Case Report

DOI: https://dx.doi.org/10.18203/2349-2902.isj20243553

Midgut volvulus secondary to congenital malrotation in an adult with Ladd's band: a case report

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Received: 24 September 2024 **Accepted:** 18 October 2024

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ABSTRACT

Adult midgut volvulus is a rare surgical disease, with an estimated incidence of less than 0.15%, and more common in Asia and Africa as compared to Europe and the US. It is a developmental anomaly characterized by failure to complete normal fetal rotation of the midgut around the Superior Mesenteric Artery (SMA) inside the peritoneal cavity. Though rare, adult midgut volvulus has the potential for bowel ischemia-related complications, and thus clinicians should have a reasonable index of suspicion to diagnose this. Early computed tomography (CT) scan with diagnostic accuracy of more than 83% would help in early recognition and management of these rare causes of intestinal obstruction. In this study, we reported the case of an adult primary midgut volvulus in a 30-year-old male who presented with features of acute bowel obstruction with a history of similar recurrent episodes and renal transplant. Diagnosis of midgut volvulus was made based on contrast-enhanced CT findings, and Ladd's procedure was performed.

Keywords: Case report, Adult, Midgut volvulus, Ladd's band, Malrotation

INTRODUCTION

Midgut volvulus secondary to intestinal malrotation is a partial or complete failure of 270-degree anticlockwise rotation of the midgut around superior mesenteric vessels at the 10th week of fetal life. It is a rare condition, with an estimated incidence of 0.2% of live births typically presenting within the 1st week of life. Midgut volvulus in an adult is even rarer, with an estimated incidence of less than 0.15%. Most adult cases are silent throughout life and are seldom unintentionally detected. Sometimes they present with acute or chronic abdominal pain and are diagnosed by a radiologist during a CT scan or by the surgeon in the operation theater. We report a rare case of a 30-year-old male presented with features of intestinal obstruction secondary to midgut volvulus. This case

report has been reported according to the revised SCARE guidelines, 2018.²

CASE REPORT

A 30-year-old male presented to the emergency department with the sudden onset of colicky epigastric pain associated with multiple episodes of bilious vomiting for 2 days with multiple similar episodes since childhood. He underwent renal transplant surgery 8 years ago because of End Stage Renal Disease (ESRD). Physical examinations revealed a scar of a modified Gibson incision over the right iliac fossa (RIF) with tenderness over the epigastric region. Further systemic examinations and blood parameters were unremarkable. His chest and plain abdominal X-ray films were normal.

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The upper gastrointestinal contrast study showed most of the small and large bowel loops to be on the left side with relatively empty central and right lower region (Figure 1). CECT was done, which showed twisting of the root of the mesentery with rotation of small bowel loops and swirling appearance of the superior mesenteric vein (SMV) around SMA (whirlpool sign) (Figure 2). DJ flexure was noted towards the right side of the midline and a transplanted kidney was noted in RIF (Figure 3). Diagnosis of midgut volvulus made.

A diagnostic laparoscopy was planned. At laparoscopy, the transplanted kidney was in RIF (Figure 4A) with rotated small bowel loops. Laparoscopy converted to midline laparotomy due to poor vision and technical difficulties. Ladd's band was present from the duodenum to the ascending colon (Figures 4B and C) with 180-degree anticlockwise rotation of the small bowel around the Ladd's band and appendix at the right upper quadrant (RUQ). Ladd's band was released, derotation of the small bowel and restoration of normal mesenteric orientation was done which was followed by appendectomy. His postoperative course was uneventful with expected recovery and discharged on the fifth postoperative day. All his follow-up visits in the outpatient department were in the expected course.

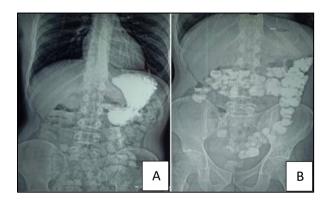


Figure 1 (A and B): Upper gastrointestinal contrast study shows the normal position of the stomach with most of the small and large bowel loops appearing to be on the right upper and left side of the abdomen, with relatively empty central and right lower region.

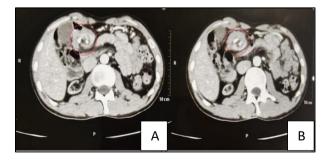


Figure 2 (A and B): CECT finding: axial view showing distinctive whirlpool-like pattern due to encircling bowel loops around the SMA (dotted circle).



Figure 3: CECT finding: coronal view showing transplanted kidney (red arrow) in RIF.

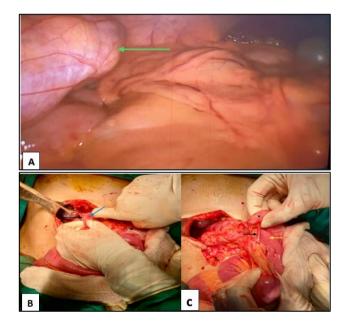


Figure 4: Intraoperative pictures: (A) laparoscopic view showing transplanted kidney in RIF (green arrow). (B) Ladd's band (black arrow) extending from ascending colon (asterisk) to duodenum (blue arrow) (C) Ladd's band attachment between ascending colon (asterisk) and jejunal mesentery entrapping jejunum (yellow arrow) in between.

