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Case Report

Congenital Malrotation of the Midgut Presenting as Acute Duodenal Obstruction in a Teenager — A Case Report

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Abstract

Background: Intestinal malrotation, a congenital anomaly, typically presents in neonates with midgut volvulus. However, recent studies suggest it may occur in adolescents and adults at a higher rate than previously thought. This case report investigates the presentation and management of malrotation with volvulus in an adolescent.

Case Presentation: A 14-year-old boy presented with acute onset abdominal pain, vomiting, and a history of intermittent abdominal pain for several years. Initial diagnosis was gastritis, but imaging revealed malrotation with volvulus. He underwent a successful Ladd's procedure.

Conclusion: This case emphasizes that malrotation can present with chronic or acute symptoms in adolescents. A high index of suspicion for malrotation is crucial, particularly in adolescents with recurrent abdominal pain. Early diagnosis and Ladd's procedure can prevent life-threatening complications.

Keywords: *Congenital Malrotation; Midgut; Teenager; Ladd's Procedure*

Introduction

Intestinal malrotation affects 1 in every 500 neonates, and 65–85% of malrotation cases are present within the neonatal period. [1,2] Malrotation is a congenital anomaly, in which the midgut fails to rotate either completely or partly during the early period of embryological development. This leads to the presence of duodenojejunal flexure (DJ) along with the small bowel toward the right of the spine. A majority of these cases, approximately 88-90% present within the first year of life.[3].

Malrotation in adolescents and teenagers is relatively rare. Most of these cases are asymptomatic and diagnosed incidentally on radiological imaging for unrelated issues or during surgery for other conditions. Rarely, some of these patients may present acutely or with chronic symptoms. Acute symptoms include nausea, vomiting, abdominal pain, abdominal distension, constipation, or obstipation. Whereas, chronic symptoms include intermittent dull aching or colicky abdominal pain and altered bowel habits (diarrhea/ constipation) with general malaise and failure to thrive.

In 1932, Ladd was the one to discover the peritoneal bands responsible for midgut malrotation, the eponymous Ladd’s bands.[4]

Here we present the case of acute small bowel obstruction caused by midgut malrotation in a 14 year old boy.

Case Presentation

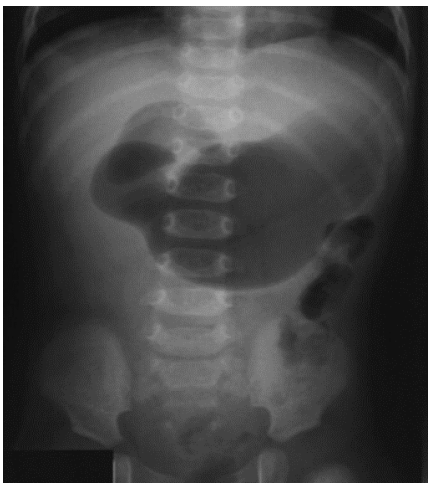
A 14 year-old boy was presented to the emergency department of our hospital, with acute onset upper and central abdominal pain and multiple episodes of bilious vomiting for 3 days. For 5-6 years, he had been having on and off episodes of intermittent dull aching upper abdominal pain, which had been treated with over the counter antacids and painkillers. He also had a recent history of having eaten out prior to the onset of the above symptoms.

The child did not have other symptoms like fever, abdominal distension or altered bowel habits. He had not undergone any previous surgeries. On examination: he was vitally stable, with only tachycardia as the predominant finding. Abdominal examination revealed upper abdominal fullness, mild epigastric tenderness without guarding, and an empty rectum on DRE. The bowl sounds were high pitched.

The child was admitted for further evaluation with a provisional diagnosis of acute intestinal obstruction. He was kept nil by mouth (NBM), given bowel rest by continuous nasogastric tube aspiration, and started on intravenous fluids, proton pump inhibitors and anti-emetics. His radiological investigations were ordered, and blood was drawn for routine workup. His symptoms started progressively worsening despite the conservative medical management. Hence, he was investigated further. Routine blood investigations were unremarkable.

