Hypertrophic pyloric stenosis in a 15-year-old male

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While hypertrophic pyloric stenosis (HPS) is a common condition of infancy that is usually identified within the first 2–4 weeks of life, it is rare in children after infancy [1,2]. In an older child, causes of gastric outlet obstruction (GOO) such as primary acquired GOO, peptic ulcer disease, pyloric stricture (PS) due to granulomatous or eosinophilic gastroenteritis, ingestion of caustic substances, or neoplasia, such as gastrinoma or primary gastric tumors, must be ruled out in order to diagnose idiopathic HPS [3,4]. In addition, it is important to distinguish HPS of infancy from late-onset HPS, which has also been described [3].

Herein, we report an atypical presentation of perforated gastric ulcers in an adolescent with HPS. While HPS in children after infancy is a rare occurrence, the diagnosis should be considered in order to identify cases that were not diagnosed early on and to prevent long-term complications.

1. Case report
1.1. Presentation

A twelve-year-old Caucasian male presented to an outside hospital following ten days of severe abdominal pain, fever, emesis, watery diarrhea and inability to eat or drink. He had a history of projectile, non-bilious emesis during the first month of life. For 3 months, his mother fed him formula with an eyedropper until he was able to drink from a bottle with minimal emesis. Throughout his childhood, he experienced chronic non-bilious emesis, gas bloat, early satiety, and abdominal pain and failure to thrive. He lagged far behind his fraternal brother on the growth curve.

Upon admission to the hospital, he was febrile and had a rigid abdominal wall with rebound tenderness. His white blood cell count was $13.6 \times 10^3$ cells/$\mu$L ($N: 4.5–13 \times 10^3$ cells/$\mu$L) and he had an elevated lipase of 620 IU/L ($N: 146–200$ IU/L) and amylase of 129 IU/L ($N: <106$ IU/L). He was treated for pancreatitis with bowel rest and intra-venous fluids. The serum lipase value decreased to 107 IU/L; however, abdominal symptoms progressively worsened. He was then transferred to our institution to for surgery due to persistent abdominal pain and rigidity in the face of reductions of pancreatic enzyme levels.

Upon admission to the hospital, he was febrile to 38.2 C, tachycardic to 114 beats per minute, and tachypneic at 30 breaths per minute. On physical examination the patient had peritoneal signs with a rigid abdominal wall and rebound tenderness. His hemoglobin (Hgb) was 14.6 g/dL ($N: 13–16$ g/dL), hematocrit (Hct) 44% ($N: 37–49$%) and white blood cell count (WBC) was $15.7 \times 10^3$ cells/$\mu$L ($N: 4.5–13 \times 10^3$ cells/$\mu$L). Electrolytes were within normal limits except for a bicarbonate value of 28 mmol/L ($N: 10–25$ mmol/L). Initial amylase and lipase levels were 58 IU/L and 148 IU/L, respectively. An abdominal ultrasound did not
show evidence of gallstones or biliary obstruction. On admission, abdominal CT findings of diffuse bowel thickening, ileus, a large volume of fluid in the stomach, and pneumoperitoneum were consistent with peritonitis. Exploratory laparoscopy revealed a perforated pre-pyloric ulcer that was repaired with a Graham patch. After surgery patient was determined to be positive for *Helicobacter pylori* (*H. pylori*) by enzyme immunoassay and was treated with Metronidazole and Amoxicillin for 3 weeks. Pantoprazole was discontinued after 6 weeks. At 8 weeks following surgery, endoscopy with a biopsy to test for *H. pylori* was negative and he reported modest improvement of intestinal symptoms.

Two years later, the patient returned with a contained perforated pre-pyloric ulcer that was treated with bowel rest, total parental nutrition (TPN), Pantoprazole, Metoclopramide, and Piperacillin-Tazobactam for 7 days. An extensive work up was performed and is described in the investigation below. He was discharged after 10 days and was tolerating a normal diet. Oral Lansoprazole was continued for 6 months.

1.2. Investigation

During the patient’s second admission, the Hgb was 14.5 g/dL, Hct was 43% and WBC was 10.9 \( \times 10^{9} \) cells/µL, and gastric pH was 3 (N: 1–3). The serum gastrin value was 79 pg/ml (N: < 115 pg/ml) and the levels of Chromogranin A 70 ng/ml (N: < 95 ng/ml), calcium 9.7 mg/dL (8.4–10.4 mg/dL), and parathyroid hormone 13 pg/ml (N: 111–80 pg/ml) were normal. An abdominal CT scan did not reveal a pancreatic tumor. Based on these results, a gastrinoma, carcinoid tumor, and parathyroid etiologies were excluded.

