesentery.[1] Involvement of intraabdominal organs is uncommon and accounts for only about 1.6% of cases.[2] Lymphangiectasia involving the intestine may be focal or diffuse. Diffuse dilatation of intestinal lacteals has been documented to cause the loss of lymphocytes and proteins into the gastrointestinal tract or peritoneal cavity leading to a varied range of signs and symptoms, sometimes causing severe lower limb edema.[3] Lymphangiectasia may further be typified as primary or secondary depending on the absence or presence of an organic cause of the lymphatic vascular dilatation.[4]

We present herein a case of primary ileal lymphangiectasia occurring in a 12-year-old boy diagnosed with intussusception. Histopathological analysis further revealed ectatic lymphatic channels consistent with lymphangiectasia. This report, in addition to documenting an exceptionally rare presentation, reviews existing relevant literature.

Case Report

A 12-year-old boy presented to the Out-Patient Department (OPD) complaining of abdominal pain, diarrhea and vomiting for one week, fever for three days and two days constipation. The child did not have episodes of chronic diarrhea, edema, or failure to thrive prior to presentation. On examination, a palpable mass in the right upper quadrant along with abdominal tenderness was noted. On auscultation, bowel sounds were absent. A provisional clinical diagnosis of acute intestinal obstruction was made.

Biochemical investigations revealed hypoproteinemia (4.9 g/dL), hypoalbuminemia (3.0 g/dL), mildly elevated serum bilirubin (2.64 mg/dL), and markedly elevated C-reactive protein (148.8 mg/L).

Abdominal X-ray showed multiple air fluid levels with ultrasound findings being equivocal due to excessive bowel gas and overlapping loops, which limited accurate delineation of a lead point. Although contrast-enhanced computed tomography (CT) was considered, an emergency laparotomy was undertaken in view of the child's acute intestinal obstruction and clinical condition. This was also necessary for evaluating the cause of intestinal obstruction and intra-operatively an ileocecal intussusception was identified. As both the intussusceptum and intussuscepiens appeared pale and bulky despite attempts at reduction, a segment of distal ileal intussusceptum with part of caecum was sent for histopathological examination to rule out the possibility of a malignant etiology. The remainder of the small and large intestine appeared grossly normal. A single mesenteric lymph node was also sent.

On gross examination, the mucosa appeared markedly pale and edematous. The telescoped segment was noted 15 cm from the ileocecal junction. Microscopic examination from the telescoped segment showed a preserved mucosal lining with both the lamina propria and submucosa showing many ectatic and dilated lymphatic channels filled with lymph (Figure 1). A few congested capillaries along with a sprinkling of chronic inflammatory cells were seen in

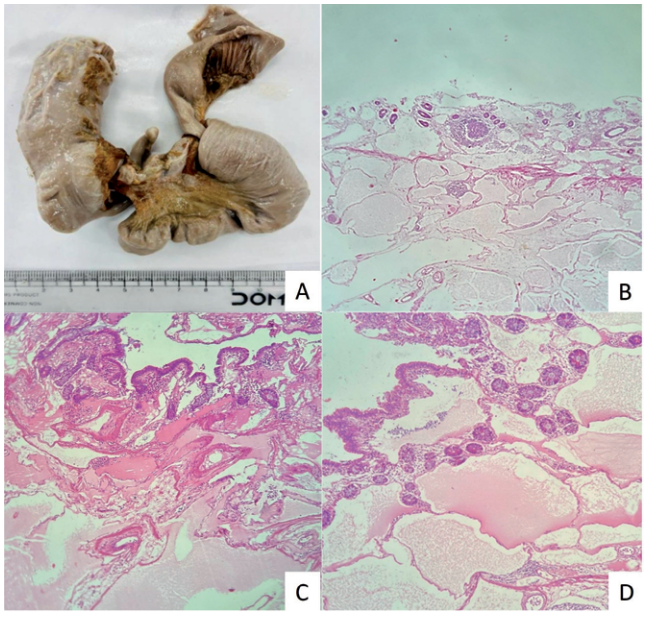

Figure 1. A) Gross findings revealing intussuscepted ileal segment displaying classic telescoping, B-D) Ileal mucosa along with dilated lymphatic channels in the lamina propria of resected small intestine reaching up to the muscularis. Hematoxylin & Eosin, (B) 40X, (C) 100X (D) 400X.

the submucosa. The muscularis propria was mildly thickened and the serosa showed focal congestion. Resection ends were viable and congested. No evidence of dysplasia or any other significant pathological finding was seen after a thorough examination of the resected tissue. Sections from the mesenteric lymph node showed reactive changes. A diagnosis of ileal lymphangiectasia leading to intussusception was made. The karyotype of the child was also performed after the histopathological diagnosis was made, which showed a normal 46, XY complement. The postoperative period was uneventful and at one year of follow-up, the child demonstrated clinical improvement with no recurrence of symptoms, normalization of serum albumin levels, and no further evidence of protein-losing enteropathy.

