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CLINICAL VIGNETTE

Intestinal Malrotation in an Adult Patient

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The patient is a 51-year-old female who developed acute onset of severe midepigastric pain that doubled her over while on vacation 18 months ago. The pain abated after a few hours, and she went out to dinner. After dinner, the pain recurred and got progressively worse. She presented to the local emergency room and received IV fluids and pain medication. A CT scan was done that was reported as normal. Between midnight and two o'clock in the morning, she was well enough to go back to her hotel room. She returned to the ER in the morning and had an abdominal ultrasound, which was also normal. Since returning home, she has experienced episodes of nausea and early satiety and continuous abdominal pain. Over the next few months, she lost 20 lbs. because of inability to eat without ensuing pain. She had no change in bowel habits and relates normal stools. There has been no overt gastrointestinal bleeding. She was given Omeprazole 40 mg per day with minimal relief of her symptoms.

She has a negative family history for GI disease and is not diabetic. She currently takes paroxetine, ondansetron, omeprazole, and OTC decongestants as needed. She awakens in the morning with some discomfort but manages to eat some oatmeal and a cup of coffee. On weekends, she may have some eggs. Lunch and dinner are sparse because of increased pain if she eats too much. She was seen by a Gastroenterologist who performed an endoscopy and a colonoscopy, which was described as normal. Biopsies for H. Pylori were negative. The review of systems was otherwise normal.

Physical exam revealed a well-developed, well-nourished female in no acute distress. Blood pressure was 120/70, pulse 68 and regular, height 5 feet 4 inches, and weight was 129 lbs. BMI was 23.86. The exam was otherwise unremarkable.

An UGI series from an outside facility revealed all of the small bowel was in the right side of the abdomen with the colon confined to the left side of the abdomen.

The findings were compatible with a complete intestinal malrotation. She was referred to Gastrointestinal Surgery at UCLA. At the time of surgery, she was found to have a malrotation of the bowel with a duodenal obstruction due to Ladd's Bands. She underwent a Ladd's procedure, which included lysis of dense adhesions causing partial obstruction of the duodenum, Kocherization of the duodenum, and an

appendectomy. Since her surgery, she has been pain free and has resumed a normal diet.

Discussion

The surgical approach to intestinal malrotation was first published in the NEJM in 1936 by William E. Ladd. He presented a paper on feeding problems in infants and children with various congenital gastrointestinal problems including pyloric stenosis and malrotation with duodenal obstruction.¹ In 2003, a paper in Current Surgery by Gamblin et al³ described a case of intestinal malrotation in an adult.² The patient was a 36-year-old woman who presented with abdominal pain, nausea, and vomiting who had undergone a laparoscopic cholecystectomy 3 years prior for abdominal pain. That surgery failed to relieve her symptoms. A CT scan from 6 months earlier was read as normal. After a trial of nasogastric suction, she felt better but had immediate pain when fed. A UGI series was consistent with intestinal malrotation, and she underwent a Ladd's procedure. Multiple articles in the literature have documented the presence of intestinal malrotation in adults.^{3,4}

Intestinal midgut malrotation usually presents in the first month of life and is a rare occurrence in adulthood. It is estimated to occur in 0.2-0.5% of the adult population. Symptoms usually occur post-prandially, and the patient may present with vague abdominal pain or with an acute bowel obstruction and intestinal ischemia. The most catastrophic event is a midgut volvulus. The syndrome occurs due to a lack of or incomplete rotation of the fetal intestines around the axis of the superior mesenteric artery during fetal development. During fetal development, the rapidly growing midgut herniates through the umbilical orifice. The duodenum rotates 90 degrees counterclockwise around the superior mesenteric artery. At this time, the cephalad limb elongates forming multiple loops. The caudad limb remains straight. During the second stage, the abdominal cavity grows large enough to accommodate the growing intestines, and they return to the abdomen and rotate another 180 degrees. The hindgut gets pushed to the left. During the third stage, there is some resorption of parts of the intestine, and the duodenum and the lower left colon become retroperitonealized. Malrotation can interfere with any of the normal embryologic rotations.

In a retrospective review of 170 patients diagnosed with intestinal malrotation at the Massachusetts General Hospital, 31% were infants, 21% were children, and 48% were adults. Infants usually presented with emesis (93%), and adults most commonly presented with abdominal pain (87%), less often with nausea or emesis. The incidence of volvulus declined with age. Fifty-nine percent of children and 32% of adults experienced symptoms for years before diagnosis. The average age at diagnosis in adults was 48 and the oldest patient 89.⁵

Intestinal malrotation has been a diagnosis most often made in infancy, but it is apparent that it exists to a significant degree in adults. In this patient after careful re-questioning of her past history, it became obvious that she was having episodic abdominal distress for many years as a teenager and young adult. She grew up in a small town with limited care available. Her CT scan done while on vacation did not report classic findings of malrotation, although after making the radiologist aware of the patient's presentation, the interpretation was changed to reflect the accurate diagnosis.

Images

Image 1. CT scan originally read as normal, but it can be seen that the small bowel is confined to the right side of the abdomen and colon to the left consistent with malrotation.



Image 2. UGI series confirming that the small intestine is isolated to the right side of the abdomen, and the colon, with the exception of the cecum, is on the left. The duodenum does not cross the midline.



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