



NON ROTATION OF INTESTINE WITH RARE PRESENTATION AS CHRONIC UPPER ABDOMINAL SYMPTOMS IN AN ADULT - A CASE REPORT

Radiodiagnosis

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ABSTRACT

Intestinal malrotation is a rare congenital abnormality caused by disorganized development of the gut which occurs between the 4th and the 10th week of gestation. Malrotation abnormalities depending on the extent of rotation achieved, may be classified as nonrotation, incomplete rotation or reversed rotation. These usually present in the paediatric population with rare presentation in the adult age group where it may remain asymptomatic and be discovered incidentally or present with chronic abdominal pain or with complications. We describe a rare case of a 48 year old female who presented with recurrent upper abdominal symptoms without any other relevant clinical finding and was diagnosed to have nonrotation of the bowel on radiological investigations with the small bowel loops situated on the right side of the abdomen, the large bowel loops on the left side and reversal of the superior mesenteric vessels relationship. This case emphasizes the fact that the diagnosis of malrotation be kept in mind when evaluating an adult patient with chronic upper abdominal symptoms in the absence of other clinical clues for appropriate radiological investigation and further management.

KEYWORDS

Intestinal Nonrotation, Intestinal Malrotation, CECT, Ladd procedure, Volvulus, Intestinal obstruction

INTRODUCTION:

Intestinal malrotation is a congenital abnormal position of the bowel within the peritoneal cavity which occurs secondary to the arrest of normal rotation of any part of the intestinal tract. It is usually accompanied with abnormal bowel fixation by the mesenteric bands or absence of fixation of portions of the bowel and consequently is associated with increased risk of bowel obstruction and midgut volvulus. Rotational abnormalities are a recognised cause of obstruction in the paediatric age group, however these are infrequently seen in adults.[1] We present a rare case of intestinal non-rotation in an adult patient who presented with chronic upper abdominal symptoms with a description of the findings associated with the case, followed by a brief narrative of the embryology, its clinical associations and the management of such patients.

Case report:

A 48 year old female, presented with history of recurrent episodes of upper abdominal pain and vomiting since past 7-8 years. Pain was intermittent, mild to moderate in intensity, non-radiating and localized to the epigastric & the umbilical region. Vomiting was episodic, containing food particles without any blood. Clinically, there was no evidence of weight loss or abdominal distension. There was no change in the bowel habits. The general and systemic examination was unremarkable and bowel sounds were present on auscultation. Patient was being managed conservatively. There was no other significant past medical or surgical history.

Ultrasound (USG) of the abdomen was done which revealed large bowel loops to the left and the small bowel loops to the right of midline. Superior mesenteric vein (SMV) was located anterior to the Superior mesenteric artery (SMA) and hence suspicion of intestinal rotational anomaly was raised.

Contrast enhanced Computerized Tomography (CECT) of the abdomen was performed to confirm the findings which revealed:

- SMV located anterior to the SMA instead of the normal posterior relationship (Fig 1)
- Duodeno-Jejunal junction located to the right of midline, failing to cross the midline onto the left side (Fig 2).
- Caecum and Ileo-caecal junction located in the midline (Fig 3a).
- Entire small bowel located to the right of midline (Fig 3a and 3b).
- Majority of the large bowel loops to the left of midline (Fig 3a and 3b).

There was no evidence of bowel obstruction. There were no other associated abnormalities noted. The findings of CECT were consistent with non-rotation of the bowel.

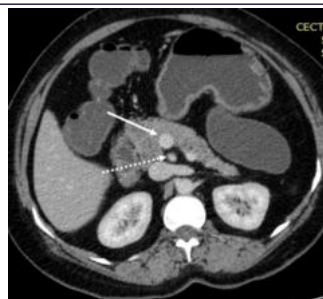


Fig 1: Axial CECT image showing SMV (solid arrow) anterior to the SMA (dotted arrow).

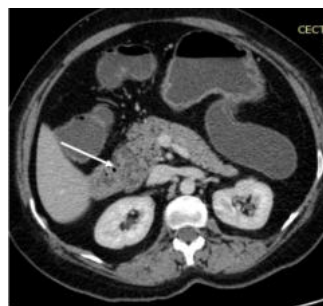


Fig 2: Axial CECT image showing duodeno-jejunal junction to the right of midline (solid arrow).

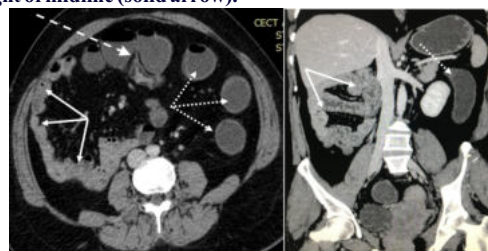


Fig 3: (a) Axial section and (b) coronal section showing small bowel loops to the right of midline (solid arrow), large bowel loops to the left of midline (dotted arrow) and midline ileo-caecal junction (dashed arrow).