DISCUSSION

Midgut volvulus is a rare condition, and rarer in adults. Incidence and clinical presentation of midgut volvulus in adult patients differ by geography. In the US and Europe, the incidence of midgut volvulus has been estimated to be around 1.7 to 5.7 per 100,000 population annually, and 3-6% of patients present with intestinal obstruction.³ Whereas in Asia and Africa, the annual incidence of midgut volvulus is much higher, about 24–60 per 100,000 population, among which 20–50% of patients seek medical care due to acute or chronic intestinal obstruction.⁴The umbilical sac herniates into the midgut

during embryonic development. At 4th week, the embryo's gut is in the form of a straight tube. A bowel loop develops with the SMA at its axis during the 5th week. Intestinal rotation primarily involves the midgut. The rotation involves three stages, the first stage occurs in weeks 5-10, which is a 90-degree anticlockwise rotation of the midgut, and the bowel returns to the abdominal cavity. In the 11th week, the second stage begins, which involves a further 270-degree anticlockwise rotation within the abdominal cavity so that the duodenal "C" loop and small bowel attach to the posterior abdominal wall with the ascending colon to the right side, the transverse colon above, and the descending colon to the left of the abdomen. In the third stage, the cecum descends, and the ascending and descending colon attaches to the posterior abdomen, with fusion and anchoring of the mesentery.^{5,6}

Intestinal anomalies can, thus, be categorized by the stage of their occurrence. The first stage anomaly includes omphaloceles caused by failure of the gut to return to the abdomen. Second-stage anomalies include nonrotation, malrotation, and reversed rotation. Stage three anomalies include an unattached duodenum, a mobile caecum, and an unattached small bowel mesentery. Adult patients typically have thick, fibrous, and vascular Ladd's band usually traveling from the caecum to the lateral abdomen, which predisposes to symptoms of intestinal obstruction.

There are usually three modes of presentation in adult patients with midgut volvulus. Chronic symptoms are the most common mode of presentation, characterized by recurrent episodes of vomiting with crampy abdominal pain in epigastric and periumbilical regions and sometimes early satiety. Less commonly, patients may have acute symptoms of intestinal obstruction, which may be life-threatening. The third mode of presentation is atypical, due to peculiar intestinal anatomy. For instance, there may be RUQ peritonitis due to acute appendicitis. Patients at risk of midgut volvulus, such as those with congenital midgut malrotation, are often asymptomatic. Many remain so, with the anomaly discovered incidentally during a CT scan performed for another reason. 9

The gold standard test for diagnosis of malrotation is upper gastrointestinal endoscopy combined with a barium study. An upper gastrointestinal contrast study often reveals a duodenal-jejunal junction located to the right of or overlying the spine and below the level of the duodenal bulb. In midgut volvulus, a corkscrew taping of the duodenum or jejunum might be present. The accuracy of the upper gastrointestinal series is reported to be around 80%. A noninvasive but equally effective test to diagnose intestinal malrotation is the CECT abdomen; hence, it is the investigation of choice with a diagnostic accuracy of more than 83%. CECT often shows right-sided small bowel, left-sided cecum, an inverse relationship between SMA and the superior mesenteric vein (SMV), aplasia of the uncinate process, and a

distinctive whirlpool-like pattern due to encircling of the loops of the bowel around the SMA. ^{10,11} However, in patients with features of bowel perforation, ischemia, or generalized peritonitis, diagnostic imaging will only act as the cause of delay in the definitive management, causing unnecessary morbidity.

The classical treatment for intestinal malrotation is Ladd's procedure, as first described by Ladd in 1932. ¹² Spigland et al, recommended that all patients with intestinal malrotation, even if asymptomatic, should undergo exploratory laparotomy. ¹³ Mazziotti et al, recently reported a series of malrotated patients who were managed laparoscopically. ¹⁴ Ladd's procedure includes anticlockwise detorsion of the volvulus, division of the abnormal peritoneal band (Ladd's band) overlying the duodenum, widening of the narrowed root of the small bowel mesentery, and placement of the small bowel to the right and the caecum to the left to avoid future malrotation. Appendectomy is an integral part of Ladd's procedure to avoid diagnostic dilemmas. ^{12,15}

CONCLUSION

Even though midgut volvulus is a rare anomaly, clinicians should have a reasonable index of suspicion in patients with features of small intestinal obstruction with no obvious immediate cause. In the event of a diagnostic dilemma, CECT abdomen would be the imaging modality of choice. Management is primarily surgical, depending upon the timing and severity of the presentation, from simple detorsion to resection and anastomosis of the bowel loop.

Funding: No funding sources Conflict of interest: None declared Ethical approval: Not required

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Cite this article as: Bhatta BR, Shrestha S, Shakya N, Shrestha M, Thapa KS. Midgut volvulus secondary to congenital malrotation in an adult with Ladd's band: a case report. Int Surg J 2024;11:2099-2102.