Discussion

Intestinal obstruction due to acute intussusception is most frequently observed in infants, aged 3 to 9 months, with the majority being of unknown origin.[5,6] However, various small bowel abnormalities can serve as a lead point for intussusception. Rare occurrences of lymphoblastic lymphoma of the ileum and small bowel hemangiomas have been reported to present in this manner.[7] The precise cause of lymphangiectasia remains uncertain, although it has often been linked to congenital issues affecting visceral lymphatic channels, resulting in obstruction of lymphatic flow and subsequent dilatation of intestinal lymphatics.[4] In our case, lymphangiectasia in the small intestine acted as the trigger for intussusception, leading to further intestinal obstruction.

PIL rarely presents as intestinal obstruction in children. PIL is a histopathological diagnosis and in the present case, the diagnosis was established following surgical resection, based on characteristic histological findings. Classically, it manifests with features of protein-losing enteropathy, including chronic diarrhea, peripheral edema, failure to thrive, hypoalbuminemia, and lymphopenia.[8] Definitive diagnosis requires histopathological examination, which characteristically demonstrates widened villi with dilated lacteals in the mucosa and enlarged submucosal lymphatic channels, in the absence of significant inflammation or mucosal atrophy. Although most cases occur sporadically, some may be associated with genetic disorders like Turner and Noonan syndromes.[9] Turner syndrome was excluded based on karyotype analysis, while Noonan syndrome was considered unlikely based on clinical evaluation.

Increased expression of Vascular Endothelial Growth Factor Receptor-3 (VEGFR-3) and Lymphatic Vessel Endothelial Hyaluronan Receptor-1 (LYVE-1) has been observed on the mucosal surface in patients with intestinal lymphangiectasia, corresponding to widely dilated lymphatic vessels in affected regions.[10] While only a

Table 1. Pediatric cases of lymphangiectasia presenting as intussusception and intestinal obstruction.

Case	Age	Clinical presentation	Histopathological diagnosis	Gross preoperative finding	Primary/ Secondary
Katoch *et al.* (2008)[12]	6 months male	Abdominal pain, palpable mass, intussusception	Numerous dilated and anastomosing lymphatic channel in the mucosa, containing mature lymphocytes. Lamina propria contain engorged blood vessels and infiltration by chronic inflammatory cells	Ileocecal mass with attached appendix measuring 7x3x1 cm. appendix was grossly normal	Primary intestinal lymphangiectasia
Kumar *et al.* (2019)[13]	17-year female (diagnosed case of Diffuse Large B Cell Lymphoma DLBCL)	Developed intestinal obstruction after induction chemotherapy along with vomiting and colicky abdominal pain	Biopsy showed ectatic lymphatic channels with widening of submucosal region with unremarkable adjacent mucosa and muscular is propria	Mass measuring 2.5x2.5x0.5 cm located 110 cm proximal to the ileocecal junction	Secondary intestinal lymphangiectasia
Our case report	12-year male	Abdominal pain, diarrhoea, vomiting	Biopsy showed preserved mucosal lining with the submucosa showing many ectatic and dilated lymphatic channels filled with lymph	Ileocecal intussusception present	Primary intestinal lymphangiectasia

few cases of intussusception due to lymphangioma or lymphangiectasia have been reported, it is essential to exclude secondary causes of intestinal lymphangiectasia, including lymphoma, other malignancies, and infections such as tuberculosis, before establishing a diagnosis of primary intestinal lymphangiectasia.[11,12] In the present case, secondary causes of intestinal lymphangiectasia were excluded postoperatively based on detailed histopathological examination of the resected bowel, which showed no evidence of malignancy, lymphoma, granulomatous inflammation, or other secondary pathology.

In addition, other causes of intussusception were evaluated intraoperatively by careful inspection and palpation of the bowel to identify a pathological lead point, such as a polyp, Meckel's diverticulum, mass lesion, or inflamed segment. These were further excluded based on operative findings, histopathology, and relevant clinical evaluation, including the absence of prior abdominal surgery, drug-related etiologies, viral illness, or congenital anomalies.

