Case study

Midgut Volvulus with Intestinal Malrotation in an Adult: A Case Report

Abstract

Background: Intestinal malrotation is a rare congenital disorder characterized by non-rotation or partial rotation of the small bowel around the superior mesenteric arteries during the embryonic stage, resulting in mid-gut volvulus. Approximately 64–90% of patients' symptoms manifest as acute intestinal obstruction due to midgut volvulus before the age of one year, but the presentation in adulthood is infrequent (0.2–0.5%). Case report: A 26-year-old lady came to us complaining of intermittent colicky discomfort, vomiting, and constipation for the previous three months. The patient's vitals were stable clinically, with 112/64 mmHg blood pressure, 72 bpm heart rate, 16 rpm respiration rate, 98.4 °F (36.9 °C) temperature, and 100% O_2 saturation on room air. The abdomen was mildly distended but had no tenderness or guarding. Abdominal Contrast CT(CECT) revealed a reversal of the relationship between the superior mesenteric vessels, a whirling appearance of the mesentery, and a twisting of the mesentery vessels (whirlpool sign), indicating mid-gut volvulus and intestinal malrotation. Emergency surgical intervention was scheduled for the patient. Conclusion: Although malrotation is frequent in childhood, symptomatic midgut malrotation can occur in adults with non-specific symptoms. Imaging and a high suspicion level are critical in diagnosing.

Keywords: Adult intestinal malrotation, malrotation, midgut volvulus, whirlpool sign.

Background

Intestinal malrotation is a rare congenital condition defined by a partial or complete failure of the small bowel's 270-degree counter clockwise rotation around the superior mesenteric arteries during embryonic development, resulting in midgut volvulus. The symptoms of 64–90% of patients begin before the age of one year as acute intestinal obstruction due to midgut volvulus, although they are only present in 0.2–0.5% of adults.[1,2,3] Intestinal malrotation affects about 1 in 500 live newborns. [4,5] Up to 40% of malrotation patients

appear within the first week of infancy, and 75% to 85% are diagnosed by the age of one year.[6] The remaining 15% to 25% of people are diagnosed during childhood, but this can extend until late adulthood. However, estimating the true incidence of malrotation in the adult population is challenging because the majority will stay asymptomatic throughout their lifetimes. [7] Adults may experience a delayed diagnosis as a result of a lower degree of suspicion, which may increase morbidity and mortality. Adults require a high level of suspicion to be diagnosed with the condition. Normal embryological rotation of the intestine starts at six weeks and goes through three stages. The intestines exceed the abdomen during stage 1. The bowel contents are extruded out of the umbilicus at 8 to 10 weeks of gestation, followed by a 90-degree counter-clockwise rotation along the axis of the superior mesenteric artery and return to the belly. Stage 2 is followed by another 180-degree counter-clockwise rotation of the midgut about the superior mesenteric artery (SMA) for a total of 270 degrees. The final step is to anchor the mesentery. The duodenum becomes retroperitoneally fixed in its third section, and the cecum becomes retroperitoneally attached to the right lateral abdominal wall. The upshot of this rotation pattern is a large base mesentery.[8] Intestinal malrotation refers to any deviation from the standard 270-degree counter-clockwise rotation. The entire small bowel is on the right due to non-rotation, whereas the colon is on the left. Aberrant mesenteric bands (Ladd's bands) develop and extend from the caecum to the lateral abdominal wall, crossing the duodenum and causing duodenal blockage as a result of correct rotation and fixation of the ceco-colic limb.[9]

We present the case of a 26-year-old lady who underwent a laparoscopic Ladd’s operation for malrotation manifested as midgut volvulus.

Case Presentation

A 26-year-old lady had complained of sporadic colicky discomfort, vomiting, and constipation for the past three months. The patient had no surgery or significant medical history. The patient's vitals were stable clinically, with 112/64 mmHg blood pressure, 72 bpm heart rate, 16 rpm respiration rate, 98.4 °F (36.9 °C) temperature, and 100% O_2 saturation on room air. The abdomen was mildly distended but had no tenderness or guarding.

