A Malrotated, Incarcerated Appendix within an Epigastric Hernia

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### Abstract

**Background:** A male patient with previously undiagnosed intestinal malrotation presented with symptomatic appendiceal incarceration within an epigastric hernia.

**Summary:** A 40-year-old male who presented with non-reducible ventral abdominal mass that was tender to palpation on physical examination. He was found to have appendiceal incarceration within an epigastric ventral hernia defect secondary to congenital malrotation of the bowel, as initially identified on computed tomography. The patient was taken to the operating room emergently, where reduction of the hernia sac, appendectomy, and proper rotation of the colon was performed. Pathologic findings were consistent with early inflammation within the appendix and a normal appearing hernia sac. Intestinal malrotation is an embryologic defect known to put patients at risk for serious complications, starting from birth and continuing throughout life. Though, typical management involves intervention in the neonatal and childhood periods, the rate and variety of complications that result from foregoing surgical correction in the absence of clinical symptoms is largely unknown. This case illustrates a potential complication associated with failure to correct both congenitally malrotated bowel and an epigastric hernia.

**Conclusion:** Intestinal malrotation can manifest and present throughout adult life - as seen in this rare complication in the setting of another developmental defect. This case highlights the need for appropriate identification, embryological understanding and surgical management of problems such as these in the acute setting.

**Keywords**

Malrotation, Epigastric hernia, Hernia, Incarceration

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**Case Description**

While uncommonly found pathologically together in the adult patient, visceral herniation and gut rotation go hand-in-hand during embryologic development. Rapid growth of the small bowel around week 6 of gestation results in physiologic herniation of the gut through the umbilical ring; the small bowel then rotates a full 270 degrees in a counter-clockwise direction to arrive at its normal anatomic position within the peritoneal cavity. Defects and missteps in these processes can have broad consequences for patients of all ages, as seen in our patient here.

A 40-year-old gentleman with a history of an epigastric hernia presented with two days of abdominal pain, discomfort, and a palpable lump in the anterior abdominal wall; his primary care doctor had promptly sent him to the Emergency Room after a brief office evaluation. He denied any fevers, chills, nausea, vomiting, bowel changes or failure to move his bowels at that time. He also denied any previous surgeries. On physical examination, a tender, supra-umbilical hernia was noted with palpable loops of bowel present in hernia sac; the hernia was not reducible even after abdominal relaxation and narcotics administration. No peritoneal signs were noted, and the abdomen was not distended. A CT scan obtained in the emergency room was consistent with colonic malrotation along the midline; the appendix and mesentery were also identified within the her-
sis or cecopexy was performed. Pathologic analysis of the appendix was consistent with early inflammatory changes. The patient progressed as expected and was discharged the next day.

Discussion

Often a concern for the pediatric population, intestinal malrotation is considered a surgical emergency because of the risk of vascular compromise to the abdominal viscera. Clinical presentation with bilious emesis is almost always assumed to be malrotation until proven otherwise in children, and it frequently prompts rapid evaluation and intervention. The prevalence of malrotated guts within the general population is difficult to assess however, especially since not all present symptomatically; it is estimated that 1 case out of 6000 live births are identified out of clinical necessity, approximately 90% of which present during infancy [1]. The remaining 10% present in a variety of different ways in both childhood and adulthood. Malrotated patients beyond the neonatal period have been known to present with intestinal obstruction, chronic abdominal pain, peritonitis, malabsorption, biliary obstruction, delayed small bowel transit, and even midgut volvulus (A common presentation in neonates as well) [2,3]. Abdominal pain is the most common complaint that brings adults to medical attention, while the incidence of midgut volvulus (a serious concern in the neonatal period) declines with age [4]. Diagnosis in adults relies much more heavily on computed tomography for symptomatic malrotation rather than the contrast studies used in infants; serendipitous diagnosis intra-operatively is also a common occurrence. Surgical management in both adult and pediatric populations traditionally includes the Ladd’s Procedure, or division of peritoneal bands, reduction of any gut volvulus, appendectomy and proper positioning of the gut with or without fixation within the peritoneum [5].
Lessons Learned

Adults presenting with sequelae of intestinal malrotation should undergo surgical management for acute issues, though it is unclear whether or not these patients would benefit from pre-emptive intervention in the setting of congenital malrotation. Co-existing conditions such as ventral hernias should be managed accordingly.

Conflicts of Interest/Sources of Funding

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