Postoperatively, our patient is being followed up with clinical assessment for recurrence of symptoms /signs along with six monthly biochemical evaluation, including serum albumin and total protein levels which have been normal at each instance. Radiological imaging at our centre is reserved for cases with clinical suspicion of recurrence or complications and since the patient is currently doing well, it has not been undertaken.

A thorough review of the current literature (Table 1) has revealed only two prior cases of lymphangiectasia-associated intussusception presenting as intestinal obstruction. In one of this two cases, lymphangiectasia was found to be secondary to intestinal Diffuse large B-cell lymphoma (DLBCL). on chemotherapy treatment. This report thus highlights the extreme rarity of this condition, warranting a broader index of suspicion while evaluating pediatric cases of intestinal obstruction. Interestingly, all cases reported till date have been described from the Indian subcontinent, which may be a subject for further study.

Conclusions

Primary intestinal lymphangiectasia is an exceptionally rare pathological lead point for intussusception in children. Although it does not typically present with features of intestinal obstruction, PIL should be considered in older children presenting with intussusception when common lead points are not identified, particularly in the presence of biochemical evidence of protein-losing enteropathy such as hypoalbuminemia and hypoproteinemia. While clinical and laboratory findings may raise suspicion, definitive diagnosis relies on histopathological examination, which is usually established following surgical intervention. Awareness of this rare entity may aid in appropriate diagnostic consideration and postoperative management.

References

1. Ashley DJ. Tumours of vasoformative tissue. In: Evans' Histopathological Appearance of Tumours. 4th ed. London: Churchill Livingstone; 1990. pp 77-108.
2. Chaturvedi KU, Singh N, Sakhuja P. Intra-abdominal lymphangioma: Clinicopathological profile of rare lesions. Indian J Pathol Microbiol 1997;40:563-66.
3. Abramowsky C, Hupertz V, Kilbridge P et al. Small intestinal lymphangiectasia in children: A study of upper GI endoscopic biopsies. Paediatr Pathol 1989;9:289-91.
4. Isa HM, Al-Arayedh GG, Mohamed AM. Intestinal lymphangiectasia in children: a favorable response to dietary modifications. Saudi Med J 2016;37:199-204.
5. Winslet MC. Intestinal obstruction. In: Russell RC, Williams NS, Christopher JK, editors. Bailey and Love's Short Practice of Surgery. 24th ed. London: Edward Arnold Ltd; 2004. pp 1186-02.
6. Coran AG. Intussusception in adults. Am J Surg 1969;117:735.
7. Shah P, Ramakantan R. Ileocaeco-colic intussusception produced by lymphoblastic lymphoma. Indian J Gastroenterol 1989;8:302-3.
8. Freeman HJ, Nimmo M. Intestinal lymphangiectasia in adults. World J Gastrointest Oncol 2011;3:19-23.
9. Wan YL, Lee TY, Hung CF et al. Ultrasound and CT findings of a caecal lymphangioma presenting as intussusception. Eur J Radiol 1998;27:77-9.
10. Miura S, Sakurai H, Nishikawa Y et al. Expression of lymphangiogenic regulatory factors in intestinal lymphangiectasia. Ann Vasc Dis 2012;5:275-81.
11. Matsuba Y, Mizuiri H, Murata T, et al. Adult intussusception due to lymphangioma of the colon. J Gastroenterol. 2003;38:181-5.
12. Katoch P, Bhardwaj S. Lymphangiectasia of small intestine presenting as intussusception. Indian J Pathol Microbiol 2008;51: 411-12.
13. Kumar S, Raja K, Gnanasekaran S et al. Intestinal lymphangiectasia: a rare cause of intussusception in an adolescent. Ann R Coll Surg Engl 2019;101:e43-e44.

Received: 19 July 2025; Accepted: 29 January 2026.

Funding: none.

Contributions: Kavita Gaur, Mukul Choudhary, Abu Suraih, considerable contribution to study design, data gathering and analysis and interpretation of data, preparation of article draft and revising it, approval of final version of manuscript to be published. Kiran Agarwal, Smita Singh, Shankar Roy Choudhary, considerable contribution to study design, analysis and interpretation of data, revision of article draft, approval of final version of manuscript to be published.

Conflicts of interest: the authors declare no conflict of interest.

Ethics approval and consent to participate: no ethical committee approval was required for this case report by the Department, because this article does not contain any studies with human participants or animals. Informed consent was obtained from the patient included in this study.

Patient consent for publication: the patient's guardians gave their written consent to use the patient's personal data for the publication of this case report and any accompanying images.

Availability of data and materials: all data underlying the findings are fully available.