

Laparoscopic Ileocolic Resection for Crohn's Disease Associated With Midgut Malrotation

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ABSTRACT

Midgut malrotation is an anomaly of fetal intestinal rotation. Its incidence in adults is rare. A case of midgut malrotation in a 51-year-old man with complicated Crohn's disease of the terminal ileum is presented. Symptoms, diagnosis, and treatment are reviewed. Preoperative workup led to correct surgical planning that ultimately allowed a successful laparoscopic resection.

Key Words: Midgut malrotation, Crohn's disease, Laparoscopic surgery.

INTRODUCTION

Midgut malrotation is an anomaly of the fetal intestine that usually presents early in childhood. The underlying cause is an abnormal rotation of the small bowel along the axis of the superior mesenteric artery, which can occur at any stage of embryogenesis, involving either part of or the entire gut.

The incidence in adults is difficult to estimate because most patients remain asymptomatic. Clinical presentation in adulthood can vary, with intestinal malrotation often being an incidental finding in otherwise healthy individuals who present with a history of recurring episodes of abdominal pain and/or bowel subobstruction. Others present with an acute onset of obstructive symptoms, and diagnosis can be made during or immediately before surgical exploration.

We hereby describe a rare case of intestinal malrotation associated with complicated Crohn's disease (CD) of the terminal ileum.

CASE REPORT

A 51-year-old male patient with known CD and severe abdominal pain was seen in the gastroenterology outpatient clinic of our hospital. The diagnosis of CD of the terminal ileum had been established by his local gastroenterologist 10 years before this episode. At the time, the diagnosis was based solely on the colonoscopy and retrograde ileoscopy, and the patient was placed on a treatment of oral mesalazine, 200 mg twice daily. In addition, steroids were also prescribed, leading to incomplete remission. No imaging of the small bowel was performed. There was no history of previous abdominal surgery or any knowledge of other significant pathologic condition. The patient complained of recurrent episodes of mild abdominal pain in the left abdomen for the past few years but denied changes in bowel habits, evidence of rectal bleeding, or febrile episodes.

Clinical examination revealed a generally healthy individual in good nutritional status (body mass index, 27); abdominal examination was substantially normal, with moderate tenderness noted in the lower quadrants.

Although blood test results revealed only a mild increase in inflammatory markers (C-reactive protein and white cell count), the patient was admitted to the gastroenterology ward for further studies because he had not been regularly

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Figure 1. Small bowel follow-through. Almost the whole jejunum is lying in the right flank—enormous dilatation of terminal ileum and consensual elevation of the left diaphragm.

followed by a specialized gastrointestinal clinic. Colonoscopy revealed an edematous and inflamed ileocecal valve with otherwise normal mucosa throughout the entire colon. It was not possible to enter the terminal ileum. The subsequent small bowel follow-through (SBFT) demonstrated a midgut malrotation associated with a severe stricture of 20 cm of the terminal ileum. As shown in **Figures 1** and **2**, the small bowel was predominantly localized in the left side of the abdomen, and the dilated loop of ileum preceding the diseased intestine was positioned in the upper left quadrant, beneath the diaphragm.

Given the patient's clinical findings—10-year history of CD, fibrotic type of stricture, partial response to conventional therapy, and presence of malrotation—and after a multidisciplinary review, it was decided to proceed with a surgical resection. The patient was fully informed of his clinical situation and was offered a laparoscopic exploration routinely performed in all cases of primary CD at our center.¹

Despite the presence of the anatomical abnormality, in association with typical inflammation of complicated CD, a successful

