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"MALROTATION OF MIDGUT" EMBRYOLOGICAL BASIS AND ITS CLINICAL SIGNIFICANCE

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ABSTRACT
The object of this communication is to call attention to the practical importance of knowledge of abnormalities of the midgut. Intestinal malrotation refers to the partial or complete failure of rotation of midgut around the superior mesenteric vessels in the embryonic life. Arrested midgut rotation results due to narrow based mesentry and increases the risk of twisting midgut and subsequent obstruction and necrosis.

Introduction

The midgut extends from the apex of the duodenal loop, which is fixed to the large liver primordium via the bile duct, to the last third of the transverse colon. Parts are: Inferior part of the duodenum with the Duodenojejunal bend, Jejunum, Cecum with vermiform appendix, Ascending colon, and Transverse colon [1-5]. The midgut is supplied with blood by the superior mesenteric artery and innervated by the vagus nerve (CN X). Within the whole midgut and rectum unit there exists only one dorsal mesenteries, the ventral being reabsorbed. Differentiation occurs in a cranio caudal sequence within a time window of roughly. Congenital malrotation of the midgut often presents within the first month of life. Pediatric radiologists are, therefore consciously attuned to this malady and its associated imaging features. The overall incidence of malrotation however is unknown because some patients will present years later or remain asymptomatic for life. Because presentation is nonspecific and the index of suspicion for malrotation progressively decreases in the older population, the clinical diagnosis is usually not considered in the initial evaluation [6-10]. At least some of the surgical literature, however, seems to favor surgery for malrotation regardless for patient age. This recommendation further underscores the importance of recognizing this unsuspected diagnosis on imaging. We review the imaging features of malrotation in adolescents and adults in the context of various clinical sceneries in which it may be encountered. Abdominal CT findings will be emphasized because abdominal CT is a frequent means of detection in patients with malrotation.

Incidence

Malrotation occurs in approximately 1 in 500 live births. Approximately 90% of patients with malrotation w are diagnosed within the first year of life [11-13]. Recurrent bowel obstruction in patients with previous abdominal operation for midgut malrotation is mostly due to adhesions but very few reported cases have been due to recurrent Volvulus [14-18].

Ontogenesis of the normal rotation of midgut

The midgut loop lies outside the abdominal cavity of the embryo, in a part of the extra-embryonic coelom that persists near the umbilicus [19-22]. The loop has a pre-arterial or proximal, segment and post-arterial, or distal, segment. Initially, the loop lies in the sagittal plane, its proximal segment being cranial and ventral to the distal segment. The midgut loop now undergoes rotation. This rotation plays a very important part in establishing the definitive relationships of the various parts of the intestine. The steps of the rotation must, therefore be clearly understood [23-27].

a. Viewed from the ventral side the loop undergoes an anticlockwise rotation by 90º, with the result that it now lies in the horizontal plane. The pre-arterial segment comes to lie on the right side and the post-arterial segment on the left [28-30]

b. The pre-arterial segment now undergoes great increase in length to form the coils of the jejunum and ileum. These loops still lie outside the abdominal cavity, to the right side of the distal limb.

c. The coils of jejunum and ileum (pre-arterial segment) now return to the abdominal cavity. As they do so, the midgut loop undergoes a further anticlockwise rotation.

As a result, the coils of jejunum and ileum pass behind the superior mesenteric artery into the left half of the abdominal cavity. The duodenum, therefore, comes to lie behind the artery and the coils of jejunum and ileum occupy the posterior and left part of the abdominal cavity [31-33].

a. Finally, the post- arterial segment of the midgut loop returns to the abdominal cavity. As it does so, it also rotates in an anticlockwise direction within the result that the transverse colon lies anterior to the superior mesenteric artery, and the caecum comes to lie on the right side. Note that all rotation has taken place in an anticlockwise direction [34].

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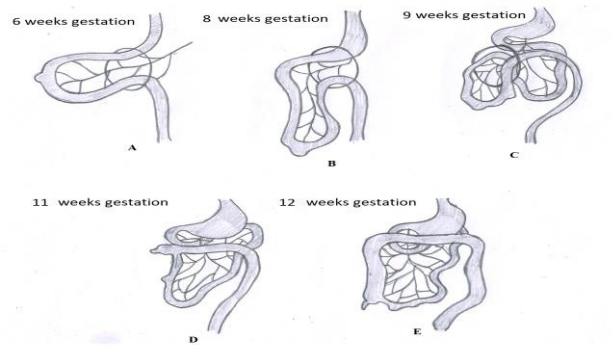
b. At this stage the caecum lies just below the liver, and an ascending colon cannot be demarcated. gradually, the caecum descends to the iliac fossa, and the ascending, transverse and descending parts of colon become distinct [35].



A- FIRST STAGE
B,C- SECOND STAGE
D,E – THIRD STAGE

Fig 1. Schematic representation of normal development of midgut.