The patient was given the option for surgical correction versus conservative management and explained about the future risk of volvulus and intestinal obstruction that may require surgical intervention. The patient opted to be managed conservatively than undergo surgical correction for the abnormality.

DISCUSSION:

The incidence of mid-gut rotational abnormalities is around 1 in 6000 live births. Approximately 30 percent of these cases present in the neonatal period, 60 percent by one year and over 75 percent by five years of age. Presentation in adulthood accounts for a meagre 0.2 to 0.5% of all cases. Of these presenting in the adult age group, about 15% present with midgut volvulus.[2,3]

These rotational abnormalities may also be associated with abdominal wall defects, polysplenia or asplenia, congenital heart disease, congenital diaphragmatic hernia, intestinal atresia's or complex anorectal malformations.[4,5]

Suitable knowledge of the composite steps involved in the embryology of the gastro-intestinal tract is essential in understanding the rotational abnormalities, briefly described as follows. Between the 4th and the 10th week of gestation, the gut undergoes a series of rotations. First the duodenum rotates 90° counterclockwise and comes to lie to the right of the SMA, while the colon rotates 90° and comes to lie to the left of the SMA. At 6 weeks the midgut herniates and duodenum rotates counterclockwise another 90°. At approximately 10 weeks the gut returns to the abdominal cavity, the duodenum completes its final 90° counterclockwise rotation and colon also rotates 180° counterclockwise so that the caecum and ileo-caecal junction come to lie in the right lower quadrant.[6,7] Based on the degree of rotation achieved, malrotation anomalies are classified as:

- Nonrotation - only the initial 90° rotation occurs with the failure of the subsequent counterclockwise rotation of the small gut. The small gut is ultimately located on the right side, while the colon confined mainly to the left side.
- Incomplete rotation – characterised by failure during any stage of the final 180° counterclockwise rotation of the small bowel and/or of the colon. Spectrum of findings vary from near normal to that of non-rotation.
- Reversed rotation - rarest of these, occurs when duodenum rotates clockwise rather than counterclockwise. Duodenum courses anterior to SMA and the colon (ascending and transverse) course posterior.

Peritoneal bands known as Ladd's bands with faulty fixation of the mesentery can be seen with all the above, predisposing to volvulus or intestinal obstruction.[8]

Most of the above anomalies are symptomatic during neonatal/infantile period and present with vomiting which may be bilious or nonbilious and abdominal distension. Hemodynamic instability, signs of peritonitis, hematochezia indicate presence of associated complications. Adults however may be asymptomatic or present with symptoms of intermittent and chronic abdominal pain with or without vomiting.[3] Hence diagnosing such a case in adults requires a keen inkling for the same and performing the appropriate investigation, keeping in mind the hemodynamic stability of the patient.

Abdominal radiography is useful essentially in the acute setting in aiding the diagnosis of intestinal obstruction and complications including pneumatosis intestinalis and pneumoperitoneum. Upper GI Barium studies in a case of non-rotation may show abnormal position of duodeno-jejunal junction and corkscrew appearance of the duodenum and proximal jejunum in setting of a volvulus. Contrast enema may reveal abnormal position of the cecum. USG is not the usual investigation of choice but may show inverse relationship of the superior mesenteric vein and artery. In the acute setting, 'whirlpool' sign caused by vessels twisting around the base of the mesenteric pedicle is a tell-tale sign of volvulus. CECT of the abdomen shows the findings required for diagnosing a case of nonrotation which include large intestine predominantly on the left side, small bowel to the right side of midline, reversal of SMA/SMV relationship and duodeno-jejunal junction to the right of midline. CECT abdomen is also useful in the acute setting and may show the level of obstruction, complications including volvulus and the associated whirlpool sign. Magnetic Resonance Imaging is only helpful in cases where ionizing radiation is contraindicated such as in pregnancy. [9-10]

Since nonrotation and other rotational abnormalities are difficult to diagnose; a high index of suspicion must be maintained both clinically and radiologically. Once diagnosed, asymptomatic adult patients may be managed conservatively while Ladd procedure using either the open or laparoscopic approach is the usual surgical line of management in these patients. Appendicectomy is a part of the surgery to avoid any mis-diagnosis later. [3,8]

CONCLUSION:

This rare case of a nonrotation of gut in an adult patient with chronic upper abdominal symptoms reinforces the importance of keeping a high level of suspicion for this condition in adult patients with chronic symptoms. Early recognition and management of such rare abnormalities is imperative since this anomaly predisposes the patient to an increased risk of developing bowel obstruction or/and volvulus. Timely radiological diagnosis and intervention can reduce the patient morbidity and mortality.

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