

Procedure for Duodenal Stricture With Malrotation Masquerading as Superior Mesenteric Artery Syndrome in an Adult Patient

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ABSTRACT

Introduction: Duodenal stricture with malrotation is rare in adults. Anatomical anomalies of malrotation include narrowing of the root of the mesentery associated with volvulus and the presence of Ladd's band, which causes duodenal obstruction. Two case reports of duodenal stricture not associated with Ladd's band have been published in the English literature.

Case Presentation: A 36-year-old thin man was admitted for vomiting after meals and was diagnosed with superior mesenteric artery (SMA) syndrome by endoscopy and computed tomography.

Management and Outcomes: He received conservative treatment but returned 3 more times with the same symptoms. He underwent laparoscopy during his fifth admission after he was diagnosed with duodenal stricture with malrotation based on preoperative examinations. A membrane-like adhesion affecting the retroperitoneum to the duodenum and several cordlike adhesions wrapping around the right dorsal side of the SMA to the anal side of the duodenum were dissected to divide the adhesions. Ladd's band was not observed. No recurrence has been observed for 1 year and 7 months.

Discussion: The jejunum was located on the dorsal side of the SMA and the duodenum was pulled by the cordlike adhesions wrapping around the right dorsal side of the SMA, and a concomitant duodenal obstruction was identified; therefore, the jejunum appeared to be compressed between the SMA and the aorta. Consequently, this case masqueraded as SMA syndrome. We successfully performed laparoscopic surgery for an adult patient with malrotation, especially with regard to dissection of the cordlike adhesions originating from the root of the SMA. This minimally invasive procedure is feasible for duodenal strictures with malrotation.

Key Words: Duodenal stricture, Ladd's band, Malrotation, Superior mesenteric artery syndrome.

Citation Takahashi N, Gyoten K, Narita K, Sato R, Suzuki H, Machishi H, Okada Y. Procedure for duodenal stricture with malrotation masquerading as superior mesenteric artery syndrome in an adult patient. CRSLS e2018.00056. DOI: 10.4293/CRSLS.2018.00056.

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Disclosure: none reported.

Conflicts of Interest: All authors declare no conflict of interest regarding the publication of this article.

Informed consent: Dr. Takahashi declares that written informed consent was obtained from the patient for publication of this study/report and any accompanying images.

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INTRODUCTION

Malrotation is a rotational anomaly of the midgut and is a rare condition in adults.¹ The typical presentation of intestinal malrotation is sudden onset of bilious vomiting in a previously healthy infant.² The typical complications associated

with this condition are midgut volvulus, which occurs around the narrow pedicle of the superior mesenteric artery (SMA), obstruction of the proximal jejunum, an ischemic midgut, and duodenal stricture due to Ladd's band.³ It is treated surgically by placing the bowel in a position of nonrotation, widening the mesenteric base, and dividing

Ladd's band (Ladd procedure).⁴ Laparoscopic surgery is used to treat intestinal malrotation in children⁴ and adults.⁵

SMA syndrome is a rare acquired disorder in which acute angulation of the SMA causes compression of the third part of the duodenum between the SMA and the aorta, leading to obstruction.⁶

This patient was initially diagnosed with SMA syndrome due to vomiting after meals, but duodenal stricture with malrotation was discovered in preoperative examinations. Cordlike adhesions wrapping around the right dorsal side of the SMA to the anal side of the duodenum were found via laparoscopy. The jejunum was located on the dorsal side of the SMA and was pulled by the adhesions, accompanied by concomitant duodenal obstruction; consequently, the jejunum appeared to be compressed between the SMA and the aorta. Therefore, this case masqueraded as SMA syndrome.

CASE PRESENTATION

A 36-year-old thin man was admitted for epigastric pain after meals and vomiting for 2 weeks. Gastrointestinal endoscopy and computed tomography (CT) revealed a stricture of the duodenum, and the patient was referred to our department for further examination. On examination, his height was 175 cm, his weight was 57 kg, and his body mass index (BMI) was 18.2 kg/m². His abdomen was distended, but no tenderness was noted. CT revealed a stricture of the duodenum on the oral side of the SMA (Fig. 1A). The aorta SMA angle was 16 degrees, and the aorta SMA distance was 8 mm (Fig. 1B). The upper gastrointestinal series revealed dilatation of the stomach and duodenum (Fig. 1C), and the patient was diagnosed with SMA syndrome. He received conservative therapy with a nasogastric tube and an infusion. He was discharged on the hospital day 24. Then, he was admitted 3 more times every 2–9 months. Two years and 3 months after his first admission, he presented with abdominal fullness and vomiting and was admitted to our hospital. His symptoms disappeared on the second hospital day, and he decided to undergo surgery. Preoperative enhanced CT, upper gastrointestinal series, and enema revealed the SMV rotation sign, a right-sided small bowel, and a left-sided entire colon; therefore, he was diagnosed with stricture of the duodenum with malrotation (Fig. 2). Laparoscopic surgery was performed on the ninth hospital day. Under general anesthesia in the supine position, a 20-mm skin incision was made at the umbilicus, a 12-mm trocar was inserted, and a pneumoperitoneum was established. Then, three 5-mm trocars were inserted into the bilateral upper abdo-

