

ADULT INTESTINAL MALROTATION PRESENTING AS MIDGUT VOLVULUS; CASE REPORT.

Barsak malrotasyonuna bağlı olarak gelişen incebarsak volvulusu; Olgu sunumu.

Shantanu Kumar Sahu, Shailendra Raghuvanshi, Arvind Sinha, Praveendra Kumar Sachan

Himalayan Institute of Medical Sciences, Department of General Surgery, Dehradun / India

Cer San D (J Surg Arts), 2012;5(1):18-21.

ABSTRACT

Intestinal malrotation is a rare congenital condition caused by the absence of or incomplete rotation of the small bowel during the embryonic period and is defined as any deviation from the normal 270 degree counterclockwise rotation of the midgut. 64-80% of cases present primarily during the neonatal period prior to the first month of life, as acute intestinal obstruction due to volvulus of small intestine. The presentation of intestinal malrotation in adults is rare, and occurs in approximately 0.2-0.5%. Contrast enhanced CT can show the abnormal anatomic location of a right sided small bowel, a left-sided colon and an abnormal relationship of the superior mesenteric vein (SMV) situated to the left of the superior mesenteric artery (SMA) instead of to the right and the characteristic 'whirlpool or whirl sign' describing the swirling appearance of bowel and mesentery twisted around the superior mesenteric arterial axis with the presence of midgut volvulus. Ladd procedure is the surgical treatment of choice. Here in, a 20-year-old man presenting with midgut volvulus has been discussed in view of the literature.

Key words: Intestinal malrotation, volvulus, CT scan, whirlpool or whirl sign, Ladd procedure.

ÖZET

İntestinal malrotasyon terimi embriyoner hayatta midgutta olması gereken satin tersi yönündeki 270 derecelik rotasyonun olmaması veya eksik olmasıyla ortaya çıkan tabloyu ifade eder. Vakaların %64-80'i hayatın ilk aylarında gelişen akut incebarsak volvulusu tablosu ile kendini gösterir. Yetişkinlerde nadirdir ve intestinal malrotasyona bağlı volvulus tablosu olguların sadece %0.2-0.5'inde görülür. Çekilen kontrastlı karın tomografilerinde sağ tarafa yerleşmiş incebarsaklar, sol tarafa yerleşmiş kalın barsak ve mezenterik arterin soluna doğru yer değiştirmiş olan superior mezenterik ven görünümü volvulus için karakteristiktir ve "girdap bulgusu-belirtisi" olarak tanımlanır. Bu belirti mezenterik ve barsak anslarının arter aksı etrafında dönerek midgut volvulusu olduğunu ifade eder. Tedavide uygulanacak tedavi Ladd prosedürüdür. Burada, 20 yaşındaki bir erkek hastada görülen midgut volvulusu literatur verileri ışığında sunulmuştur.

Anahtar kelimeler: Malrotation, volvulus, BT, girdap belirtisi (işareti), Ladd prosedürü.

INTRODUCTION

Intestinal malrotation is a rare congenital condition caused by the absence of or incomplete rotation of the small bowel during the embryonic period.

64-80% of cases present primarily during the neonatal period prior to the first month of life, as acute intestinal obstruction due to volvulus of small intestine. The

presentation of intestinal malrotation in adults is rare, and occurs in approximately 0.2-0.5% (1-3).

Case

A 20 year male presented to the surgical emergency with features of upper abdominal pain, vomiting and constipation since 15 days. On examination the patient was dehydrated. Abdominal examination showed distension with a sluggish bowel sound. Routine hematological investigations and renal function tests were within normal limits. Plain abdominal roentogram was inconclusive with few air-fluids levels present in the small bowel. Contrast-enhanced CT scan of abdomen was planned which showed the swirl appearance of bowel loops starting from 3rd part of duodenum with twisting of the intervening mesenteric vessels involving the superior mesenteric arteries and vein (whirlpool sign) suggestive of mid gut volvulus. There was also proximal dilatation of duodenum and stomach suggestive of duodenal obstruction (Figure 1 and 2).

