CASE REPORT

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A Rare Cause of Ileus in Adulthood: Congenital Midgut Malrotation and Performing the Ladd Procedure

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ABSTRACT Intestinal malrotation is a rare entity in adults. It may present with non-specific findings and we may also encounter acute clinical findings due to obstruction and ischemia. The gold standard surgical treatment for intestinal malrotation is the Ladd procedure. A 24-year-old male patient was admitted to our medical center with sudden onset of abdominal pain, nausea and vomiting. Surgical treatment was performed after an urgent evaluation and examination. We started the Ladd procedure laparoscopically and then finished it with laparotomy. Even though it is rare in adults, it should be kept in mind by surgeons in differential diagnoses. The Ladd procedure can be performed laparoscopically in elective or acute settings. In cases accompanied by ileus or volvulus, operation may not be complete laparoscopically. As clinical experience is shared, algorithms for adult malrotation patients will appear.

Keywords: Adult; ileus; intestinal volvulus; laparoscopy

Midgut malrotation occurs as a lack of rotation of the intestinal tract during embryologic development. As a result, positional abnormalities are observed in the intestines. It is often present in the 1st week of life. Intestinal malrotation is rarely seen in adults, with an incidence of approximately 0.2-0.5%.[1]

In adults, malrotation may be acute and life-threatening. Narrow mesenteric base predisposes to midgut malrotation and Ladd's bands cause to small bowel obstruction.[2] It may also present chronically with nonspecific and subtle symptoms such as intermittent abdominal pain, food intolerance, bloating and vomiting. Computed tomography (CT) is preferred in adults due to the limited visualization ability of ultrasonography. The typical finding in imaging modalities is the whirlpool sign caused by the rotation of the intestines with the mesentery.[3]

The treatment of the patient presenting with acute conditions is to manage complications and normalize the anatomy. There is controversy over the appropriate management of patients with chronic symptoms. Currently, there are no predictive features for progression and complications.[4] The fact that elective procedures lead to fewer complications than emergency surgical procedures is considered as a benefit. Treatment of malrotation at any age is treated surgically with the Ladd procedure. Ladd procedure named after it was first performed by Dr. William Ladd in 1936. This operation is aimed both to treat acute problems and to reduce the potential for future volvulus. Stages of the operation include reduction of the midgut volvulus; complete lysis of the adhesive collo-duodenal (Ladd) bands; dilation of the mesenteric floor; and optionally prophylactic appendectomy.[5]

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CASE REPORT

A 24-year-old male patient was admitted to our hospital with sudden onset of severe abdominal pain, nausea and vomiting. The patient complained that the pain aggravated after eating. He pointed out that it was the first time in his life that he had experienced such abdominal pain.

The physical characteristics of the patient were evaluated as tall and normal body mass index. Vital signs were regular. On physical examination, bowel sounds were heard as hyperactive with auscultation. Palpation examination revealed abdominal tenderness. There was no history of surgery and no comorbidities. As for the laboratory values, complete blood count showed an increase in white blood cells. There was no significant increase in CRP and lactate levels. The diagnosis of midgut volvulus was made by CT. The image shows approximately three turns of the small intestine around itself and a whirlpool sign, which is the classical finding for volvulus (Figure 1).

Operation was planned immediately and started laparoscopically. Laparoscopical view was not adequate, it was not feasible to reverse the volvulus. Laparotomy was performed. The ileocecal region was seen in the upper middle part of the abdomen instead of the normal location. Small intestines which rotated three turns around itself, was reversed in a counterclockwise direction. There was no evidence of intestinal necrosis or ischemia. Fibrous band was seen distal to the 2nd part of the duodenum. It was considered as the cause of ileus. Sigmoid colon was collapsed. Mesenteric artery and especially venous structures were obviously congested and dilated. Ladd's and intermesenteric bands were resected (Figure 2). Appendectomy was performed. Colonic segments were repositioned to their usual localizations and suture-pexy was applied.

Postoperative follow-up was uneventful and the patient was discharged on the 4th day. The patient's complaints completely regressed and no complications were observed.