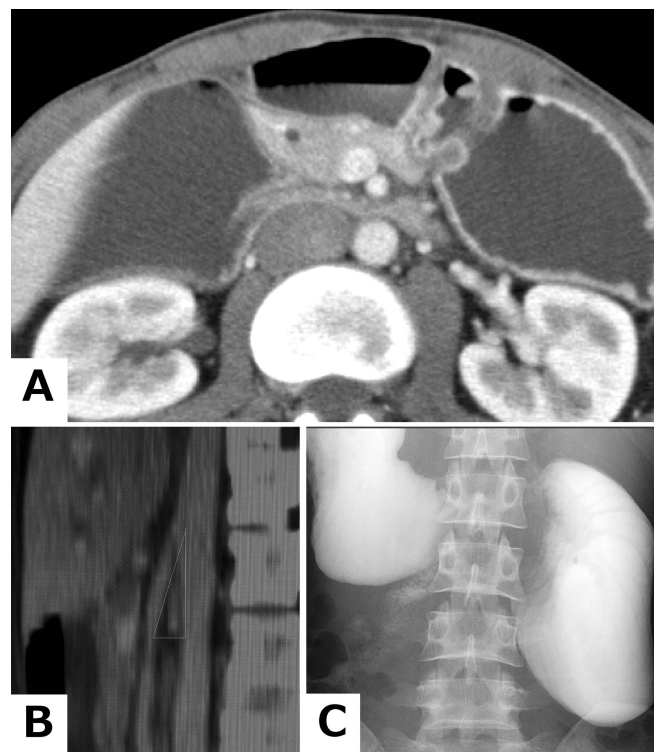


Figure 1. Imaging study during first admission. **A.** Coronal plane of the enhanced CT: a stricture of the duodenum on the oral side of the SMA. **B.** Sagittal plane of the enhanced CT: the aorta SMA angle was 16 degrees, the aorta SMA distance was 8 mm. **C.** Upper gastrointestinal series: dilatation of the stomach and duodenum was seen.

men and the right lower abdomen. A right-sided small bowel and a left-sided colon were noted. The dilated descending part of the duodenum was observed and the anal side of descending part of the duodenum, which ran to the right caudate side, was also observed. A membrane-like adhesion was identified from the retroperitoneum on the right side of the inferior vena cava (IVC) to the end of duodenum on the lower side of the liver (Fig. 3A). As the membrane-like adhesion was dissected, adhesions extending to the surface of the IVC and the dorsal side of the hepatoduodenal ligament were identified and sharply dissected (Fig. 3B). Several cordlike adhesions wrapping around the right dorsal side of the SMA to the anal side of the duodenum were observed on the dorsal side of the hepatoduodenal ligament (Fig. 3C), which were dissected to complete the division of the duodenal adhesions (Fig. 3D). Ladd's band was not observed. Last, prophylactic appendectomy was performed. The postoperative course was uneventful. Meals were initiated on the second day after surgery, and the patient was discharged on the sev-

