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Perforated Gastric Volvulus in a Patient with Prior Surgery for Hirschsprung's Disease

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Background	A male patient with previous surgery for Hirschsprung's disease as an infant presented with a perforated gastric volvulus.
Summary	Our patient, a young adult with a history of Hirschsprung's disease, presented with severe abdominal pain. Previous surgeries include colostomy formation, pull-through procedure, ostomy reversal, and lysis of adhesion as a teenager due to small bowel intruction. Prior to presentation, the patient's pain had worsened over 24 hours without relief. Diagnostic imaging revealed gastric volvulus with pneumoperitoneum, leading to emergent surgery. Resection of the perforation and gastropexy were performed successfully, but the patient developed an intraabdominal abscess postoperatively, necessitating percutaneous drainage. While surgical intervention is indicated for many childhood conditions, patients remain at risk for long-term sequela afterward. This case highlights the possibility of rare but emergent abdominal pathology following surgical procedures in individuals with congenital disorders.
Conclusion	Our case underscores the lifelong risk of complications that patients with congenital diseases requiring surgical management carry into adulthood, extending beyond the time of their initial operation.
Key Words	Hirschsprung's disease; gastric volvulus; adhesions

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Case Description

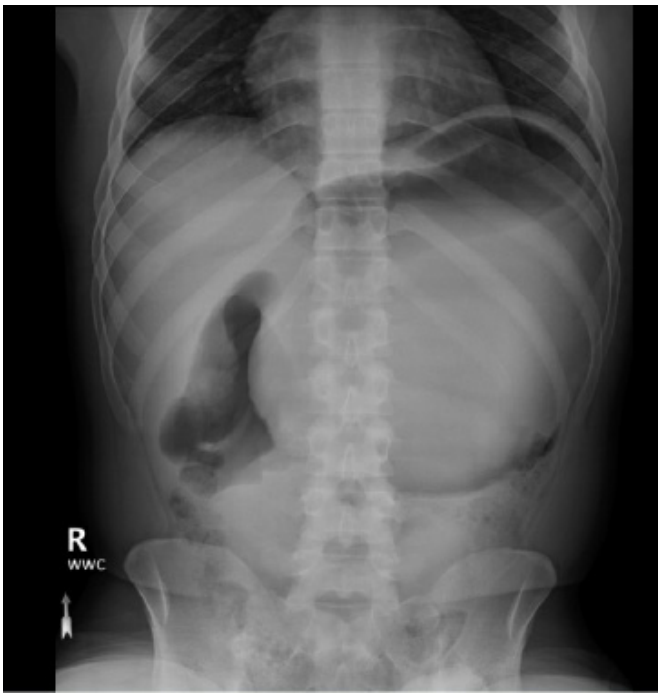
Hirschsprung's disease (HD) is a uncommon development disorder affecting the enteric nervous system, characterized by the absence of ganglion cells in the distal colon, causing functional obstruction. It occurs in approximately 1 in 5000 live births. Patients with HD often undergo surgery in infancy to address the issue. While surgery is necessary for definitive treatment, it carries long-term risks. We present a case of a young adult with HD who developed gastric volvulus and subsequent gastric perforation due to adhesions from previous abdominal surgeries.

A 26-year-old man presented to the emergency department complaining of increasingly severe abdominal pain lasting more than 24 hours. He has a medical history of Hirschsprung's disease and has undergone several surgeries, including a diverting colostomy as a newborn, a pull-through procedure at four months, and colostomy reversal at one year. Additionally, he had an exploratory laparotomy with adhesion removal at 16 years due to small bowel obstruction. Initially, the pain was in his lower abdomen but had shifted to the area around his periumbilical area by the time of presentation. He described the pain as sharp and constant, 10/10 in intensity, and associated with nausea, but no vomiting, fever, chills, or other symptoms.

At the time of exam, the patient displayed signs of moderate distress, exhibiting hypertension (166/98 mmHg) and tachypnea (22 breaths per minute), with other vital signs within normal ranges. Notably, two well-healed surgical scars were observed upon inspection: one transverse scar in the right lower quadrant and another midline scar. Lab work was significant for an elevated white blood cell count of 14.89×10^9/L.

