Case Report

Left-sided cecal diverticulitis associated with midgut malrotation

Jia-Hui Chen a, b

Introduction

Midgut malrotation is a congenital anomaly in the embryological development of intestinal rotation. It has been estimated to affect approximately one infant in 500 live births [1, 2]. However, the true incidence is difficult to determine as a substantial number of cases go undetected throughout life. The vast majority of complications associated with midgut malrotation present in the 1st month of life and 60–85% of cases are diagnosed in this age group [1, 2]. It is reported that more than 90% of patients present by their first birthday [3]. Adult midgut malrotation is very rare, and its incidence has been reported to be between 0.0001% and 0.19% [3, 4]. Most adult diagnoses of midgut malrotation are made in asymptomatic patients; either on imaging investigations for unrelated conditions or during surgery for other pathology. This scenario of incidental diagnosis is becoming increasingly common, particularly with the improvements and increased use of diagnostic imaging techniques in modern practice. Due to the abnormal cecal position caused by malrotation, a patient presenting with left-sided cecal diverticulitis will demonstrate atypical symptoms, with pain projecting to the left of the midline. We present the first reported case of ruptured left-sided cecal diverticulitis associated with midgut malrotation.

Case report

A 26-year-old male had an emergency admission with a 2-day history of left iliac fossa (LIF) pain associated with nausea in the absence of vomiting and no change in bowel habits. He did not have any significant medical history and no history of abdominal surgery. On examination, his body temperature was 38.3°C, and he had tachycardia (105 beats/min) with a normal blood pressure (132/80 mmHg). Abdominal examination revealed tenderness in the LIF, normal bowel sounds and a normal digital rectal examination. The urinalysis was normal. Laboratory tests showed an elevated white cell count (18.4 × 10^9 cells/L) and C-reactive protein concentration (64 mg/L). A plain abdominal radiograph showed no abnormalities. Abdominal computed tomography (CT) revealed a thickened, inflamed appendix with fecalith images in the LIF and some features compatible with midgut malrotation [Figure 1]. An emergency laparotomy performed through a midline incision confirmed midgut malrotation with the small bowel located on the right side of abdomen and ruptured cecal diverticulitis with abscess formation in the LIF [Figure 2]. A cecectomy was performed with no attempt

Access this article online

Quick Response Code:

Website: www.tcmjmed.com

DOI: 10.4103/tcmj.tcmj_190_17

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

How to cite this article: Chen JH. Left-sided cecal diverticulitis associated with midgut malrotation. Tzu Chi Med J 2018;30(1):47-50.
Colonic diverticulosis is etiologically related to a chronic low-fiber diet [5]. The increased incidence of diverticulosis has coincided with the increasing refinement of cereals by the food industry, and the disease is more prevalent in countries with a high degree of industrialization and a generally low intake of dietary fiber. Diverticulosis has been considered a rare condition in patients younger than 40 years old and is thought to affect approximately 50%–70% of patients over 80 years old. Previous estimates of the percentage of patients with diverticular disease younger than 40 years old are in the order of 10% [5].

The presence of diverticula also varies according to anatomic location. More than 90% of the diverticula in Western societies are on the left side, and they can be found in the sigmoid and descending colon. Isolated right-sided colonic diverticula are found more commonly in patients from Asian countries, although some recent small studies have suggested an increase in left-sided disease in some areas of Asia [6].

To date, no population-based studies have been able to identify a specific group of patients at higher risk of diverticulitis [7]. Acute diverticulitis conventionally has been considered a disease of patients over 50 years old by many authorities [8]. Less than 20% of patients who present with diverticulitis are younger than age 45, based on a large retrospective review of hospitalizations for this condition [7]. In one report, acute diverticulitis was considered more aggressive in younger and obese patients than in older adults [8]. The patient was diagnosed with cecal diverticulitis intraoperatively. This raises the question of why did our patient develop diverticular disease at the age of 26. The patient had a high body mass index of 37 kg/m² and low intake of dietary fiber, which may have led to diverticular formation.

