Midgut malrotation with volvulus - a case report

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Abstract

Midgut malrotation is a rare anatomic anomaly that complicates the diagnosis and management of acute abdominal pain. It is a congenital anomaly that arises from incomplete rotation or abnormal position of the midgut during embryonic development. A two years old boy was brought with recurrent abdominal pain and vomiting. On ultrasonography, the intestinal malrotation was suspected and thus contrast enhanced computed tomography (CECT) was done. On CECT, abnormal relationship between the superior mesenteric artery (SMA) and superior mesenteric vein (SMV), colon on the left side of the abdominal cavity, ‘Whirlpool sign’ etc., were found. All findings were suggestive of midgut malrotation with volvulus. Detection of uncomplicated malrotation should not be trivialized because such patients might experience a future complication.

Key words: faulty rotation, fixation, mesenteric bands.

Introduction

Malrotation and midgut volvulus is one of the major paediatric surgical emergencies. Delay in diagnosis can result in infarctive necrosis of the entire small bowel and is potentially fatal. Malrotation is an abnormal position of the bowel within the peritoneal cavity due to faulty rotational or fixational evolution of the gut, usually involving both the small and large bowel. Abnormal bowel fixation by mesenteric bands (Ladd’s bands) or absence of fixation of portions of the bowel leads to increased risk of bowel obstruction with or without volvulus.

Midgut malrotation has been estimated to occur in approximately one in 500 live births. However, it is difficult to ascertain the true incidence because this condition will go undetected during childhood in a substantial subset of patients. Reportedly, 64-80% of patients present within the first month of life, but these figures generally do not take into account those who remain undiagnosed after childhood.

Detection of uncomplicated or quiescent malrotation should not be trivialized because no reliable means exist to predict which patients might experience a future complication. Furthermore, some patients with ostensibly asymptomatic malrotation may, in fact, relate a history of episodic abdominal pain or vomiting.

Case report

A two years & ten months old boy was brought with presenting symptoms of recurrent abdominal pain and vomiting. On ultrasonography, the intestinal malrotation was suspected and evaluated further. A CECT was performed and serial axial sections of abdomen were studied with and without the use of Intra Venous(IV) contrast. Bowel loops were opacified with prior use of oral and rectal contrast. The following observations were made:

1. Midgut malrotation with volvulus were noted.
2. There was abnormal relationship between the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) with SMV located to the left of SMA. (fig. 1)
3. There was swirling of superior mesenteric vessels. (fig. 2)
4. 'Whirlpool sign' was noted suggestive of intestinal volvulus. (fig. 2)

5. The large bowel loops were located in the left side and small bowel loops in the right side of the abdominal cavity. (fig. 3)

6. Other organs like liver, pancreas, kidneys, spleen, urinary bladder were normal except gall bladder was distended.

7. The duodeno-jejunal junction was seen abnormally low on the left side. The rest of the distal jejunum and ileal loops were located to the right of the spine. (fig. 4)

8. Major vessels were grossly normal.

Discussion

The typical findings of midgut malrotation are:

- A duodeno-jejunal flexure in an abnormal position (usually to the right of the midline but occasionally just to the left).
- Obstruction of the second or third part of the duodenum by the Ladd’s bands.
- A small bowel volvulus if present.
- The small bowel lying entirely on the right side of the abdomen and
- An abnormally positioned caecum.

In the present study duodeno-jejunal flexure is located just left to midline, small bowel volvulus is present and the small bowel lies on the right side of the abdomen.

The normal anatomical position of the SMV is on the right side of the SMA, but sometimes also in front. When the SMV is located on the left side of SMA, intestinal malrotation is frequently associated with complete absence of embryological rotation of the primitive intestinal loop (non rotation). The malposition of SMV and SMA is not only specific of midgut malrotation, but can also be included with tumors or tumor like lesions.

The classic presentation of malrotation is of bilious vomiting within the first year of life, usually within the first month, but symptoms may present at any age and the diagnosis should always be considered. In older children, intermittent obstruction can occur, with chronic or recurrent abdominal pain and vomiting. Occasionally, a malabsorption syndrome results from chronic venous and lymphatic obstruction.

Deviation from the normal relationship between the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) is a useful indicator of malrotation.

Midgut volvulus is a complication of malrotation in which clockwise twisting of the bowel around the SMA axis occurs because of the narrowed mesenteric attachment. This life-threatening condition is a clear indication for emergent surgery.

Intestinal malrotation is usually an isolated abnormality but can be associated with congenital heart disease or situs problems.

A midline abdominal mass of whirling small intestinal loops and the whirlpool pattern of the superior mesenteric vein around the superior mesenteric artery in midgut volvulus is termed as the "Whirlpool Sign". Imaging findings in cases of midgut malrotation, is so specific that in the appropriate clinical setting, familiarity with the ultrasound and CT findings of "Whirlpool sign may establish the specific preoperative diagnosis before the occurrence of serious and fatal complications.

In 1992, Dufour et al examined over 300 patients with ultrasonography. Finding the superior mesenteric vein (SMV) to the left of the SMA was highly suggestive of malrotation, while an anterior SMV was questionable.

Children with congenital abnormalities of the abdominal wall (exomphalos, gastroschisis and diaphragmatic hernia) have some degree of malrotation but rarely develop clinical symptoms.
Embryological Significance

At approximately six week of gestation the duodenoejejunal and ileococcal segments of the primitive gut herniate into the extraembryonic coelom in the umbilical cord. Both loops elongate and rotate 270° anticlockwise around the axis of the superior mesenteric artery. By the end of the third month of gestation the bowel loops are returned to their final positions in the abdominal cavity, with their mesenteries becoming fixed to the parietal peritoneum at several sites. The duodenal loop is fixed with the duodenoejejunal junction (DJJ) in the left upper quadrant at the ligament of Treitz and the ileoacccal junction fixed in the right lower quadrant. The normal small bowel mesentery therefore has a broad diagonal base across the abdomen.

Any arrest in the normal 270° anticlockwise rotation occurring during physiological umbilical herniation results in malrotation and malfixation of the small bowel. The duodenojejunal junction will be displaced medially and inferiorly and/or the caecum will be displaced medially and superiorly. The length of the small bowel...
mesentery is consequently shortened and the risk of the entire small bowel twisting on its narrow pedicle is increased.

The surgical management of malrotation is the Ladd's procedure. Any volvulus is reduced and peritoneal bands are divided. The small bowel is returned to the right side of the abdomen and the large bowel to the left.

**Conclusion**

Midgut malrotation with volvulus is a surgical emergency and there may be dreaded consequences if it is neglected. The radiologist may encounter this important diagnosis in several different clinical settings such as an incidental imaging finding, the cause of acute abdominal symptoms etc.

**References**


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