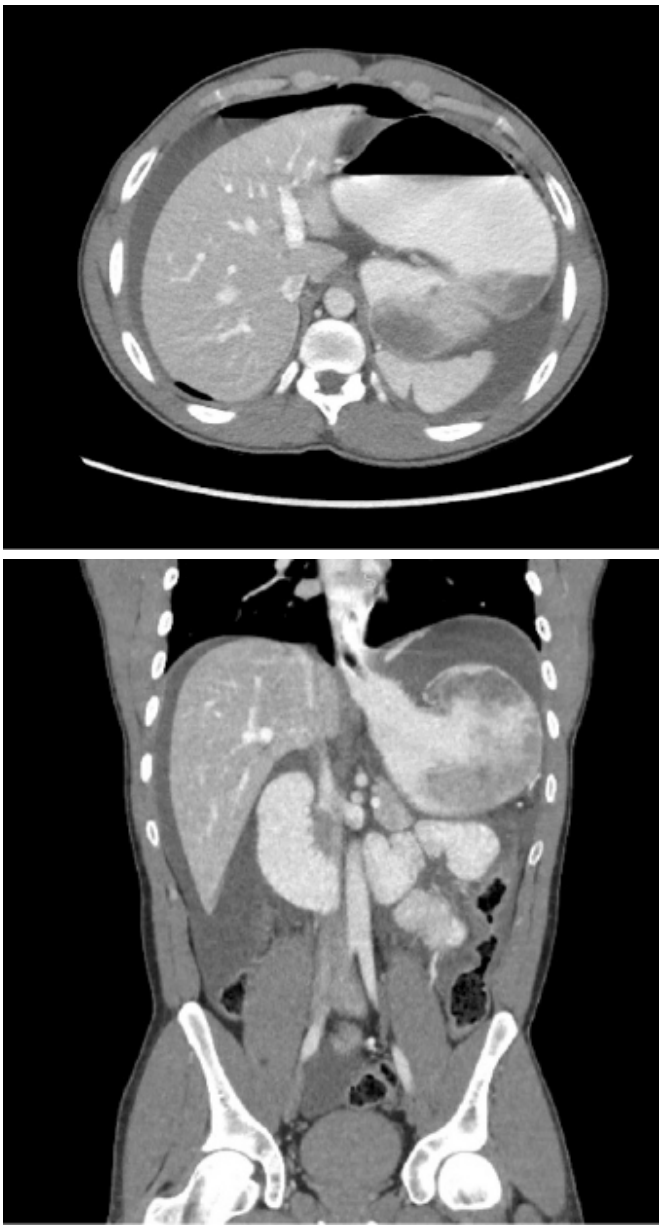
A chest X-ray revealed a mass-like opacity in the left mid abdomen, extending to the right paraspinal region and causing displacement of bowel loops inferiorly and to the right side (Figure 1). This prompted further evaluation with a computerized tomography (CT) scan, which indicated findings suggestive of gastric volvulus (likely mesentero-axial) with perforation, along with a large amount of pneumoperitoneum and free fluid (Figure 2).

Figure 1. Mass-like Opacity in Left Upper Quadrant. Published with Permission



At this point, the surgical team was consulted. The patient was evaluated and quickly taken to the operating room for an emergent exploratory laparotomy. Preoperatively, the patient received piperacillin-tazobactam. During the surgery, significant adhesions were noted in the abdomen. The small bowel was adhered to the previous midline incision, with a thick band resembling a clothesline extending from the stomach to the abdominal wall. Additionally, 1.5 liters of gastric contents containing visible food particles (such as grains of rice) were evacuated from the abdomen. After careful lysis of adhesions, a 1.5 cm area of necrotic gastric tissue with ulcers was found in the fundus. The surrounding gastric tissue was soft and appeared healthy and viable without induration. The necrotic area was clamped with an allis clamp and the healthy tissue below the clamp was retained using a TA stapler while the ulcerated tissue was excised sharply above the staple line. Subsequently, a gastropexy was performed by suturing the anterior stomach to the anterior abdominal wall in two points of fixation using permanent sutures. The abdomen was then copiously irrigated.

Figure 2. Pneumoperitoneum and Free Fluid with Suspected Mesentroaxial Gastric Volvulus. Published with Permission



The patient did well overall in the postoperative period. He was continued on piperacillin-tazobactam due to the perforation with gross contamination of the abdomen. Despite this, he developed an intraabdominal abscess that was subsequently drained percutaneously by interventional radiology. The patient improved following the drainage and was discharged home on postoperative day 9, tolerating a regular diet.