An upper gastrointestinal contrast study and nuclear gastric emptying scan indicated delayed gastric emptying. Esophagogastroduodenoscopy (EGD) with biopsy revealed a normal esophagus, a large amount of undigested food noted in the fundus, hyperemic gastric mucosa with normal rugae, scarring at the pre-pyloric ulcer, a narrow pyloric channel, and normal duodenal bulb. An endoscopic ultrasound revealed circumferential thickening of the pyloric muscle resulting in narrowing of the gastric outlet. An antroduodenal motility study revealed normal gastro-duodenal motility and excessive intra-gastric pressures after administration of erythromycin. Collectively, these findings suggested a mechanical gastric outlet obstruction due to a hypertrophic pyloric muscle.

1.3. Treatment

Seven months after his second admission, an EGD guided Botox injection into the pylorus produced temporary relief of gastric bloating and emesis but symptoms gradually recurred. Six months later, another Botox injection failed to relieve symptoms.

Two weeks after the second Botox injection into the pylorus, he returned with abdominal pain and emesis due to a gastric wall abscess in the area of the second pre-pyloric ulcer. He was given bowel rest, TPN, Ceftriaxone and Metronidazole for 12 days. Percutaneous aspiration of a gastric wall abscess by interventional radiologists grew mixed enteric flora. On hospital day 12, he was discharged home on a full liquid diet and Pantoprazole. A central line was inserted to continue IV Ceftriaxone and Metronidazole and provide TPN at home until inpatient medical stabilization. He was gradually advanced to a normal diet. One year after surgery, he was completely free of nausea, vomiting, early satiety, epigastric pain, gas bloat or flatulence. He gained 14.5 kg and increased his weight from the 25th to the 90th percentile.

2. Discussion

Hypertrophic pyloric stenosis is a rare cause of gastric outlet obstruction (GOO) in children and adolescents, though the frequency is unknown [3–5]. Classically, HPS is seen in infants younger than four months, occurring at a rate of 3–4 in 1000 live births and presenting with non-bilious emesis and metabolic alkalosis, which our patient had at presentation [1,2]. Multiple genetic and environmental risk factors, including early exposure to macrolide antibiotics, have been associated with infantile HPS [6–12]. This patient’s only known risk factor for HPS was being a first-born male child. The delay in diagnosis of HPS resulted in failure to thrive and gastric ulcer perforation with peritonitis. As a reconstruction was performed. Pathologic examination of the distal stomach and pylorus confirmed pyloric muscle hypertrophy negative for neuroendocrine markers, a gastric pseudo-diverticulum surrounded by scar tissue, and no malignancy (Fig. 1). Following surgery, he received IV Ceftriaxone and Metronidazole for 3 days and was discharged home after 8 days.

1.4. Outcome and follow-up

At the time of discharge, the patient was able to tolerate a soft, bland diet. He was sent home on Pantoprazole 40 mg twice daily. He was gradually advanced to a normal diet. One year after surgery, he was completely free of nausea, vomiting, early satiety, epigastric pain, gas bloat or flatulence. He gained 14.5 kg and increased his weight from the 25th to the 90th percentile.

![Fig. 1. Pylorus with muscle hypertrophy with 70 mm × 60 mm × 50 mm red-white firm gelatinous encapsulated nodule abutting the serosal surface. Chromogranin and synaptophysin neuroendocrine markers negative.](https://example.com/image-url)
result, a pyloroplasty was no longer possible and he required a distal gastrectomy.

Idiopathic HPS has been rarely reported in older children but consistently presents with projectile, recurrent, and episodic vomiting as well as weight loss and chronic abdominal pain [3,13]. In addition, several patients have presented with coffee ground emesis due to gastric ulceration [3]. One case was described in 2013 in which an eight-year-old boy had long standing episodes of non-bilious emesis and failure to thrive until he was treated with pyloromyotomy [1]. In contrast, those with late-onset HPS do not have symptoms in infancy and those with complete GOO are often diagnosed within one month of development of symptoms [3,4,13,14]. Similarly, idiopathic primary acquired GOO, also known as Jodhpur disease, as described in 1997 by Sharma et al., is distinct from HPS in that the pylorus is normal, lacks muscular hypertrophy, and does not consistently present with symptoms in infancy. Primary acquired GOO is rare and the etiology is not yet known [3,4,15].