The blood results were all within normal levels. The patient was evaluated using a contrast-enhanced CT scan of the abdomen. On the CT scan, the small intestine was located to the right of the abdominal midline, whereas the large intestine was located to the left of the abdominal midline. The relationship between superior mesenteric vessels was reversed, resulting in a swirling appearance of the mesentery as well as twisting of the mesentery vessels (whirlpool sign), indicating midgut volvulus and intestinal malrotation **(*Figure 1*)**. The patient was hospitalized and then taken for Ladd's operation.

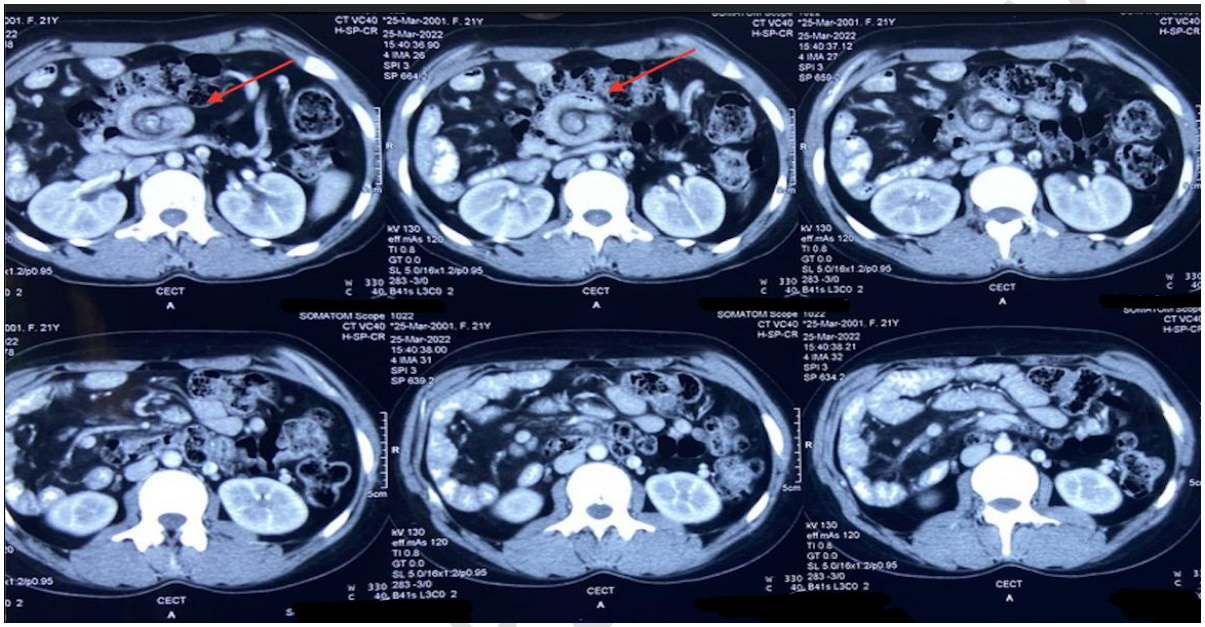

Figure 1: The swirling look of the mesentery as well as the twisting of the mesentery vessels (Whirlpool sign) (marked with red arrow)

Volvulus of the small bowel and intestinal malrotation were found during the laparoscopy. The whole of the small intestine was on the right side, and the colon was on the left. Volvulus detorsion counter clockwise, division of Colo duodenal Ladd's bands ***(Figure 2)***, straightening of the duodenum's loop, mesenteric base widening, and appendectomy were all performed laparoscopically. The Patient's postoperative recovery was uneventful, and she was discharged on the fifth postoperative day. During subsequent visits, the patient appeared to be in good health, with none of the initial symptoms appearing.