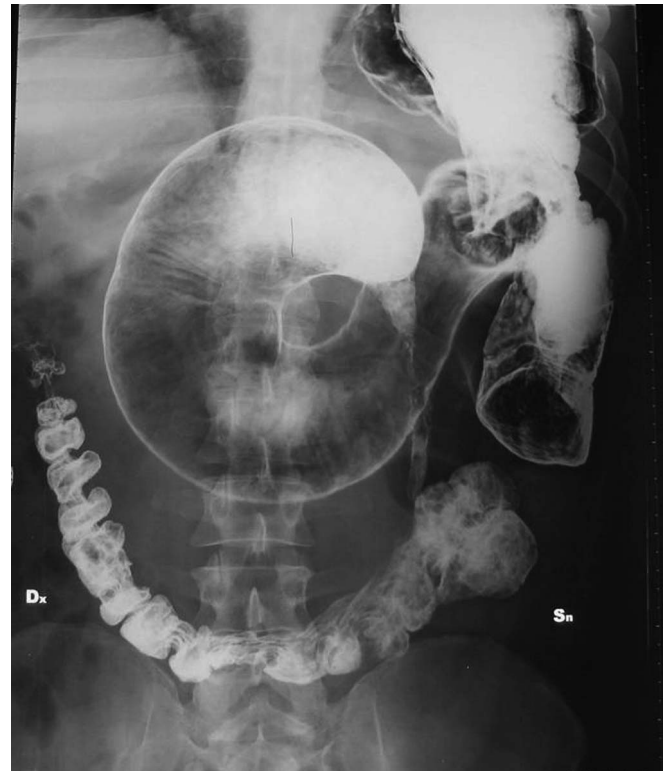


Figure 2. Small bowel follow-through. Chrons disease structure of a left sided terminal ileum.

fully laparoscopic ileocecal resection was done. Midgut malrotation had resulted in both the colon and ileum to become central intraperitoneal structures. This anatomical abnormality therefore required adjustment of trocar positioning and a modified surgical approach compared with our routine surgical technique for CD (**Figure 3**).

Pneumoperitoneum was induced with an open technique through a navel incision. Because of the dilated bowel, and given the intestine displacement, an additional 10-mm trocar was placed just cranial to the pubis. Laparoscopy confirmed the presence of malrotation and evidence of a severely inflamed stricture of the terminal ileum; the small bowel showed proximal dilatation for approximately 100 cm, which was more pronounced for the first 40 cm before the known stricture. The right colon was in a fully medial position without peritoneal cover, as was expected. However, it was possible to proceed to a standard ileocolic resection. Because of the severe prestenotic dilatation, 60 cm of the terminal ileum was resected. The mesentery, containing the vessels of the terminal ileum and cecum, was divided using the ultrasonic dissector; the small and large bowel were resected using the 60-mm

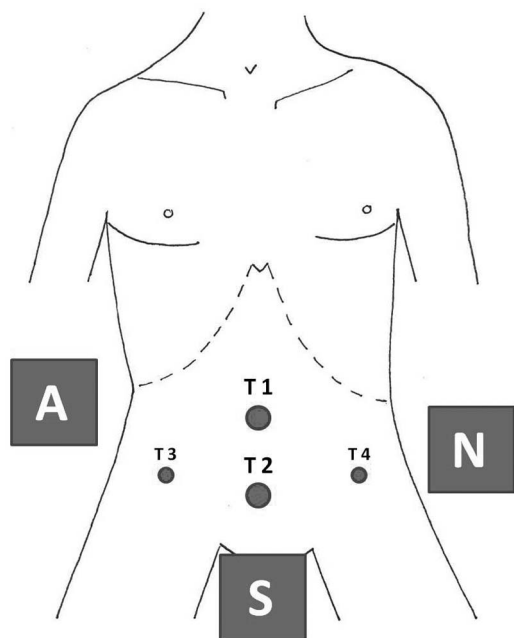


Figure 3. Patient position and trocar placement during surgery. S = surgeon, A = assistant, N = scrub nurse; T1 = Hasson first access, T2 = camera, T3–4 = 5-mm operative trocars.

Echelon Flex (Ethicon, San Angelo, TX) carrying the vascular cartridge for the ileum and the blue cartridge for the colon. Suprapubic access was enlarged to a final incision of 4 cm to retrieve the specimen and to staple (blue cartridge) a side-to-side ileocolic anastomosis. The patient had an uneventful postoperative recovery and was discharged home on the fifth postoperative day.

He is well 1 year after his operation and on a mesalazine recurrence prevention protocol.²

DISCUSSION

A thorough review of the literature of intestinal malrotation in adults did not reveal other published articles of midgut malrotation associated with CD. However, in 1954, a case report was published describing intestinal malrotation in a patient who underwent a surgical resection of 40 cm of distal ileum, “where it crossed the midline to enter the cecum, involved by regional ileitis.”³ More recently, a case of concurrent CD and intestinal malrotation was presented at the Southeastern Surgical Congress in 2011 and is briefly reported in *The American Surgeon*.⁴

Midgut malrotation is a congenital abnormal rotation of the small bowel along the axis of the superior mesenteric artery, which is usually associated with laxity and loss of fixation of the gut in the abdomen. It can occur at any stage of embryogenesis

and involves part of or the entire gut. It is often associated with duodenal or jejunal atresia, Hirschsprung disease, gastroesophageal reflux, intestinal intussusceptions, and anorectal malformations. The incidence is between 0.0001% and 0.19% in asymptomatic adults, and the prevalence in children younger than 1 year of age is 3.9 per 10,000.

