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Malrotation with Abnormal Fixation of Gut

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Abstract: Malrotation is a spectrum of anatomic abnormalities of the incomplete rotation and fixation of the intestinal tract during development of the fetus. Malrotation affects approximately 1 in 500 live births. It is seen mostly in midgut because of fault in 2nd and 3rd stage of midgut rotation. Though rare these are seen accidentally during routine investigations or not diagnosed at all throughout life. The incidence of malrotation in adults is rare and reported in between 0.0001% to 0.19%. Malrotation if significant can cause problems like volvulus, torsion in midgut or can lead to wrong diagnosis of the condition. So, surgeons should keep this in mind during operation for acute abdomen. Non-significant malrotation or malfixation does not cause any problem so remain mostly unidentified. Here is discussed a case of malrotation with malfixation diagnosed during routine dissection of abdomen of a cadaver.
Keywords: Malrotation of gut, Abnormal fixation of gut.

INTRODUCTION

The normal adult position of gut in abdominal cavity results after complex embryological process called 'rotation of midgut loop' and 'zygosis' [1]. Due to complexity, errors in location of alimentary canal are mostly confined to midgut loop [2]. Malrotation is a spectrum of anatomic abnormalities of incomplete rotation and fixation of the intestinal tract during development of the fetus. [3]. Most of the adults diagnosed of mid gut malrotation are asymptomatic. They are detected during imaging investigations for unrelated conditions or during the operation for other pathology in body [4]. The caecum is unusually positioned in 80% of patients with malrotation [5-7]. The overall incidence of malrotation, is unknown because some patients will present years later or remain asymptomatic for life [8]. Thus it becomes important for the surgeons and radiologists to be aware of the anatomical location of intestines due to errors in rotation of midgut loop [2].

CASE REPORT

During routine dissection of 65 yr old embalbed female cadaver, abdomen was opened by incising anterior abdominal wall. After retracting greater omentum following observations were noted.

Caecum and appendix were in right lumbar region and were of normal caliber (Fig.1). Ascending colon was shorter in length than normal. As a result sigmoid colon was in right iliac region (Fig. 2). The relations of transverse colon, superior mesenteric vessels and duodenum were normal (Fig. 3). Mesentry attachment to post abdominal wall was from D-J flexure to ileocaecal junction and was smaller than normal. The lower attachment of mesentry was not upto right sacro-iliac joint but above it. The attachment of sigmoid mesocolon was below and parallel to mesentry attachment, from left sacro-iliac joint to S_2 midline. The structures crossed by sigmoid mesocolon were left common iliac vessels, left ureter, inferior mesenteric vessels (Fig. 4 & 5). Superior mesenteric artery was deviated towards left instead of right . Branching of both superior and inferior mesenteric artery was normal. Portal vein was formed by union of superior mesenteric vein, splenic vein as normal but inferior mesenteric vein drained into superior mesenteric vein instead of splenic vein (Fig. 6). Jejunum coils were in left upper quadrant whereas ileum coils were in umbilical region. As a result of right lumbar position of caecum there was abnormal fixation of small intestine and large intestine. As right iliac region was empty sigmoid colon occupied that region (Fig.7).

