

Adult-Onset Midgut Volvulus Due To Intestinal Malrotation: Case Report and Literature Review

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How to citation this article: Dr Manish Kumar Pradhan S, Dr Ranjith K B, Dr Gopinatha Pai, Dr Nikhil N G, “Adult-Onset Midgut Volvulus Due To Intestinal Malrotation: Case Report and Literature Review”, IJMACR- July - 2025, Volume – 8, Issue - 4, P. No. 252 – 258.

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Type of Publication: Case Report

Conflicts of Interest: Nil

Introduction

Intestinal malrotation is a congenital anomaly found in 0.2%–1% of the population, posing various clinical challenges ¹. Intestinal malrotation occurs when there is incomplete rotation of the midgut by 270 degrees counterclockwise around the superior mesenteric vessels during embryonic development ¹. This condition leads to positioning of distal duodenum and jejunum to the right side of the columna vertebralis in affected individuals ². The narrow-based mesentery that is associated with arrested midgut rotation increases the likelihood of twisting of midgut, potentially predisposing to obstruction and necrosis. The majority of cases (60–80%) manifest as an acute surgical emergency, most commonly due to midgut volvulus during the neonatal period.

Beyond the neonatal period, the clinical presentation becomes more diverse. Recent studies indicate a declining frequency as patients mature, with symptoms ranging from acute obstruction to chronic intermittent colicky pain and bile-stained vomiting ^{1,4-6}. It is quite rare in adults and poses a significant challenge to promptly diagnose and manage intestinal malrotation. This is exacerbated by nonspecific symptoms and limited familiarity amongst healthcare provider ^{4,5,7}. These challenges contribute to higher morbidity and mortality rates in adults suffering from this condition. We present here a unique case of a 34-year-old male with delayed presentation of intestinal malrotation, presenting with acute on chronic symptoms of midgut volvulus with unique anatomic findings. A review of the literature of intestinal malrotation in adults will follow.

Objective

This poster aims to discuss a rare case of intestinal malrotation undiagnosed until adulthood, highlighting the diagnostic journey, clinical presentation, and surgical management.

Case report

A 34-year-old male patient presented to the casualty with complaints of pain in the upper abdomen of 1 week duration. It was associated with abdominal distension and bloating sensation which was relieved following vomiting. No history of altered bowel habits was present. He reported multiple episodes of similar history for the past 1 year. On examination, his vitals were stable. Abdomen was mildly distended with vague fullness noted in upper quadrants. On palpation, abdomen was soft without any sign of guarding and rigidity. Tenderness was noted in the epigastric region. Per rectal examination revealed formed stools. Erect X Ray of abdomen showed colonic loops loaded with stool and displaced towards the left (Figure 1).



Figure 1: Erect X ray Abdomen showing loops of colon loaded with stools displaced to the left

CECT Abdomen and pelvis revealed dilated stomach, duodenum and dilated proximal jejunum measuring 7 cm with evidence of twisting of mesenteric vessels with change in relationship of SMV and SMA, S/O mid gut

volvulus with transition point near the mid-point of jejunum.



Figure 2: CECT Abdomen showing dilated stomach and duodenum



Figure 3: CECT Abdomen showing transition point of midgut volvulus at the proximal jejunum

Endoscopy revealed Reflux esophagitis, dilated duodenum with normal mucosa.

Patient was taken up for emergency exploratory laparotomy. Intraoperatively, grossly distended duodenum and stomach with twisting of the proximal jejunum around 8cm away from the D-J flexure was noted. Band like adhesions were noted at point of twist. Malrotation of the gut was noted with small bowel loops to the right side and ascending and descending colon to the left. Caecum and appendix were found lying in the left iliac fossa. Adhesions were noted between ascending and descending colon. All of bowel was viable without any signs of ischemia. After derotation of the jejunum, adhesiolysis with appendicectomy was performed. Postoperatively, patient recovered well without any

complications. Patient passed stools on POD2. On follow up, patient recovered well.