The true incidence in adults is difficult to estimate because the condition remains mostly asymptomatic. In the non-pediatric population, midgut malrotation can be found in association with irritable bowel syndrome or psychiatric-related generic abdominal symptoms.⁵

It is possible to distinguish between two different models of presentation of malrotation in adults. One type of presentation is characterized by chronic symptoms and a history of recurring episodes of bowel subobstruction (nausea, vomiting, and intermittent abdominal pain). Abdominal distention is usually not a characteristic feature because the obstruction tends to be located proximally. These patients may be seen by several physicians and undergo many investigations before the correct diagnosis is made. The other group of patients presents with an acute onset of obstructive symptoms. With careful questioning, these patients may also describe a prior history of intermittent, but milder, symptomatology. Malrotation may, therefore, be an unexpected finding at surgery.⁶

The knowledge of the embryology of the intestinal rotation is a key point for understanding its anomalies. The gut is in the form of a straight tube at the fourth week of life. It is during the fifth week, when a vascular pedicle develops, that the gut is divided into foregut, midgut, and hindgut. The midgut forms the small bowel and the colon up to the splenic flexure. Intestinal rotation primarily involves the midgut and can be divided into 3 stages: stage I (weeks 5–10), characterized by extrusion of the midgut into the extraembryonic cavity, a 90° counterclockwise rotation, and return of the midgut into the fetal abdomen; stage II (week 11) involves further counterclockwise rotation within the abdominal cavity completing a 270° rotation that 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; and stage III involves fusion and anchoring of the mesentery. The cecum descends, and the ascending and descending colon attach to the posterior abdomen.⁷

Anomalies occurring during stage I include omphaloceles caused by failure of the gut to return to the abdomen. Stage II anomalies include nonrotation, malrotation, and reversed rotation, whereas anomalies that occur during the third stage include an unattached duodenum, mobile cecum, and an unattached small bowel mesentery.⁸

The best imaging technique for the diagnosis of midgut malrotation is the SBFT, which usually allows identification of the position of the duodenojejunal flexure and small bowel loops. The position of the duodenojejunal flexure is highly accurate in predicting malrotation. In particular cases, a barium enema may help to determine the position of the cecum and colon. Malrotation is usually suspected on computed tomography scans when there is a reversal of the normal superior mesenteric artery and vein relationship. Several authors have reported this finding and suggest that when reversal is present, the child should undergo further tests to confirm or exclude malrotation. However, inversion of the superior mesenteric artery and vein relationship may be present in normal rotation, and a normal relationship may be present in children with malrotation. Therefore, a computed tomography scan cannot be used as a screening procedure for intestinal malrotation.^{9–11}

As stated before, intestinal malrotation often is an incidental finding and generally does not require surgical intervention. In the case reported here, malrotation was not clinically suspected, given the established diagnosis of CD. It is likely that if the small bowel was investigated at the time when diagnosis of CD was made, the malrotation could have been detected earlier. In fact, the workup for CD should always include a study of the small bowel by means of one or more of the available imaging techniques: small intestine contrast ultrasonography, SBFT, magnetic resonance or computed tomography scan with study of the bowel.

It is difficult to establish how many of this patient's symptoms were related to CD and what the impact was of the malrotation. However, as soon as the diagnosis was confirmed, the decision to proceed with surgical intervention was made, and laparoscopic ileocecal resection was undertaken. Because of midgut malrotation, the right colon and ileum become the central intraperitoneal structures; therefore, planned adjustment of port positioning is a prerequisite for a successful operation. The laparoscope was moved to a suprapubic position, and the surgeon stood between the patient's legs; from a low central position, it was easier to gain visualization of the entire abdominal cavity and to start the operation with the division of loose congenital adhesions between the right colon, in a fully medial position and without peritoneal cover, and the retroperitoneum covering the sacrum and the right iliac vessels. The resection of the mesentery, containing the vessels of the ileum and right colon, was approached from a medial to lateral position, as is done for standard laparoscopic small bowel resection. The operation was not associated with increased intrasurgical risk nor did the surgeon encounter any intraoperative complication. In fact, because of the malrotation, mobilization of the right

colon was easier than usual. Furthermore, the remnant right colon reached the pubis easily and without tension.

CONCLUSION

The coincidental discovery of CD in a patient with malrotated gut is most likely fortuitous. No cause–effect relationship could be demonstrated.

In case of surgery, in the presence of intestinal malrotation, one must consider that the right colon is centralized and intraperitoneal and the ileocolic mesentery is shared with the small bowel.

However, based on our experience, no specific technical difficulties for a laparoscopic approach were identified.

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