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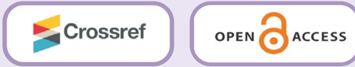

Case Report

A case of Total Non-rotation of gut - A case Report

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Abstract

Around one in every 500 infants exhibit intestinal malrotation. It is a congenital abnormal position of the bowel within the peritoneal cavity due to failure of or improper rotation of the gut tube in embryonic life. Classic clinical presentation of malrotation in a newborn is bilious vomiting with or without abdominal distention. In paediatric patients the typical presentation is bilious vomiting patients. Here we report a case of a four year old male child with complaints of recurrent vomiting and constipation since infancy. Though malrotation mainly presents in infancy, delayed diagnosis is possible. The importance of radiologic investigation is discussed in the context of avoiding potentially fatal intestinal ischemia.

Introduction

Intestinal malrotation is defined by a congenital abnormal position of the bowel within the peritoneal cavity, involving small and large intestines and the duodenojejunal junction.[1] It might result in a narrow-based mesentery-related mid-gut volvulus.[2] A volvulus is an axial rotation or twisting of a segment of the bowel around the mesentery. The rotation obstructs the lumen and if tight enough also causes vascular occlusion in the mesentery.[3] It is critical to rule out this diagnosis when a baby vomits bile because, if left untreated, mid-gut infarction develops very quickly.[2] Around one in every 500 live-born babies exhibit malrotation.[1] However, it is significantly more prevalent as an anatomical entity, occurring in 0.2% – 1% of the general population.[2] In the 1950s and 1960s, the mortality rate for affected neonates was around 30%, but it has subsequently sharply declined to between 3 to 5%. In older children and adults, malrotation and midgut volvulus can be present.[1]

Case Report

A four year old male child was brought to the Paediatrics OPD of North Bengal Medical College and Hospital with complaints of recurrent vomiting and constipation since infancy. He had similar episodes of such events in the past and was treated symptomatically by a local physician. On examination the general survey and systemic examination were unremarkable. An ultrasonography (USG) whole abdomen was ordered. The USG revealed an altered relationship between the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) with the artery lying to the right of the vein. There was a swirling appearance of the mesentery and superior mesenteric vein around the superior mesenteric artery. There were a few proximally dilated loops of the small gut. A barium meal follow-through (BMFT) was ordered. BMFT revealed the corkscrew appearance of the duodenum and jejunum. Duodenojejunal flexure was located below and right of the midline. Jejunal and ileal loops were identified on the right side with almost no small gut loop to the left of the midline. On lateral films, there was an overlap of the second and fourth parts of the duodenum. A diagnosis of total nonrotation of the gut with midgut volvulus was made. The patient was managed

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by the paediatric surgery department and underwent Ladd's procedure. The findings were confirmed intraoperatively. The immediate postoperative period was uneventful. The patient is asymptomatic and under regular follow-up to date.

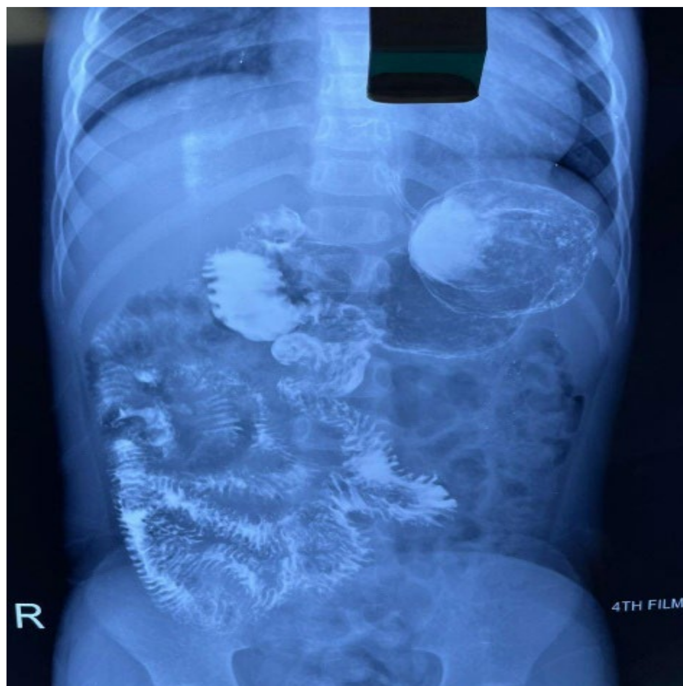

Figure 1: Barium meal follow-through double contrast study of the abdomen showing stomach and proximal duodenum in its normal expected position with D2 segment of duodenum showing corkscrew appearance.