Ontogenesis of the abnormal fate of Mullerian duct

A. Non-rotating: quite common; called "left-sided colon" and generally is asymptomatic, but twisting or volvulus can occur

a. Midgut does not rotate after it enters the abdomen. Thus, the caudal limb enters before the cranial limb

b. Small intestines lie on the right side and the entire large intestines on left. May cause obstruction of vessels and gut if kinking or twisting occurs.

B. Volvulus and mixed rotation: cecum lies below the pylorus and is fixed to the posterior abdominal wall by peritoneal bands that cross over the duodenum

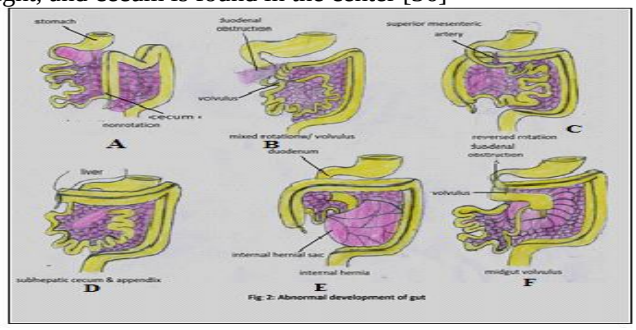
a. Usually causes duodenal obstruction

b. Due to a failure of the midgut loop to complete final 90 degrees of rotation, thus, terminal ileum enters the abdominal cavity first

C. Reversed rotation: rare, clockwise rotation (not counterclockwise)

a.Duodenum lies in front of the superior mesenteric artery and transverse colon behind it, which may obstruct the latter due to pressure from the artery

b. Small intestines lie on the left; large intestines lie on the right, and cecum is found in the center [36]



A – NONROTATION
A- MIXED ROTATION /VOLVULUS
B- REVERSED ROTATION
C- SUBHEPTIC CECUM & APPENDIX
D- INTERNAL HERNIA
E- MIDGUT VOLVULUS

Fig 2. Schematic representation of abnormal development of midgut.

Discussion

Most of the malrotatiion cases are observed in the first month of life. Yet, it may be seen in adults. Even though clinical symptoms are obscure, adult patients visit hospital mostly with complaints such as vomiting and recurrent abdominal pain, probably due to chronic partial obstruction. Some may present with malabsorption due to inability to eat and protein loss associated with diarrhea caused by chronic volvulus [37]. Imaging studies such as plain radiography, contrast enhanced radiograph stomach-duodenum radiography; ultrasonography and computerized tomography scan can help diagnose malrotation. Contrast enhanced radiograph has been shown to be the most accurate method. Typical radiological signs corkscrew sign, which is caused by the dilation of various duodenal segments at different levels and the relocation of duodenojejunal junction due to jejunum folding [38]. In the ultra-sonography, the superior mesenteric artery. Doppler USG may shoe the whirlpool sign with rotation of SMV around SMA which is typical for malrotation. Besides, jejunal arteries lie to the right instead of to the left in computerized tomography scan as another diagnostic sign of malrotation. Since malrotation commonly causes intestinal obstruction all patients deserve elective laparotomy [39]. Ladd's procedure has been the standard procedure of elective treatment of intestinal malrotation. This procedure consists of following steps: first, midgut volvulus is untwisted; bands causing obstruction are divided; segments of colon and small bowel and large bowel ischemia,Ladd's procedure was not an option is our cause and resection of ischemic segments was mandatory. In some patients, extensive small bowel syndrome and subsequent complications may be unavoidable in those patients. Other alternatives such as cecopexy, endoscopic untwisting and laparoscopic management have been used in previous cases in literature. Laparoscopic Ladd's procedure for elective cases has been shown effective, even superior to conventional procedure in some aspects[40]. Yet, it is clear that superior to conventional procedure in some aspects. Yet, it is clear that surgical options should be patient based [41].

The present case focuses attention at this critical rare subject by several points. First, presentation of adult malrotation cases can be obscure, even though whirlpool sigh in abdomen computed tomography scan may give suspicion of bowel twisting. Yet, an acute presentation is commonly associates with extensive bowel necrosis and may give suspicion of bowel twisting [42]. Our patients were left with a short segment of large bowel, therefore refeeding distally was thought to be not contributive to reabsorptions of nutritional elements. Despite vigorous effort to maintain her fluid-electrolyte balance and to supply nutritional requirement via parenteral formulae, developed intraabdominal abscess and subsequent enterocutaneus fistula, which both worsen her medical condition. Finally, ended up with multiple organ failure due to uncontrolled sepsis [43]. This shows us timely recognition of malrotation is the key to save life if possible, but massive small bowel resection is sometimes unavoidable and may associate with fatal consequences.

Conclusion

Malrotation should be considers in differential diagnosis in patients presented with acute abdomen and intestinal ischemia. Patients may be asymptomatic or have obscure symptoms. Therefore, only anamnesis, physical examination and imaging may lead the surgeon to accurate diagnosis.

Surgical intervention should be prompt to limit morbidity and mortality [44].

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