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Case Series

Midgut malrotation: case series

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ABSTRACT

Mal rotation of midgut is associated with other anomalies usually encountered in neonatal period or early childhood. If undetected in childhood it presents in adulthood with small bowel obstruction, repeated appendicitis or chronic abdominal symptoms. CECT abdomen is mandatory for diagnosis though it often presents as surgical surprise on abdominal laparotomy. Hereby, we presented 3 cases where it was undetected till adulthood though CECT was mandatory for definitive diagnosis. Case 1 patient presented with chronic abdominal pain on left abdomen was actually malrotation with appendix lying on left hypochondrium and stenosed fourth part duodenum adding to vomiting off and on. Case 2 patient in adulthood presented with repeated sub-acute intestinal obstruction because of midgut mal-rotation. Relieved after Ladd band was cut and obstructive symptoms relieved. Case 3 patient had inflammed appendix in subhepatic position was cause of chronic pain with para duodenal hernial sac adding to intestinal obstruction with malrotation of midgut.

Keywords: Midgut, Malrotation, CECT, USG, Ladd

INTRODUCTION

Malrotation of midgut is encountered in paediatric age groups but usually comes as surgical surprise in adults. The incidence of rotational anomalies of the midgut is approximately 1 in 6000 live births.[1] The midgut comes out of coelomic cavity via umbilical ring at fourth month of embryonic development. The intestine begins rotation by tenth week of gestation and completed by twelve weeks. Reversal or arrest of normal process of rotation; results in the anomalies. Presentation of mal-rotation encountered in adults is rare with incidence ranging between 0.0001% to 0.19%.[2,3] Such low incidence in adults with added diagnostic dilemma adds to delay in diagnosis and management.[4] Adults intestinal mal-rotation often presents with chronic non-specific symptoms which are ignored or confused with other disease presentation includes; intermittent abdominal pain, vomiting, bloating, mal-absorption and alternating constipation and diarrhoea.[5-7] Gold standard for diagnosis remains an oral and intravenous contrast CECT with cross-sectional images, but in reality it is often encountered as intra-operative surprise although definitive management includes Ladd's procedure via open or laparoscopic approach.[8]

CASE SERIES

Case 1

A thirteen years female presented to the surgical emergency with acute pain left upper abdominal quadrant aggravated with meals associated with bilious vomiting of one day duration. The patient had history of bouts of multiple episodes of bilious vomiting in the past and gradually subsiding by itself. Clinically distended abdomen with tenderness and ill-defined suspicious lump in left hypochondrium was palpated. X-ray abdomen revealed a double air bubble sign. USG abdomen revealed left malrotated kidney with bilateral cortical echo. There was distended stomach, proximal duodenum with collapsed distal bowel loops. The jejunal loops were

clumped together in right upper abdomen. CECT showed whirlpool appearance along with twisted mesentery involving duodenum, proximal jejunum with dilated proximal stomach and duodenum. There was mal-rotation of duodenum and proximal jejunum with duodenum-jejunum flexure not crossing the midline. Exploratory laparotomy revealed stenosis at fourth part duodenum with caecum and ascending colon occupying left side abdomen. The appendix lying on left side was tense inflamed and distended with no obvious ischemic changes. Ladd's band was noted and small bowel was enclosed in a sac. Ladd's procedure performed with side to side duodeno-jejunal anatomosis. The patient recovered uneventfully in post-operative period and under regular follow up in surgical clinics with no obvious signs of obstruction or vomiting till date.

