CLINICAL VIGNETTE

Adult Presentation of Acute Midgut Malrotation

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Case Presentation

A 29-year-old female with no significant PMH presented to ER with severe sudden onset generalized abdominal pain and vomiting. She was in her usual state of health two hours prior to symptom onset and had shared a meal with a friend who was asymptomatic. She denied any recent change in bowel habits, bleeding, fevers, or chills. She reported 1.5 years of mild-to-moderate episodes of post-prandial lower-mid abdominal bloating and cramping that generally resolved with rest or bowel movement. Her symptoms had not improved with elimination diets or with a low FODMAPD diet. She had no previous surgeries and past medical and family history were unremarkable. She takes an oral contraceptive daily, does not smoke and is not currently sexually active. She drinks 2-5 glasses of wine a week.

Vitals on presentation: HR 58, RR 20, BP: 114/73, Sp02 of 99% and pain 10/10. On exam she is AAO x 3 and writhing in pain. She has a distended abdomen with generalized tenderness, guarding and rebound tenderness. CBC, chemistries lipase, amylase, urine pregnancy tests are unremarkable. CT of the abdomen shows SBO with distended right and transverse colon and dilated segments of small bowel with an ill-defined transition point. CT also shows right-sided duodenum (consistent with midgut malrotation) and an unremarkable appendix. Seven hours after presentation, she underwent emergent explorative laparotomy. Findings included cecal volvulus with healthy, non-ischemic bowel. The volvulus was detorsed and patient was admitted. After further review of her CT and improved understanding of the anatomy, the decision was made to return to the OR for surgical correction of the malrotation. On Post-op Day 3, she underwent laparoscopic Ladd procedure, ileocecectomy with primary anastomosis. Her post-op course was complicated by prolonged ileus, and she was discharged home on hospital day 12.

Discussion

Midgut malrotation is a congenital anomaly that typically presents in the first 4 weeks of life. During normal embryonal development, the 3 divisions of the GI tract herniate out from the abdominal cavity, where they undergo a 270 degree counterclockwise rotation around the superior mesenteric vessels. The bowel then return to the abdominal cavity with fixation of the duodenojejunal loop to the left of the midline and the cecum in the right lower quadrant. Any variation in this process is known as intestinal malrotation. This embryonic phenomenon was first described in 1923 and the surgical treatment was described in 1936, by William Ladd. The Ladd procedure remains the cornerstone of treatment today.

Intestinal malrotation occurs between 1 in 200 and 1 in 500 live births in the U.S, with symptomatic malrotation occurring in 1 in 6000 live births.[1] Age of presentation is variable; with 50%-85% presenting in the first month of life and 15% presenting in adulthood.[1] Adult presentation can be categorized as incidental, acute, or chronic. Incidental malrotation can be found on imaging or laparotomies for other health conditions. Patients with chronic presentation of malrotation have non-specific gastrointestinal symptoms including intermittent colicky abdominal pain, bloating, weight loss, and nausea or vomiting. These symptoms may arise from intermittent bowel obstruction from Ladd bands fibrous stalks of peritoneal tissue that arise from malrotation or intermittent midgut volvulus.

Acute presentation with volvulus is reported to be 0.2-0.5% of adult cases of midgut malrotation. One recent literature review of 92 adult cases of acute presentation of intestinal malrotation reported the average age of presentation to be 40 and a 1.7:1 ratio of male to female.[2] There is a high risk of ischemia to bowel supplied by the SMA and a 5% mortality association of midgut volvulus. Of the 92 adult midgut volvulus cases, 19% required bowel resection with an average resection length of 121 cm. The case being reported had 25 cm of bowel resection. The treatment of intestinal malrotation is the Ladd procedure, which involves detorsion of the volvulus, division of Ladd's bands, broadening of the small bowel mesentery, and then divisions of adhesions around the SMA.

This case highlights the importance of maintaining a high index of suspicion for anatomical anomalies for non-specific intermittent chronic abdominal pain that often presents to the primary care doctor. The diagnosis of malrotation is often delayed, as it was in this case. Our patient had 1.5 yrs of intermittent pain prior to presenting to the ER. She presented multiple times to her primary care doctor office and was appropriately referred to gastroenterology and gynecology. She had an unremarkable abdominal and pelvic ultrasound. Her symptoms were minimally improved with dietary changes, probiotics, or laxatives. She reported moderate symptom improvement with decreased stress, further supporting a diagnosis of IBS and delaying the malrotation diagnosis. Had the diagnosis been established prior to ER presentation, her

treatment would have remained the same. Since there is no way to predict who will develop catastrophic bowel necrosis, all midgut malrotation (incidental, chronic, acute) is treated with the surgical Ladd procedure. Fortunately, this patient is now 3 months post-operative and is recovering well.

REFERENCES

1. **Torres AM, Ziegler MM**. Malrotation of the intestine. *World J Surg*. 1993 May-Jun;17(3):326-31. Review. PubMed PMID: 8337878.
2. **Butterworth WA, Butterworth JW**. An adult presentation of midgut volvulus secondary to intestinal malrotation: A case report and literature review. *Int J Surg Case Rep*. 2018;50:46-49. doi: 10.1016/j.ijscr.2018.07.007. Epub 2018 Jul 11. PubMed PMID: 30077833; PubMed Central PMCID: PMC6083817.

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