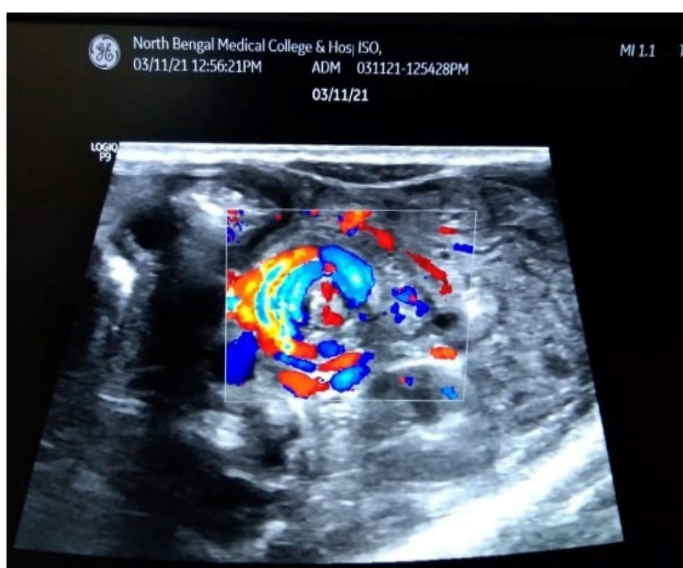

Figure 2: Whirlpool sign demonstrating the altered relationship between SMA and SMV

Discussion

Before four weeks of gestation, the gut is a short, straight tube. The duodenum, jejunum, ileum, and colon are forced to herniate into the umbilical cord as the straight tube starts to elongate more quickly than the embryo during the fourth to fifth week. The duodenum first turns 90 degrees anticlockwise, moving to the right of the SMA, while the colon undergoes 90 degrees rotation to rest at the right of the artery. Over the next three weeks, the duodenum continues to rotate anticlockwise so that, by the end of eight weeks, it has completed another 90 degrees while the colon undergoes no rotation. The embryonic bowel returns to the abdominal cavity at 10 weeks and the colon now rotates 180 degrees anticlockwise, placing the cecum in the right lower quadrant of the abdomen, while the duodenum completes its last 90 degrees of anticlockwise rotation, positioning the duodenojejunal junction to the left of the spine.[1,4] This mechanism fails in classical intestinal malrotation; the caecum is in the center and the duodenojejunal flexure is to the right of the midline, making the small bowel mesentery's base narrow, increasing the risk of midgut volvulus.[3] The section of the midgut's mesenteric attachment that runs from the duodenojejunal junction to the cecum is unusually short in those with malrotation. The gut is therefore prone to twist around the SMA and SMV. This may cause intermittent abdominal distention and pain or acute bowel necrosis.[1]

Bilious vomiting in a newborn infant is the hallmark symptom of malrotation, and until proven differently, malrotation should be assumed in any kid who experiences bilious vomiting. Early diagnosis (or exclusion) of malrotation is the aim of first bowel imaging to avoid volvulus and potentially fatal intestinal ischemia.[4] Malrotation mainly presents in infancy but delayed diagnosis like in our case is possible through radiology, which is easy and cheap.

