



Malrotation of the Gut: An Uncommon but Surgically Important Condition in Adults

Zamri Zuhdi, Krishna Kumar Katheraveloo

Abstract

Congenital intestinal obstruction is rare in adulthood. It usually occurs in infants. An unusual presentation of this condition in an adult has led to difficult preoperative diagnosis. We present a case of a 48-year-old gentleman who presented with proximal jejunal obstruction by a congenital band causing gut malrotation. He successfully underwent Ladd's procedure to release the obstruction. We will discuss the non-specific presentation of this rare congenital condition, its diagnosis and management together with the literature review.

Key words: Congenital band, gut malrotation, Ladd's procedure

Introduction

Midgut malrotation is an anomaly of fetal intestinal rotation and fixation that usually presents within the first month of life. It is rare for malrotation to present in adulthood and it is mostly asymptomatic and incidentally discovered later in life during surgery for other conditions [1,2]. However, some adults may acutely present with bowel obstruction and intestinal ischaemia due to volvulus or chronically present with

symptoms of bowel obstruction.

Because presentation is nonspecific and because the index of suspicion for malrotation progressively decreases in the older population, the clinical diagnosis is usually not considered in the initial evaluation. In this report, we present a case of malrotation in an adult who presented with cramping abdominal pain and who subsequently underwent emergency laparotomy.

Department of Surgery
Universiti Kebangsaan
Malaysia Medical Centre
(UKMMC)
Kuala Lumpur, Malaysia

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Corresponding author:
Dr.Zamri Zuhdi
Department of Surgery
Universiti Kebangsaan
Malaysia Medical Centre
(UKMMC)
Jalan Yaakob Latif, 56000
Bandar Tun Razak, Cheras
Kuala Lumpur, Malaysia
zamriz7582@gmail.com

Case Report

A 48-year-old gentleman presented to our center with cramping generalised abdominal pain. His abdominal pain began five days prior to admission. The cramping pain came shortly after eating and recurred intermittently. The pain occurred every 2 to 3 hours and was about 30 to 40 minutes in duration. He vomited twice prior to admission, and his last bowel movement was 2 days before the admission date. He denied any history of pancreatitis, kidney stones, peptic ulcer disease or colitis. He also had no previous history of abdominal surgery.

His vital signs were all normal. His abdomen was mildly distended on inspection. Bowel sound was normal on auscultation. He exhibited no peritoneal signs. However, mild diffuse tenderness to deep palpation was appreciated. His rectal examination was normal.

An abdominal x-ray revealed dilated loops of small bowels that were sequestered to the right side of the abdomen, and absence of a caecal shadow and a faecal-loaded colon were confined to the left side of the abdomen (Figure 1).

A computed tomography (CT) scan of the abdomen showed a dilated and displaced small bowel to the right side of the abdomen with a collapsed colon to the left side. Orientation of the superior mesenteric vein in relation to the superior mesenteric artery was noted to



Figure 1. Abdominal x-ray showed dilated loops of small bowels which were sequestered to right side of the abdomen (arrow).



Figure 2. Superior mesenteric vein (white arrow) situated on the left of the superior mesenteric artery (black arrow).

be to the left of the artery rather than the normal right ventral position (Figure 2).

Based on the diagnosis of malrotation, the patient consented to exploratory laparotomy and Ladd's procedure. Upon entering the abdomen, we noted the caecum being in the left together with the appendix. The ileocaecal junction was also in the left. There were caecal bands notably attached to the jejunum, which made the proximal jejunum dilated. The bands were lysed, the caecum returned to the left iliac fossa and an appendicectomy was performed. Post-operatively, the patient's recovery was uneventful. He tolerated a regular diet on postoperative day 2, and was discharged home on postoperative day 3.

