Case Report



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Intestinal malrotation in an adult: case report

Erişkinlerde intestinal malrotasyon: Olgu sunumu

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Intestinal malrotation is a developmental anomaly of the midgut in which the normal fetal rotation of intestines around the superior mesenteric artery and their fixation in the peritoneal cavity fail. Rotational anomalies of the midgut are rare in adults. Operative intervention is required generally when they are symptomatic. While difficult to diagnose, prompt recognition and surgical treatment usually lead to a successful outcome. Intestinal malrotation is rarely asymptomatic and generally diagnosed incidentally in adults. In the present report, a case of incidental intestinal malrotation with clinical findings of small bowel obstruction is discussed with a literature review.

Key Words: Embryology; intestine; ischemia; malrotation;

Intestinal malrotasyon, orta bağırsak (*midgut*) bölümünün peritoneal kavitede arteria mesenterica superior etrafında normal fetal rotasyonunu yapamaması ve fiksasyon bozukluğu ile seyreden bir gelişimsel anomalidir. Midgut rotasyon anomalilerine erişkinlerde nadir rastlanır. Bunlar, genellikle, klinik bulgulara sebep olduklarında cerrahi girişimi gerektirirler. Tanısını koymak güç olsa da erken tanı ve tedavi başarılı sonuç verir. İntestinal malrotasyon nadir olarak asemptomatik seyredip tanısı genellikle insidental olarak konur. Bu yazıda, kliniğimize ince bağırsak tıkanıklığı bulguları ile başvuran bir insidental intestinal malrotasyon olgusu literatür eşliğinde irdelendi.

Anahtar Sözcükler: Embriyoloji; intestine; iskemi; malrotasyon.

Congenital midgut malrotation, a rare anatomic anomaly that can lead to duodenal or small bowel obstruction, is observed rarely beyond the first year of life. Symptomatic patients present with either acute bowel obstruction/intestinal ischemia with a midgut or cecal volvulus or with chronic vague abdominal pain.^[1]

CASE REPORT

A 60-year-old man presented with acute epigastric pain and bilious vomiting. He had a long history of constipation, opening his bowels 2-3 times a week with laxatives. On physical examination, his vital signs were pulse 90, blood pressure 126/67, and respiratory rate 16. The abdomen was not distended; however, he had a palpable, tender mid-epigastric mass. His rectal examination was normal.

Hemoglobin, white blood cell count, basic biochemistry panel, and arterial blood gases were all within normal values. Plain radiographs suggested bowel obstruction with the localization of small intestinal loops predominantly on the right side. Chest radiography did not reveal any signs of perforation of a hollow viscus. At surgery, the small bowel was found to be twisted several times around the superior mesenteric artery. Surgical examination showed a duodenum crossing the spine and entering the jejunum in the left upper quadrant. The fourth duodenal segment and the normal duodenojejunal junction were not developed. All colon segments with the cecum were found to the left of the spine (Fig. 1a).

The small intestine lay on the right side of the abdomen and the large intestine on the left side. The duodenum ran caudally in a straight line from its first part onwards. The cecum lay on the left side of the abdomen and the ileum entered it from the right. The mobile mesentery was fixed. At laparotomy, midgut volvulus in a clockwise direction was found. The volvulus was untwisted completely in a counter-clockwise direction and then the viability of the bowel was assessed. Segmented massive gangrene of the small bowel was

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Fig. 1. (a) The cecum were found to the left of the spine. (b) An area of gangrenous small intestine was resected.

present. An area of gangrenous small intestine (30 cm of necrotic small bowel) was resected and the abdomen was then closed (Fig. 1b). After a difficult post-operative period, the patient recovered satisfactorily.

DISCUSSION

Malrotation of the midgut is an abnormality in the embryological development of the gastrointestinal tract. By the 4th intrauterine week, the gastrointestinal tract is in the form of an endoderm-lined tube. Approximately during the 5th week, a vascular pedicle develops and the gut can be divided into foregut, midgut and hindgut. The superior mesenteric artery supplies blood to the midgut. Intestinal rotation primarily involves the midgut. The rotation of intestinal development has been divided into three stages. Stage 1 occurs in weeks 5-10. It includes extrusion of the midgut into the extra-embryonic cavity, a 90° counterclockwise rotation, and return of the midgut into the fetal abdomen. Stage 2 occurs in week 11 and involves further counter-clockwise rotation within the abdominal cavity, completing a 270° rotation. This rotation brings the duodenal "C" loop behind the superior mesenteric artery with the ascending colon to the right, the transverse colon above, and descending colon to the left. Stage 3 involves fusion and anchoring of the mesentery. The cecum descends, and the ascending and descending colon attach to the posterior abdominal wall. According to this concept, cases of failure of rotation will involve the entire midgut, and a classical and severe malposition will result, with the small bowel located on the right side and the colon on the left side of the peritoneal cavity. Stage 1 anomalies include omphaloceles caused by failure of the gut to return to the abdomen. Stage 2 anomalies include nonrotation, malrotation, and reversed rotation. Stage 3 anomalies include an unattached duodenum, mobile cecum, and an unattached small bowel mesentery.^[2-4]

Midgut mal- and nonrotation refers to a failure in the counter-clockwise rotation of the midgut, which

results in the misplacement of the duodenojejunal junction to the right of midline.

Midgut volvulus is rare in adults.^[5] Most acute presentations occur in the first month of life. In the adult with malrotation, midgut volvulus is the most common cause of bowel obstruction.^[6] Acute presentation is with volvulus of the midgut or ileocecum occurring most frequently in the neonate, with the likelihood decreasing with age.^[7,8] The chronic presentation is more challenging, with symptoms including chronic abdominal pain, bloating, vomiting, constipation, and diarrhea all being reported.^[9] The volvulus occurs around the primitive dorsal mesentery, and thus constricts and compresses the superior mesenteric vessels. This process will particularly affect the venous drainage and the involved bowel will become filled with blood. The infarcted bowel will bleed into its lumen, and if the volvulus is then relieved spontaneously, the patient will pass blood-stained diarrhea signifying the end of the attack.

At operation, the entire mass of the small bowel must be delivered through the wound, the volvulus completely untwisted in an counter-clockwise direction, and then the viability of the bowel assessed. If gangrene is evident, the affected gut is resected and the bowel continuity restored. When the viability of the gut is uncertain, reoperation is carried out 24 to 48 hours later, during which time adequate resuscitation is continued.^[10] Limited resection may then be possible.

In conclusion, at operation, the mesentery must be sutured to the posterior abdominal wall to prevent further episodes of volvulus. Emergency exploration with resection of the gangrenous bowel is vital for the patient's survival.

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