Hypertrophic Pyloric Stenosis in Adults: A Rare Entity

Idiopathic hypertrophic pyloric stenosis (IHPS) is predominantly a disease of infants with an incidence between 0.1% and 0.8%. The adult form is a rare entity with very few cases reported. We present a 34-year-old woman with Autism and Tourette’s syndrome who presented to our Tertiary center with a chief complaint for 2 years of progressive post-pandrial nausea and vomiting of undigested food. She was found to have stenosis of the pylorus on upper endoscopy examination with failure to have a sustained response to pyloric dilation treatment on two different occasions upon follow up (FU). Robotic-assisted laparoscopy pyloroplasty was performed successfully achieving resolution of symptoms over a 6 month FU. We report a case of IHPS in an adult and review the diagnosis and management approaches as well as the latest advances in treatment approaches and future directions.

INTRODUCTION

Idiopathic hypertrophic pyloric stenosis (IHPS) is usually diagnosed and treated during the first few months of life. The adult form of IHPS is a rare entity and most physicians nowadays are not aware of it. This disease was first described in 1922 by Oliver a surgeon from Ohio in the Annals of Surgery. We report a case of IHPS in an adult and then we review the experience in the literature, diagnosis and management approaches.

Case Presentation

A 34-year-old woman with Autism and Tourette’s syndrome presented with a chief complaint for 2 years of progressive post-prandial nausea and vomiting of undigested food accompanied by mild diarrhea. Her parents described that mainly solids are regurgitated and sometimes this can be food ingested from a few
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days before. The patient had previously been evaluated for gastroparesis versus gastric outlet obstruction by another gastroenterologist in the community prior to referral to our Tertiary Gastroenterology Center. Prior endoscopy 4 months ago at another hospital showed pyloric stenosis and a balloon dilation with 12 mm balloon catheter was performed. Parents reported improvement of symptoms but only for a short period of approximately 1 month. Pertinent past history was Helicobacter pylori infection 2 years ago with no ulcers present and treated with an antibiotic regimen. A 20 to 30 pounds weight loss is reported within the 2 years of symptoms. Laboratory data were reviewed and the relevant findings were TSH of 2.5, HgA1c 5.7, Hg 13.5 and albumin 4.1. A Gastric emptying study (GES) was done and there was a retention percentage at 4 hours of 85% (normal <10%). Upper GI series showed significantly delayed emptying of contrast from a somewhat dilated stomach into the small bowel with no masses or ulcers apparent. (See Figure 1) A repeat Upper endoscopy was performed at our center. Pyloric stenosis was encountered with nodular and congested pre-pyloric mucosal inflammation present and smooth narrowing of the pylorus with stenosis and no masses, ulcers or extrinsic deformity identified. (See Figure 2) Biopsies were taken. A Through the Scope (TTS) balloon dilation was performed at 15 mm, 16mm, and 18 mm maximal dilation. After dilatation the pylorus was traversed and some nodular mucosa of the proximal duodenum was seen and biopsied. Pathology results of gastric tissue showed Helicobacter pylori negative chronic active gastritis and the duodenal pathology showed acute duodenitis with preserved villous architecture and no eosinophils identified. However interestingly it was noted in the prepyloric biopsy there was extensive intestinal metaplasia likely reflecting her prior history of H. pylori infection/PUD. (see Figure 3) Some eosinophils were identified in the antral mucosa biopsy, however no eosinophils were seen in the duodenum and the patient’s eosinophils blood level was normal not supporting the diagnosis of Eosinophilic gastroenteritis.