Informed consent was obtained from the patient for the case presentation.

DISCUSSION

Congenital intestinal malrotation's occurrence in adults extremely rare, with 90% of cases observed in newborns. It arises in the embryologic period with a disruption in intestinal rotation. Embryologic development of the small intestine begins at 4 weeks of

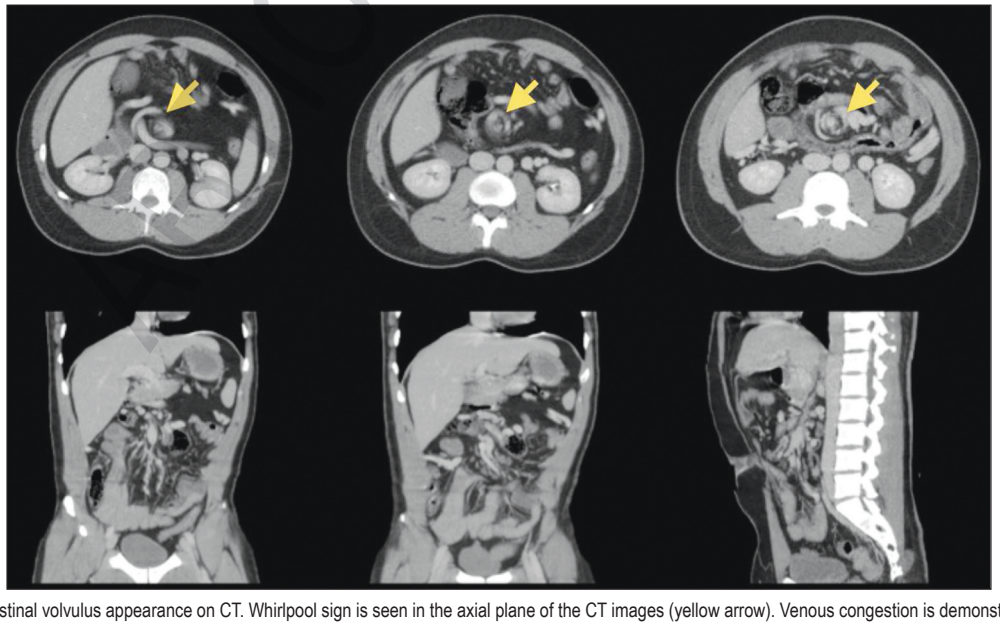

FIGURE 1: Intestinal volvulus appearance on CT. Whirlpool sign is seen in the axial plane of the CT images (yellow arrow). Venous congestion is demonstrated in the coronal plane and mesenteric rotation in the sagittal plane.

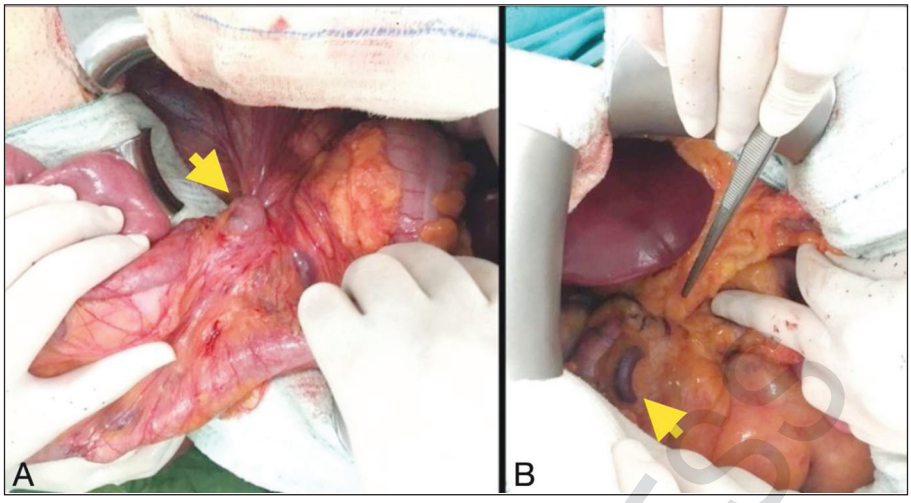

FIGURE 2: A) Perioperative appearance of the Ladd band (Yellow arrow), **B)** Mesenteric venous congestion (Yellow arrow)