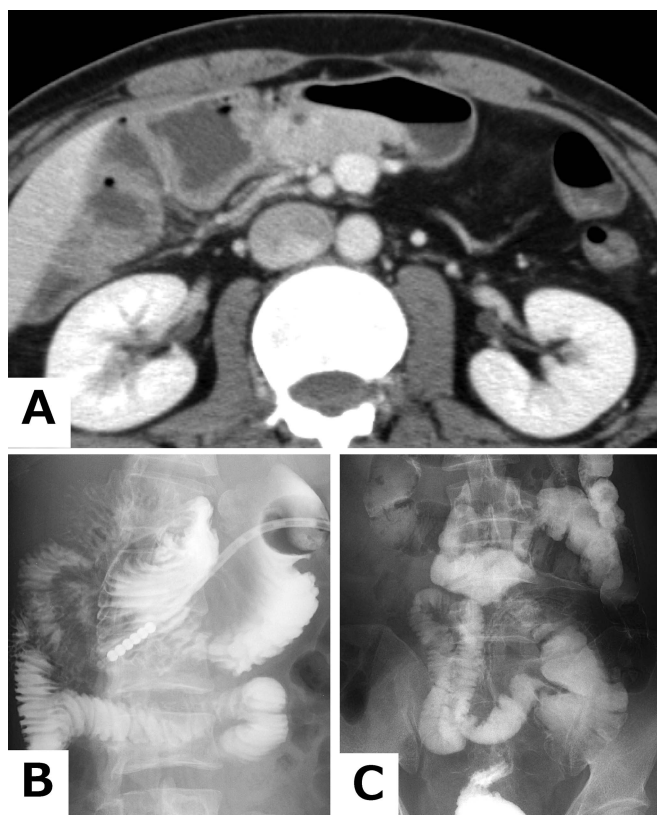


Figure 2. Preoperative imaging study during fifth admission. **A.** Enhanced CT: SMV rotation sign. **B.** Upper gastrointestinal series (right anterior oblique position): a right-sided small bowel. **C.** Enema (supine position): a left-sided entire colon.

enth postoperative day. No recurrence has been observed for 1 year and 7 months after the operation.

DISCUSSION

Malrotation describes a rotational anomaly of the midgut and is a rare condition in adults.¹ During the fetal period, the midgut portion of the intestines undergoes normal rotation during the first 4–6 weeks of gestation. The intestine herniates into the umbilical cord and back into the abdominal cavity with 270-degree counterclockwise rotation around the SMA during the next 10–11 weeks of gestation. The fourth portion of the duodenum and the proximal jejunum of the intestines form a C-loop, including development of the ligament of Treitz. The cecal portion of the intestine becomes fixed to the retroperitoneum.^{3,9} Incomplete rotation results in the 2 typical abnormalities associated with malrotation symptoms. A narrowed mesenteric root is caused by a reduced distance between the ligament of Treitz and the cecum,² and a

fibrotic band known as Ladd's band crosses over the duodenum, which connects the cecum to the lateral abdominal wall and fixes the ascending colon from the cecum to the retroperitoneum.³ These anatomical anomalies lead to midgut volvulus associated with a narrowed common mesentery,^{3,4} often causing catastrophic loss of the small intestine due to strangulation.⁷ Additionally, duodenal obstruction occurs due to compression of Ladd's band.^{3,8} Rotation anomalies are classified into non-rotation, mixed rotation, or malrotation and reverse rotation.⁸ This patient exhibited the nonrotation type, and Ladd's band was absent.

Diagnosing malrotation in adults is considered difficult.^{5,7,8} Sudden onset of bilious vomiting in infants is a typical condition² due to midgut volvulus and duodenal stricture associated with Ladd's band. In older children and adults, symptoms include postprandial bloating, intermittent cramping, and occasional vomiting.² Many individuals remain asymptomatic, and the condition is found incidentally later in life.⁸ The diagnosis is often delayed or misjudged because adult patients with malrotation present to the hospital infrequently, lack obvious indications, and develop symptoms suggestive of obstruction.⁵ For this patient, it was difficult to notice malrotation due to only vomiting after meals.

Midgut malrotation is easily recognized when imaging studies are carried out. The double-bubble sign on radiography can represent stricture of the duodenum by Ladd's band in infants.¹¹ The superior mesenteric vein (SMV) rotation sign, in which the SMV lies on the left ventral aspect of the SMA rather than the right ventral aspect,¹⁰ and the whirlpool sign, which appears as “whirling” of the mesenteric vessel trunk,¹² are useful radiographic findings on CT and ultrasound for identifying malrotation, especially with volvulus. Malrotation is also diagnosed by upper gastrointestinal series and enema use,^{8,10} revealing the duodenojejunal flexure at the caudal side of the gastric outlet,² the absence of the normal horizontal third part of the duodenum, a right-sided small bowel, and a left-sided colon.^{8,10,11} The diagnostic rate on upper gastrointestinal series is 100% in adults.⁵ This patient was diagnosed with duodenal stricture of the midgut malrotation by enhanced CT, upper gastrointestinal series and enema, and surgery was subsequently carried out.