Discussion

Midgut malrotation has been estimated to occur in approximately one in 500 live births. Approximately 85 percent of malrotation cases present in the first two weeks of life [2]. However, it is difficult to ascertain the true incidence because this condition will go undetected during childhood in a substantial number of patients. In adulthood, intestinal malrotation is estimated to occur in 0.2 to 0.5 percent [3].

Intestinal malrotation can be broadly defined as any deviation from the normal 270-

degree counterclockwise rotation of the midgut. During fetal development, the midgut

supplied by the superior mesenteric artery grows too rapidly to be accommodated in the peritoneal cav-

ity. Prolapse into the umbilical cord occurs around the sixth week. Between the tenth and twelfth week, the midgut returns to the abdominal cavity, having undergone a 270-degree counterclockwise rotation around the superior mesenteric artery.

The rotation of intestinal development has been divided into 3 stages: Stage 1 occurs in weeks 5 to 10. It includes extrusion of the midgut into the extraembryonic cavity, a 90-degree counterclockwise rotation, and return of the midgut into the fetal abdomen. Stage 2 occurs in week 11 and involves further counterclockwise rotation within the abdominal cavity, completing a 270-degree 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 the descending colon to the left. Stage 3 involves fusion and anchoring of the mesentery. The caecum descends, and the ascending and descending colon attach to the posterior abdomen.

Adult presentation of malrotation is a difficult diagnosis because of the low incidence of the disorder. Patients with intestinal malrotation, who were not diagnosed until adulthood, may present with a variety of chronic symptoms, including nausea, vomiting, vague abdominal pain, dyspepsia and peptic ulcer disease. The location of the pain may vary from epigastric pain to left upper abdominal pain, and the pain may be described as either intermittent cramping pain or persistent aching pain [4]. Others have described severe abdominal pain followed by diarrhea that is suggestive of chronic volvulus. Vomiting may or may not be bilious and it is variable in duration and frequency. Our patient presented with worsening abdominal pain and acute intestinal obstruction; thus, a suspicion of volvulus was there during initial presentation.

The diagnosis of the rotational anomaly can be made by radiographic studies. In the absence of volvulus, a plain x-ray of the abdomen is of little diagnostic value. The absence of a cecal gas shadow or the localization of small intestinal loops predominantly in the right side should arouse the suspicion of malrotation. The standard upper gastrointestinal series may show a vertical duodenum that does not cross the midline. The accuracy of the upper gastrointestinal series is reported to be over 80 percent [1]. On barium examination, the

entire colon is seen in the left half of the abdomen with the ileum entering the cecum from the right. Malrotation can be diagnosed on CT by the anatomic location of a right-sided small bowel, a left-sided colon, an abnormal relationship of the superior mesenteric vessels and aplasia of the uncinata process. Our patient's abdominal x-ray and CT findings were highly suggestive of gut malrotation.

Surgical therapy remains the mainstay of treatment in symptomatic patients regardless of age at presentation. All patients with malrotation are candidates for laparotomy, even if they are asymptomatic, because the complications associated with intestinal malrotation are based on anatomical reasons that do not alter with age; the potential to develop a sudden onset of acute midgut volvulus in an asymptomatic patient at any age therefore persists [5]. The most commonly used approach is the Ladd procedure, which was done in our patient. This procedure involves counterclockwise reduction of the volvulus if present, division of any coloduodenal bands, widening of the mesenteric base to prevent repeated volvulus and prophylactic appendectomy [3].

Conclusion

Adult presentation of malrotation is a difficult diagnosis because of the rare incidence of the disorder. Diagnosis requires a high index of suspicion. Specific findings that are diagnostic of malrotation can be detected through the use of both upper and lower gastrointestinal tract barium studies, a CT scan and often emergency laparotomy.

Treatment remains the same as it was originally described by Ladd in 1936. Complete resolution of acute obstruction or chronic abdominal pain is the result of a high index of suspicion, appropriate diagnostic studies and aggressive definitive treatment.

Conflict of interest statement

The authors have no conflicts of interest to declare.

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