Figure 4: DJ flexure with adhesions distal to it

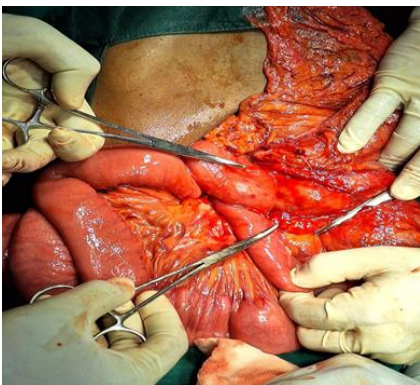


Figure 5: Upper artery pointed to the proximal dilated duodenum; lower artery pointed to the distal collapsed duodenum. Twist of the duodenum can be appreciated between the two



Figure 6: Interbowel loop adhesions



Figure 7: Showing malrotation of gut with small bowel loops to the left and colon to the right, caecum and appendix in the left side of abdominal cavity

Discussion

Development of gut occurs serially in three stages of rotation. It begins with Stage I, occurring between 5th and 10th gestational weeks. It begins with extension of midgut into the extra embryonic cavity, followed by a 90° counterclockwise rotation around the superior mesenteric artery (SMA) and retraction of bowel loops into the foetal abdomen. Stage II occurs in the 11th week, entails further 180° counterclockwise rotation within the abdominal cavity, completing a 270° rotation. During this stage, the duodenum descends below the SMA, following its C-loop trajectory, while the transverse and ascending colon demonstrate the path of rotation with the cecum moving upward towards the artery. Stage III involves mesentery fusion and anchoring. The duodenum becomes retroperitoneally fixed in its third portion, emerging at the ligament of Treitz, and the cecum is anchored to the lateral abdominal wall by peritoneal bands. The branches of the superior mesenteric artery originate and stabilize along a line extending from its origin from the aorta to the cecum in the right lower quadrant ⁴.

Intestinal malrotation encompasses any deviation from the typical counterclockwise rotation of the midgut in stage II. When this rotation fails, the entire small bowel remains positioned on the right side of the abdomen, and the colon on the left, with absence of the ligament of Treitz, resulting in incomplete intestinal rotation. This abnormal positioning, along with anomalous adhesions and a shortened mesentery, predisposes the bowel to volvulus—a clockwise twisting around the axis of the superior mesenteric artery—leading to potential ischemia and necrosis of the small intestine. Malrotation is further characterized by the presence of peritoneal or Ladd's bands that extend from the cecum to the lateral abdominal wall and across the duodenum, which can contribute to duodenal compression and obstruction⁵. The shortened mesentery of the displaced midgut forms a pedicle around which the midgut can rotate, predisposing to volvulus and subsequent ischemia of the midgut³. Various types of Intestinal Malrotation include Non rotation, Incomplete rotation, Reverse rotation and anomalous fixation of mesentery.²

The incidence of mid-gut malrotation is estimated to be around 1 in 6000 live births.

BCL6 gene, a part of signalling pathway is deemed essential for normal intestinal rotation. Genetic mutations in this gene leads to abnormal positioning of the cecum and formation of fibrotic bands

While most cases of malrotation get diagnosed within the first month of life, it can manifest in adults as well.². Adults typically present with symptoms attributable to Chronic bowel obstruction such as, vomiting and recurrent abdominal pain^{1,6,7}. Approximately 30% of patients exhibit vomiting, while 20% experience recurrent nonspecific abdominal pain. These symptoms can stem from either acute or chronic intestinal

obstruction, often attributed to the presence of Ladd bands and/or volvulus, which may lead to ischemic necrosis and perforative peritonitis. Malabsorption may also manifest, characterized by diarrhoea and nutritional deficiencies due to bowel lymphedema caused by chronic volvulus, resulting in protein loss into the bowel lumen. Uncommon presentations of chronic volvulus include obstructive jaundice due to mechanical compression of the biliary tract, chylous ascites, and superior mesenteric vein thrombosis, secondary to prolonged lymphatic and venous obstruction⁶⁻⁹.

The clinical presentation of midgut malrotation often resembles various other gastrointestinal conditions including necrotizing enterocolitis in neonates and intussusception in older infants. Due to rarity of condition in adulthood, diagnosis of adult intestinal malrotation relies on imaging studies. Absence of the caecal gas shadow or predominance of small intestinal loops on the right side should raise suspicion of malrotation. Upper gastrointestinal barium series remains highly accurate (over 80% sensitivity) for detection, typically revealing the duodeno-jejunal junction failing to cross the midline and positioned below the duodenal bulb. Contrast enema typically shows right colon malpositioning, with the ileum entering the cecum from the right. However, due to risk of perforation in cases of midgut volvulus of malrotated gut, contrast bowel studies are not preferred in recent times. Contrast-enhanced CT, the investigation of choice, can delineate the anatomical positioning of right-sided small bowel, left-sided colon, and abnormal relationship of the superior mesenteric vein (SMV) positioned left of the superior mesenteric artery (SMA) instead of the typical right-sided configuration, diagnostic of malrotation. Additionally, the "clockwise

whirlpool sign" observed on colour Doppler ultrasonography, showing the clockwise wrapping of the SMV and mesentery around the SMA, has been recognized as a crucial diagnostic feature in midgut volvulus¹⁰⁻¹⁴.