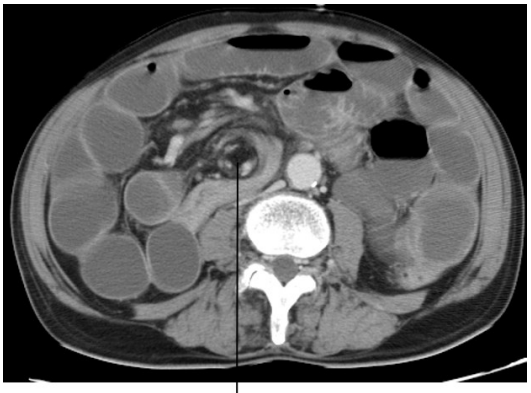
Investigations

1. X ray abdomen: Gross distension of the stomach and proximal duodenum.

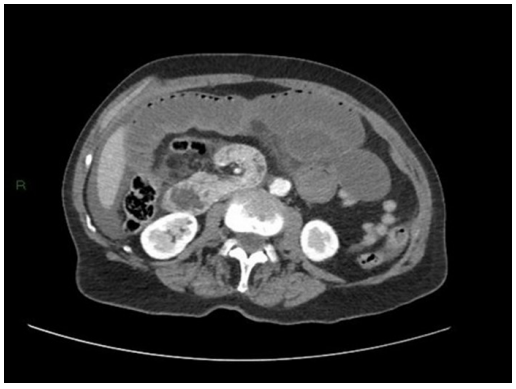


2. X ray abdomen: Contrast enhanced CECT of the abdomen revealed :

- Caecum to the left of the spine.
- Swirling of the root of the mesentery and superior mesenteric vein (SMV) seen around the super mesenteric artery (SMA) in a counterclockwise fashion, suggestive of “whirlpool sign” classical of midgut malrotation with volvulus.
- stomach with a paucity of bowel gas?? Small bowel volvulus.



Whirlpool sign of midgut volvulus. (SMA/SMV reversal)



Axial + Portal venous phase showing midgut volvulus.

In view of the above radiological findings the child was posted for emergency laparotomy and Ladd's procedure. (Laparoscopy was offered but refused by parents in view of financial constraints). At laparotomy a note was made of ladd's bands traversing from caecum laterally and compressing the duodenum. These were divided. The bowel was eviscerated and a counter-clockwise detorsion was made. The mesentery was broadened. Appendectomy was not done. The small bowel was placed in the right para colic gutter with the colon to the left. The child recovered well and was started on oral feeds on the third post operative day and discharged on the fifth day. At follow-up after a week he was doing well, able to eat full meals without pain or vomiting and had no other issues.

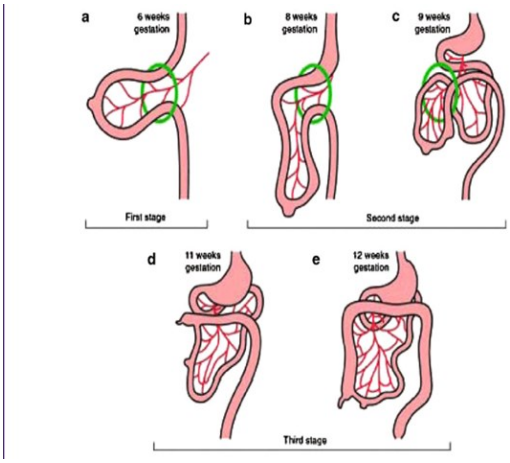
Discussion

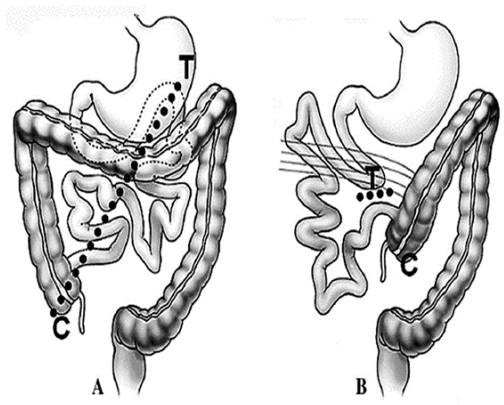
Traditionally, most cases of malrotation with midgut volvulus occur in infants within the first year of life. However, recent studies suggest that malrotation can be present in older adolescents and adults at a higher rate than previously thought [5].