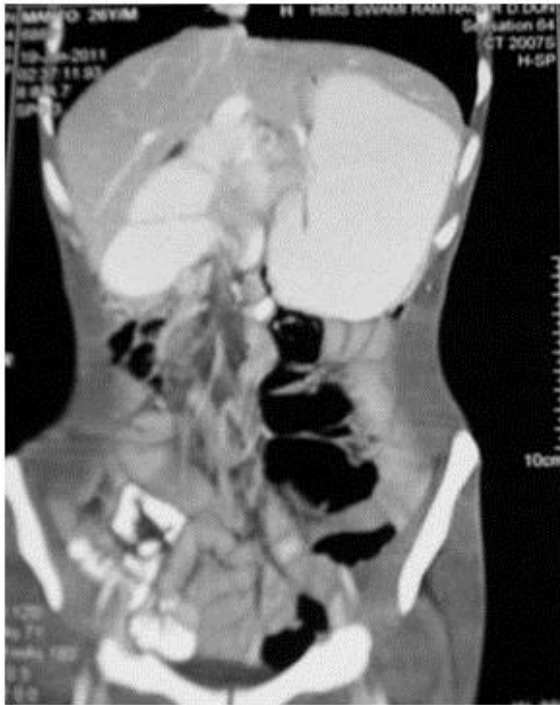


Figure 1: Contrast-enhanced CT scan of abdomen showing proximal dilatation of duodenum and stomach suggestive of duodenal obstruction.

Exploratory laparotomy was done which revealed volvulus of small intestine which was viable. Malrotation of intestine was seen with the entire small bowel remaining on the right side of the abdomen and the colon on the left along with the absence of ligament of Treitz. Ileum was seen entering the caecum from the right side. Ladd's bands extending from the caecum to

the lateral abdominal wall and crossing the duodenum was also seen causing duodenal obstruction (Figure 3).

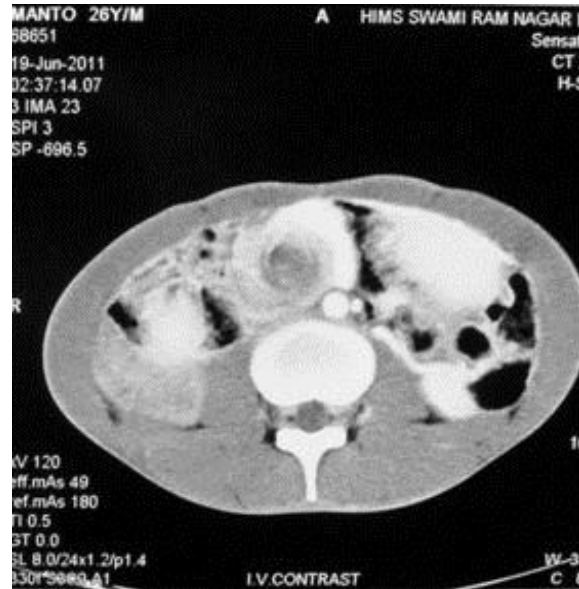


Figure 2: Contrast-enhanced CT scan of abdomen showing "whirlpool sign" suggestive of midgut volvulus.

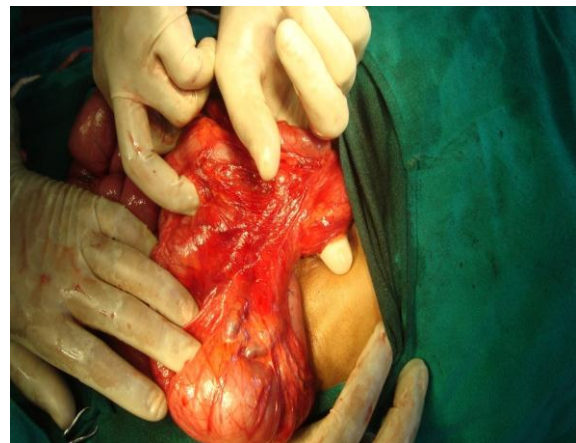


Figure 3: Intraoperative photograph of the patient with malrotation of bowel with a viable midgut volvulus.