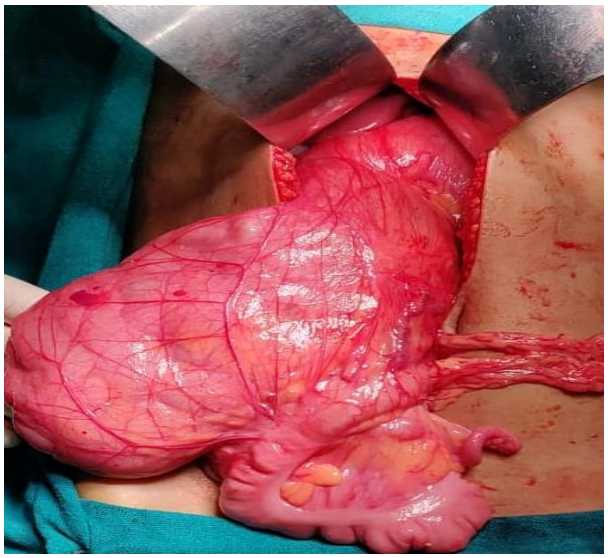

Figure 1: Mal rotation with small bowel content in sac.

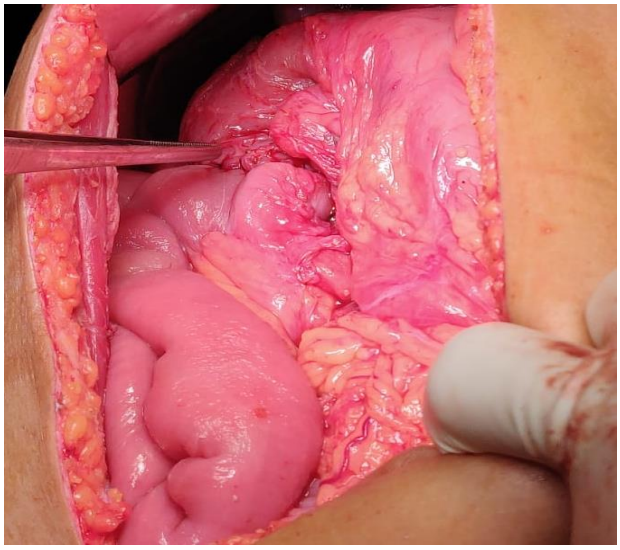

Figure 2: Appendix was tense, inflamed in (L) abdomen.

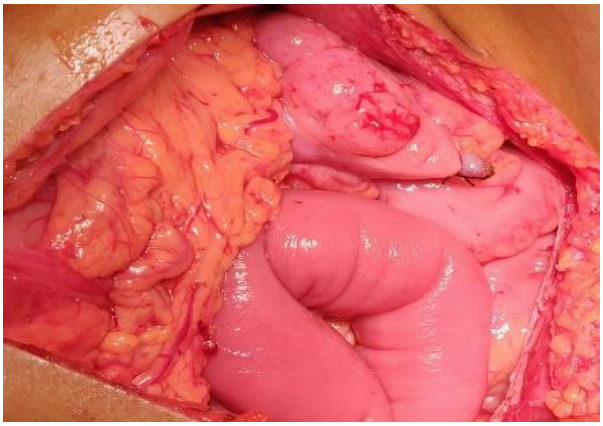

Figure 3: Ladd band excised with appendectomy.

Case 2

A 22 years male presented to the surgical emergency with pain right upper abdomen of 12 days and bilious vomiting of 10 days duration with features of complete intestinal obstruction of one day only. Past history revealed no tuberculosis or contacts at home nor previous surgical intervention. Clinically, abdomen was distended with tenderness in right hypochondrium, lumbar region with no obvious guarding or rigidity. Hyper peristaltic bowel sounds were present.

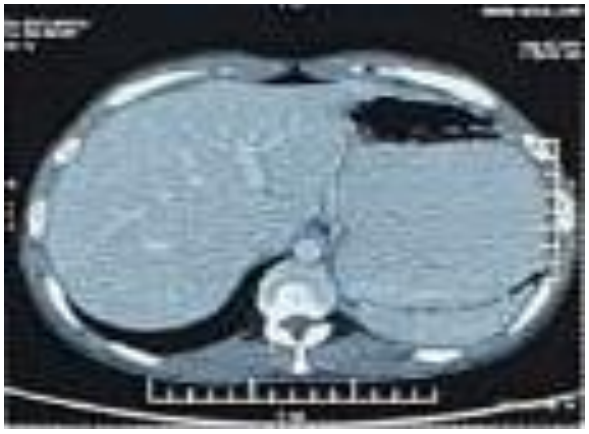

Figure 4: Distended stomach.