Why was this patient initially diagnosed with SMA syndrome? SMA syndrome is a rare condition in which the third portion of the duodenum is intermittently compressed between the aorta and the SMA.¹³ The symptoms are obstructive manifestations and weight loss. As the

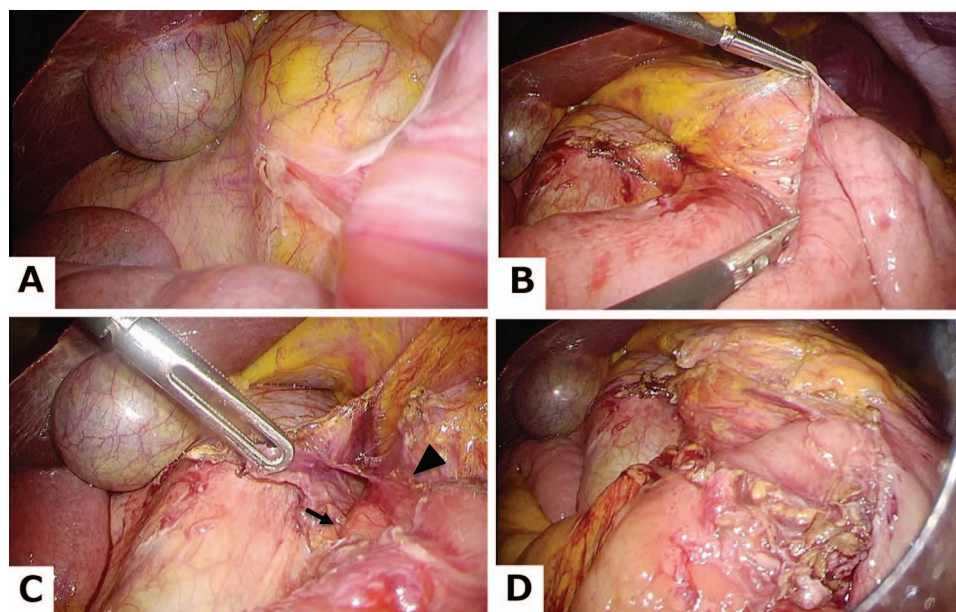


Figure 3. Operative findings. **A.** Membrane-like adhesion from the retroperitoneum on the right side of the IVC to the duodenum. **B.** Adhesions extending to the surface of the IVC and the dorsal side of the hepatoduodenal ligament were dissected. **C.** Cordlike adhesions (arrowhead) wrapping around the right dorsal side of the SMA (arrow) to the anal side of the duodenum. **D.** Completing the division of the duodenal adhesions.

normal aorta SMA angle is 38–65 degrees, decreasing the angle to < 25 degrees will decrease the aorta SMA distance to < 10 mm and cause compression of the third part of the duodenum.¹⁴ Konen et al reported that the mean aorta SMA angle of patients with SMA syndrome was 9 degrees (range 8–10 degrees) and that the mean aorta SMA angle of healthy volunteers was 44.4 degrees (range 28–65 degrees) in their study using CT angiography with 3-dimensional reconstructions.¹³ This patient was referred to our hospital with epigastric pain after meals and vomiting, and CT revealed stricture of the duodenum on the oral side of the SMA, dilatation of the stomach and duodenum (Fig. 4A), narrowing of the aorta SMA angle, the intestine positioned behind the SMA, the colon positioned from the right side to the left side in the abdominal cavity (Fig. 4B), and no SMV rotation sign. Therefore, he was diagnosed with SMA syndrome again at the time of his fifth admission. When he decided to undergo surgery, previous CT scans were reviewed retrospectively. Evidence suggestive of malrotation was noted when his symptoms disappeared during his second admission. Subsequently, preoperative enhanced CT performed after his symptoms disappeared revealed loss of duodenal obstruction, a right-sided small bowel and left-sided colon, and the SMV rotation sign. These phenomena at the time of onset suggested a 180-degree counterclockwise rotation