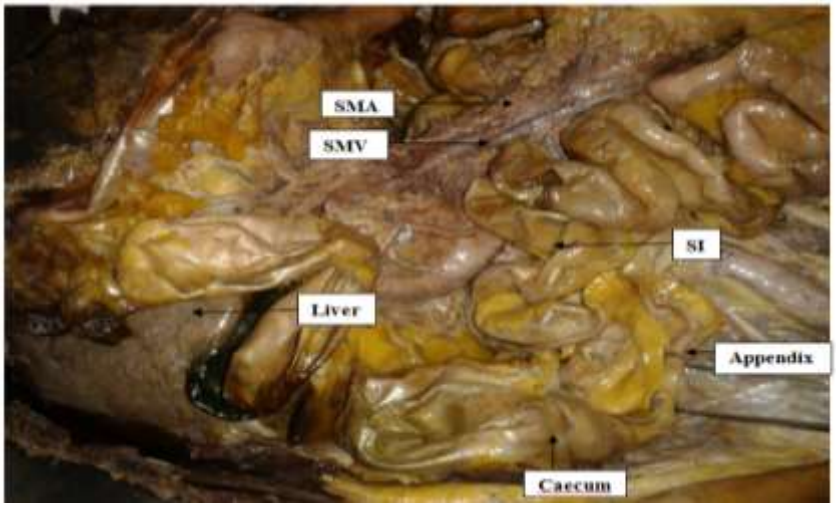

Fig. 1: Showing caecum and appendix in right lumbar region

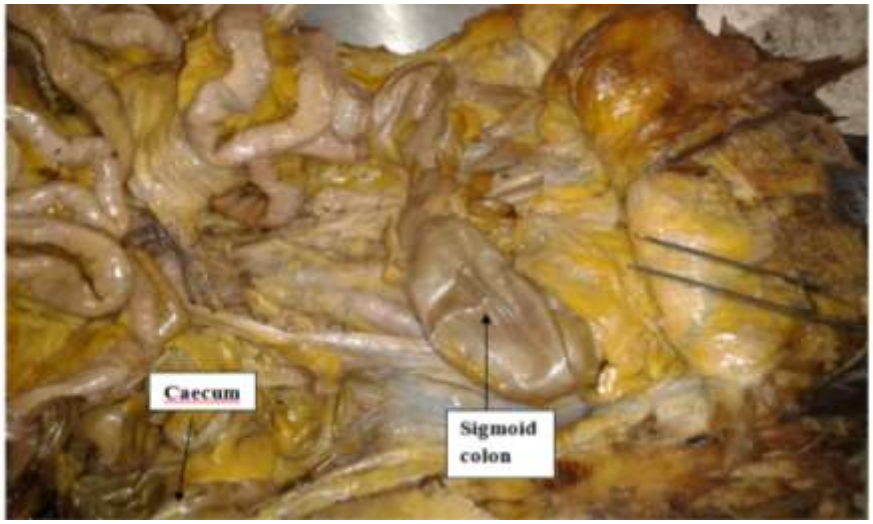

Fig. 2: Showing short ascending colon, sigmoid colon in right iliac region

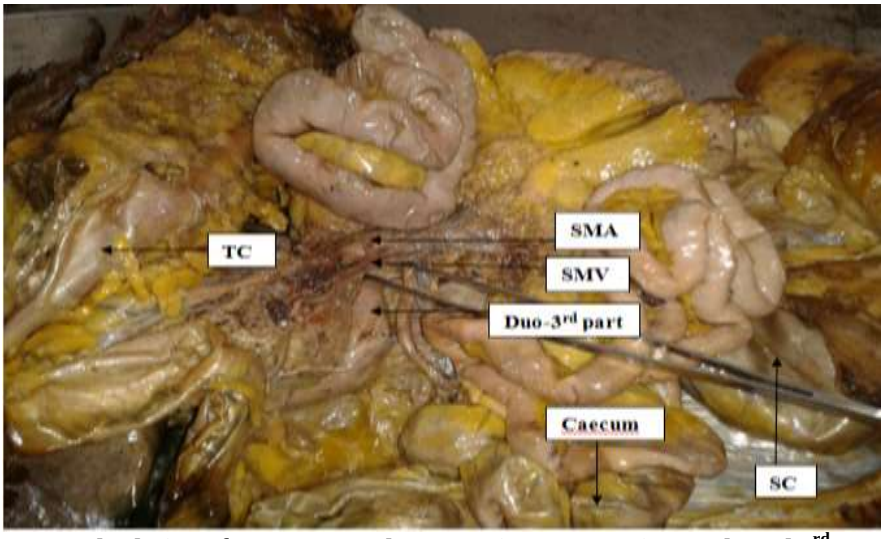

Fig. 3: Showing normal relation of transverse colon, superior mesenteric vessels and 3rd part of duodenum

Fig. 4: Showing structures crossed by sigmoid mesocolon. Sigmoid mesocolon dissected.