Two months after the pyloric dilation due to persistence of symptoms patient was referred for surgical evaluation and a Robotic-assisted laparoscopic pyloroplasty with the Heineke Miculicz approach was performed at our center with full thickness biopsy of pylorus and antrum. At surgery the pylorus was divided in an axial direction with extension approximately 2 cm on the stomach and duodenum. This incision was then closed in a transverse fashion with an inner layer of running suture with full thickness mucosal to serosal bites. The second layer was created with interrupted Lembert stitches (See Figure 4). Pathology specimens from surgery confirmed hypertrophic fibroadipose tissue in the pylorus consistent with the diagnosis of Idiopathic Hypertrophic Pyloric Stenosis. Pathology of the antrum of the stomach showed normal numbers of interstitial cells of Cajal (ICC) were present based on c-kit staining. There was an average of 12 ICC per high power field which is normal. Also there was a normal population of enteric neurons. There was no inflammation, necrosis, fibrosis or evidence of malignancy identified in the muscularis propria ruling out GIST and neoplastic processes. The patient responded well to surgery, regaining weight slowly and symptoms including nausea/vomiting and diarrhea have completely resolved during last follow up 6 months after surgical intervention. The family and patient report being very satisfied with the results.

We will now focus on the discussion and review of literature for adult Idiopathic hypertrophic Pyloric Stenosis, a rare entity but important diagnosis of exclusion for investigating the explanation of nausea and vomiting.

Discussion
Pyloric stenosis in the adult is classified as primary and secondary. The most common type is secondary
and the main focus is scaring and distortion of distal pre-pyloric antrum and pylorus due to active peptic ulcer and accompanying scarring from healed ulcers. Zollinger-Ellison syndrome as a rare cause of recurrent peptic ulcer disease should always be considered and the serum gastrin level in our case was normal. Hypertrophic gastritis (Ménétrier’s disease), adenocarcinoma, gastrointestinal stromal tumors, carcinoid tumors, lymphomas including MALT, eosinophilic gastroenteritis, bezoars and extrinsic effects of postoperative adhesions or ectopic pancreas tissue are in the differential diagnoses and were excluded in our patient. No malignancies were identified on imaging or pathology. No ulcers were identified on endoscopy and mechanical obstruction secondary to food or external lesion/adhesions was not identified on imaging or endoscopic examination. The history of previous peptic ulcer disease and the extensive
histological metaplasia in the antrum was assumedly due to past Helicobacter pylori infection. This was not associated with active ulcers or pyloric scarring.

Development of HPS without obvious predisposing factors is defined as primary or Idiopathic. This type is characterized mainly by hypertrophy and hyperplasia of the pyloric muscle with evidence of hypertrophied fibroadipose tissue in the pathology sample. This entity is mainly described in the first few weeks and months of life typically in the first born male child with a higher prevalence in the Jewish population. The adult type is extremely rare with only about 200 cases described in the literature. For unknown reasons Idiopathic HPS seems to be more prevalent in middle-aged males (80%), although our case is in a female, and there are reports of middle-aged female presentations.

The etiology remains uncertain. Genetic factors have been proposed and a familial form of HPS has been described. Some authors have hypothesized that Adult IHPS is a persistence of the milder form of infantile IHPS, since anatomical and histological changes are similar. This theory implies that there is a relatively asymptomatic phase for some years until inflammation, edema or spasm precipitate occlusion of the pylorus triggering symptoms. In our patient her mental state challenges with Tourette’s syndrome and Autism clearly limited understanding from her history whether some symptoms may have been present over the years.

Clinical presentation in the adult is similar to gastric outlet obstruction with symptoms including epigastric pain, nausea, vomiting, and early satiety. In children an “olive” sized abdominal mass may be palpated in the right upper quadrant but has not been reported in adults.

