

A Rare Case of Isolated Hindgut Malrotation with Internal Hernia and Coexisting Enteric Duplication Cyst

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Abstract

An 8-year-old boy presented with a history of intermittent episodes of spastic abdominal pain accompanied by bilious vomiting. Imaging revealed an abnormal position of the large bowel loop, which was associated with an abnormal vascular course. Additionally, a mesenteric cystic lesion was found in relation to the small bowel. A possibility of isolated hindgut malrotation with enteric duplication cyst was suggested based on these findings. The surgical team confirmed this diagnosis during the procedure. Furthermore, they discovered an internal hernia intraoperatively, with a defect in the colonic mesentery through which the terminal ileum, IC junction, cecum, and appendix herniated.

Keywords: Intestinal Malrotation; Duplication cyst; Internal Hernia.

Introduction

Intestinal malrotations are one of the most interesting yet challenging cases for clinicians. They can be broadly defined as any deviation in the position of the intestines from their normal anatomical location.¹ This can include the small bowel, the large bowel, or both, and can be associated with abnormal fixation or non-fixation of the bowel, which predisposes to obstruction.¹

Classically, small bowel malrotations have been extensively studied and classified, while isolated hindgut malrotations are rare. Even rarer are hindgut malrotations associated with an internal hernia and enteric duplication cyst, which is the case we present here.

Case Report

An 8-year-old boy was admitted with severe acute abdominal pain that was spastic and intermittent in nature, associated with multiple episodes of bilious vomiting. The child was malnourished, but his physical and blood investigations were unremarkable. He gave a history of similar intermittent episodes in the past, which had been managed conservatively in multiple other hospitals.

Ultrasonography of the abdomen revealed that the small bowel loops were predominantly on the left side of the abdomen, and large bowel loops were on the right side. Additionally, on the left side of the abdomen, a cyst with internal echoes having a gut signature was seen in relation to the bowel, indicating an intestinal duplication cyst.

The contrast-enhanced CT abdomen findings were as follows: The D3 segment of the duodenum was seen to cross to the left side with the duodeno-jejunal junction in its expected location. The SMA-SMV relationship was maintained with the SMV lying on the right side of SMA. The small bowel loops were predominantly on the left side of the abdomen and the entire colonic loops were positioned on the right side of the abdomen [Figure 1]. The

ileocecal junction, cecum, and appendix were noted in the right lumbar region [Figure 2]. The IMA was seen coursing to the right side of the midline, instead of its normal left-sided course [Figure 3]. A tubular cystic lesion was observed in relation to the small bowel loops on the left side of the abdomen [Figure 4]. Intrabdominal solid organs were essentially normal, with normal situs.



Figure 1: Coronal sections of the contrast-enhanced computed tomography (CECT) scan of the abdomen, showing the abnormal positioning of the small bowel loops on the left side (green arrow) and the large bowel loops on the right side (orange arrow), suggesting the possibility of hindgut malrotation.



Figure 2: Coronal sections of CECT abdomen showing the position of ileocecal junction (orange arrow), cecum and appendix (green arrow) in the right lumbar region.



Figure 3: 3D volume-rendered CECT arterial phase images showing the abnormal right sided course of Inferior mesenteric artery (green arrow).

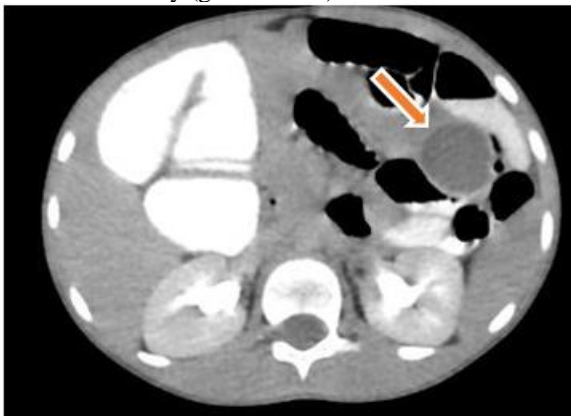


Figure 4: Axial sections of CECT abdomen showing a well-defined fluid filled cyst (orange arrow) on the left side of abdomen in relation to the small bowel loops.

A diagnosis of isolated hindgut malrotation associated with an enteric duplication cyst was suggested.

Surgical findings confirmed the diagnosis, with the colonic loops lying on the right side of the abdomen and small bowel loops on the left side. A tubular duplication cyst was observed along the distal jejunum and proximal ileum. An additional and interesting finding was that the ascending colon had mesentery and there was a defect in the ascending colon mesentery, through which the terminal ileum, IC junction, cecum, and appendix were herniating [Figure 5]. The ascending colon was also noted to be twisted along its vascular axis. The twisted portion of the ascending colon was resected and the remaining ends were re-anastomosed. The position of the IC junction and terminal ileum were corrected. A part of the small bowel loop, specifically the distal jejunal-proximal ileal loops containing the duplication cyst, was excised [Figure 6].



Figure 5: Intraoperative image showing the herniation of IC junction, cecum and appendix through the mesenteric defect of ascending colon.



Figure 6: Resected surgical specimen of small bowel and the associated intestinal duplication cyst (along the mesenteric border) Arrows shows the duplication cyst.

Histopathology examination confirmed the diagnosis of an enteric duplication cyst, which also contained ectopic gastric mucosa.

Following the procedure, the child showed complete symptomatic relief. He is now on a normal diet and is gaining weight.

Discussion

Malrotation limited to the colon is much rarer than that of the small bowel alone or that of both the small and large bowel together.² It is suspected when an abnormal position of the colon is seen in conjunction with a normal position of the small bowel. The presence of associated complications with an abnormal course of the inferior mesenteric artery helps to confirm the diagnosis.^{3,4} Colonic volvulus is its most common complication of such malrotations.^{5,6}

Internal hernias are defined as the herniation of viscera through an orifice in the peritoneum or mesentery.⁷ Although the most common clinical manifestation is small bowel obstruction, they can remain clinically silent or cause intermittent obstruction in cases where they are self-reducing.^{7,8} Radiological features such as an abnormal location or crowding of small bowel loops, evidence of intestinal obstruction, features of apparent fixation, and mesenteric vessel abnormalities guide us to the diagnosis of an internal hernia.⁷ Preoperatively, a radiological diagnosis of an internal hernia could not be made in our case. It is presumed that the herniations were transient and self-resolving, and one such hernia was not present during the scan. The clinical history intermittent self-resolving abdominal pain of the child also supports this postulation.

Enteric duplication cysts are rare congenital malformations of the intestinal tract closely attached to the gut wall, lined with intestinal epithelium.⁹ Complications include recurrent abdominal pain, intussusception, obstruction, and bleeding.¹⁰ Ultrasound typically shows the presence of a cyst in relation to the gut with a double-wall gut signature.⁹ CT and MRI can depict the location and extension of the cyst, complications, associated anomalies, and anatomical relationships.¹⁰

Conclusion

Isolated hindgut malrotations are rare entities. Whenever an abnormal position of a bowel is encountered, its vascular course and other related pathologies predisposing to bowel obstruction should be thoroughly checked. A mesenteric fluid-filled structure in relation to the bowel should raise the possibility of an enteric duplication cyst

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