Case study

Midgut Volvulus secondary to intestinal malrotation in adult male – A case report and Mini review

Abstract

Intestinal malrotation is a congenital anomaly that occurs due to abnormal rotation and fixation of the intestine when it returns to the abdominal cavity during fatal development. There have been only 92 documented cases of this condition in medical literature. The incidence of malrotation is approximately 1 in 6,000 live births. Most malrotation cases, about 60-80%, become evident in the first month of life, with 90% being diagnosed within the first year. Adult presentations of malrotation are exceedingly rare, accounting for only 0.2 to 0.5% of cases, and among these, only 15% present with midgut volvulus. The incidence of midgut malrotation in adults has been reported to be between 0.00011 and 0.19%. Most diagnoses of midgut malrotation in adults occur in asymptomatic patients, often through imaging investigations or incidental findings during surgeries for other medical issues.

Diagnosis of midgut volvulus is primarily established through various methods, with CT abdomen being the most common (67%), followed by ultrasonography (15%), and during exploratory laparotomy (18%). It's important to note that midgut volvulus is associated with a high risk of bowel ischemia and necrosis, as it affects the blood supply to the intestines through the mesenteric artery, resulting in a significant 5% mortality rate.

We are reporting an unusual case involving a 20-year-old male who was found to have midgut volvulus affecting his jejunum and small bowel. This condition was a consequence of congenital malrotation of the bowel. The patient underwent an exploratory laparotomy, during which the volvulus was corrected by detorsion, inter-mesenteric bands adhesiolysis, and an appendectomy was performed.

Key words

Intestinal malrotation, midgut volvulus, Ladd’s procedure.

Introduction

Midgut malrotation is a condition marked by an abnormality in the regular embryonic rotation of the gut, which can manifest as acute abdominal obstruction with sudden onset or as chronic gastrointestinal symptoms. It necessitates prompt diagnosis and treatment to prevent severe complications like small bowel necrosis and gangrene. [1,2]

This developmental anomaly involves the rotation of the embryonic bowel. Malrotation can present acutely, intermittently, or remain asymptomatic. The diagnosis of malrotation with midgut

volvulus is a critical surgical emergency. During embryonic development, the small bowel typically undergoes a 270-degree counter clockwise rotation around the axis of the superior mesenteric artery (SMA) between the 8th and 12th weeks of gestation, followed by the retraction or fixation of the bowel mesentery into the abdominal cavity. [2,3,4]

There are several types of midgut malrotation:

1. Non-rotation.
2. Incomplete rotation.
3. Reverse rotation.
4. Anomalous fixation of the mesentery.

Genetic mutations in genes like BCL6 can affect the normal intestinal rotation pathway, resulting in an abnormal position of the cecum and the formation of Ladd's bands connecting the cecum to the retro-peritoneum and duodenum, leading to obstruction. The shortened mesentery of the midgut can create a pedicle around which the midgut may twist, causing volvulus and subsequent midgut ischemia and gangrene. Midgut malrotation is often associated with congenital anomalies, including congenital heart disease (CHD), congenital diaphragmatic hernia, omphalocele, intestinal and biliary atresia. [1,3,4]

Case Report

On May 22, 2010, a 20-year-old male patient was admitted to our centre, reporting severe abdominal pain, two episodes of vomiting, and a two-day history of constipation. Upon physical examination, the patient exhibited generalized abdominal tenderness with voluntary guarding but showed no signs of peritonitis. His abdomen appeared mildly distended, and all laboratory tests returned normal results. A CT scan of the abdomen confirmed the presence of midgut volvulus, with a positive "whirlpool sign."

A decision was made to proceed with an open exploratory laparotomy. During the surgery, a jejunal and small bowel volvulus was discovered, with the small bowel exhibiting signs of ischemia, characterized by a purple-blue discoloration due to a lack of blood supply. The midgut was carefully exposed in the surgical field, and the volvulus was successfully untwisted in a counter-clockwise direction. The previously bluish superior mesenteric vein (SMV) was engaged and had returned to a healthy pink colour. It was noted that the SMV was positioned to the left of the superior mesenteric artery (SMA), and the cecum was observed in the right upper quadrant. Additional congenital bands within the intestines were separated during the procedure, and adhesinolysis was carried out up to the broadened base of the mesentery. The Ladd's procedure was completed, including an appendectomy. The small bowel was positioned without tension in the right hemi-abdomen, while the large bowel was placed in the left abdomen. The patient's post-operative recovery progressed without any complications, and they were discharged on the eighth day following the surgery. The patient has remained asymptomatic for the past five years. **(Fig 1-8)**