Midgut malrotation is, by comparison, is relatively uncommon. Depending on the location of the cecum, patients with acute cecal diverticulitis and malrotation may present atypically with left-sided abdominal pain. Left-sided abdominal pain most commonly raises the question of possible sigmoid colon diverticulitis, creating a diagnostic dilemma in these patients. Furthermore, a trend may be developing where diverticulitis, once a disease primarily of older adults, may be becoming more prevalent in younger adults [8] and may exhibit a somewhat different demographic and clinical course [9]. This phenomenon may further bias a clinician confronted with a young patient with an acute abdomen and left-sided symptoms.
Malrotation is a consequence of the altered embryologic development of the midgut [10,11]. During normal development, the liver undergoes a period of rapid growth during the fifth gestational week which causes a displacement of the midgut into the umbilical stalk and a 90° counterclockwise rotation (when viewed anteriorly) around its vascular pedicle. During the 10th week of gestation, the midgut returns to the abdominal cavity and undergoes an additional 180° counterclockwise rotation. Finally, during the 11th and 12th weeks of gestation, the cecum descends from below the liver into its final position in the right iliac fossa. Altered midgut development can result in a variety of anatomic configurations [12,13]. Of these, the most common is nonrotation [14]. In nonrotation, the preretal segment of the midgut is located exclusively on the right side of the abdomen, and the postretal segment of the midgut is located on the left side. This variant results from early return of the cecal bud, which comes to rest in the left lower quadrant of the abdomen [15]. The resultant anatomic configuration can also include a reversed relationship of the SMA and SMV as well as reversed location of the DJ junction anterior to the SMA [16]. However, not all of these anatomic anomalies were noted in our patient.

Image studies of midgut malrotation include plain radiography, ultrasonography, CT, and fluoroscopy. Abdominal radiographs, in the absence of midgut volvulus, are neither specific nor sensitive. They may show right-sided jejunal markings and absence of a stool-filled colon in the right lower quadrant. Ultrasonography may show an inversion in the SMA/SMV relationship with the SMA on the right and the SMV on the left. CT may also show an abnormal SMA/SMV relationship and the large bowel predominantly on left and small bowel predominantly on the right. An upper GI contrast study is the examination of choice when the diagnosis is suspected. The key finding of malrotation is an abnormally located DJ junction. In the frontal view, the DJ junction does not cross the midline to the left of the left-sided vertebral body pedicle, and it lies inferior to the duodenal bulb. In the lateral view, the second and third duodenum are not located posteriorly in a retroperitoneal position. Although not specific criteria for malrotation, the jejunum is commonly located to the left of the spine. In our patient, CT showed that the DJ junction did not cross the midline to the left of the left-sided vertebral body pedicle and straight duodenum. The SMA was on the left side, and SMV was on the right side [Figure 1]. From the anteroposterior view on the upper GI series, the DJ junction did not cross the midline to the left of the left-sided vertebral body pedicle and was inferior to the duodenal bulb [Figure 3].

In general, adult patients with malrotation can exhibit three different clinical presentations [17]. Some patients express symptoms and signs of acute obstruction, while other presentations are related to chronic abdominal manifestations such as pain and intermittent obstruction. Other patients complain of atypical symptoms unrelated to midgut malrotation. In the present case, malrotation was incidentally discovered in a previously asymptomatic adult patient presenting with an acute abdomen, confounding the diagnosis and treatment. A surgical operation called a “Ladd’s procedure” is performed to alleviate intestinal malrotation. The procedure involves surgical division of Ladd’s bands, widening of the small intestine mesentry, an appendectomy and correction of placement of the cecum and colon. In the medical literature, surgery is recognized as the appropriate treatment for symptomatic disease directly related to malrotation [18-20]. However, the role of surgery as a treatment option for patients with asymptomatic disease – especially adult patients – is debated.