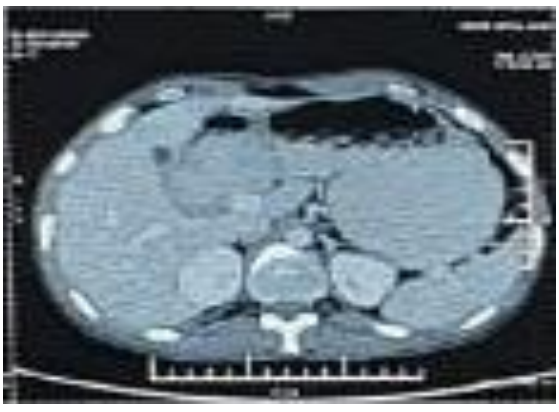

Figure 5: Distended stomach and duodenum.

X-ray abdomen showed multiple air fluids levels. The CECT abdomen revealed markedly dilated loop of duodenum in the right flank with suspicion of volvulus. The duodenum-jejunum transition appeared in midline with right sided rotation. Ileo-caecal junction was pulled up at right sub-hepatic region. On exploration grossly dilated stomach and duodenum was encountered. Duodenum was enormously dilated out of proportion with extending from right hypochondrium, lumber region and occupying right iliac fossa. Caecum, jejunum, descending colon and omentum were densely clumped together in left lumbar region. A band was noted arising from lateral abdominal wall to the duodenum which was excised and Ladd's procedure done. Bowel continuity established after adhesion-lysis with repositioning of bowel contents back. The patient was followed up and no signs of obstruction were noted.

Figure 6: Bowel loops clumped in (L) side.

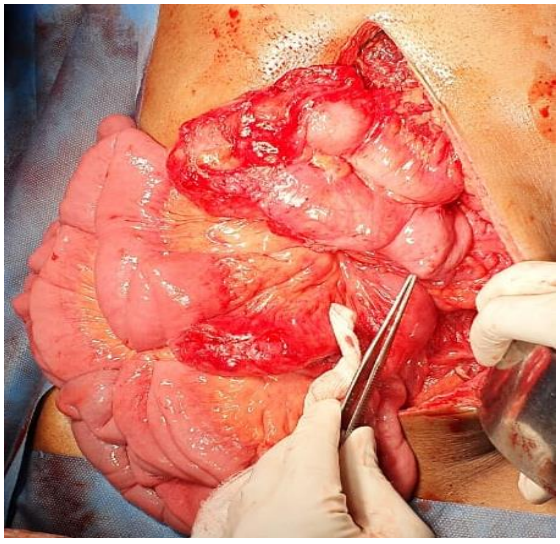

Figure 7: Band of Ladd compressing the bowel.

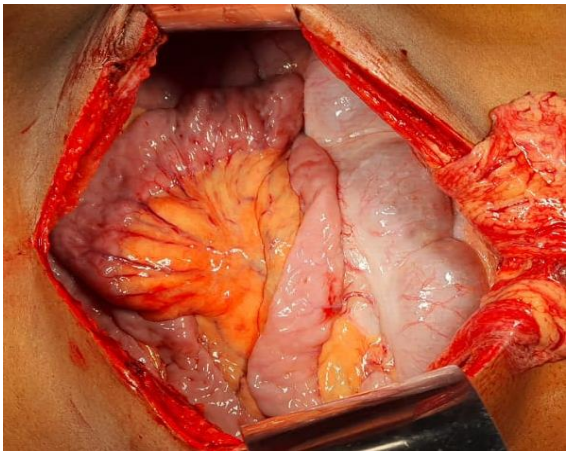

Figure 8: Caecum, jejunum, illium in (L) abdomen.