Patients with malrotation often have other congenital anomalies, such as diaphragmatic hernia, congenital heart defects, or omphalocele [6, 7]. These conditions should be considered during evaluation, especially in asymptomatic patients with other malformations.

During normal embryonic development, between the 4th and 8th week, the intestines protrude outside the abdominal cavity through the umbilicus while undergoing a 90-degree counterclockwise rotation. As they return to the abdomen between the 8th and 10th week, they undergo an additional 180-degree rotation, resulting in a total of 270-degree counterclockwise rotation and a broad fan-shaped mesentery [8]. This positions the duodenum in its characteristic C-shape, with the jejunum and ileum located centrally within the abdomen, traveling from the upper left quadrant to the ileocecal junction in the lower right quadrant.

Phases of Intestinal rotation in the embryological life





A. Normal position of small and large bowel.

B. Malrotation: Small bowel is on the right and caecum is on the left.

Malrotation occurs when the complete rotation of the intestines is disrupted. Incomplete rotation of the duodenojejunal limb and partial rotation of the cecocolic limb can result in a stalk of bowel and mesentery susceptible to twisting (volvulus) [8, 9]. Additionally, fibrous bands called Ladd's bands can form, attaching the cecum to the abdominal wall and potentially compressing the duodenum, causing obstruction [6].

Non-rotation is a variant where the small bowel remains on the right side and the cecum lies on the left or midline of the lower abdomen. In this case, Ladd's bands typically don't form, and the risk of volvulus or duodenal compression is lower [10].

When the intestines twist around the malrotation stalk (midgut volvulus), it creates a surgical emergency. This twisting cuts off blood supply to a large portion of the small intestine, leading to ischemia and necrosis (tissue death) if not promptly corrected. Symptoms of volvulus include bilious vomiting, severe abdominal pain, and significant morbidity and mortality if diagnosis and surgery are delayed [9]. In older children, presentation can be atypical, with less severe symptoms like non-bilious vomiting and vague abdominal pain [11]. Ladd's bands compressing the duodenum can also cause small bowel obstruction.

Adolescent patients with malrotation may present with acute or chronic symptoms or be incidentally discovered during imaging for other reasons. Acute presentations suggestive of midgut volvulus include sudden-onset abdominal pain, bilious vomiting, or constipation. Chronic presentations can include dull aching or cramping abdominal pain, altered bowel habits, and malabsorption, which are less specific symptoms [12]. For adults, a CT scan of the abdomen is the gold standard for diagnosis, while an upper GI series is typically used in the pediatric population. CT can reveal abnormalities like inversion of the superior mesenteric artery and vein (SMA and SMV), bowel position and viability, and signs of volvulus (whirlpool sign). Ultrasonography and MRI are less commonly used diagnostic tools [13].

Ladd's procedure is the surgical treatment for malrotation. The core principles remain the same regardless of open or laparoscopic techniques. The surgery involves six key steps:1) Entering the abdominal cavity & exposing the abdominal contents (evisceration) 2) Detwisting the bowel (counterclockwise in acute cases) 3) Dividing Ladd's bands 4) Broadening the small intestine mesentery 5) (Optional) Appendectomy (less common today) 6) Repositioning the small intestine along the right flank and the colon along the left flank of the abdomen.

Conclusion

A noteworthy aspect of this case is the patient's history of vague abdominal pain previously misdiagnosed as chronic gastritis. This case highlights the importance of considering intestinal malrotation and volvulus in adolescents presenting with chronic abdominal symptoms. Early and prompt diagnosis can prevent life-threatening complications like volvulus with bowel ischemia and gangrene. Malrotation can be effectively treated with open or laparoscopic Ladd's procedure, which remains the gold standard surgical approach.

Acknowledgements

None.

Conflict of Interest

No potential conflicts of interest to disclose.

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