gestation with the gut tube, which is formed from the endoderm. At the 5th week of gestation, the intestinal tube divides into 3 with the formation of blood vessels: Foregut, midgut and hindgut. Intestinal rotation occurs mainly in 3 stages and is mostly associated with the midgut. In the 1st stage, the midgut protrudes from the embryonic cavity. It rotates 90 degrees counterclockwise around the superior mesenteric artery and returns to the cavity. The 2nd stage starts at 11 weeks and the midgut rotates another 180 degrees counterclockwise. With the completion of these 2 stages, the ascending colon is positioned on the right side, the transverse colon on the upper side and the descending colon on the left side. In the 3rd stage, the midgut mesentery, small intestines, ascending colon and descending colon are fixed to the posterior abdominal wall. Also, the cecum descends to the right side. Adult malrotations are seen acute conditions as volvulus or small bowel obstruction. Although there is no consensus on the treatment of asymptomatic cases, keeping intestinal malrotations in mind among the differential diagnoses in cases presenting with acute abdomen is associated with better outcomes. Delayed diagnosis may warrant fatal extensive small intestinal necrosis. We diagnosed our patient with volvulus by finding the small intestine rotating 3 turns on its own axis on radiologic examination. The patient was immediately prepared for the gold standard Ladd procedure.[6] We started the operation laparoscopically but due to technical difficulties, we completed as open surgery. There is no standardized difficulty scale with radiological findings, so we preferred a minimally invasive procedure to start the surgical treatment.

As a result of the growing prevalence of minimally invasive surgery, the laparoscopic Ladd procedure has proven to be feasible. In Matzke and colleagues published data, they demonstrated that the laparoscopic Ladd procedure is equally safe, effective and feasible as the standard open Ladd procedure in adults with intestinal malrotation without midgut volvulus.[7] Mazziotti et al. and Panda et al. showed results in the same direction with their studies.[8,9] Coe et al. reported that laparoscopic Ladd procedure can also be performed in patients presenting with volvulus concomitant with intestinal malrotation.[10] In our basic surgical knowledge, it is obvious that elective procedures will cause fewer complications than emergency interventions. Neville and colleagues also found that there was no statistically significant difference in symptom relief between patients undergoing emergency or elective procedures and laparoscopic or open surgery.[11]

Intestinal malrotations are usually treated within the 1st year of life because they represent an emergency. Pediatric surgeons are more experienced in the diagnosis and treatment of intestinal malrotation. It may occur in adults with subtle symptoms. Therefore,

when there is clinical suspicion, the support of pediatric surgeons may be requested, as in the case series of Kotobi et al.[12]

Congenital intestinal malrotation is an uncommon entity in adults and results in significant diagnostic and therapeutic challenges due to the lack of specific symptoms and clinical experience. For these reasons, late diagnosis can lead to serious and life-threatening consequences for the patient. Our diagnosis was made quickly and treatment was initiated urgently. This situation ensured that complications such as ischemia and necrosis did not occur during the case. We also aimed to show that radiologic findings may have an effect on the feasibility of laparoscopic Ladd procedure. According to the degree of volvulus detected on tomography, it may help us to choose our technique for the case. In cases accompanied by ileus, laparoscopic manipulation is not sufficient. Laparoscopic interventions should not be insisted in cases as in our surgical treatment.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Sami Acar;* ***Design:*** *Çağıl Karaevli;* ***Control/Supervision:*** *Çağıl Karaevli, Sami Acar;* ***Data Collection and/or Processing:*** *Çağıl Karaevli, Sami Acar;* ***Analysis and/or Interpretation:*** *Sami Acar;* ***Literature Review:*** *Çağıl Karaevli, Sami Acar;* ***Writing the Article:*** *Çağıl Karaevli;* ***Critical Review:*** *Çağıl Karaevli, Sami Acar.*

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