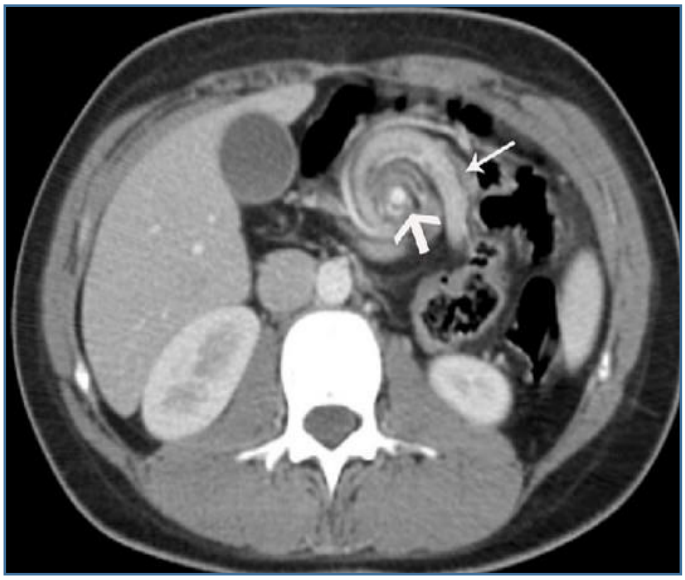

Fig-1 CT abdomen showing “whirlpool sign” (white arrows)

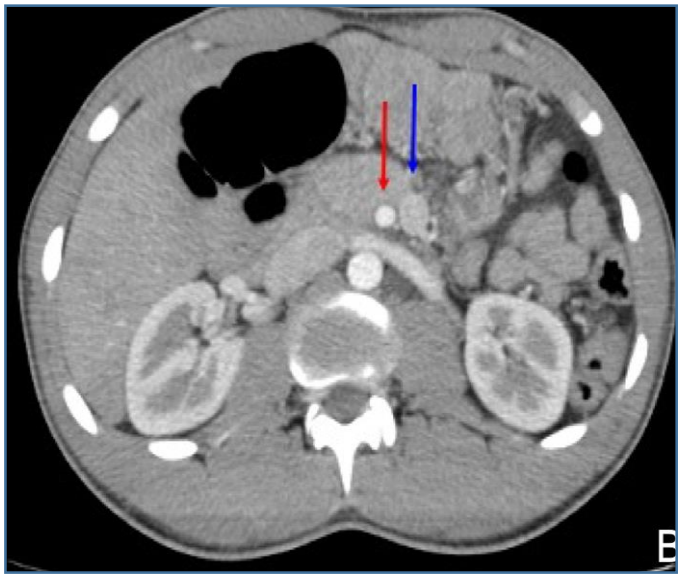

Fig-2 CT abdomen showing reversal sign of the SMA (Red arrow) and SMV to the left (blue arrow)

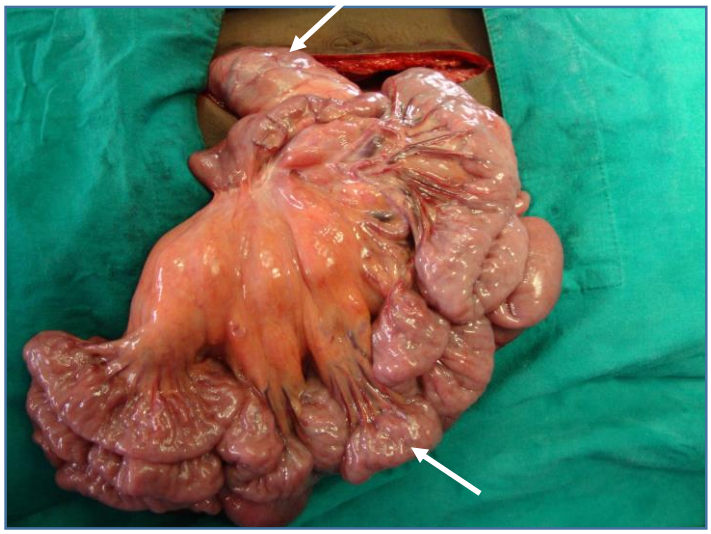

Fig-3 Intraoperative photograph showing midgut malrotation (“whirlpool sign”)