Considering the rarity of idiopathic HPS in older children, other causes of GOO should be investigated. The initial work up for GOO should first aim to rule out more common diagnoses, such as acute or chronic pancreatitis, biliary obstruction, appendicitis, and thyroid dysfunction. All of these conditions were ruled out in our patient.

Pyloric sticture (PS) may present similarly to HPS and is seen more commonly in the adolescent patient [3]. A pyloric sticture in the older child may be due to peptic ulcer disease, ulceration from gastrinoma or autoimmune disease, primary gastric neoplasia including carcinoma and lymphoma, granulomatous diseases, eosinophilic gastroenteritis, amyloidosis, ingestion of caustic substances, or iatrogenic causes [3,4,16]. Diagnosis of PS is based primarily on the patient’s history and associated symptoms. Endoscopically guided biopsy with histological analysis may diagnose or exclude eosinophilia, amyloidosis, granulomatous disease, or neoplasia, while anti-nuclear antibodies and rheumatoid factor may rule out autoimmune processes. Hypertrophy of the pyloric muscle detected on abdominal ultrasound with HPS is not seen with PS [17,18].

Our patient had symptoms highly suggestive of HPS during the first month of life. Persistent emesis and failure to thrive throughout childhood suggests a chronic partial gastric outlet obstruction that began during infancy, such as HPS. It is unclear why a surgical evaluation was not performed during infancy or why the HPS did not resolve spontaneously. If left untreated, mortality of HPS is high. The frequency of spontaneous resolution of HPS in mild cases is unknown [2,19].

Perforation of a gastric ulcer is also an uncommon event in children, however, possible causes include chronic H. pylori gastritis, gastrinoma causing Zollinger-Ellison (ZE) syndrome, ingestion of caustic substances or foreign bodies, primary gastric neoplasia, and iatrogenic causes, such as chronic steroid or NSAID use or instrumentation [20–22]. While no correlation has been reported between H. Pylori and HPS in the literature, an H. Pylori infection superimposed on impaired gastric emptying and increased gastric pressure may have caused gastric ulceration and perforation in our patient. However, the gastrointestinal tract is not yet colonized by bacteria until after infancy and, therefore, chronic gastritis alone cannot account for his early symptoms [3,18,23]. In this patient, a normal gastric pH while not receiving antacids, normal gastric levels and an EGD without classic gastric hypertrophic mucosa rugae eliminated ZE syndrome as the etiology for a perforating gastric ulcer [20,21]. In addition, he had no history of iatrogenic exposure such as esophageal dilatation or endoscopy and histopathology of gastric mucosa excluded other causes such as gastritis and connective tissue or autoimmune diseases.

Endoscopic injection of botulinum toxin A (Botox) into the pyloric has not been shown to be an effective treatment option in HPS. Unlike esophageal achalasia, a condition that responds to Botox injection, release of acetylcholine is not impaired in HPS [12]. This patient received Botox injection before HPS was identified on ultrasound. A second Botox injection was requested by the family to alleviate symptoms and attend a family event prior to surgery.

The Ramstedt pyloromyotomy, first described in 1912, is the standard treatment for infantile HPS [2,19].

Alternative procedures such as gastroduodenostomy (Billroth I) or gastrojejunostomy (Billroth II) have been utilized in patients with peptic ulcer induced gastric outlet obstruction. Pyloromyotomy or pyloroplasty were considered in this patient [3,20,24]. However, the presence of a gastric pseudo-diverticulum and scaring in the distal stomach from recurrent perforated ulcers required a Billroth I to restore gastric emptying.

3. Conclusion

We present an unusual case of a 15-year-old male who had symptoms of a partial gastric outlet obstruction since infancy and developed a perforated gastric ulcer. Exploratory laparotomy revealed hypertrophic pyloric stenosis that likely persisted since infancy. Symptoms were relieved at age 15 years by a distal gastrectomy and Billroth I reconstruction. This case presentation underscores the importance of early identification and surgical treatment of HPS in order to avoid chronic symptoms, failure to thrive, more extensive surgery and greater morbidity and mortality.

References


