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Case Report

Laparoscopic Management of Intestinal Malrotation With Cocoon Deformity: A Case Report

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Intestinal malrotation is diagnosed and treated mostly in infancy and childhood, but it is rarely encountered in adults. Here, we present a case of adult intestinal malrotation with cocoon deformity that was managed by a laparoscopic procedure. A 71-year-old man presented with intermittent abdominal pain, nausea, and vomiting. Preoperative findings from abdominal computed tomography, upper gastrointestinal contrast imaging, and enema examination showed intestinal malrotation without volvulus. Diagnostic laparoscopy revealed nonrotation of the midgut, and most of the small bowels were contained in a large peritoneal sac without strangulation. With a definitive diagnosis of intestinal malrotation with cocoon deformity, further laparoscopic repair was performed by widely opening the peritoneal sac, and widening the base of the mesenteric pedicle to prevent future volvulus. There were no postoperative complications. The laparoscopic approach for intestinal malrotation with cocoon deformity can be a safe and useful technique. With the advantage of its minimal invasiveness, it can be an alternative to laparotomy.

Key words: Intestinal malrotation – Cocoon deformity – Adult – Laparoscopy

Intestinal malrotation is a congenital anomaly resulting from errors in fetal intestinal rotation and fixation. Disturbances during any of the individual stages of midgut rotation result in anomalies. Intestinal malrotation can cause shortness of the mesenteric root and allows the small bowel to twist

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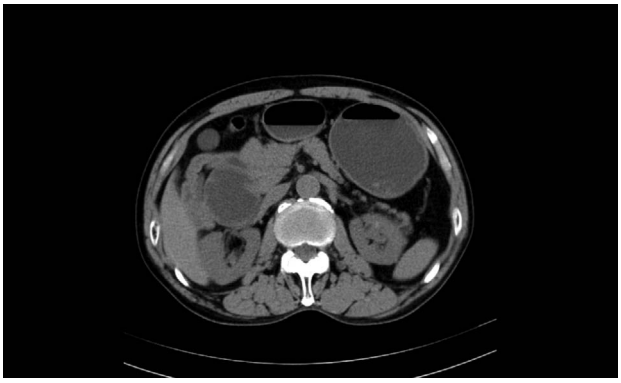

Fig. 1 Abdominal CT showing gastroduodenal dilatation without any signs of strangulation.

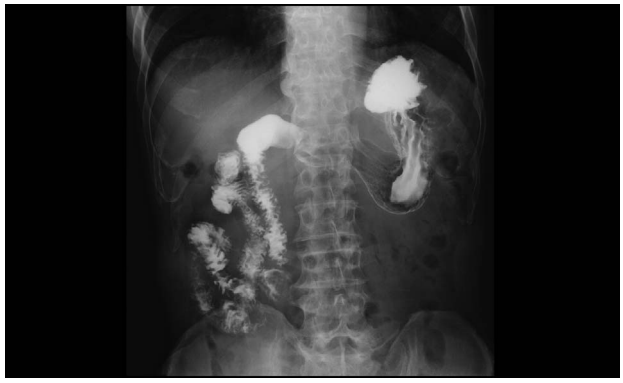

Fig. 2 UGI contrast imaging showing a right-sided duodenojejunal junction and a duodenum failing to cross the midline.

around the pedicle of the superior mesenteric artery, which places the patient at high risk of volvulus of the midgut and intestinal ischemia. Although the volvulus is absent, congenital adhesive bands (Ladd's bands) may compress the duodenum and cause chronic obstruction.[1] It can be classified into various subtypes according to its rotation and the fixation of the duodenojejunal limb and the cecocolic limb of the midgut. The 4 different subtypes include nonrotation, incomplete rotation, reverse rotation, and mesocolic hernia.

The peritoneal sac that encapsulates the bowels, the so-called cocoon deformity, is the consequence of Ladd's bands forming chronically and attaching the bowel to their cavity.[2] Although classified as the nonrotation subtype, its management rarely has been described previously because of its rarity.

Due to the lifetime risk of midgut volvulus, all cases of intestinal malrotation, including those that are identified incidentally or are asymptomatic, should be surgically treated.[3] Classically, intestinal malrotation was managed operatively via laparotomy. Recently, however, laparoscopy has proven to be safe and effective not only as a diagnostic tool but also as an alternative approach to definitive surgical treatment in both infants and adults.[1,4]

Here, we report a typical case of adult onset intestinal malrotation with cocoon deformity that was managed by laparoscopic surgery.

