Malrotation of gut with associated variations

Case Report

Introduction

During development the abdominal portion of alimentary canal may suffer a large variety of perversion. In the three main divisions of alimentary tract namely foregut, midgut, hindgut; incidence of essential errors in disposition is almost confined to the midgut [1]. Since the work of Mall [2] and Frazer [3] et al., it is generally believed that the normal position of the gut inside the abdominal cavity results after a complex embryological process called “rotation of the midgut loop”. Consequently, disorders of the positioning and fixation of the gut are called malrotation. Malrotation of gut has got surgical importance as failure to recognize the nature and characteristic features of the misplacement may lead to grave errors in exploratory laparotomy or to injurious prolongation of operation [1].

Malrotation occurs in approximately 1 in 500 live births [4]. The caecum is unusually positioned in 80% of patients with malrotation [5, 6]. 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. Unusual conditions on rotation and fixation of the intestines are of intense interest to the pediatric surgeon, as it is a frequently encountered condition and most surgeons will know of at least one case where unusual rotation and fixation of the midgut result in a narrow base to the small bowel mesentery, volvulus may occur, often with catastrophic consequences [5, 7].

Case Report

During routine dissection of abdominal cavity of 75-year-old embalmed male cadaver with no previous history of abdominal operation, malrotation of gut was observed.

• The caecum and appendix was present in right upper quadrant of the abdomen under visceral surface of liver instead of right iliac fossa. The caecum was mobile, dilated, having dimensions of 7 cm in length and 9.5 cm in width. Other associated structures were in usual presentation (Figure 1).
• Ascending colon was absent.
• The duodenum was mobile, redundant and showed kinking with lack of fixity to posterior abdominal wall. The C-shaped loop of duodenum with four parts was present (Figure 2).

Key words [subhepatic caecum] [mobile duodenum] [malrotation] [superior mesenteric vessels]

Dr. Poorwa Baburao Kardile
Department of Anatomy
Shri Vasantrao Naik Government Medical College
Yavatmal, Maharashtra, INDIA.

+91 940 4947887
drpoorwakardile@yahoo.com

Received November 1st, 2012; accepted May 20th, 2013

Abstract

Malrotation of gut is generally regarded as pediatric pathology. It has been estimated that it affects approximately 1 in 500 live births. Detection of adult midgut malrotation is rare and its incidence has been reported to be between 0.0001% to 0.19%. On careful dissection of abdominal cavity of 75-year-old embalmed male cadaver, malrotation of gut was observed. The variants were seen in of caecum, ascending colon, duodenum and superior mesenteric vessels. The caecum was mobile, dilated and subhepatic in position. The dimensions of caecum were 7x9.5 cm. Ascending colon was absent due to non-descent of caecum in right iliac fossa. The duodenum was mobile, redundant and it showed kinking with lack of fixity to posterior abdominal wall. There was inverse relation between superior mesenteric vessels. The superior mesenteric vein was lying to the left side of artery. The variant was seen due to failure of completion of rotation in stage 3. Such presentation are rare in adults is usually nonspecific and this often leads to diagnostic and treatment delay with possible bowel ischemia and necrosis. Subhepatic caecum may predispose to intussusceptions. An understanding of the vascular variants is critical in performing pancreaticoduodenectomy in malrotated patients as well as helpful in identifying adult patient with this congenital condition.

side of artery having usual branching pattern. The other structures were in usual presentation (Figure 3).

Embryology
In order to understand malrotation, a brief review of the embryology of intestine is required. The anatomical development of the intestinal tract is a complex process. In 1898 Mall [2] first described the embryology of malrotation. Dott [1] described relation between anatomy of malrotation and its clinical outcome. Frazer and Robbins [3] described the process of rotation and fixation in three stages. Stage-1 occurs in 5-10 weeks. There is physiological umbilical herniation. At sixth week the herniating bowel rotates 90° around the superior mesenteric artery axis counterclockwise. Stage 2 occurs in 11th week. There is reduction of midgut hernia back into abdomen. There is 180° counterclockwise rotation such that duodenum courses inferior and posterior to the superior mesenteric artery. The colon courses anterior to the superior mesenteric artery with the caecum located to the right and subhepatic in position. Stage 3 occurs in 12th week. The subhepatic caecum descends into the right iliac fossa of the abdomen forming ascending colon. There is fixation of intestine to posterior abdominal wall.

Interruption of typical intestinal rotation and fixation during fetal development can occur at a wide range of locations depending on stage of rotation. If anomaly occurs in Stage 1 it results in omphaloceles caused by failure of gut to return to the abdomen. In stage 2 there is non-rotation, incomplete rotation, reverse rotation, internal hernias. If in stage 3 as in our case then there is subhepatic caecum, unattached duodenum, and mobile caecum [8].