According to a PubMed database literature search using the MeSH keyword “malrotation,” operative intervention is favored for patients with asymptomatic diseases. One reason cited for favoring surgery is the belief that if a thorough history is obtained, truly asymptotic disease is rare [18]. Furthermore, surgery provides complete symptom relief for two-thirds of patients and at least partial symptom relief for nearly all patients [21]. In addition, there is an up to 20% lifetime risk of acute volvulus or ischemia in older children and adults with malrotation, and emergency surgery is associated with a 2.5-fold increased risk of complications compared with patients who have elective Ladd’s procedures [19,22]. Given these benefits, most authors have concluded that the potential benefits of surgery appear to outweigh the risks.

One formal risk-to-benefit analysis on the management of asymptomatic malrotation with a Ladd’s procedure concluded otherwise [23]. In their statistical model, Ladd’s procedure was associated with improvement in the quality of life only for patients younger than 20 years. A mortality benefit for surgery was also limited to pediatric patients. Additional two-and three-way analyses suggested that observation alone is beneficial for patients as young as 12 years. These results led the authors to conclude that Ladd’s procedure is not indicated for asymptomatic disease in adults.

McVay et al. also suggested observation of asymptomatic malrotation [22]. In their retrospective review, 36 patients had the asymptomatic disease at initial presentation, and only six patients eventually developed symptoms. In addition, none of the initially asymptomatic patients developed volvulus or other acute complications of malrotation. The authors concluded that
nonoperative management can be an appropriate treatment strategy for asymptomatic patients, as it was rare for them to progress to symptomatic diseases or develop acute abdomens.

Recurrent volvulus as a cause of bowel obstruction following Ladd’s operation for midgut malrotation is very rare both in children and adults and very few of these cases have been reported in the literature [24-27]. Recurrent symptoms in such cases are usually considered due to adhesions, and one may be inclined to adopt a nonoperative approach. Fu et al. [24] reported only two recurrences in a series of 12 adults treated for symptomatic malrotation with one of them requiring reoperation and the other managed conservatively. It is believed that the increasing use of CT enables preoperative diagnosis with certainty as this has the overall advantage of detecting the abnormal location of the midgut as well as any other intra-abdominal anomalies. The finding of midgut malrotation should lead to suspicion of intestinal volvulus which may require early surgical intervention to prevent bowel ischemia and infarction.

While the patient described herein was diagnosed with asymptomatic nonrotation, he was not symptomatic before the acute presentation. Given the potential lack of benefit, we did not perform Ladd’s procedure in the first operation. However, he had a poor surgical outcome. Furthermore, the patient’s particular variant of malrotation, namely nonrotation, meant his anatomy was already in a post–Ladd’s procedure configuration except that the ascending colon was trapped. While the appendix was removed and the mesentery was widened after the cecectomy, as would occur with a Ladd’s procedure, these steps did not result in measurable improvement. He did well after a second operation with a right hemicolectomy. We herein suggest a right hemicolectomy might be an optional rescue procedure for left-sided diverticulitis-associated midgut malrotation with postoperative complications like our case. Radiologists should also be aware of this anatomic variant since they are most likely to make the diagnosis.

