



ADULT MIDGUT MALROTATION - A REPORT OF TWO CASES IN CONTRAST

General Surgery

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ABSTRACT

Intestinal malrotation is usually a disease of the pediatric population. This entity is rare in adults. It may present with a wide variety of symptoms ranging from acute abdominal emergency to non-specific and vague chronic gastrointestinal symptoms in adults. The chronic abdominal symptoms causes diagnostic dilemma with the older population because of its low incidence. We report two cases of intestinal malrotation. A 26 year old male presenting with vague gastrointestinal symptoms. The diagnosis of intestinal malrotation was established preoperatively with the help of radioimaging. Ladd's procedure was performed. This case emphasizes the importance of keeping high index of suspicion in patient of chronic abdominal pain for the diagnosis of malrotation in adult age group. The second case is that of asymptomatic gut malrotation in a 32 year old male who presented with acute abdomen due to peptic perforation. Peptic perforation in asymptomatic gut malrotation has not been reported in literature.

KEYWORDS

Adult midgut malrotation; intestinal malrotation; ladd's band; ladd's procedure, peptic perforation, acute abdomen

INTRODUCTION

Midgut malrotation is a congenital anomaly due to abnormal or incomplete rotation and fixation of the midgut during embryonic development. Its rate of occurrence is about 1 in 500 live births [1]. 60-85% cases present in the first month of life. About 90% of these patients are diagnosed in first year of life [2]. Rest of the cases are diagnosed in childhood, adulthood or even remain undiagnosed. Gastric perforation in association with intestinal malrotation in a newborn has been described and may be associated with mechanical obstruction and increased intraluminal pressure[10]. Adult midgut malrotation is very rare with prevalence of about 0.2%[3]. It usually remains asymptomatic throughout life[4]. When symptomatic it poses a diagnostic challenge because the incidence of adult malrotation decreases with increasing age. We report two cases of adult midgut malrotation, one who presented with vague abdominal symptoms and the other who presented with peptic perforation and acute abdomen.

CASE-1

A 26 year old male presented to us with recurrent attacks of epigastric and periumbilical pain since 18 months. Pain was associated with bilious vomiting. The pain usually subsided within 2 hours. This time however, pain and vomiting were continuous since 2 days. The patient is otherwise a healthy young man with no history of any chronic illness. His vital signs were within normal limits. Abdomen was distended with mild tenderness in the epigastrium. Tympanic note all over the abdomen on percussion. Rectum was empty. He was admitted for further evaluation. His routine hematological and biochemical profile including serum electrolytes was normal. Plain radiograph of chest was normal. Erect abdominal radiograph showed dilated jejunal loops. No abnormality was detected in ultrasound scan of abdomen. Upper GI endoscopy was done to rule out peptic ulcer disease which revealed dilated duodenum without any evidence of ulcer. Contrast CT abdomen revealed reverse relation of SMA & SMV. The stomach and duodenum (upto third part) was dilated suggesting obstruction due to midgut malrotation. 'Whirlpool' sign was present. In view of midgut malrotation and clinically increasing distension, exploratory laparotomy was performed. Intraoperatively, duodenum was found dilated. Duodenojejunal flexure was found to the right of midline with one clockwise rotation of small bowel around the SMV. No evidence of small bowel gangrene was seen. Ladd's procedure with cecopexy was performed. Post operative recovery was uneventful and patient was discharged on 6th postoperative day. Patient is asymptomatic at 6 months follow up.