Imaging examination does not provide pathognomonic findings; but excludes neoplasm and extrinsic masses. Upper GI series will typically show barium stopping at the pylorus and can define a smooth symmetric appearance of pylorus while ruling out bezoars, ulcers, masses and other disease. Abdominal CT scan can be helpful by showing distal stomach thickening and perhaps dilatation of proximal stomach. Upper endoscopic (EGD) examination is required to rule out peptic ulcers and neoplastic processes plus obtaining biopsy tissue for other possible etiologies as performed in our case. Pyloric biopsies are important as there is a case of GI stromal tumor or a spindle cell neoplasm presenting as IHPS in the literature. Our pyloric surgical tissue also excluded this possibility. On EGD examination the pylorus has a fixed appearance with smooth borders previously described by Schuster and Smith as the “cervix sign”. Definitive diagnosis is done by pathology of the surgical specimen. Typical microscopic findings include hypertrophy and hyperplasia of inner circular muscle layer of the pylorus. Gross examination seen mainly by surgeons at the time of surgery includes an elongated and thickened pylorus. The normal defined thickness of the pylorus muscle is 3 to 8 mm with an average of 4 mm. In adult hypertrophic pyloric stenosis the thickness is increased to as much as 1 to 1.5 cm on average and measurements as high as 3 cm have been reported in the literature.

The treatment for IHPS is surgery. A possible correlation with long-term hypertrophic pylorus and the development of gastric carcinoma has been reported. An older surgical approach is gastric resection and Billroth I anastomosis. The current recommendation (continued on page 34)
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(continued from page 32)

is pyloroplasty, either open or laparoscopic. The less invasive surgical approach of laparoscopic pyloroplasty has become the procedure of choice by most surgeons as it is highly effective and safe. A further advancement is robotic-assisted laparoscopic pyloroplasty which was performed in our patient. The Heineke Miculicz surgical pyloroplasty technique entails dividing the pylorus in an axial direction with extension approximately 2 cm on the stomach and duodenum. This incision is then closed in a transverse fashion with an inner layer of running suture with full thickness mucosal to serosal bites. The second layer is created with interrupted Lembert stiches (See Figure 4).

Other proposed therapeutic interventions include aggressive dilation of pyloric stenosis, an acceptable treatment which has a high recurrence rate only providing temporary relief. Intra-pyloric injection of botulin toxin into the muscle layer to relax the muscle is another approach. This is not a definitive treatment but may provide brief improvement as described in infantile hypertrophic pyloric stenosis. Upon literature review a recent publication proposed a partially covered self-expandable metallic stents as a safe and favorable outcome in the treatment of benign pyloric obstruction and in salvage after balloon dilation failure. The authors report an overall rate of 90% being symptom free at a median of 11 months follow-up with no serious adverse events reported after stent placement. It would be assumed that stent dislodgement would occur over time.

New improvements in flexible endoscopic instrumentation are allowing for a less invasive endoscopic pyloromyotomy approach. A recent report in the Journal of GI Endoscopy showed results of treatment in four pigs with submucosal dissection of the circular muscle layer achieving a successful pyloromyotomy. Endoscopic pyloromyotomy has been successfully described in the literature as early as 2005 in 10 patients with congenital pyloric stenosis with no complications and 100% success rate. The same authors propose this technique as an equally effective modality as laparoscopic pyloroplasty. There is also a case report of gastric peroral endoscopic myotomy in a young woman with refractory gastroparesis who underwent endoscopic pyloromyotomy with a triangular-tip knife (KD 640L; Olympus) and submucosal dissection technique cutting into muscularis propria extending from the pylorus sphincter to 0.5 cm into duodenal bulb. The patient tolerated the procedure without complications and 12 weeks post procedure follow up showed significant improvement of gastroparesis symptoms. This technique of endoscopic pyloromyotomy raises the possibility that in the near future this can be an alternative to surgery for both Pediatrics and Adult hypertrophic pyloric stenosis treatment.

Take Home Message

The entity of idiopathic hypertrophic pyloric stenosis in an adult should be considered when patients present with nausea and vomiting which has not responded to empiric attempts to control symptoms with antiemetics and proton pump inhibitors to address the accompanying heartburn from vomiting gastric acid. The vomiting profile may suggest gastroparesis in that contents are vomited some hours after eating and food from previous meals may be recognized. Weight loss may also become a concern. Gastroenterologist should be sensitized by our review that there is the entity of adult idiopathic hypertrophic pyloric stenosis and that there is a surgical solution, currently best achieved by laparoscopic pyloroplasty with robotic assistance.

References