Discussion

Hirschsprung's disease is a rare congenital disorder characterized by the absence of ganglion cells along a variable portion of the distal large intestine, resulting in gastrointestinal dysmotility and obstruction. Diagnosis often occurs in neonates who fail to pass meconium within 24 hours after birth.[1] Surgical intervention involving removal of the aganglionic segment and anastomosis of innervated bowel to the anus, known as a "pull-through" procedure, is the primary treatment. While traditionally performed as multiple staged open operations with a diverting colostomy, advancements have led to laparoscopic and transanal single-stage repairs becoming the mainstay of treatment.[2,3]

Not only does our patient have a history of a rare congenital disorder, but he also presented with an uncommon gastrointestinal disorder–gastric volvulus (GV). GV occurs when the stomach rotates at least 180° on its mesentery, leading to obstruction, strangulation, necrosis, and/or perforation. It affects both sexes and all races equally, with the highest incidence in the fifth decade of life. Gastric perforation is a life-threatening complication contributing to the 42-56% mortality rate associated with untreated GV.[4] There are three broad subtypes of GV: organo-axial, mesentero-axial, and combined. The organo-axial subtype involves rotation along the cardio-pyloric axis (long axis), while the mesentero-axial subtype occurs along the short axis connecting the lesser and greater curvatures. Our patient exhibits the mesentero-axial subtype, which is less common overall but more frequent in children and young adults.[5]

Gastric volvulus can be further classified based on etiology as either primary (25-30%) or secondary (70-75%). Primary (or idiopathic) GV occurs in the absence of diaphragmatic or intra-abdominal abnormalities predisposing patients to volvulus formation. The most common cause of primary GV is laxity of the ligamentous attachments of the stomach, including the gastrohepatic, gastrocolic, gastrosplenic, and gastrophrenic ligaments.[6] Secondary (or acquired) GV is related to underlying conditions such as paraesophageal and diaphragmatic hernias, connective tissue disorders, anterior abdominal wall defects, gastric tumors, or abdominal adhesions.[4,7]

Abdominal adhesions are fibrous bands that span two or more organs within the abdomen and/or the inner abdominal wall. Etiologic studies show that almost 90% of adhesions develop following previous surgeries, occurring much more frequently after laparotomy as compared to laparoscopy.[8] In our patient's case, adhesion formation

played a crucial role in the development of GV. During the exploratory laparotomy, we discovered a thickened adhesive band tethering the stomach to the anterior abdominal wall, likely the point around which the stomach rotated. Of note, this was our patient's second operation due to adhesions, with the previous one performed at 16 years old to address a small bowel obstruction caused by adhesions in the right lower quadrant, resulting in obstruction of the terminal ileum. While it is unclear which surgery led to the specific adhesion formation resulting in the stomach's tethering, all previous procedures were related to the initial diagnosis of HD or related sequela. Given that these surgeries were open abdominal procedures, the likelihood of intra-abdominal adhesion formation was increased. These procedures and the resulting adhesions set the stage for our patient's GV.

Up to 70% of patients with acute GV present with Borchardt's triad of intractable vomiting, severe epigastric pain, and an inability to pass a nasogastric tube due to obstruction of the gastric inlet. In cases with nonspecific symptoms like ours, radiologic modalities are useful in making a definitive diagnosis. These include plain chest and upright abdominal radiographs, upper GI contrast study, barium swallow study, and/or abdominal CT scans.[6] Imaging may also detect pneumoperitoneum and ascites, indicative of gastric perforation, as demonstrated in our case. The presence of pneumoperitoneum and free fluid seen on our patient's imaging prior to the surgical consult was consistent with the abdominal rigidity, distension, and diffuse tenderness found during physical examination and confirmed during surgery.

Surgical intervention via laparoscopy or laparotomy is the current gold standard of treatment for acute GV. Decompression or endoscopic reduction may be attempted in patients with significant comorbidities or contraindications. However, it is important to bear in mind that most acute GV cases are secondary to underlying conditions that require surgical intervention, including paraesophageal hernias and adhesions. Furthermore, the presence of gastric necrosis or perforation requires surgical resection and irrigation of the peritoneal cavity. The surgical approach should include the following key steps: reduction of the volvulus, resection of any nonviable gastric segments, and prevention of recurrence through anterior gastropexy using either suture gastropexy or gastrostomy tube placement.[9] Close monitoring in the postoperative period and prompt recognition of complications are also of paramount importance.

Conclusion

Our case highlights the unique association between rare conditions like Hirschsprung's Disease and gastric volvulus, exacerbated by prior abdominal surgeries and resulting adhesions. Minimally invasive laparoscopic and transanal approaches for HD management may mitigate similar morbidity in the future.

Moreover, diagnosing acute GV remains challenging yet crucial for preventing life-threatening complications. With vigilant clinical suspicion and appropriate imaging, clinicians can intervene promptly to prevent necrosis and perforation-related mortality. Surgical repair with anterior gastropexy stands as the gold standard, accompanied by vigilant postoperative monitoring to address potential complications like intra-abdominal abscesses in the setting of gastrointestinal perforation promptly.

Lessons Learned

Abdominal surgery for congenital disease may result in long-term sequela due to adhesion formation. Our case highlights the lifelong risk carried by patients with major abdominal operations in childhood. When possible, laparoscopic and transanal approaches should be undertaken to reduce the likelihood of adhesion formation and subsequent related morbidity.

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