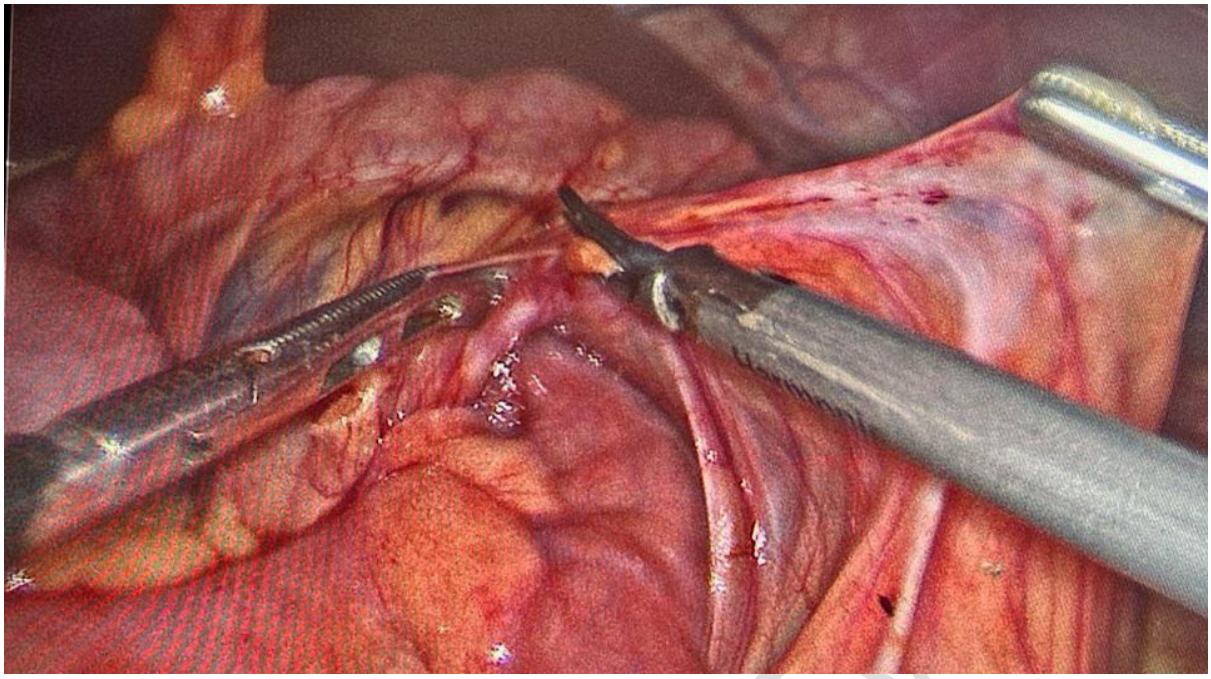

Figure 2: Division of Ladd's bands

Discussion

Malrotation is not always accompanied by symptoms. Many people live without complaint, and the anomaly is only revealed at autopsy. Some may have chronic and unexplained abdominal pain, while others may have acute episodes of severe abdominal pain. [10] Acute or chronic intestinal obstruction can cause symptoms. This can be caused by abnormal peritoneal bands (like Ladd's bands) or a volvulus. There is no standard set of symptoms associated with this clinical condition. Pain can be felt anywhere from the epigastric region to the upper left quadrant of the abdomen, and it can be described as either a cramping pain that comes and goes or constant aching discomfort. It typically occurs after eating and can last anywhere from a few minutes to an hour. Patients with chronic complaints caused by malrotation are likely to remain symptomatic until the anatomic condition is surgically addressed. [11] The lack of specific physical symptoms, as well as the low adult prevalence, make diagnosis challenging. Adult patients' symptoms are frequently misdiagnosed as irritable bowel syndrome, peptic ulcer disease, biliary and pancreatic disease, and psychiatric disorders.[12] Obstructive jaundice produced by mechanical biliary tract compression, chylous ascites, and superior mesenteric vein thrombosis caused by long-term lymphatic and vein occlusion is all rare examples of chronic volvulus.[13,14,15,16,17,18] This syndrome may also manifest acutely due to midgut volvulus, and it may lead to intestinal

ischemia and gangrene. Both of these complications may occur. If the patient survives, the effects are severe, often resulting in major intestinal gangrene, death, and small bowel syndrome.[19] The key to survival is early detection of the problem [9].