Declaration of patient consent

The authors certify that an appropriate patient consent form has been obtained. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Torres AM, Ziegler MM. Malrotation of the intestine. World J Surg 1993;17:326-31.
2. Matzke GM, Moir CR, Dozois EJ. Laparoscopic Ladd procedure for adult malrotation of the midgut with cocoon deformity: Report of a case. J Laparoendosc Adv Surg Tech A 2003;13:327-9.
3. von Flüe M, Herzog U, Ackermann C, Tondelli P, Harder F. Acute and chronic presentation of intestinal nonrotation in adults. Dis Colon Rectum 1994;37:192-8.
4. Wang CA, Welch CE. Anomalies of intestinal rotation in adolescents and adults. Surgery 1963;54:839-55.
5. Jacobs DO. Clinical practice. Diverticulitis. N Engl J Med 2007;357:2057-66.
6. Ozick LA, Salazar CO, Donelson SS. Pathogenesis, diagnosis, and treatment of diverticular disease of the colon. Gastroenterologist 1994;2:299-310.
7. Ettioni DA, Mack TM, Bear RW Jr., Kaiser AM. Diverticulitis in the United States: 1998-2005: Changing patterns of disease and treatment. Ann Surg 2009;249:210-7.
8. Zaidi E, Daly B. CT and clinical features of acute diverticulitis in an urban U.S. Population: Rising frequency in young, obese adults. AJR Am J Roentgenol 2006;12:2932-5.
9. Lahat A, Menachem Y, Avidan B, Yanai H, Sakhnin E, Bardan E, et al. Diverticulitis in the young patient – Is it different? World J Gastroenterol 2006;12:2932-5.
10. Dott NM. Anomalies of intestinal rotation: Their embryologic and surgical aspects with report of five cases. Br J Surg 1923;11:251-86.
11. Mall FT. Development of the human intestine and its position in the adult. Bull Johns Hospkins Hosp 1898;9:197-208.
12. Plackett TP, Myers J, Gigliano RA Jr. Constipation-predominant irritable bowel syndrome complicating asymptomatic nonrotation of the midgut. J Am Osteopath Assoc 2010;110:437-40.
13. Kapfer SA, Rappold JF. Intestinal malrotation—not just the pediatric surgeon’s problem. J Am Coll Surg 2004;199:628-35.
14. Schey WL, Donaldson JS, Sty JR. Malrotation of bowel: Variable patterns with different surgical considerations. J Pediatr Surg 1993;28:96-101.
15. Cherney LS. Non-rotation of mid- and hindgut with preservation of the middle mesentry; case report. Ann Surg 1954;139:241-3.
16. Pickhardt PJ, Bhalla S. Intestinal malrotation in adolescents and adults: Spectrum of clinical and imaging features. AJR Am J Roentgenol 2002;179:1429-35.
17. Welte FJ, Grosso M. Left-sided appendicitis in a patient with congenital gastrointestinal malrotation: A case report. J Med Case Rep 2007;1:92.
18. Durkin ET, Lund DP, Shaaban AF, Schurr MJ, Weber SM. Age-related differences in diagnosis and morbidity of intestinal malrotation. J Am Coll Surg 2008;206:658-63.
19. Moldrem AW, Papaconstantinou H, Broker H, Megison S, Jeyarajah DR. Late presentation of intestinal malrotation: An argument for elective repair. World J Surg 2008;32:1426-31.
20. Gohl ML, DeMeester TR. Midgut nonrotation in adults. An aggressive approach. Am J Surg 1975;129:319-23.
21. Masson RT, Franklin PA, Wagner CW. Malrotation in the older child: Surgical management, treatment, and outcome. Am Surg 1995;61:135-8.
22. McVay MR, Kokoska ER, Jackson RJ, Smith SD. Jack Barney award. The changing spectrum of intestinal malrotation: Diagnosis and management. Am J Surg 2007;194:712-7.
23. Malek MM, Burd RS. The optimal management of malrotation diagnosed after infancy: A decision analysis. Am J Surg 2006;191:45-51.
24. Fu T, Tong WD, He YJ, Wen YY, Luo DL, Liu BH, et al. Surgical management of intestinal malrotation in adults. World J Surg 2007;31:1797-803.
25. Biko DM, Anupindi SA, Hanhan SB, Bliiman T, Markowitz RI. Assessment of recurrent abdominal symptoms after Ladd procedure: Clinical and radiographic correlation. J Pediatr Surg 2011;46:1720-5.
26. Panghaal V, Levin TL, Han B. Recurrent midgut volvulus following a Ladd procedure. Pediatr Radiol 2008;38:471-2.
27. El-Gohary Y, Alagtal M, Gillick J. Long-term complications following operative intervention for intestinal malrotation: A 10-year review. Pediatr Surg Int 2010;26:203-6.