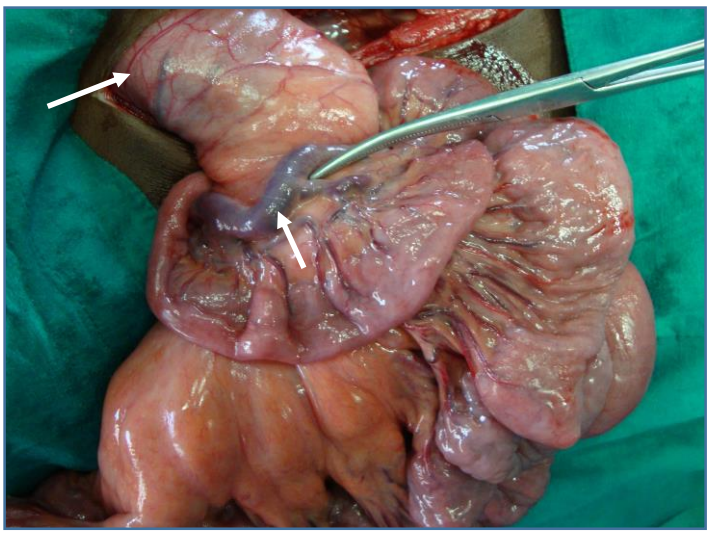

Fig-4 Intraoperative photograph showing midgut malrotation with engorged SMV and short mesentery

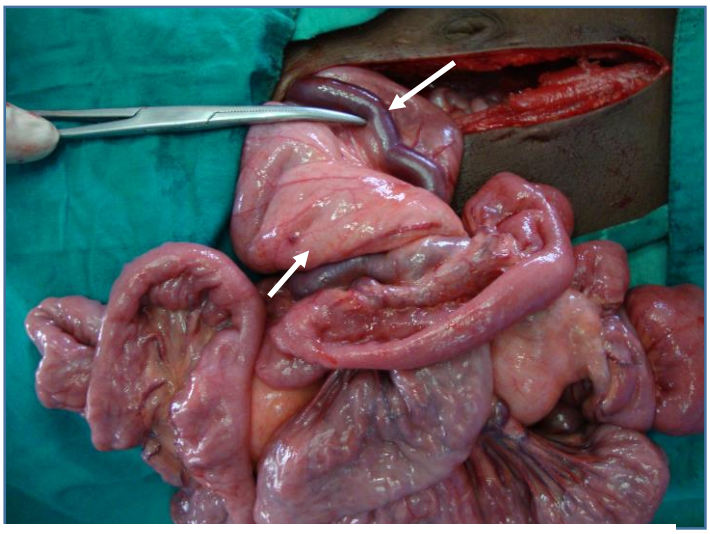

Fig-5 Intraoperative photograph showing SMA vein on left side of the SMA artery accompanied by bluish discoloration of the bowel.

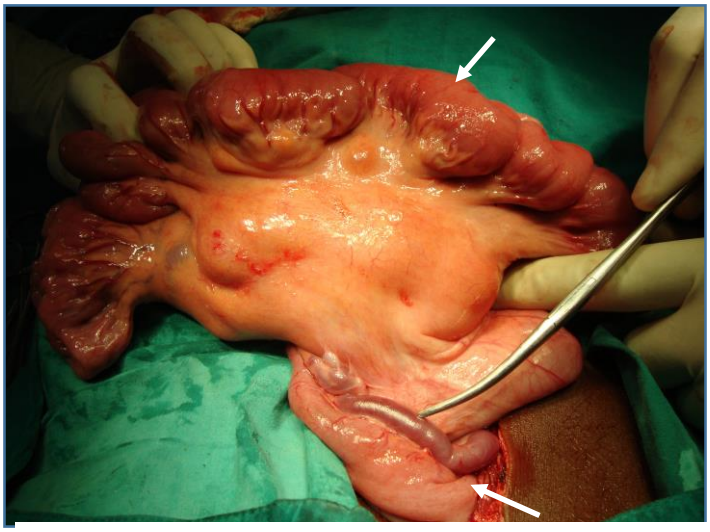

Fig-6 Intraoperative photograph showing complete detorsion resulting in a healthy pink mesentery and bowel.

