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Case Report

Intestinal malrotation with midgut volvulus: associations and role of elective surgery in adults

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ABSTRACT

Intestinal malrotation, a congenital anomaly of rotation of midgut, with paediatric dominant presentation is increasingly being observed in adult population. Here, the severity of presentation ranges from asymptomatic to life threatening midgut volvulus. Presentation with midgut volvulus, managed by an emergency surgery with definitive Ladd's procedure is the current standard. Presently, there is no data to support elective repair in stable patients with chronic midgut volvulus. Management of asymptomatic adult with malrotation is also a topic of controversy with no recommended guidelines. We herein, present two cases of malrotation with midgut volvulus with initial conservative management followed by elective Ladd's procedure. Rare associations like nutcracker phenomenon and pancreatitis were also observed in one of our patients. Our study aims to correlate our findings with the published literature, to understand these associations better and to review the need for an emergent operation for chronic midgut volvulus.

Keywords: Intestinal malrotation, Ladd's procedure, Midgut volvulus, Nutcracker syndrome, Pancreatitis

INTRODUCTION

Incidence of malrotation is approximated at 1 in 6000 live births. 64-80% of these are encountered in the neonate period and 90% in the infancy. Only 0.2-0.5% of cases present in adults with 15% presenting with midgut volvulus.[1] The Stringer classification has divided malrotation into three types that is; non-rotation, duodenal malrotation and duodenal plus caecal malrotation.[2] Presentations in an adult may vary from asymptomatic and incidental, chronic vague abdominal complaints to acute abdomen. Where acute abdomen necessitates emergency exploratory laparotomy, the other two may be managed conservatively or with elective surgery. Unlike in neonates, the need for operative correction in adult malrotation is a topic of controversy with no recommended guidelines. Malrotation presenting with complications decreases with advancing age. On the other hand, complications after operative intervention increase with advancing age. Adults often have coexisting pathologies which might be the actual cause of discomfort. Hence an individualised planned approach should be preferred in the adult population.

CASE REPORT

Case 1

A 28-years-old female, presented with complaints of pain in the abdomen, vomiting and postprandial fullness for 1 week without features of intestinal obstruction. She was hemodynamically stable with normal biochemical parameters. On evaluation, she was diagnosed to have non-rotation of the gut with midgut volvulus without intestinal obstruction. She was managed conservatively and symptoms resolved. She was discharged with advice regarding high-risk symptoms and the need for follow up. She presented ten months later with bilious vomiting, obstipation and pain in the abdomen for 1 day. She was hemodynamically stable with mild epigastric tenderness

with normal biochemical parameters. X-ray abdomen revealed multiple air-fluid levels (Figure 1). Computed tomography (CT) scan revealed non-rotation with midgut volvulus with small and large bowel obstruction with an uncomplicated umbilical hernia (Figure 2). There was reversal of relation between superior mesenteric artery (SMA) and superior mesenteric vein (SMV). The small bowel was on the right side and the large bowel was on the left side (Figure 3). Duodenojejunal (DJ) flexure was on the right side. Ladd's band were compressing the duodenum with proximal duodenal dilatation. She was again managed conservatively initially which resolved her symptoms. In view of recurrent episodes and midgut volvulus decision for elective operative intervention was made. Intraoperatively there was a clockwise twist of the midgut around the SMA (Figure 4).

The small intestine, caecum and ascending colon were congested, mesentery was thickened, and blood vessels were dilated with a diameter of about 1 cm. There were multiple enlarged mesenteric lymph nodes. All these features were indicative of chronic midgut volvulus. After anticlockwise derotation, Ladd's bands were released. Ladd's procedure was completed with an appendicectomy. The postoperative course of the patient was uneventful. The patient is asymptomatic for ten months now.

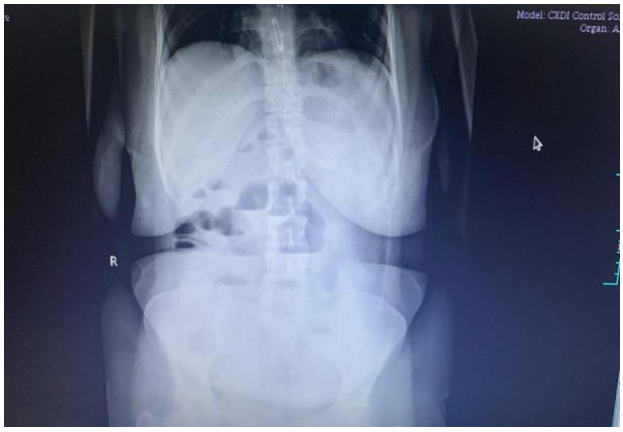

Figure 1: X-ray showing multiple air-fluid levels.