The upper gastrointestinal series (UGI) is the imaging test of choice to diagnose or exclude malrotation and midgut volvulus. Normally, the duodenojejunal junction is located at the level of the ligament of Treitz to the left of the spinal pedicle. A rotational disorder is present when the duodenojejunal junction is not in this location. A controlled injection of barium or iodinated contrast is made into the stomach to avoid overfilling.[5,6] Performance of a UGI in the presence of malrotation with a volvulus reveals a corkscrew appearance or coiled spring appearance as the contrast passes into the distal duodenum and proximal jejunum. There may also be a bird-beaked appearance at the level of the obstruction from luminal narrowing.[6] The false positive rate of this test may reach 15%, and a false negative rate of 3 - 6%.[1]

On ultrasound imaging, malrotation is detected when the normal relationship between the SMA and SMV is reversed.[5]

The literature suggests ultrasound imaging can be useful in screening for malrotation, but its efficacy has been questioned.[7] "Whirlpool" sign (WS) corresponds to a clockwise wrapping of the superior mesenteric vein and the mesentery around the superior mesenteric artery. The WS should be routinely researched at all ages of the paediatric population because it is sensitive (81%) enough for its diagnosis.[8,9] Infants presenting with bilious vomiting must undergo USG for studying the SMA-SMV relationship and a barium study to rule out the diseases of malrotation.

Conclusions

Malrotation is a relatively common anomaly that can lead to life-threatening volvulus and bowel ischemia. Most cases of intestinal malrotation in infancy can be treated successfully with early diagnosis and intervention. Malrotation and midgut volvulus may also manifest in older children and adults where the symptoms are generally vague chronic abdominal pain and vomiting. The corkscrew appearance of the distal duodenum and proximal jejunum is indicative of the presence of malrotation with volvulus. Ultrasonography can be used for screening. Ladd's procedure is performed for the correction of malrotation. After surgical therapy most patients are healthy, and the recurrence rate is low.

References

1. Applegate KE, Anderson JM, Klatte EC. Intestinal malrotation in children: a problem-solving approach to the upper gastrointestinal series. Radiographics. 2006 Sep;26(5):1485-500.
   DOI:10.1148/rg.265055167
   PMID:16973777

2. Adams SD, Stanton MP. Malrotation and intestinal atresias. Early Hum Dev. 2014 Dec 1;90(12):921-5.
   DOI: 10.1016/j.earlhumdev.
   PMID: 25448782

3. O'Connell PR, McCaskie AW, Sayers RD. Bailey & Love's Short Practice of Surgery. 28th ed. CRC Press, Taylor & Francis Group; 2022.
   DOI:10.1201/9781003106852

4. Langer JC. Intestinal rotation abnormalities and midgut volvulus. Surg Clin North Am. 2017 Feb 1;97(1):147-59.
   DOI:10.1016/j.suc.2016.08.011
   PMID: 27894424

5. Applegate KE. Evidence-based diagnosis of malrotation and volvulus. Pediatr Radiol. 2009 Apr;39(SUPPL.2).
   DOI:10.1007/s00247-009-1177-x
   PMID: 19308378

6. Morris G, Kennedy A, Cochran W. Small bowel congenital anomalies: a review and update. Curr Gastroenterol Rep. 2016 Apr;18:1-2.
   DOI:10.1007/s11894-016-0490-4
   PMID: 26951229

7. Binu V, Nicholson C, Cundy T, Gent R, Piotto L, Taranath A, et al. Ultrasound imaging as the first line of investigation to diagnose intestinal malrotation in children: safety and efficacy. J Pediatr Surg. 2021 Dec 1;56(12):2224-8.
   DOI:10.1016/j.jpedsurg.2021.04.009
   PMID: 34030880

8. Esposito F, Vitale V, Noviello D, Di Serafino M, Vallone G, Salvatore M, et al. Ultrasonographic diagnosis of midgut volvulus with malrotation in children. J Pediatr Gastroenterol Nutr. 2014 Dec 1;59(6):786-8.
   DOI:10.1097/MPG.0000000000000505
   PMID: 25023580

9. Pracros JP, Sann L, Genin G, Tran-Minh VA, Morin de Finfe CH, Foray P, et al. Ultrasound diagnosis of midgut volvulus: the "whirlpool" sign. Pediatr Radiol. 1992 Apr;22:18-20.
   DOI:10.1007/BF02011603
   PMID:1594304