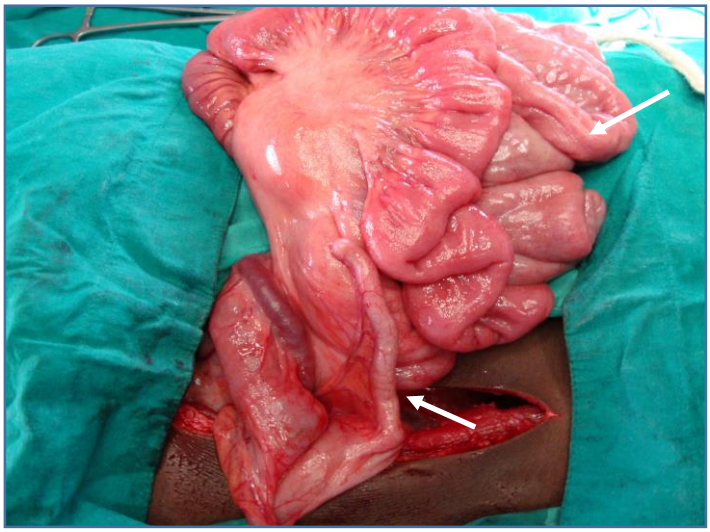

Fig-7 Intraoperative photograph showing after complete detorsion healthy pink mesentery and bowel with appendix

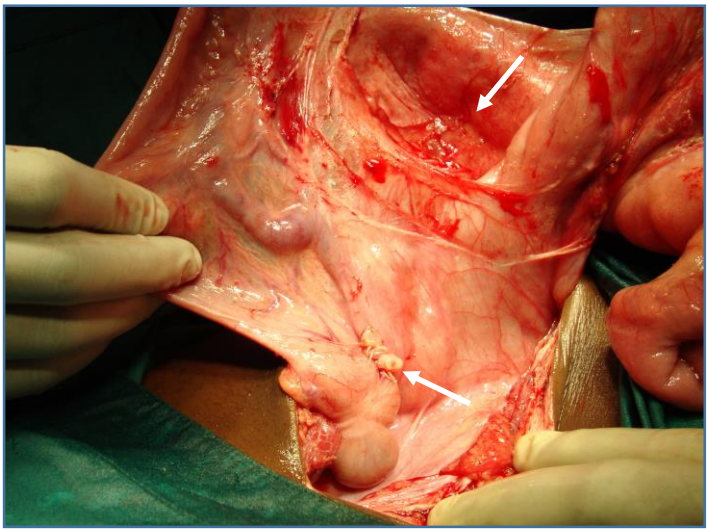

Fig-8 Intraoperative photograph showing Ladd's band excision and adhesiolysis with appendectomy

Discussion

Intestinal rotation and fixation begin at the 6th gestational week with a total of 270-degree anticlockwise rotation at the 12th week, the intestinal segment were fixed as the ascending colon on right sided, descending colon on left sided and cecum on right iliac fossa, according to Stringer's classification, there are three different types of midgut malrotation, as

1. Non-rotation
2. Duodenal malrotation
3. Duodenal and cecal malrotation

Mal-fixation of the mesentery results in a narrow pedicle predisposes to volvulus. Abnormal fibrous band arises from cecum compression of the duodenum called Ladd's band. Ladd's band named after William Ladd, show first described the surgical removal of these fibrous band in 1932. [1,2,3]

The exact aetiology of malrotation remains uncertain and martin et al identified mutation in forehead box transcription factor Foxf1 and L-R asymmetry genes and nutation associated with multiple gastrointestinal tract atresia, pancreatic and biliary atresia. Clinical presentation of intestinal malrotation of infants commonly presenting with midgut volvulus. Adults presentation of malrotation can be incidental on laparotomies, chronic and acute as midgut volvulus. [2,4,6]

These congenital Ladd's bands extend from the right lateral abdominal wall, across the duodenum and attached to the undescended cecum. Ladd's band compress the duodenum causes duodenal obstruction. Abnormal location of the cecum produces a narrow superior mesenteric vascular pedicle. This narrow SMA pedicle and lack of posterior peritoneal fusion predispose of subsequent midgut volvulus and obstruction with potential catastrophe.

The reported gold standard for diagnosis of gut malrotation is an upper gastrointestinal contrast study in the paediatric age group. CT scan with or without GI study is the investigation of choice. The CT abdomen, short mesentery, mesenteric twist and wrap around the narrow SMA pedicle to create" Whirlpool Sign" on CT scan. This pattern was first described by Fosher in a patient with midgut volvulus. [1,2,4,7]

Mini-review

92 cases of adult intestinal malformation with acute midgut volvulus have been documented in published literature. We conducted a search covering the period from 1983 to 2012, utilizing PubMed and Web Japan Medical Abstract databases, which yielded 37 reported cases of intestinal malformation. Among these cases, 20 patients were male, and 17 were female. Notably, Flue et al and colleagues reported nine cases, Dietz et al and co-authors reported ten cases, while Choi et al collaborators described 177 patients over a 35-year span. [2,4,5]

