



MIDGUT MALROTATION WITH CAECAL VOLVULUS IN AN ADULT PRESENTING AS ACUTE INTESTINAL OBSTRUCTION – A CASE REPORT

General Surgery

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ABSTRACT

Midgut Malrotation, considered a pediatric pathology is a rarity in an adult population. We report a case of Midgut Malrotation in a 24-year-old male presenting as caecal volvulus with clinical features of acute intestinal obstruction. Pre-operative diagnosis of caecal volvulus was suspected based on CT findings. Intra-operatively, caecal volvulus with closed-loop obstruction with the abnormal location of small bowel, caecum and appendix associated with Ladd's bands was seen. Exploratory laparotomy with gangrenous bowel resected from caecum to hepatic flexure with ileo-transverse anastomosis and Ladd's procedure was done. Postoperatively there were no complications. Surgeons face a dilemma regarding the diagnosis of midgut malrotation as it rarely presents with specific symptoms resulting in the scenario of incidental diagnosis. A high index of suspicion with the prompt investigation and surgical intervention is required to prevent delay in treatment and poor prognosis.

KEYWORDS

Midgut Malrotation, Ladd's Bands, Caecal Volvulus

INTRODUCTION

During embryological development, any abnormal deviation from the normal 270 degrees counter clockwise rotation of the midgut can be described as Midgut malrotation¹. It is usually considered a pediatric pathology & vast majority of the complications associated with midgut malrotation present in the first month of life. The reported incidence of adult midgut malrotation is between 0.0001 % and 0.19 % showcasing its rarity^{2,3}. Most of the patients are asymptomatic and are incidentally diagnosed on imaging investigations or intra-operatively. Two distinct clinical patterns, acute and chronic presentations are seen in adults.

Chronic presentation is a common feature in adults & is characterised by intermittent abdominal cramps, bloating, nausea and vomiting over several months or years. The symptoms may be highly nonspecific⁴.

Acute Presentation in an adult typically presents with symptoms of acute bowel obstruction. These patients may or may not report a previous history of abdominal symptoms⁴.

Surgeons usually characterise another differential diagnosis for an acute abdomen as they have a low index of suspicion considering midgut malrotation. We report a case of an adult patient with an acute presentation of midgut malrotation as caecal volvulus and its operative management.

CASE REPORT

A 24-year-old male patient arrived in the emergency with chief complaints of acute onset of abdominal pain for 2 days accompanied with nausea and bilious vomiting. There was h/o of obstipation for 24 hours. The patient had no prior history of similar complaints or surgical history and neither co-morbidities.

On examination, the general condition of the patient was afebrile, normotensive, tachycardia and tachypnea. The abdominal examination revealed moderate distension with tenderness in the epigastric region. There was no evidence of peritonitis.

Routine blood investigations revealed a haemoglobin of 14.2g/dl with WBC count of 10.3×10^3 while the remaining investigations including serum electrolytes, urea, amylase, lactate, liver function tests (LFTs), clotting profile, C-reactive protein (CRP) and arterial blood gas (ABG) were normal. The flat plate abdomen-erect did not reveal air under diaphragm but showed dilated bowel loops in the central and upper abdominal regions. Chest radiograph showed no significant findings.

The decision for further investigation of computed tomography was taken. The CT scan revealed a caecal volvulus with close loop obstruction.

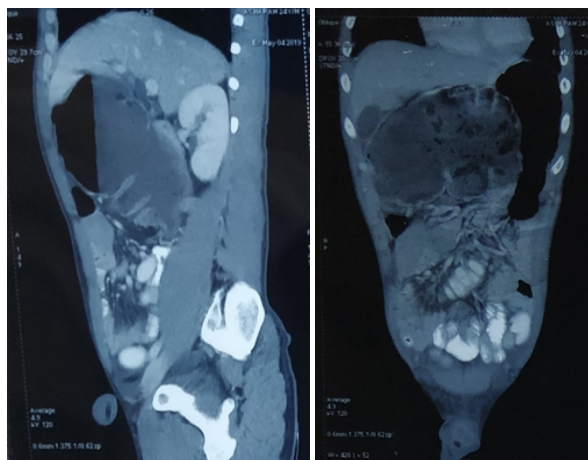


FIGURE 1

FIGURE 2

FIGURE 1 & 2- SHOW VOLVULUS OF CAECUM FORMING A CLOSED LOOP OBSTRUCTION

The patient was resuscitated with intravenous fluids, antibiotics and analgesics. A nasogastric tube was inserted for decompression. Catheterization was done to evaluate the urine output. The patient was posted for an emergency exploratory laparotomy.

Intraoperative findings revealed that bowel extending from the ileocaecal junction to hepatic flexure of the colon was distended with gangrenous changes and caecal volvulus was present on the left side of the abdomen. Small bowel had collapsed (proximal part collapsed) occupying the right paracolic gutter and the right iliac fossa. Fibrous bands were present over the distal part of the duodenum, on the right side of the abdomen. All these findings confirmed midgut malrotation.

The operative procedure consisted of resection of caecal volvulus from ileocaecal junction to hepatic flexure and Side to side Ileo- Transverse anastomosis. The congenital Ladd's bands were divided- Ladd's Procedure. The small bowel was placed on the left side of the abdomen. Midgut Malrotation was corrected.