Figure 2: Midgut volvulus as seen on CECT scan with evidence of swirl sign (arrow).

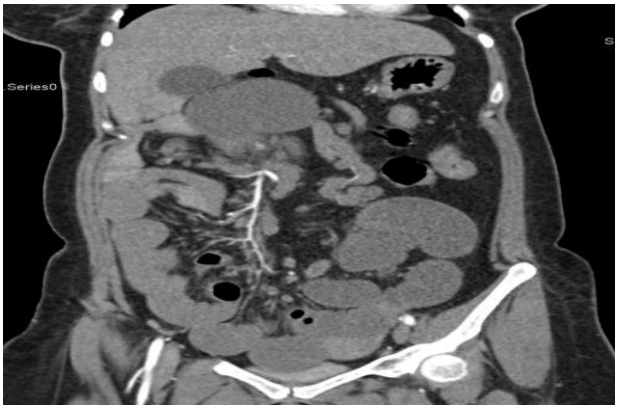

Figure 3: Non-rotation with small bowel on the right and large bowel on the left with maintained vascularity.

Figure 4: Intra-operative midgut volvulus.

Case 2

A 39-year-old male patient was referred from a private hospital, where an emergency exploratory laparotomy was done in view of intestinal obstruction two months back. No details were available of the operative procedure. The patient developed symptoms of intestinal obstruction again on the fourth postoperative day for which a contrast-enhanced CT scan of the abdomen was done. It revealed midgut volvulus. Nasojejunal (NJ) tube was inserted for the same and the patient was on NJ feeding for 2 months. He developed pain in the abdomen with recurrent vomiting for one week for which he was referred to us. He was passing liquid stools and flatus inconsistently. On examination he was hemodynamically stable, per abdomen was soft with no tenderness. Biochemical parameters were within normal range. He was underweighting and malnourished. X-ray abdomen showed an abnormal position of the DJ flexure without signs of intestinal obstruction (Figure 5). CT scan was consistent with midgut volvulus with dilated duodenum with SMV anterior to SMA (Figure 6). There was no intestinal obstruction. It was also associated with chronic calcific pancreatitis and the incidental nutcracker phenomenon was seen (Figure 7). The patient was optimised with continued NJ feeding and total parenteral

for an elective exploratory laparotomy. Intraoperatively DJ flexure was on the right side of SMA. Ladd's bands were extending from the caecum to the proximal jejunum causing its compression with proximal dilatation of the duodenum. There were also peritoneal bands between the small intestinal loops, between the entering and exiting loop of the volvulus (Figure 8). These were indicative of a chronic volvulus. The rest of the intestine was in a normal position. Ladd's procedure was performed. Appendicectomy was not performed on this patient. The postoperative course was uneventful. The patient is asymptomatic with adequate weight gain for six months now.

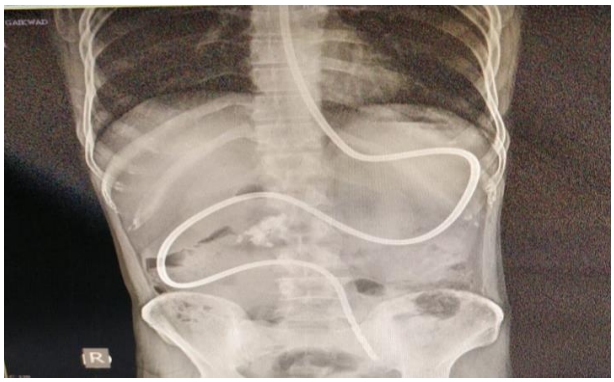

Figure 5: X-ray with NJ tube in situ with calcific pancreatitis.

Figure 6: CECT scan showing dilated duodenum.

Figure 7: Nutcracker phenomenon.

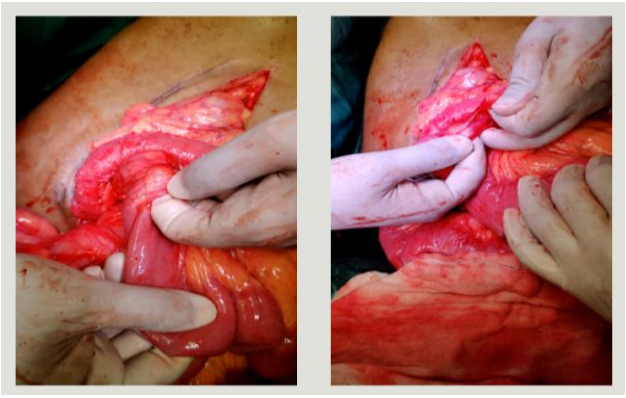

Figure 8: Interbowel bands.