Case 3

A 17 years male presented to surgical emergency with acute intestinal obstruction with severe sharp cutting pain at umbilical and para umbilical region of one day duration. The patient has been experiencing mild dull tolerable pain in the same region for past 6 months. X-ray abdomen revealed dilated bowel loops with air fluid levels. CECT revealed abnormal bowel orientation with suspicion of paradoudenal hernia/internal hernia which was non-compressible adding to suspicion of obstruction. On exploration caecum and appendix were pulled up in sub hepatic region. Hernia sac was noted beneath the caecum extending to right iliac fossa opening through inferior illeocaecal recess. Whole of the jejunum with part of ileum was entangled forming content of hernia sac. Inter-bowel adhesions with multiple mesenteric lymphadenopathy were additional findings. The hernia sac was opened, reduction and repositioning of bowel contents done with excision of sac, adhesiolysis of inter-bowel loops done. Mesenteric lymph nodes proved non-specific granulomatous infection on histo-pathological examination. Post-operative period was uneventful. Patient is under regular follow up in surgical clinics.

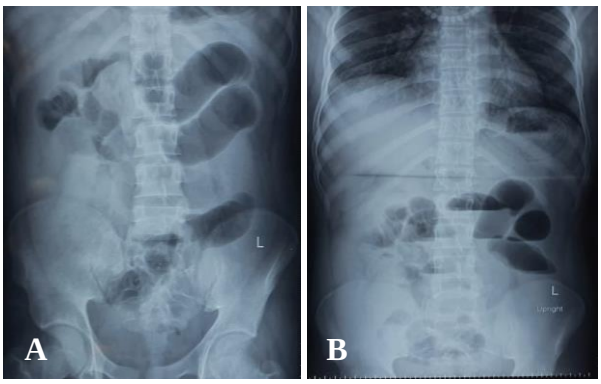

Figure 9: X-ray (A) Supine- with illeal loop obstruction; (B) Erect- abdominal with multiple air fluid level.

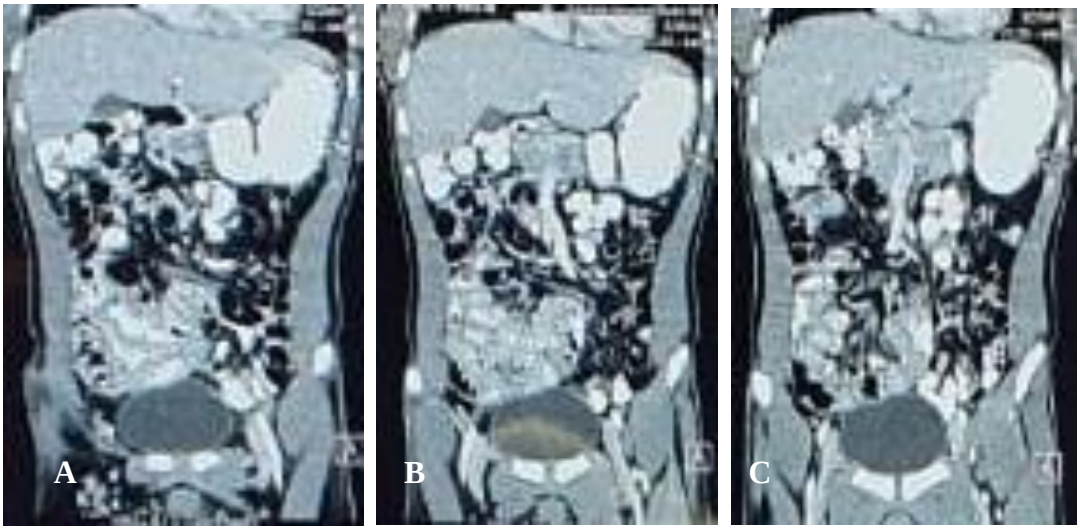

Figure 10: CECT (A) Distended stomach; (B) Caecum and appendix below liver; (c) Sac with bowel contents.