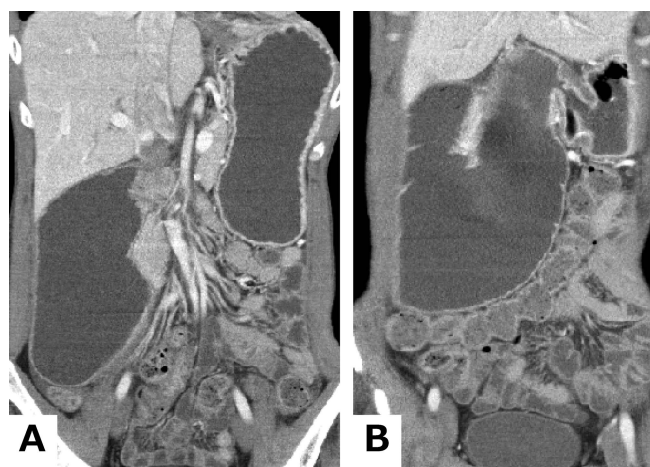


Figure 4. Enhanced CT during first admission. **A.** Duodenal stricture and dilatation of the stomach and duodenum. **B.** Colon positioned from the right side to the left side in the abdominal cavity.

around the SMA due to something pulling on the anal side of the duodenum, which was useful for the operation.

The Ladd procedure is the standard surgical treatment for malrotation and involves placing the bowel in a position of nonrotation, widening the mesenteric base, and dividing Ladd's band, a congenital adhesive band compressing

Table 1.
Successful Operative Management for Duodenal Stricture in Adult Patients With Malrotation Without Ladd's Band

Year	Author	Age (years)	Sex	Symptom	Operative Findings	Procedures	Type of Malrotation
2014	Saxena ¹⁸	55	M	Abdominal pain with distention and vomiting	The third part of the duodenum and the terminal ileum located posterior of SMA were compressed by SMA pedicle.	Duodenojejunostomy, ileocolic anastomosis, appendectomy	Nonrotation
2013	Motomura ¹⁹	66	F	Vomiting after meal	The duodenum was surrounded with cordlike structures and bending.	Division of adhesions, fixing duodenum with retroperitoneum	Nonrotation
2017	Current case	36	M	Vomiting after meal	Cordlike adhesions wrapping around the right dorsal side of SMA to anal side of the duodenum were found.	Laparoscopic division of adhesions, appendectomy	Nonrotation

the duodenum and causing chronic obstruction. The Ladd procedure is used for children,¹⁵ but it is also suitable for adult malrotation patients.¹⁶ Furthermore, Matzke et al⁵ reported that the laparoscopic Ladd procedure was as feasible and safe as the open Ladd procedure for adults with malrotation without volvulus. They reported that the laparoscopic group, which included 11 patients, resumed oral intake earlier, had a shorter hospital stay, and required less intravenous narcotic medications compared with the open group, which included 10 patients. They also reported that no patients required a second operation related to volvulus or recurrent symptoms. In this patient, our preoperative diagnosis was stricture of duodenum with Ladd's band due to malrotation. However, the intraoperative findings of our patient revealed that Ladd's band was not present; adhesions from the retroperitoneum to the duodenum, especially cordlike adhesions wrapping around the right dorsal side of the SMA, were identified. Preoperative assessment was very useful for the decision to dissect these cordlike adhesions and anastomosis was avoided. The patient has had no recurrence for 1 year and 7 months after the operation.

Jit and Grewal¹⁷ examined 100 cadavers and reported that the suspensory muscle of the duodenum could be defined as a triangular muscle with its base attached caudally to the duodenum and its apex located cranially in the region of the origins of the superior mesenteric and coeliac arteries. Additionally, the muscle was attached to the third and fourth parts of the duodenum or to the duodenojejunal flexure. They also reported that it contains smooth muscles, collagen fibers, nerves, and blood vessels. This patient's cordlike adhesions, originating from the root of the SMA to the anal side of the duodenum, pulled duodenum to have fallen into place as a normal rotation at the time of onset. If cordlike adhesions were collected during surgery, then histological examination was carried out to confirm the suspensory muscle.

Laparoscopy is a beneficial and minimally invasive technique for adult patients with malrotation.⁵ Two cases of successful operative management for duodenal stricture in adult patients with malrotation without Ladd's band have been published in English-language journals, which were retrieved from a PubMed search using the keywords "malrotation," "duodenum," and "adult" (Table 1). As both cases underwent open laparotomy, our case represents the first report of a laparoscopic procedure for duodenal stricture in an adult patient with malrotation without Ladd's band.

CONCLUSION

We successfully performed laparoscopic surgery on an adult patient with malrotation, especially in terms of dissection of the cordlike adhesions originating from the root of the SMA. As the jejunum was located on the dorsal side of the SMA and malrotation was accompanied by duodenal obstruction at the time of attack, this case masqueraded as SMA syndrome. The minimally invasive procedure described here is feasible for the treatment of duodenal stricture with malrotation.

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