Case Presentation

A 71-year-old man presented with intermittent abdominal pain, nausea, and vomiting. Physical examination revealed only mild tenderness in the right lower quadrant without peritoneal signs. Abdominal ultrasonography and laboratory studies demonstrated no abnormalities. Abdominal computed tomography showed gastroduodenal dilatation without signs of strangulation (Fig. 1). Diagnosed with small bowel obstruction, he was admitted to our hospital for a conservative follow-up. His symptoms spontaneously resolved soon after admission. Further examination of upper gastrointestinal contrast imaging (UGI) revealed a right-sided duodenojejunal junction, and a duodenum that failed to cross the midline (Fig. 2). Enema examination showed cecal malposition and a left-sided colon (Fig. 3). Diagnosed with small bowel obstruction secondary to intestinal malrotation, laparoscopic surgery was subsequently performed.

After general anesthesia and with the patient in the supine position, a 12-mm umbilical trocar was placed for a pneumoperitoneum with a pressure of 12 mmHg. Three additional 5-mm trocars were placed in the left lower abdomen, right upper, and lower abdomen. Intraoperative findings showed

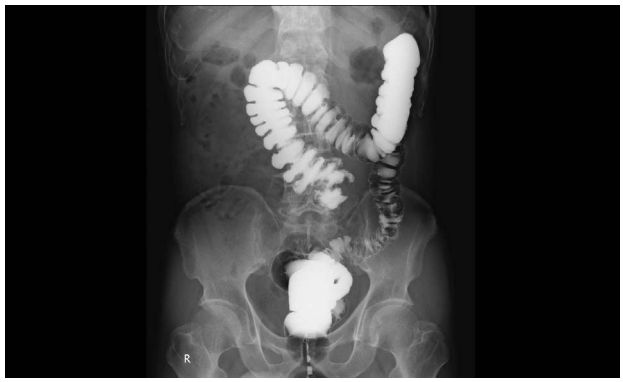

Fig. 3 Enema examination showing cecal malposition and a left-sided colon.

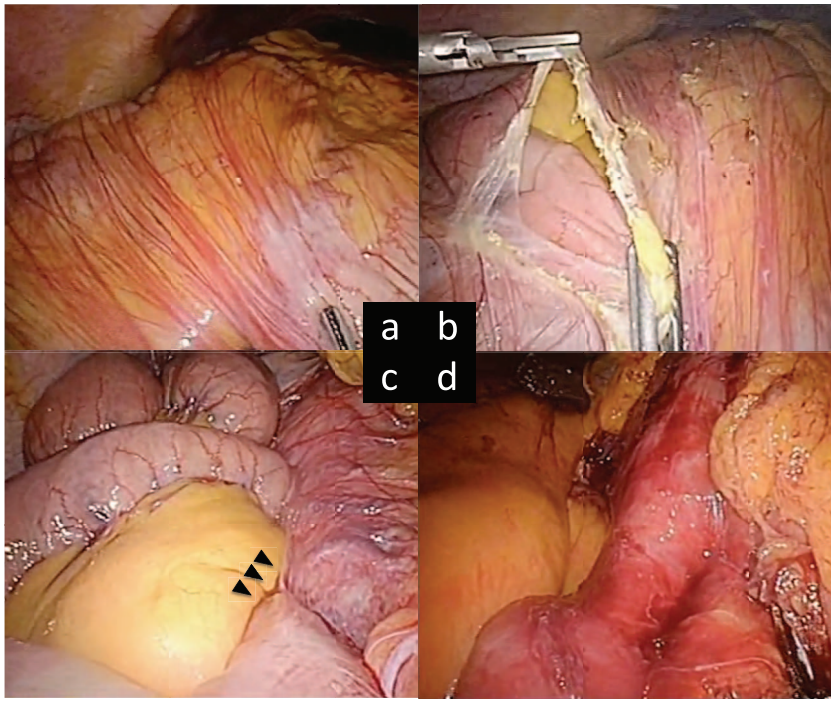

Fig. 4 Operative findings and process. (a) Large transparent peritoneal sac, so-called the cocoon deformity, containing most of the small bowels. (b) Opening the peritoneal sac. (c) Black arrows showing the Ladd's bands that obstructed the second portion of the duodenum. (d) Ladd's bands opened widely.

lack of adherence of the ascending colon to the retroperitoneum, resulting in cecal malposition and a left-sided colon, which were found during preoperative enema examination. Most of the small bowels were contained in a large transparent peritoneal sac, the so-called cocoon deformity, which obstructed the second portion of the duodenum. There was no distention or strangulation in the encased bowels. The sac resulted from the Ladd's bands that chronically fixated and maintained the malpositioned bowel in place. With a definitive diagnosis of intestinal malrotation, further laparoscopic repair was subsequently performed. The peritoneal sac was opened to widen the base of the mesenteric pedicle and prevent future volvulus (Fig. 4). After confirming the absence of gangrenous changes in the entire small bowel, the skin incisions were closed. The total operation time was 65 minutes, with minimal bleeding. No complications were encountered during the postoperative course and the patient discharged from the hospital after 4 postoperative days. He has remained free of any symptoms after 2 postoperative years.

