A case of midgut malrotation presenting as subacute intestinal obstruction in an adult

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Keywords: Malrotation; ladd band; intestinal obstruction

Introduction
Midgut malrotation occurs due to variations of rotation and fixation of the intestine during fetal development. Over 90% of cases present during the neonatal period [1]. The reported incidence of adult midgut malrotation is between 0.0001% and 0.2% [1, 2]. Adults with intestinal malrotation present with atypical symptoms [1] requiring a high degree of clinical suspicion in diagnosis.

Case presentation
A 68 year old male presented with nausea and abdominal bloating of three months duration. His bowel habits were normal. Patient was previously well. Clinical examination was unremarkable except for a right sided uncomplicated direct inguinal hernia. Patient underwent upper gastrointestinal endoscopy (UGIE) twice after optimal preparation. Food particles were seen in the pyloric region. Scope could not be negotiated beyond the distal second part of the duodenum. Ultrasound scan demonstrated dilated 1st and 2nd part of the duodenum. Ultrasound scan demonstrated dilated 1st and 2nd part of the duodenum with to and fro movements. Upper gastrointestinal contrast studies demonstrated midgut malrotation (Figure 1).

Intravenous Iohexol, oral and rectal contrast enhanced Computed Tomography scan showed gross dilatation of 1st and 2nd parts of duodenum with wall thickening (Figure 2). No extrinsic compression was seen. Small intestine was not filled with contrast. A diagnostic laparoscopy was performed. Midgut was found to be malrotated. The 3rd part of the duodenum was atretic with multiple Ladd's bands. Fibrotic bands were noted also around caecum and ascending colon. Adhesions were surgically divided. A side to side diversion gastrojejunostomy was created with 60 mm endoscopic stapler device. Postoperative period was unremarkable. Patient was asymptomatic after 24 months follow up.

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Received: 03-03-2018 Accepted: 17-03-2018
DOI: http://doi.org/10.4038/sljvs.v36i1.8485

Discussion
The midgut rotates 2700 counter clockwise around the axis of superior mesenteric vascular pedicle during the embryonic life. In midgut malrotation, peritoneal fibrous bands known as Ladd's bands [3] fix the small intestine and undescended caecum to the posterior abdominal wall. These Ladd's bands compress the duodenum and can potentially cause duodenal obstruction [4]. It can present as acute or chronic intestinal obstruction [4]. However, adults commonly present with chronic intestinal obstruction, characterized by intermittent crampy abdominal pain, bloating, nausea and vomiting over several months or years [1, 2, 4].

Plain radiographs may show absence of stool filled colon in right lower quadrant [1, 5]. But plain radiographic evidence is neither specific nor sensitive [5]. Twisting of the intestine and the mesentery around the axis of the superior mesenteric artery may be detected ultrasonically as “whirlpool sign” [5]. Malposition of bowel loops can be accurately diagnosed by CT [1, 4, 5]. Nowadays, UGI contrast studies are increasingly used to diagnose midgut malrotation presenting with chronic intestinal obstruction [5].

Surgical division of these adhesion bands in infants, known as “Ladd's Procedure” was first reported in 1936 [3]. According to the current evidence, elective Ladd's procedure is considered as the gold standard in midgut malrotation presenting with chronic intestinal obstruction in adults [5]. Chronic intestinal obstruction due to intestinal malrotation which required diversion of the intestine as in our case are reported sparsely. Data on long term post-operative outcome between open and laparoscopic approach are limited in adults. However, there is increasing evidence to suggest that the Laparoscopic Ladd's procedure can be performed safely in selected patients without increasing short term complications [5].

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

References

Learning Points:
- Midgut malrotation rarely presents in adult life.
- Commonest presentation of midgut malrotation in adults is chronic intestinal obstruction.
- Computed tomography combined with upper gastrointestinal contrast studies can diagnose adult midgut rotation accurately.
- Open and laparoscopic Ladd's procedure is the treatment.