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# Hypertrophic pyloric stenosis in a 15-year-old male

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ABSTRACT

We present the case of a fifteen-year-old male with hypertrophic pyloric stenosis (HPS) thought to have originated in early infancy when chronic non-bilious emesis, early satiety and failure to thrive were evident. He developed two episodes of perforated pre-pyloric ulcer at ages 12- and 14-years. Work up for chronic *Helicobacter pylori* and Zollinger Ellison syndrome were negative. An upper GI study revealed a massively enlarged stomach and delayed gastric emptying. Endoscopic ultrasound revealed a thickened pylorus and a narrow pyloric canal. Electrogastrogram and antroduodenal motility studies supported a mechanical gastric outlet obstruction. A distal gastrectomy with Billroth I reconstruction was performed. One year after surgery, the patient's symptoms resolved and he had gained 14.5 kg in weight. This is a very rare case of HPS presenting after infancy and emphasizes the importance of early diagnosis of HPS to prevent long term complications including failure to thrive and death.



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 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 weeks, 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, abdominal pain and failure to thrive. He lagged far behind his fraternal brother on the growth curve.

On presentation to a community hospital, he was febrile and had a rigid abdominal wall with rebound tenderness. His white blood cell count was 13.6 × $10^3$ cells/μL (*N*: 4.5–13 × $10^3$ cells/μL) 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 × $10^3$ cells/μL (*N*: 4.5–13 × $10^3$ cells/μL). 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

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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 $10.9 \times 10^3$ 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 bloat 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 inflammation around the distal stomach had resolved and surgery could be performed.

Three weeks later, an exploratory laparotomy revealed a thickened, narrow pylorus with scarring around a pseudo-diverticulum in the area of the contained pre-pyloric perforation. The presence of the pre-pyloric pseudo-diverticulum and scarring precluded a pyloroplasty. Therefore, a distal gastrectomy with a Billroth I 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.

## 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

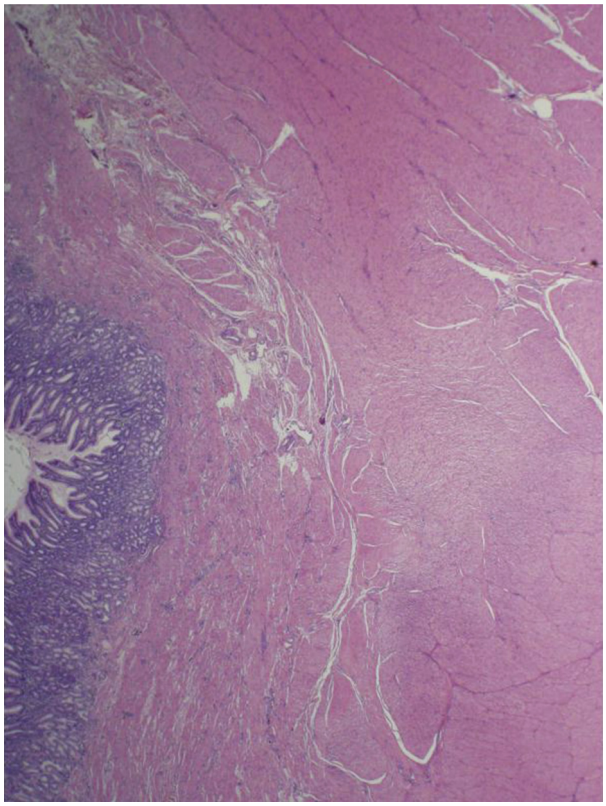

**Fig. 1.** Pylorus with muscle hypertrophy with 70 mm × 60 mm × 50 mm red-white firm gelatinous encapsulated nodule abutting the serosal surface. Chromogranin and synaptophysin neuroendocrine markers negative.