Fig. 5: Showing structures crossed by sigmoid mesocolon. Sigmoid mesocolon dissected.

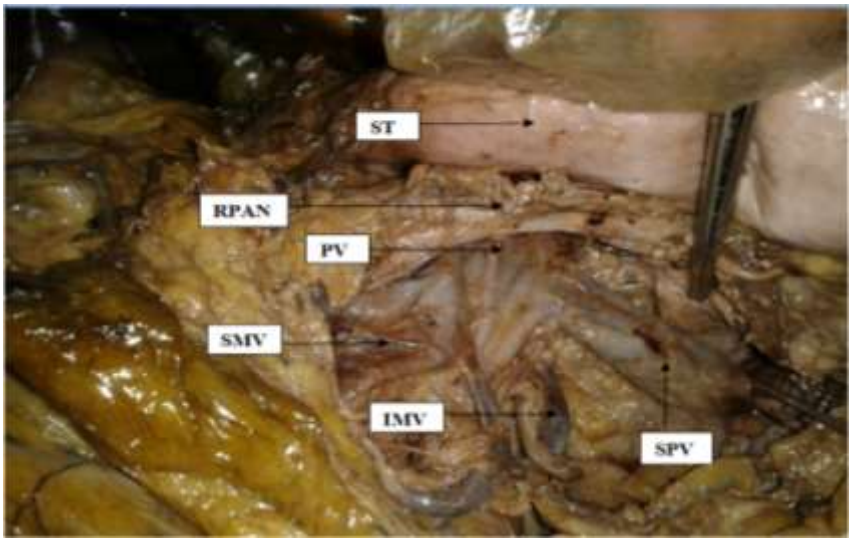

Fig. 6: Showing portal vein formation. IMV draining into SMV instead of Splenic vein.

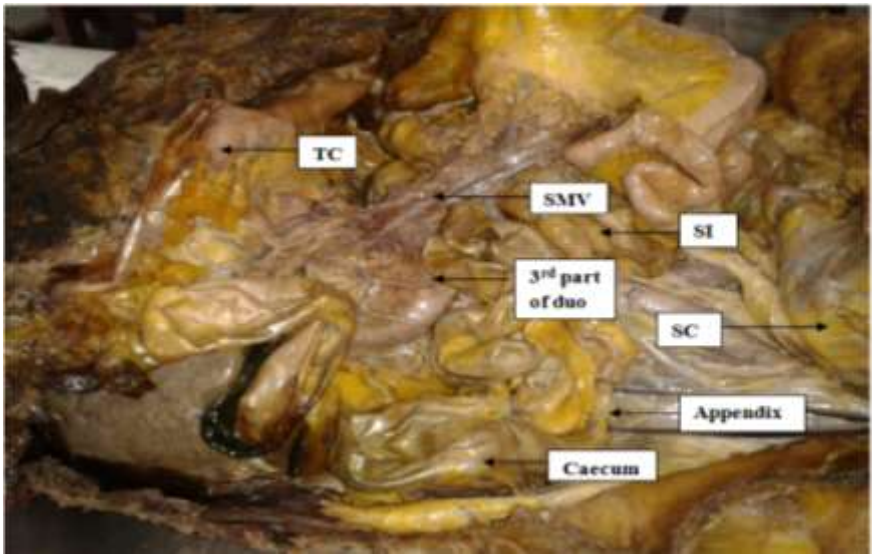

Fig. 7: Showing the overall arrangement of all organs in abdomen

DISCUSSION

The development of the intestinal tract is a complex process [9]. The midgut rotation is described in three stages [7].

Stages in the rotation of midgut loop

A) 1st stage; 5 – 10 weeks

The midgut loop lies in umbilical cord. There is physiological umbilical herniation of midgut loop, largely brought by development of liver. Here the loop rotates through 90 degrees in the anticlockwise direction around superior mesenteric artery as an axis.