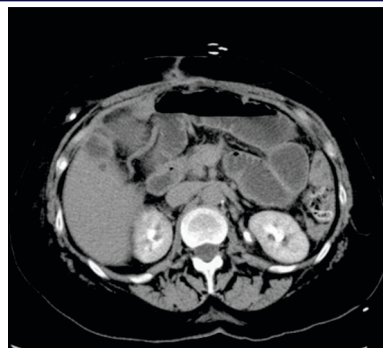


IMAGE 1: CT axial section showing dilated duodenum to the left of midline

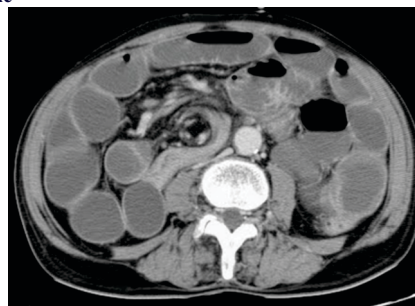


IMAGE 2: CT axial section showing "whirlpool" sign



IMAGE 3: Entero-enteric fibrous bands-Ladd's bands

CASE-2

A 34 year old male presented with sudden onset abdominal pain since last 24 hours. He had two episodes of vomiting associated with low grade fever. He had no significant past medical or surgical history. He was a cigarette smoker. On examination, he was febrile, had a pulse of 110 beats per minute, Blood pressure-100/70 mm Hg. Per abdomen showed features of peritonitis. Plain Chest radiograph revealed pneumoperitoneum. USG also showed free air in the peritoneal cavity, moderate free fluid with echoes strongly suggestive of hollow viscus perforation. After Resuscitation patient was taken up for an emergency laparotomy. Cecum was noted to be lying in the left hypochondrium. The relation between ileum and caecum was reversed. An 8 mm size perforation was noted in the anterior wall of stomach in antral region. Duodenojejunal flexure was to right of midline and several entero-enteric fibrous bands (ladd's) were noted. Midgut malrotation with gastric perforation was confirmed. After thorough peritoneal lavage, ladd's procedure with cecopexy, appendicectomy and Graham's omentoplasty was performed. Post-operatively, the patient showed steady improvement and was started on oral diet on 4th post-operative day. He was discharged in stable condition on 6th post-operative day. Patient is asymptomatic at 6 months follow-up.

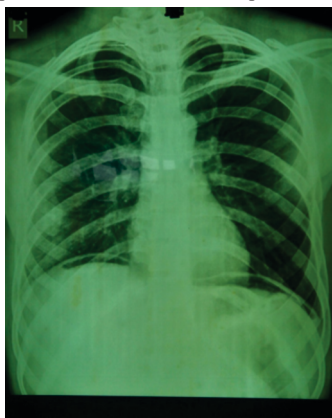


IMAGE 1: Plain chest radiograph showing pneumoperitoneum

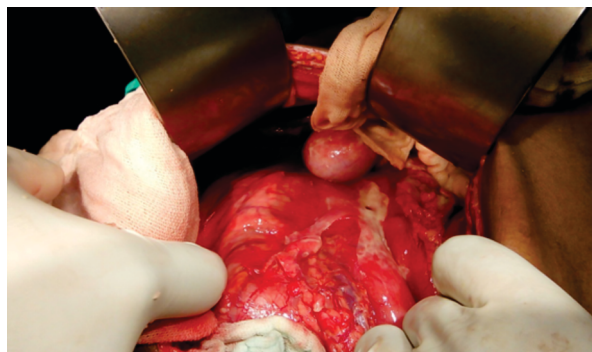


IMAGE 2: Intra-operative image of small gastric antral perforation

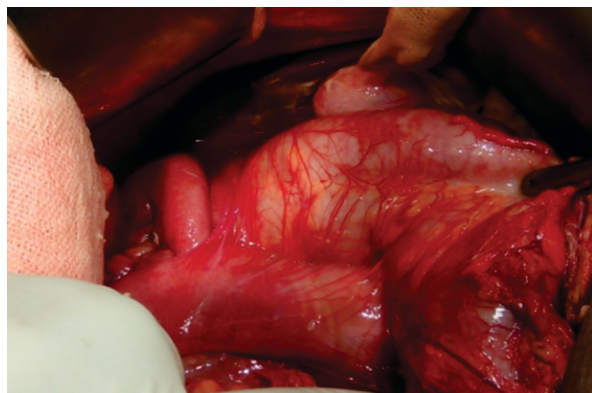


IMAGE 3: Caecum in left Hypochondrium and ileocecal relation reversed. Ileum entering the cecum from right to left. Plenty of entero-enteric band adhesions noted