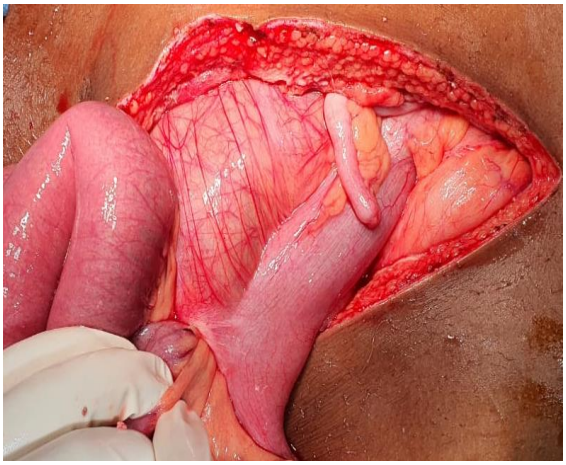

Figure 11: Inflamed appendix in subhepatic position.

DISCUSSION

Congenital anomalies of mid gut rotation are rarely encountered in adult age group.[9] Bilious vomiting is often the common presentation in neonates. Neonatal intestinal obstruction with bilious vomiting includes duodenal atresia, jejunum/ileal atresia, midgut malrotation and volvulus, necrotizing enterocolitis, and meconium ileus.[10] Acute obstruction or volvulus is common presentation, but incidental surgical surprise on laparotomy is very rare.

Adults presentations vary from asymptomatic to acute episode of intestinal obstruction requiring emergency surgical intervention with various spectrum of malrotation halted in utero. Since, duodenum is displaced from normal position to the superior mesenteric artery, the mesentery fails to get fixed to the posterior abdominal wall resulting in intestinal torsion. Adult's presentation may be asymptomatic or present with nonspecific symptoms. Two distinct patterns of adult presentations includes: acute and chronic.[11-13] Chronic cases may present as intermittent abdominal pain with nausea and vomiting over months. Since the abdominal symptoms are non-specific; high index of suspicion of midgut mal-rotation must be taken into account when investigating the cause of varied vague abdominal symptoms. The patho-physiology of these chronic symptoms relates to the compression effect of Ladd's bands running from the caecum and ascending colon to the right abdominal wall.[11,14] Acute presentation may be due to volvulus of the midgut or ileo-caecum resulting in obstruction or internal herniation caused by Ladd's bands. CT with or without contrast study is considered the investigation of choice. USG and CT scan may show the superior mesenteric vein and artery in anomalous position. It might show abnormal associated anatomy of the midgut. The shortened mesentery allows the small bowel and mesentery to twist and wrap around the narrowed SMA pedicle to create a distinctive 'whirlpool' appearance on CT scan.

Symptomatic midgut mal-rotation mandates definitive surgical intervention. Incidentally, discovered patients without symptoms and their management are controversial. Choi et al reviewed 177 patients over a 35-years period and reported that asymptomatic patients have a low risk of volvulus. Thus screening, elective surgery may not be necessary with close follow-up.[15] Several case series have recommended that elective Ladd's procedure should be performed in all patients with intestinal malrotation. The classical Ladd's procedure consists of 4 parts: division of Ladd's bands overlying the duodenum; (a) widening of the narrow root of the small bowel mesentery by mobilising the duodenum; (b) division of the adhesions around the SMA to prevent volvulus; (c) counter clockwise de-torsion of the midgut volvulus (if present) and (d) appendicectomy to prevent future diagnostic dilemma of an abnormally located appendix.[16] Procedure done by laparoscopy; an additionally benefit of minimal invasive surgery can be offered safely in the absence of volvulus. Both the patients have not developed any signs of obstruction upon follow up.

CONCLUSION

The diagnosis of mal-rotation in adulthood is difficult in view of vague symptoms mimicking many other disease profiles but should be suspected when symptoms are nonspecific yet persistent. Non-specific abdominal obstructive symptoms in adulthood presenting to surgical clinics at regular interval must be evaluated. Plain film abdomen and CECT abdomen is the minimal evaluation tool mandatory for diagnosis prior to diagnostic laparotomy by open or via laparoscope. Many times, undiagnosed cases may come as surgical surprise when laparotomy is done for other reasons. Asymptomatic or incidentally diagnosed patients of malrotation may be managed conservatively but regular follow up is mandatory.

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