Discussion
Malrotation of gut is generally regarded as pediatric pathology with the majority of patients presenting in childhood. It has been estimated that it affects approximately 1 in 500 live births [4]. Detection of midgut malrotation in adults is rare and its incidence has been reported to be between 0.0001% to 0.19%. Incidental diagnosis may occur in adulthood during cadaveric study or during imaging investigations and surgery for unrelated pathology. The evidence from post mortem studies suggests that gut malrotation may affect up to 1 in 6000 [9, 10].

Definitive ascending colon arises by relative descent of the caecum. In the condition known as subhepatic caecum the adult caecum and appendix are found close to the visceral surface of the liver and the ascending colon is absent or...
Malrotation of gut with associated variations

unusually short. The possible cause or causes may reside in failure of the caecum to reach the right lower quadrant from subhepatic position, or in an intrinsic growth defect in the ascending colon; or in failure of the caecum to become attached to the growing posterior abdominal wall. [11–13]. Recently, it was shown that intestinal malrotation results from inactivating heterozygous mutations in the forkhead transcription factor FOXF1 [14].

In present case redundancy, kinking, mobility and lack of duodenal fixation to the posterior abdominal wall was noticed. However, a single C-shaped loop of duodenum was present. In early fetal life duodenum is mobile as it is entirely covered by peritoneum. With the rotation of gut duodenum rotates to the right side and then behind the superior mesenteric artery. The peritoneum covering posterior surface of rotated duodenum comes in contact with the parietal peritoneum of posterior abdominal wall. Subsequently both layers of peritoneum disappear by the process of zygosis. Hence, definitive duodenum becomes retroperitoneal and fixed except proximal 2.5 cm of the first part. But due to incomplete rotation, the mesoduodenum may persist making it mobile. Hollinshed [15] and Long et al. [16], mentioned a long duodenum with undulation and redundancy due to lack of duodenal fixation, angularity, kinking, formation of more than one loop in the course of duodenum is indicative of malrotation similar to the findings reported in this case.

Malrotation not only affects the positioning of the midgut, but also its vascular supply. In the present case we reported inversion of superior mesenteric vessels. Inversion of the usual relationship of the superior mesenteric artery and vein has been described as a finding suggestive of malrotation by Weinberger [17], Zerin [18], Chao [19], Dufour [20], Zissin [21]. Generally, the superior mesenteric vein is to the right of the artery. In malrotation, the vein is frequently to the left of the 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.

Zerin [18] evaluated anatomic relationships between the superior mesenteric artery and superior mesenteric vein ultrasonography in malrotated subjects. The superior mesenteric artery and superior mesenteric vein were inverted in six patients (67%) and were usual in three (33%). Dufour [20], Zissen [21] and Plackett [22], noticed the most common vascular variants are inversion of the relationship between the artery and vein and vertical position of the vessels. These variants are found in over 60% of patients with malrotation in general. The inverse relation of superior mesenteric vessels with unusual location of gut is helpful in detecting gut malrotation in living subjects by abdominal color Doppler ultrasonography [23–25].

Malrotation of midgut is of surgical importance because of diagnostic problems presented by appendicitis, renal colic. Intestinal malrotation is a rare condition but is considered an important cause of bowel obstruction in adults. The presentation in adults is usually nonspecific and this often leads to diagnostic and treatment delay with possible bowel ischemia and necrosis. Subhepatic caecum may predispose to intussusceptions (Waugh’s syndrome) in early months of age however incidence of malpositioned and malfixated caecum diminishes with progressing age [26]. As truly said malrotation with its propensity for volvulus is a time bomb within [27].

An understanding of the vascular variants is critical in performing pancreaticoduodenectomy in malrotated patients.
Failure to appreciate the variant location of the superior mesenteric artery to the left of the superior mesenteric vein can result in devascularization of the short segmental vessels feeding the small bowel. As each of these small vessels is essentially an end artery, their ligation can lead to small bowel death with catastrophic outcomes [22].

Conclusion

In the present case we can conclude that gut rotation has reached Stage-2 but failed to complete Stage-3 as caecum is subhepatic in position and mobile along with duodenum. The associated vascular variant observed in the form of inversion of superior mesenteric vessels should alert sound clinician with possibility of malrotation and will be helpful in sound management of patient. These variations are frequent in pediatric setup but may rare in adult setup making this case report unique.

Acknowledgement

The authors are thankful to Dr. M. N. Ughade, Prof and HOD, ACPM Medical College, Dhule, for inspiring us for this research. Also authors are grateful to Mr. G. N. Hulke, Modeller, Department of Anatomy, Shri Vasantrao Naik Government Medical College, Yavatmal, for his artistic contribution to this paper. Last but not the least authors are grateful to the staff of the Department of Anatomy, Government Medical College, Miraj.

References