Ladd procedure was done which involved counterclockwise detorsion of the viable midgut volvulus, division of the abnormal coloduodenal Ladd bands tethering the midgut and causing extrinsic duodenal compression, mobilization of the duodenum and the right colon, widening of the mesenteric base by section of possible adhesions near the superior mesenteric vessels to prevent further volvulus and removal of the malpositioned appendix. Patient had an uneventful post-operative recovery and was discharged on 7th post-operative day.

DISCUSSION

The rotation of intestinal development has been divided into 3 stages. Stage I occurs between 5th to 10th gestation week. It includes extrusion of the midgut into the extraembryonic cavity, a 90° counterclockwise rotation and return of the midgut into the fetal abdomen. Stage II occurs in 11th week and involves further counterclockwise rotation within the abdominal cavity completing a 270° rotation. The duodenum rotates caudal to the artery, and its C-loop traces this path. The transverse and ascending colon demonstrate the path of rotation of the cecum cephalad to the artery. Stage III involves fusion and anchoring of the mesentery. The duodenum becomes fixed retroperitoneally in its third portion, emerging at the ligament of Treitz, and the cecum becomes fixed to the lateral abdominal wall by peritoneal bands. The takeoff of the branches of the superior mesenteric artery elongates and becomes fixed along a line extending from its emergence from the aorta to the cecum in the right lower quadrant (4).

Intestinal malrotation can be broadly defined as any deviation from the normal 270° counterclockwise rotation of the midgut (stage II). When it fails to rotate, the entire small bowel remains on the right side of the abdomen, the colon is on the left, and the ligament of Treitz is absent resulting in malrotation or incomplete rotation of intestines with the resultant positioning of the intestine with unusual adhesions. These aberrant adhesions in combination with a shortened mesentery, allows volvulus to occur due to clockwise twisting of the bowel around the superior mesenteric arterial axis which can lead to ischemia and necrosis of small bowel. Malrotation is also associated with peritoneal or Ladd's bands extending from the caecum to the lateral abdominal wall and crossing the duodenum, which can cause duodenal obstruction (5).

Clinical presentation of adult intestinal malrotation is usually asymptomatic. Approximately 30% of these patients present with vomiting and 20% have recurrent unspecific abdominal pain. Symptoms can arise from acute or chronic intestinal obstruction that may be caused by the presence of the Ladd bands and/or a volvulus with resultant ischemic necrosis and perforation peritonitis. Some may present as malabsorption associated with diarrhea and nutritional deficiencies caused by bowel lymphedema resulting from lymphatic obstruction by chronic volvulus and resulting in loss of proteins into the bowel lumen. Rare presentations of chronic volvulus include cases of obstructive jaundice by mechanical compression of the biliary tract, chylous ascites and superior mesenteric vein thrombosis, secondary to long-standing lymphatic and venous obstruction (6-9).

Diagnosis of adult intestinal malrotation is made by imaging studies. The absence of caecal gas shadow or the localization of small intestinal loops predominately in the right side should arouse the suspicion of malrotation. Upper gastrointestinal barium series remains accurate for detection (accuracy over 80%) with the duodeno-jejunal junction failing to cross the midline and lies below the level of the duodenal bulb. Contrast enema examination usually shows malposition of the right colon with the ileum entering the cecum from the right. Selective superior mesenteric angiogram showing abnormal curling and corkscrew appearance of vessel toward the right is also diagnostic of mid gut volvulus due to malrotation. Contrast enhanced CT can show the anatomic location of a right sided small bowel, a left-sided colon and an abnormal relationship of the superior mesenteric vein (SMV) situated to the left of the superior mesenteric artery (SMA) instead of to the right with malrotation. With the presence of mid gut volvulus, the characteristic 'whirlpool or whirl sign' describing the swirling appearance of bowel and mesentery twisted around the superior mesenteric arterial axis is seen in CT scan and with the presence of intestinal ischemia or necrosis becomes an ominous sign. Ladd's obstruction can also be additionally seen as duodenal obstruction in adult intestinal malrotation. The 'clockwise whirlpool sign' on color Doppler ultrasonography showing the wrapping of the SMV and the mesentery around the SMA in a clockwise pattern however has been described as an important diagnostic finding in midgut volvulus (10-14).