Discussion

Intestinal malrotation is usually diagnosed within the first month after birth in most patients. Although the incidence of malrotation in adulthood is 0.2%,[5,6] autopsy studies estimate the true incidence to be as high as 1% of the total population.[7] Unlike the pediatric population, most adult patients with malrotation lack other congenital anomalies and are free of symptoms related to their malrotated bowel. So, it may be incidentally discovered later in life during surgery for other conditions.[8,9] However, some adults may present acutely with midgut volvulus and intestinal ischemia or chronically with symptoms of intermittent bowel obstruction or vague abdominal complaints, which are often mistaken for irritable bowel syndrome; peptic ulcer, biliary and pancreatic disease; and psychiatric disorders.[8] In the present case, the patient had symptoms of intermittent abdominal pain, nausea, and vomiting. After taking a thorough medical history, we noticed that he had a prolonged history of vague abdominal complaints that always resolved spontaneously. It is important to keep these infrequent anomalies in mind, even in adult patients. Specific findings on UGI and barium enema (BE) imaging can contribute to the definitive diagnosis of intestinal malrotation.[10] UGI studies often reveal a duodenojejunal junction located to the right of or overlying the spine and below the level of the duodenal bulb. In the case of a midgut volvulus, corkscrew taping of the duodenum or jejunum may be visualized. BE has been used to identify the position of the cecum

Table 1 Previous reports of intestinal malrotation with cocoon deformity

Published (year)	Age	Gender	Symptom	Preoperative diagnosis	Operative approach	Peritoneal sac	Complication
Matzke GM (2003)[14]	30	Male	Epigastric pain	Intestinal malrotation	Laparoscopy	Incised widely	None
Mahalik SK (2012)[16]	2	Male	Abdominal pain with distension, bilious vomiting	Intestinal malrotation with volvulus	Laparotomy	Excised	None
Patel RV (2013)[17]	1 week	Female	Bilious vomiting	Intestinal malrotation	Laparotomy	Opened widely	None
Our case	71	Male	Intermittent abdominal pain	Intestinal malrotation	Laparoscopy	Opened widely	None

and may reveal a beak-like stenosis of the intestine in patients with midgut volvulus.[11] Malrotation can also be diagnosed on computed tomography (CT) with the presence of a right-sided small bowel, a left-sided cecum, and an inverse relationship between the superior mesenteric artery (SMA) and the superior mensenteric vein (SMV) wherein SMV is visualized to the left of, instead of the right of, SMA.[12] A distinctive whirlpool sign, the consequence of the twisting of the small bowel and mesentery around the narrowed SMA pedicle, may be seen on CT imaging of a patient with midgut volvulus. Although most patients with malrotation will not have all the specific findings described previously, the identification of any one of these abnormalities warrants closer scrutiny and consideration of other diagnostic modalities.[10] Precise knowledge of the peritoneal anatomy and its abnormalities is mandatory for a proper understanding of the findings.

Patients with intestinal malrotation are treated using the Ladd procedure, which requires mobilization of the right colon and cecum by division of the Ladd bands, mobilization of the duodenum, dissection of adhesions around the superior mesenteric artery to broaden the mesenteric base, and an appendectomy.[13–15] Classically, the Ladd procedure is managed via laparotomy. However, laparoscopic diagnosis and treatment in both infants and adults have been recently described as safe, feasible, and effective.[1,4]

According to Holcomb, Ladd bands may cause the bowel to encapsulate into a cocoon-like deformity in the presence of longstanding partial obstruction or internal herniation, which may cause partial or complete obstruction of the duodenum. Only 3 case reports referring to intestinal malrotation with cocoon deformity have been previously reported (Table 1).[14,16,17] In terms of age, patients ranged from being neonates to adults. Our case involved the oldest patient to be reported. Although all cases were preoperatively diagnosed as intestinal malrotation, the peritoneal sac that encapsulated the bowels was not identified. Lack of specific findings on radiographic images of the peritoneal sac makes it difficult to make a precise diagnosis before surgery, as demonstrated in our case. Since none of the patients suffered from any complications or recurrences, widening the opening of the peritoneal sac is adequate for the treatment of the cocoon deformity. The laparoscopic approach may be useful for clarifying the diagnosis. Furthermore, it allows the performance of subsequent surgical treatment,

as presented in our case. Midgut volvulus and severe bowel distension are the important causes of open conversion in laparoscopic management. The absence of gangrenous changes in herniated bowel loops may make the laparoscopic approach possible. A previous case was also managed by laparoscopy. The present case will be the second case that has demonstrated the safety and effectiveness of laparoscopic management for intestinal malrotation with cocoon deformity.

Conclusion

We presented a rare case of intestinal malrotation with cocoon deformity. Laparoscopic approach can be a safe and useful technique with the advantage of minimal invasiveness.

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