In adults, imaging is often utilized to identify intestinal malrotation. The upper gastrointestinal contrast series is highly accurate in demonstrating that the duodenojejunal junction does not cross the midline. Contrast-enhanced CT can show the entire small bowel to the right and the colon to the left, as well as an aberrant relationship between the superior mesenteric vein (SMV) and the SMA and the existence of midgut volvulus (whirlpool sign), which is described as twisting of the small bowel around the SMA.[20]

Our patient's CT scan revealed intestinal malrotation and midgut volvulus, with the characteristic "whirlpool sign" linked to intraoperative findings. Ladd's operation was performed laparoscopically on this patient. Dr. William Ladd's Ladd technique, performed in 1936, has long been the standard treatment for malrotation, particularly in children.[21] Adults have successfully had the same treatment as we have. Ladd's procedure consists of counter clockwise volvulus detorsion, division of the Colo duodenal Ladd's bands, straightening of the duodenum's c loop, widening of the mesenteric base, and appendectomy. An appendectomy is performed because a future appendicitis diagnosis may be difficult. In recent years, laparoscopic Ladd's surgery has led to the early resumption of oral tolerance and early discharge.[22,23] The most common complication of the treatment is small intestinal obstruction due to adhesions, with the majority of patients presenting within one year.[24] As a result, monitoring these patients is crucial. During the postoperative phase, our patient appeared to be doing well with the resolution of her complaints. Although Ladd's surgery is still the best option for patients with normal malrotation, a clinical dilemma occurs when patients are largely asymptomatic and are identified with malrotation incidentally or when patients are diagnosed with atypical malrotation ("malrotation variant"). The decision to operate on patients who may or may not be experiencing symptoms exposes them to a variety of risks and problems. There have been a few cases of adult midgut malrotation manifesting as volvulus. [25,26,27]

Conclusions

Although malrotation is common in children, symptomatic midgut malrotation can manifest in adults as non-specific symptoms. Making a diagnosis requires imaging and a high level of

suspicion. Ladd's procedure is a well-known malrotation operation. These patients must continue to be followed up on.

Consent

written informed consent was obtained from the patient for publication of this case report and accompanying images

References

1. McIntosh R, Donovan EJ: Disturbances of rotation of the intestinal tract. Am J Dis Child. 1939, 57:116-66. doi:10.1001/archpedi.1939.01990010125010

2. Frantzides CT, Cziperle DJ, Soergel K, Ste- wart E: Laparoscopic Ladd procedure and caecopexy in the treatment of malrotation beyond the neonatal period. Surg Laparosc Endosc. 1996 Feb;6(1):73-5. PMID: 8808567.
3. Mazziotti MV, Strasberg SM, Langer JC: Intestinal rotation abnormalities without volvulus: the role of laparoscopy. J Am Coll Surg. 1997, 185:172-6. doi:10.1016/S1072-7515(01)00900-0.
4. Filston HC, Kirks DR. Malrotation—the ubiquitous anomaly. J Pediatr Surg 1981;16:614–20. doi:10.1016/0022-3468(81)90015-4
5. Torres AM, Ziegler MM. Malrotation of the intestine. World J Surg 1993 May-Jun;17(3):326 –31. doi: 10.1007/BF01658699.
6. Andrassy RJ, Mahour GH. Malrotation of the midgut in infants and children. Arch Surg 1981 Feb;116(2):158–60. doi: 10.1001/archsurg.1981.01380140020004.
7. Gamblin TC, Stephens RE Jr, Johnson RK, et al. Adult malrotation: a case report and review of the literature. Curr Surg 2003 Sept-Oct;60(5):517–20. doi: 10.1016/S0149-7944(03)00030-8.
8. Kotze PG, Martins JF, Rocha JG, et al.: Ladd procedure for adult intestinal malrotation: Case report. ABCD. Arquivos Brasileiros de Cirurgia Digestiva (São Paulo. 2011, 24:89-91.
9. Aboagye J, Goldstein SD, Salazar JH, et al.: Age at a presentation of common pediatric surgical conditions: Reexamining dogma. J Pediatr Surg. 2014 Jun,49(6):995-999. doi:10.1016/j.jpedsurg.2014.01.039.