The classic treatment for incomplete intestinal rotation is the Ladd procedure, which entails counterclockwise detorsion of the viable midgut volvulus (if present), division of the abnormal coloduodenal Ladd bands tethering the midgut and causing extrinsic compression, mobilization/ kocherisation of the duodenum and the right colon, widening of the mesenteric base by section of possible adhesions near the superior mesenteric vessels to prevent further volvulus and removal of the malpositioned appendix. The aim of this procedure is to reduce the risk of acute volvulus, by locating the small intestine in a non-rotating position and widening the base of the mesentery. Appendectomy is performed due to possible difficulty in the diagnosis of future appendicitis, distant from the classic lower right quadrant position. Laparoscopic Ladd procedure is safe now days due to early discharge and oral intake (15, 16).

REFERENCES

1. McIntosh R, Donovan EJ. Disturbances of rotation of the intestinal tract. *Am J Dis Child* 1939; 57(1):116-66.
2. Frantzides CT, Cziperle DJ, Soergel K, Stewart E. Laparoscopic Ladd procedure and cecopy in

the treatment of malrotation beyond the neonatal period. *Laparosc Endosc Percutan Tech* 1996;6(1):73-5.

3. Mazziotti MV, Strasberg SM, Langer JC. Intestinal rotation abnormalities without volvulus: the role of laparoscopy. *J Am Coll Surg* 1997;185(2):172-6.

4. Gohl ML, DeMeester TR: Midgut malrotation in adults. An aggressive approach. *Am J Surg* 1975; 129(3):319-23.

5. Filston HC, Kirks DR: Malrotation: The ubiquitous anomaly. *J Pediatr Surg* 1981;16(8):614-20.

6. Wang C, Welch CE. Anomalies of intestinal rotation in adolescents and adults. *Surgery* 1963;54(12):839-55.

7. Brandt ML, Pokorny WJ, McGill CW ve ark.. Late presentations of midgut malrotation in children. *Am J Surg* 1985;150(6):767-71.

8. Spitz L, Orr JD, Harries JT. Obstructive jaundice secondary to chronic midgut volvulus. *Arch Dis Child* 1983;58(5):383-5.

9. Schwartz DL, So HB, Schneider KM ve ark. Recurrent chylous ascites associated with intestinal malrotation and lymphatic volvulus. *J Pediatr Surg* 1983;18(2):177-9.

10. Torres AM, Ziegler MM. Malrotation of the intestine. *World J Surg* 1993;17(3):326-31

11. Berdon WE. The diagnosis of malrotation and volvulus in the older child and adult: a trap for radiologists. *Pediatr Radiol* 1995;25(2):101-3.

12. Grika Lb, Popky GL. Angiography in midgut malrotation with volvulus. *Am J Roentgenol* 1980; 134(5):1055-6.

13. Nichols DM, Li DK. Superior mesenteric vein rotation: a CT sign of midgut malrotation. *AJR Am J Roentgenol* 1983;141(4):707-8.

14. Janssens F, Verswijvel G, Smits J ve ark. Midgut volvulus in an adult patient. *JBR-BTR*, 2003;86(2):74-6.

15. Ladd WE. Surgical diseases of the alimentary tract in infants. *N Engl J Med* 1936;215(10):705-8.

16. Matzke GM, Dozois EJ, Larson DW, Moir CR. Surgical management of intestinal malrotation in adults: comparative results for open and laparoscopic Ladd procedures. *Surg Endosc* 2005;19(109):1416-9.