DISCUSSION

Malrotation can be defined as any deviation from the normal 270 degree counterclockwise rotation of the midgut. The rotation of intestine in the developmental stage has been divided into three stages: Stage 1 occurs in weeks 5 to 10. It includes extrusion of the midgut into the extra embryonic cavity, a 90- degree counterclockwise rotation, and return of the midgut into the fetal abdomen. Stage 2 occurs in week 11 and involves further counterclockwise rotation within the abdominal cavity, completing a 270 degree rotation. This rotation brings the duodenal 'C' loop behind the SMA with the ascending colon to the right, the transverse colon

above, and the descending colon to the left. Stage 3 involves fusion and anchoring of the mesentery. The caecum descends, and the ascending colon and descending colon attach to the posterior abdomen. On this basis, Dott classified the various forms of intestinal malrotation. Stage 1 anomalies include exomphalos. Stage 2 anomalies include non-rotation, malrotation, and reverse rotation. Stage 3 anomalies include failure of retroperitonealisation (deficient fixation) [6].

Presentation of midgut malrotation in adult population may be acute or chronic in nature. Acute presentation manifests with acute intestinal obstruction due to midgut or ileocaecal volvulus. This type of presentation is very frequent in neonates but as the age increases, chronic presentation becomes more common. Chronic symptoms are more common in children older than 2 years of age [5]. It is characterized by abdominal colic, bloating, nausea, vomiting, constipation, diarrhoea and also symptoms mimicking irritable bowel syndrome, peptic ulcer disease and pancreatic disorders [7]. Physical signs, diagnostic of the condition are absent and patients are often labeled as functional. These chronic symptoms are due to compression by Ladd's bands [2,4].

Most of the patients of gut malrotation in adolescent and adult age are asymptomatic and are discovered incidentally when investigations or laparoscopy or laparotomy is performed for any other pathology as in our case-2.[4,8]. Diagnosis of the malrotation of gut is usually made by radiological investigations. Plain abdominal X-rays are not diagnostic of malrotation in adults and children although it may suggest the need for further studies or exclude other pathology. Upper GI contrast study is considered the gold standard for the diagnosis of gut malrotation with accuracy of over 80% [4]. It will show the duodenum and duodenojejunal flexure located to the right side of the spine.

Ultrasound with colour doppler study of the abdomen may reveal malposition of SMA, duodenal dilatation with distal tapering and fixed midline bowel and mesentery twisted around the SMA axis. These features classically present as 'whirlpool' sign [9].

CT scan with or without UGI contrast is becoming preferred investigation for children and adults with a diagnostic accuracy of about 80%. CT shows the characteristic whirl or whirlpool sign, positioning of SMV lying to the left or anterior to the artery and presence of right sided duodenojejunal flexure [2].

In case-1, presentation was chronic in nature. To rule out peptic ulcer disease, UGI endoscopy was done which showed duodenal dilatation. It prompted us to go for CECT abdomen with oral contrast which yielded the diagnosis of midgut malrotation.

Once diagnosis is made, surgical intervention (Ladd's procedure) is the standard therapy- both for the symptomatic as well as asymptomatic patients [5]. Ladd's procedure is being performed laproscopically more frequently nowadays and is comparable to open procedure when there is no acute volvulus.

Gut malrotation in adult is very rare. Only about hundred cases have been reported in last 80 years [5]. Manifestation is highly non-specific and patients may sometimes even be asymptomatic. High index of suspicion is needed to diagnose the problem. In other words, one may need to exclude gut malrotation in a patient of non specific abdominal symptoms before labelling them as functional.

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