10. Wang C, Welch CE. Anomalies of intestinal rotation in adolescents and adults. *Surgery*. 1963 Dec;54:839-855. PMID: 14087118
11. Gamblin TC, Stephens RE Jr, Johnson RK, Rothwell M. Adult malrotation: a case report and review of the literature. Curr Surg. 2003 Sep-Oct;60(5):517-20. doi: 10.1016/S0149-7944(03)00030-8. PMID: 14972216.
12. Fukuya T, Brown BP, Lu CC. Midgut volvulus as a complication of intestinal malrotation in adults. Dig Dis Sci. 1993 Mar;38(3):438-444. doi: 10.1007/BF01316496.
13. Gohl ML, DeMeester TR: Midgut malrotation in adults. An a ggressive apprach. Am J Surg. 1975, 129:319-23. doi: 10.1016/0002-9610(75)90249-4
14. Filston HC, Kirks DR: Malrotation: The ubiquitous anomaly. J Pediatr Surg. 1981, 16(1):614-20. doi: 10.1016/0022-3468(81)90015-4
15. Wang C, Welch CE: Anomalies of intestinal rotation in adolescents and adults. Surgery. 1963 Dec,54:839-55. PMID: 14087118
16. Brandt ML, Pokorny WJ, McGill CW ve ark: Late presentations of midgut malrotation in children. Am J Surg. 1985 Dec;150(6):767-71. doi: 10.1016/0002-9610(85)90426-X
17. Spitz L, Orr JD, Harries JT: Obstructive jaundice secondary to chronic midgut volvulus. Arch Dis Child. 1983, 58:383-5. doi: 10.1136/adc.58.5.383
18. Schwartz DL, So HB: Schneider KM ve ark: Recurrent chylous ascites associated with intestinal malrotation and lymphatic volvulus. J Pediatr Surg. 1983 April;18:177-9. doi: 10.1016/s0022-3468(83)80547-8
19. Zengin A, Uçar Bİ, Düzgün ŞA, Bayhan Z, Zeren S, Yaylak F, Şanal B, Bayhan NA. Adult midgut malrotation presented with acute bowel obstruction and ischemia. Int J Surg Case Rep. 2016;22:5-7. doi: 10.1016/j.ijscr.2016.03.018. Epub 2016 Mar 18. PMID: 27015011; PMCID: PMC4844668.
20. Nichols DM, Li DK: Superior mesenteric vein rotation: a CT sign of midgut malrotation. AJR Am J Roentgenol. 1983 Oct;141(4):707-8. doi: 10.2214/ajr.141.4.707
21. Ladd WE: Surgical diseases of the alimentary tract in infants: N Engl J Med. 1936 Oct:705-8. doi: 10.1056/NEJM193610152151604

22. Matzke GM, Dozois EJ, Larson DW, et al.: Surgical management of intestinal malrotation in adults: comparative results for open and laparoscopic Ladd procedures. Surg Endosc. 2005 Oct;19(10):1416-9. doi: 10.1007/s00464-004-8249-7
23. Seymour NE, Andersen DK. Laparoscopic treatment of intestinal malrotation in adults. J Soc Laparoendosc Surg 2005 Jul-Sep;9(3):298–301. PMID: 16121875
24. Biko DM, Anupindi SA, Hanhan S.B, et al.: Assessment of recurrent abdominal symptoms after Ladd procedure: clinical and radiographic correlation. Journal of Pediatric Surgery. 2011 Sep;46(9):1720-1725. doi: 10.1016/j.jpedsurg.2011.03.018
25. Bhatia S, Jain S, Singh CB, et al.: Malrotation of the Gut in Adults: An Often Forgotten Entity. Cureus 2018 Mar 12;10(3):2313. doi: 10.7759/cureus.2313
26. Victor S Alemany MD: Adult Midgut Malrotation: A Case Report. J Surg Proce Case Rep. 2020, 1:1-5. doi: 10.1016/S0149-7944(03)00030-8
27. Sahu SK, Raghuvanshi S, Sinha A, et al.: ADULT INTESTINAL MALROTATION PRESENTING AS MIDGUT VOLVULUS; CASE REPORT. Journal of Surgical Arts. 2012, 5(1):18-21. Corpus ID: 53898533