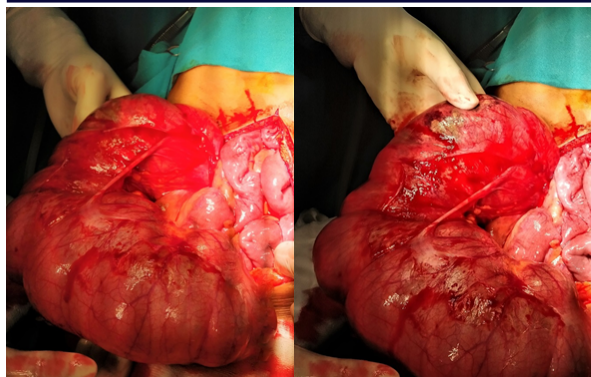
**FIGURE 3**

FIGURE 3 - collapsed small bowel present in upper abdomen and right paracolic gutter

FIGURE 4

FIGURE 4 – caecal volvulus with closed loop obstruction

Postoperatively, the patient had an uneventful recovery. He was started on oral sips on the first post-operative day and intake was gradually increased. There was no post-operative ileus and patient passed flatus and stools. Patient was discharged on the fifth post-operative day. Follow up after 10 days and one month revealed that he was well and there had been no post-operative complications.

DISCUSSION

The embryology of intestinal rotation was described by Mall in 1898 and 25 years later, Dott correlated these observations with clinical problems^{5,6}. In 1936 Ladd wrote the classic article on the treatment of the condition, describing 21 cases in which treatment was based on an understanding of the underlying embryology⁷.

During 4th week of fetal development, the gut gets divided into the foregut, midgut and hindgut according to the vascular supply. The midgut is supplied by the superior mesenteric artery (SMA). There is rapid elongation during the 5th week and the midgut outgrows the capacity of the abdominal cavity. This leads to a temporary physiological herniation into the umbilical cord at sixth week with return to the abdominal cavity 4 to 6 weeks later. The midgut undergoes a 270-degree counterclockwise rotation around the SMA axis simultaneously. This process leads to the formation of the duodenum, placing it behind the SMA in a retroperitoneal position. During the 10th week there is a progressive reduction of the herniation back into the abdominal cavity. The duodeno-jejunal flexure and jejunum reduce first and lie to the left of the SMA. The distal small bowel then follows and lies to the right of the abdominal cavity. Caecum descends from right upper quadrant to right lower abdomen. The base of the small bowel mesentery subsequently fuses with the posterior peritoneum from the ligament of Treitz at the DJF to the caecum, completing the whole process at the eleventh week of fetal development^{8,3,9,7}.

The “classic” form of malrotation or nonrotation occurs due to a total failure of both the duodenojejunal and cecocolic loops to rotate. However, failures at individual stages of rotation leads to different degrees of nonrotation. Balthazar emphasized 28 cases of adult malrotation with six possible configurations. He divided these into complete or partial failures of rotation and into abnormalities affecting the duodenojejunal loop, the cecocolic loop, or both¹⁰.

Clinically the patient either presents with acute or chronic duration of symptoms. Wang and Welch showed that 24 of 50 patients were clinically asymptomatic in their case series of adolescents and adults with malrotation³. Dietz et al studied a series of 10 adults with bowel obstruction caused by intestinal malrotation. They reported that 5 adults presented with chronic features and that the duration of symptoms extended to 30 years⁹. Moldrem et al reported that 48.5% of their 33 patients presented with an acute abdomen¹¹.

Multiple diagnostic modalities like plain abdominal radiograph, ultrasound scan (USS), computed tomography (CT) scan, magnetic resonance imaging (MRI) scan and mesenteric arteriography can be used to identify malrotation. Plain radiography is neither sensitive nor specific in the diagnosis of gut malrotation. Color Doppler USG may reveal malposition of the SMA^{11,1,13}. Pacros, first described characteristic USG findings of midgut volvulus included duodenal

dilatation with distal tapering and fixed midline bowel and mesentery twisted around the SMA axis representing as the 'whirlpool' sign¹³. Upper gastrointestinal (UGI) contrast study is the gold standard for diagnosis of gut malrotation particularly in the pediatric age group^{9,1,12}.

These generally show the duodenum and duodenojejunal flexure located to the right of the spine. Nicholis and Li were the first to identify how the relationship of SMV and SMA is a useful indicator in diagnosis but a proportion of patients with malrotation might even have a normal SMA-SMV relationship¹⁴.

The shortened mesentery allowing the small bowel and mesentery to twist and wrap around the narrowed SMA pedicle to create a distinctive Whirlpool appearance on CT scan was first described by Fisher¹⁵.

The surgical management of intestinal malrotation was first described by William Ladd in 1936.⁷ In 2003, Matzke reported a case where he performed Laparoscopic Ladd Procedure for Adult Malrotation of the Midgut with Cocoon Deformity.¹⁶

In 2005 Matzke conducted a comparative study between the open and laparoscopic Ladd's procedure in 21 patients indicating that laparoscopic is as feasible an option as an open procedure.¹⁷

CONCLUSION

Malrotation of the intestinal tract is a product of well-defined aberrant embryology. This ailment pertains mostly to the pediatric population decreasing the surgeon's exposure to this condition. Therefore, an understanding of the anatomy, diagnostic criteria, and appropriate therapy for this emergency condition is imperative. Our case report expands on the condition in which a patient presents with intestinal malrotation with caecal volvulus and further details the investigations and surgical procedure carried out.

The consequences of caecal volvulus associated with malrotation may be catastrophic leading to bowel ischemia and necrosis. Evidence of which portends a poor prognosis and death. Maintain a high index of suspicion and consider a prompt surgical intervention to prevent an abdominal catastrophe and fatality.

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