B) 2nd stage; 10 – 11 weeks

The midgut loop returns to abdominal cavity from umbilical cord in sequential manner. The preaxial limb of loop develops into small intestine and postaxial loop develops into proximal part of large intestine. There is reduction of physiological midgut hernia back into abdomen. The small intestine returns first, passing posterior to superior mesenteric artery and occupies central part of abdomen. As the large intestine returns, it undergoes further 180^0 anticlockwise rotation. Later it occupies right side of abdomen. The ascending colon becomes recognizable as posterior abdominal wall progressively elongates. Thus the duodenum passes posterior to the superior mesenteric artery. The transverse colon comes anterior to the superior mesenteric artery. The caecum is located in right loin region, subhepatic in position. The descending colon to the left flank region. The small intestine coils from left upper to right lower quadrant of abdomen.

C) 3rd stage; 11 week until shortly after birth

The subhepatic caecum descends into the right iliac fossa of the abdomen forming ascending colon. Definitive ascending colon is formed by relative descent of the caecum. The duodenum and pancreas become retroperitoneal. Ascending colon becomes retroperitoneal. The attachment of dorsal mesentry of small intestine to posterior abdominal wall is greatly modified after formation of ascending colon. The main function of the 3rd stage is efficient fixation to post abdominal wall. Too early fixation of small intestine also can lead to imperfect descent of the caecum [10].

Malrotation affects approximately 1 in 500 live births. Detection of midgut malrotation in adults is rare and incidence has been reported to be between 0.0001-0.19%. Diagnosis may occur incidentally in adulthood during cadaveric study or in imaging investigations and surgery for unrelated pathology. Post mortem studies have suggested that the gut malrotation may affect up to 1 in 6000 [7].

Here in this present case the 1st and 2nd stage of rotation has occurred normaly. In the 3rd stage the descent of caecum is at fault. So the attachment of mesentry to posterior abdominal wall is small than normal. The ascending colon is short.

The possible cause or causes may be due to failure of the caecum to reach the right lower quadrant from subhepatic position, (as arrested in right lumbar region in this case) or an intrinsic growth defect in the ascending colon; or the failure of the caecum to become attached to the growing posterior abdominal wall [7, 11-13]. Abnormal position of caecum could be because of early fixing of small intestine to posterior abdominal wall.

No functional disturbance is seen because of abnormal fixation. Excessive fixation may results in interference with mobility, kinks, and compression of the bowel. Deficient fixation may cause ptosis, torsion, volvulus which can lead to acute abdomen and wrong

diagnosis as acute appendicitis or meckle's diverticulitis because of abnormal position of caecum [14].

Malrotation not only affects the positioning of the midgut, but also the vascular supply of the midgut [15]. In malrotation, the vein is frequently to the left of artery or rotates around the artery. The superior mesenteric artery is constant. However, superior mesenteric vein anatomy reflects the development and the anatomy of the bowel [7]. But in present case the relation of both superior mesenteric artery and vein is normal i.e. vein to the right of artery. In this case the inferior mesenteric vein joins the superior mesenteric vein instead of splenic vein and the portal vein is formed by union of superior mesenteric vein and splenic vein as normal. As the right iliac fossa is empty sigmoid colon has occupied the space. Even though the sigmoid colon has occupied right iliac region, attachment of sigmoid mesocolon is normal.

CONCLUSION

Non-rotation/ malrotation or malfixation though rare to diagnose early, results in abnormal position of intestine in abdomen. The intestinal non-rotation/malrotation is important for the surgeons as location of caecum and as such appendix is different. The clinical presentation of appendicitis is different and the surgical procedure changes depending on the location. The position of Meckel's diverticulum changes due to the change in position of small intestine [2]. So surgeons should be aware of all these conditions before exploration of patient for any acute abdominal emergency as these abnormal location of organs will lead to wrong diagnosis .

Abbreviations

SMA: Superior Mesenteric Artery, SMV: Superior Mesenteric Vein, SI: Small Intestine, TC: Transverse Colon, SC: Sigmoid Colon, Duo: Duodenum, LCIV: Left Common Iliac Vein, LCIA: Left Common Iliac Artery, UR: Ureter, IMV: Inferior Mesenteric Vein, ST: Stomach, RPAN: Retracted Pancreas, PV: Portal Vein, SPV: Splenic Vein

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