The surgical management of intestinal malformation was originally detailed by William Ladd in 1936 and has since remained the primary method of treatment. The classic Ladd's procedure comprises five key components. Firstly, any bowel torsion must be carefully untwisted in a counter-clockwise fashion, and segments showing signs of ischemia necessitate resection. Secondly, adhesinolysis of Ladd's bands is performed to aid in the mobilization and detorsion of the duodenum. Further division of inter-mesenteric bands expands the mesentery's base, and appendectomy is typically carried out. Additionally, adhesions around the superior mesenteric artery (SMA) are divided to prevent future volvulus. Finally, the bowel is returned to its natural non-rotated position, meaning the small intestine is arranged on the right side of the abdomen, and the colon on the left side. Most authors are opinion that Ladd's procedure represents an effective treatment for intestinal malformation. [2,3,6]

In recent years, the laparoscopic approach for the surgical treatment of intestinal malformation, including laparoscopic Ladd's procedures in paediatric patients, has been increasingly reported in the literature. However, it is worth noting that laparoscopy approach can be technically challenging, and conversions to open procedure is common. [2,4,5]

Conclusion

Intestinal malrotation is a rare condition but is considered an important cause of bowel obstruction in adults and CT abdomen is the diagnostic tool of choice. Prompt surgical exploration is crucial for patient’s survival. Ladd’s procedure is intervention of choice which can be performed by open laparotomy or laparoscopically

References

1. William A. Butterworth, James W. Butterworth, An adult presentation of midgut volvulus secondary to intestinal malrotation: A case report and literature review, International Journal of Surgery Case Reports, https://doi.org/10.1016/j.ijscr.2018.07.007.

2. Emanuwa, O.F., Ayantunde, A.A. & Davies, T.W. Midgut malrotation first presenting as acute bowel obstruction in adulthood: a case report and literature review. World J Emerg Surg 6, 22 (2011). https://doi.org/10.1186/1749-7922-6-22

3. Prasan Kumar Hota, D Abhishek, and V Bhaskar, ADULT MIDGUT MALROTATION WITH LADD’S BAND: A RARE CASE REPORT WITH REVIEW OF LITERATURES, Bali Medical Journal (Bali Med. J.) 2014, Volume 3, Number 3: 137-142 P-ISSN.2089-1180, E-ISSN.2302-2914

4. Torres AM, Ziegler MM. Malrotation of the intestine. World J Surg. 1993 May-Jun;17(3):326-31. doi: 10.1007/BF01658699. PMID: 8337878.

5. Ramprasad Rajebhosale, Rajesh Chidambaranath, Cindy Cleto, Dylan Parmar, Najam Husain, Pradeep Thomas, EP.TH.273
Intestinal Malrotation in Adults: A case series and Literature Review, *British Journal of Surgery*, Volume 108, Issue Supplement_7, October 2021, znab309.033, https://doi.org/10.1093/bjs/znab309.033

6. Ribeiro, Tiago & Greene, Brittany & Bennett, Sean & Msallak, Haytham & Karanicolas, Paul. (2022). Variations in intestinal malrotation in adulthood: A case report of midgut volvulus and literature review for the surgeon. International Journal of Surgery Case Reports. 91. 106750. 10.1016/j.ijscr.2021.106750.

7. Mohan A, Ansari WA, Gada PB, Dandge SM, Nagwani PD.Intestinal malrotation with midgut volvulus:associations and role of elective surgery in adults.Int Surg J2022;9:1272-6.

8. Jarvis J K, Rambhajan A (December 26, 2020) Adult Intestinal Malrotation in a Non-Paediatric Hospital in Trinidad: A Case Report and Literature Review. Cureus 12(12): e12305. doi:10.7759/cureus.12305

9. Bastiaan W. Haaka, Sander T. Bodewitz , Caroline F. Kuijper, Louise M. de Widt-Levert, Intestinal malrotation and volvulus in adult life, B.W. Haak et al. / International Journal of Surgery Case Reports 5 (2014) 259–261

10. Rohit Kapoor, Narinder Singh, Shubh Kirti b¥and Abhishek Sharma, Management of Sixteen Year Old Male with Intestinal Malrotation by Ladd's Procedure: A Case Report and Review of Literature, Asian Journal of Case Reports in Surgery, 5(1): 188-192, 2022;Article no.AJCRS.87995