Isolated Hindgut Malrotation: A Rare Variant of Intestinal Malrotation

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INTRODUCTION

Malrotation is a congenital abnormal position of the bowel in the peritoneal cavity and may involve both the small and large bowel or either. This could be accompanied by abnormal bowel fixation by mesenteric bands or absence of fixation of portions of the bowel leading to increased risks of bowel obstruction, volvulus, and bowel ischemia.1,2

The term malrotation applies to a wide range of intestinal anomalies, from a readily apparent omphalocele in newborns to asymptomatic nonrotation of the large and small bowel in adults. Most people with malrotation may show clinical signs of the condition soon after birth; however, many times this condition may be diagnosed long after infancy.3,4

Malrotation limited to the colon is much rarer than that of the small bowel alone or that of both the small and the large bowel together.5,12

In this article, we present two cases of isolated inferior mesenteric artery (IMA), i.e., hindgut malrotation with normal rotation of small bowel with symptoms unrelated to this malrotation.

CASE REPORT

Case 1
A 12-year-old boy who presented with dull aching abdominal pain particularly in periumbilical location with loose motions for a month with intermittent mild fever. On examination, no specific diagnosis was made. On investigation, he was anemic, no other biochemical abnormality was found.

He was then referred to our department with a provisional diagnosis of small bowel diarrhea. Ultrasonography (USG) was done and no significant abnormality was found. A computed tomography (CT) enterography was then performed.

The CT enterography was performed on a 64-detector row CT scanner (Brilliance CT; Philips, Best, the Netherlands) with 300 ml of 20% mannitol in 3 L of plain water administered orally. 70 ml of intravenous non-ionic iodinated contrast material (300 mg I/ml, Omnipaque: GE Healthcare, Shanghai).

The CT enterography revealed sigmoid and descending colon located on the right side of the abdomen (Figure 1a and b).
Cecum and ascending colon were also on the right side posterior to the descending colon (Figure 1b). IMA was seen coursing to right side of the midline, instead of to normal left sided course (Figure 2a and b).

Superior mesenteric artery-superior mesenteric vein (SMA-SMV) axis appeared normal (Figure 3a). Duodenoejunal (DJ) flexure was on the left (Figure 3a) and proximal jejunal loops were on left side of abdomen (Figure 3b). No abnormal bowel dilatation was noted.

In addition to abnormal location of distal bowel, there was mild enhancing circumferential wall thickening involving the sigmoid colon and the rectum with few enlarged pericolonic lymphnodes suggesting infective-inflammatory etiology.

Patient’s symptoms resolved with the antihelminthic and antibiotic course.

Case 2
A 25-year-old male who presented with dull aching abdominal pain, abdominal distension, and intermittent mild fever for 15 days. On examination, no specific diagnosis was made. On No biochemical abnormality was found except mildly raised erythrocyte sedimentation rate.

He was then referred to imaging department for USG. USG revealed no significant abnormality. Then, we decided to go ahead with CT.

CT abdomen and pelvis was performed on a 128-detector row CT scanner (GE Optima 660) with 30 ml of oral contrast in 2 L of plain water administered orally. 70 ml of intravenous non-ionic iodinated contrast material (300 mg I/ml, magnapaque).

CT showed that the sigmoid and descending colon were on the right side in right iliac fossa and right lumbar region (Figure 4a and b) with IMA on right side (Figure 5a and b). SMA-SMV axis appeared normal (Figure 5c). DJ flexure was to the left (Figure 5c) and proximal jejunal loops were on the left side of abdomen. No abnormal bowel dilatation was noted.

There was short segment homogenously enhancing mild wall thickening of rectum, cecum and mid ascending colon (approximately measuring 5 mm) s/o infective-inflammatory etiology.

In both cases, sigmoid and descending colon was on the right with IMA coursing on right side. However, Case 1 had cecum posterior to the descending colon (Figure 1b) and Case 2 had it anteriorly (Figure 4b). Both cases present with symptoms unrelated to this variation.

**DISCUSSION**

The development of the intestinal tract is a complex process. In 1898, Mall first described the embryology of malrotation. Frazer and Robbins described the process of rotation and fixation in three stages.

Stage 1: Physiological umbilical herniation at 5-10 weeks. At 6th week, the herniating bowel rotates 90° anticlockwise around the SMA.

Stage 2: There is a reduction of midgut hernia back into abdomen at 11th week with 180° anticlockwise rotation such that duodenum goes inferior and posterior to the SMA. The colon goes anterior to the SMA with the cecum located to the right and subhepatic in position.

Stage 3: The subhepatic cecum descends into the right iliac fossa of the abdomen at 12th week forming ascending colon and fixation of intestine to posterior abdominal wall.
If anomaly occurs in Stage 1, it results in omphaloceles caused. If it occurs in Stage 2, there is non-rotation, incomplete rotation, reverse rotation, internal hernias and in Stage 3 there is subhepatic cecum, unattached duodenum, and mobile cecum. If anomaly occurs in Stage 1, it results in omphaloceles caused. If it occurs in Stage 2, there is non-rotation, incomplete rotation, reverse rotation, internal hernias and in Stage 3 there is subhepatic cecum, unattached duodenum, and mobile cecum. Stinger classified several types of malrotation according to the embryologic state of development. Ia - Nonrotation of the colon and duodenum IIa - Nonrotation of the duodenum only IIb - Reversed rotation of duodenum and colon Ilc - Reversed rotation of the duodenum IIIa - Nonrotated duodenum and colon IIIb - Incomplete fixation of the hepatic flexure IIIc - Incomplete attachment of the cecum IIIId - An internal hernia near the ligament of Treitz.

Both of our cases have isolated IMA, i.e., hindgut malrotation with normal SMA, SMV relation, normal position of DJ flexure and normal position of cecum. These atypical malrotations should be managed on case by case basis. Resection should be reserved for obstructive cases.

CONCLUSION

Intestinal malrotation involving midgut is much common incidental finding on imaging, but isolated hindgut malrotation is a very rare. Abnormal position of bowel if seen, then their corresponding arterial course should be looked at and must be reported.

REFERENCES


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