A selective superior mesenteric angiogram may display abnormal curling and a corkscrew appearance of vessels towards the right, diagnostic for midgut volvulus due to malrotation.

Diagnosing intestinal malrotation in adults is challenging due to its atypical presentation and low index of suspicion among healthcare providers. Standard imaging techniques such as abdominal ultrasound and CT scans may not always clearly indicate malrotation. Advanced imaging techniques, including an upper gastrointestinal series, are often required to confirm the diagnosis. This case highlights the importance of thorough patient history and the use of appropriate diagnostic tools.

The standard treatment for incomplete intestinal rotation is Ladd procedure, which involves : counterclockwise detorsion of any viable midgut volvulus, if present followed by division of abnormal coloduodenal Ladd bands that tether the midgut and cause external compression, mobilization or Kocherization of the duodenum and right colon, widening of the mesenteric base by releasing adhesions near the superior mesenteric vessels to prevent future volvulus and removal of the malpositioned appendix. The primary goal of this procedure is to reduce the risk of acute volvulus by positioning the small intestine in a non-rotated state and broadening the mesenteric base. Appendectomy is typically performed due to the appendix's potentially atypical location, which could complicate future diagnoses of appendicitis which typically present with right lower quadrant pain. In recent times, laparoscopic

approach to the Ladd procedure is considered safe, allowing for early recovery and return to work.^{15,16}.

Patients with midgut malrotation generally have a favourable prognosis, provided there is no occurrence of midgut volvulus, necrosis, small gestational age, premature birth, or other associated abnormalities. Mortality rates following surgery for malrotation range from 3 to 9 percent. Additional risk factors such as volvulus, prematurity, or concurrent anomalies may further increase the mortality rate. Widening the mesenteric base during surgery reduces the risk of recurrent volvulus, which is estimated to occur in 2 to 8 percent of cases.

Common surgical complications include recurrent volvulus, early postoperative small bowel obstruction caused by adhesive bands, wound infections, deep vein thrombosis, and aspiration pneumonia due to nasogastric tube placement for bowel decompression.

Additional complications may involve bowel loss leading to short bowel syndrome and vitamin deficiencies. Treatment for short bowel syndrome typically involves anti-diarrheal medications, nutritional therapy such as small quantity, high-protein diets, increased hydration, and replacement of vitamins and minerals. Parenteral nutrition may be necessary if the bowel length is insufficient or during rehabilitation. Autologous bowel reconstruction and small bowel transplantation are considered as last-resort options.

The present case focuses attention at this critical rare subject by several points. First, presentation of adult malrotation cases can be obscure, even though whirlpool sign in abdomen computed tomography scan may give suspicion of bowel twisting. Yet, an acute presentation is commonly associated with extensive bowel necrosis and may lead to massive resection. We observed no

gangrenous bowel during laparotomy. Thus, adhesiolysis with appendectomy was performed, following which the patient condition improved. Management of patients with short bowel syndrome due to massive small bowel resection is a challenging situation and commonly complicating the patient's status with electrolyte disturbances, malnutrition, immune deficiency, organ failure and sepsis. Refeeding enteroclysis is a helpful option in such patients, usually requires a considerable amount of healthy distal small bowel for absorption of nutrients, bile salts and fluids ¹⁵. Our patients did not undergo any bowel resection and thus his condition improved. This case shows timely recognition of malrotation is the key to save life, if possible, but massive small bowel resection is sometimes unavoidable and may associate with fatal consequences.

Conclusion

We report a rare case of a 34 year old male presenting with midgut volvulus secondary to intestinal malrotation, subsequently requiring adhesiolysis. This case emphasises the importance of maintaining a high index of suspicion for this insidious condition in addition to closely monitoring such patients. This allows prompt identification and management of such rare cause of deteriorating surgical patient, hence optimising patient outcomes.

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