DISCUSSION

Malrotation of the gut presenting in an adult is a rare entity but should be kept as a differential in all age groups. While acute intestinal obstruction with features of peritonitis is a clear indication of emergency exploratory laparotomy, a similar approach in patients with subacute obstruction and stable vitals is not warranted.

Graziano et al recommend that consideration may be given to operating on asymptomatic patients who are younger, while observation may be appropriate in the older asymptomatic patient.[3] This being a level 3-4 evidence, grade C recommendation.[3] This was based on the following literature search (Table 1).[4-8]

Table 1: Present literature on indication and timing of surgical intervention in malrotation.

Study	Year	Conclusion
Cohen et al	2003	Surgical correction in all age groups
Spigland et al	1990	Surgical correction in all age groups
Malek et al	2005	Surgical correction in all age groups
Malek et al	2006	Ladd's procedure – justified in young; not justified in adults with malrotation for the rare occurrence of midgut volvulus
Prasil et al	2000	Surgical correction in all age groups

The majority of the published studies are in favour of surgical correction given the catastrophic effects of midgut volvulus that can happen. However, the study by Malek favours a conservative approach in the selective adult population since the incidence of midgut volvulus is rare in adults.[7]

A population-based study on adult small bowel volvulus carried out by Taylor et al in 2014 provides strong evidence that timely diagnosis and operative management should be utilized for the treatment of medically fit adult

patients presenting with small bowel volvulus (SBV) including those with SBV and intestinal malrotation.[9] Waiting for signs of peritonitis, intestinal ischemia, coagulopathy, and/or failure of nonoperative management should be avoided to improve outcomes.

In both our cases, patients presented with midgut volvulus yet an initial conservative approach was carried out. This denotes that midgut volvulus need not always require emergency exploratory laparotomy. An initial preoperative optimisation can be carried out to improve the postoperative outcome along with continued aggressive monitoring in cases with the absence of peritonitis, vascular insufficiency and pneumoperitoneum. In both our cases even when the obstruction was relieved on conservative management, midgut volvulus did not resolve. Purely conservative management was hence not possible in either of the cases. Definitive operative intervention is thus needed as early as possible after the initial optimisation of the patient even in an adult with low surgical risk.

The second case was associated with chronic calcific pancreatitis and the nutcracker phenomenon. Attributing recurrent pancreatitis to malrotation is an attractive hypothesis that remains unproven. We could find five published articles associating pancreatitis with malrotation. None of these cases was associated with a pancreatic anomaly.[10-14] Four out of five were treated with an elective Ladd's procedure. One case was treated conservatively as it was an incidental finding and was also associated with alcoholism. Similar to the last case our patient also had a history of alcoholism which could have been the cause of pancreatitis. But compared with the CT scan two months prior to presentation the pancreatic duct calcification was of recent onset. The patient was abstinent for six months prior to it. The patient had unresolved midgut volvulus for these 2 months. Ladd’s procedure was performed in view of midgut volvulus and not pancreatitis. We could find two case reports associating pancreatitis with malrotation but with obstruction due to intussusception and a phytobezoar.[15,16]

Association of nutcracker syndrome with malrotation is also very rare. We found three case reports with such an association.[17-19] Two out of three were symptomatic and were treated with Ladd's procedure and renal vein transposition. Since our patient was asymptomatic nothing specific was done for the nutcracker phenomenon. Weight loss might be contributing factor to the development of the new-onset nutcracker phenomenon in this patient.

Our second case highlights the fact that an adult patient with malrotation presenting with vague abdominal complaints can have multiple factors which can be the causative factor for his symptoms. Prompt diagnosis of the pathology and diagnosis of the primary aetiology for the pain is therefore important to relieve the symptoms of the patient.

Presence of midgut volvulus in both our patients necessitated an operative intervention. But in absence of volvulus, the need for operative correction needs to be individualised based on age, morbidity, associated pathologies and surgical risk factor of the patient.

CONCLUSION

Malrotation of the gut needs to be kept as a differential in cases of acute abdomen and patients with chronic abdominal complaints irrespective of age. Incidence of midgut volvulus in malrotation of gut decreases with increasing age. Hence elective Ladd's procedure is not justified in asymptomatic adult patients with malrotation. Midgut volvulus can present without intestinal obstruction. In patients with midgut volvulus with the absence of vascular insufficiency, preoperative optimisation followed by an elective Ladd's procedure can be attempted to decrease complications associated with emergency surgery. Adult patients with malrotation with chronic abdominal discomfort should also be investigated for other pathologies which may be the primary cause of the symptom. This is especially important in patients with the onset of symptoms in old age. Acute pancreatitis and the nutcracker phenomenon are two rare associations of malrotation of the gut. Evidence for a direct cause and effect relationship is lacking at present and needs further study.

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