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CASE REPORT

Adult idiopathic hypertrophic pyloric stenosis - a common presentation with an uncommon diagnosis

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ABSTRACT

Background and Objectives: Adult Idiopathic hypertrophic pyloric stenosis (AIHPS) is a rare but well-defined entity in adults with only 200-300 cases reported so far in the literature.We describe a case of AIHPS and the relevant literature review.

Methods and Results: The patient presented with acute onset upper abdominal pain associated with nausea, vomiting, foul-smelling black tarry stools, and anorexia. On the Esophagogastroduodenoscopy (EGD), pylorus demonstrated a unique "cervix sign." The patient had multiple endoscopic dilations with minimal relief. She then underwent a distal partial gastrectomy with a Billroth 1 gastroduodenostomy with considerable improvement in her symptoms on follow up.

Conclusion: Adult Idiopathic hypertrophic pyloric stenosis (AIHPS) is a rare disease which is also underreported due to a difficulty in diagnosis. The most common symptoms of AIHPS are post-prandial nausea, vomiting, early satiety, and epigastric pain as seen in our patient. Endoscopy usually shows ?Cervix sign? a unique sign showing a fixed, markedly narrowed pylorus with a smooth border. Multiple treatments have been proposed for AIHPS, including endoscopic dilation, pyloromyotomy with or without pyloroplasty, gastrectomy with a Billroth 1 gastroduodenostomy. Currently, there is no evidence of one surgical technique being superior to another. Further research needs to be done on AIHPS before one technique can be standardized as the standard of care.

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Adult idiopathic hypertrophic pyloric stenosis (AIHPS); pyloric stenosis; gastric outlet obstruction; cervix sign

1. Introduction

Adult idiopathic hypertrophic pyloric stenosis (AIHPS) is a rare but well-defined entity in adults with only 200–300 cases reported so far in the literature [1–4]. AIHPS pathological anatomy was first described in 1842 [5], and Maier et al. later confirmed it as a specific disease in 1885 [6]. We report a case to highlight this difficult to diagnose but potentially curable disease.

2. Case presentation

50-year-old Caucasian female with a past medical history of hypertension came into the emergency department with the chief complaint of abdominal pain. She reported acute onset constant left upper abdominal pain that started two weeks ago, progressively worsened, rated a 10/10 in intensity, non-radiating and described as sharp, burning and stabbing. It was associated with nausea, vomiting, foul-smelling black tarry stools, and anorexia. She reported experiencing chronic abdominal pain, vomiting, and diarrhea since she was 13 years old. She saw 6–8 doctors a month in her youth with most doctors attributing her symptoms to a sensitive stomach or food poisoning. Her first esophagogastroduodenoscopy (EGD) in 2013 showed a hiatal hernia. A repeat EGD was done in April 2017, which, according to the patient, was inconclusive in elucidating a cause of her symptoms. In the current admission, a gastric emptying study was done, which showed delayed gastric emptying. CT abdomen pelvis showed thickening of the distal gastric antrum (Figure 1). On subsequent EGD, 300 ml of residual oatmeal was appreciated in the stomach, and the pylorus demonstrated a 'cervix sign.' A stenotic pylorus was found, which was dilated to 15 mm (Figures 2, 3). At this time, our main differential diagnosis included AIHPS, peptic ulcer disease, malignancy, and Gastrointestinal stromal tumors (GISTs). The definitive diagnosis could only be made on the final pathology report. The patient was discharged and had multiple endoscopic dilations over the coming months with minimal relief in symptoms. After discussion with gastroenterology, general surgery, and the patient who wished for a more permanent treatment for her underlying condition, she underwent a distal partial gastrectomy with a Billroth 1 gastroduodenostomy. Pathology was significant for focally hyperplastic gastric muscularis propria. There was no evidence of surface gastric metaplasia or increased numbers of polymorphonuclear leukocytes (Figure 4). After the pathology report, the patient was

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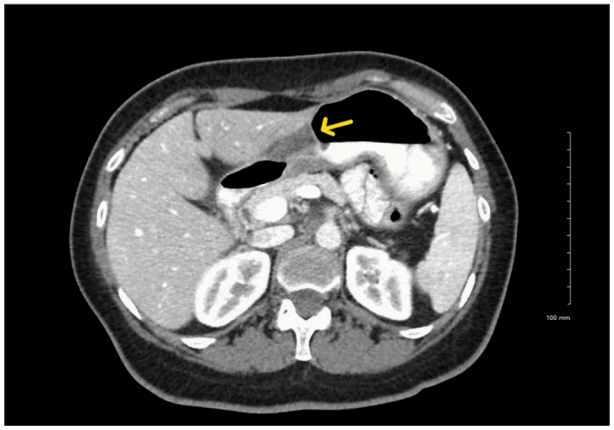

Figure 1. CT abdomen pelvis showing thickening of the distal gastric antrum (yellow arrow). Full color available online.

diagnosed with AIHPS as any evidence of malignancy or GISTs had been ruled out. Postoperatively, the patient had a gradual and slow recovery, and on her 3-month follow-up visit, she reported considerable improvement of her symptoms.

3. Discussion

Adult idiopathic hypertrophic pyloric stenosis (AIHPS) is a rare disease that is under-reported due to a difficulty in diagnosis. This challenge in diagnosis is evident by our patient, who consulted multiple doctors and underwent procedures over the years before her diagnosis of AIHPS was made. AIHPS is more commonly reported in middle-aged males while our case was a middle-aged female [7,8]. Hypertrophic Pyloric Stenosis (HPS) has been classified in various ways by multiple authors. The most universally accepted classifications are by Danikas et al. and Zarineh et al. Danikas et al. divided HPS into three types: Type 1, which is infantile HPS diagnosed at

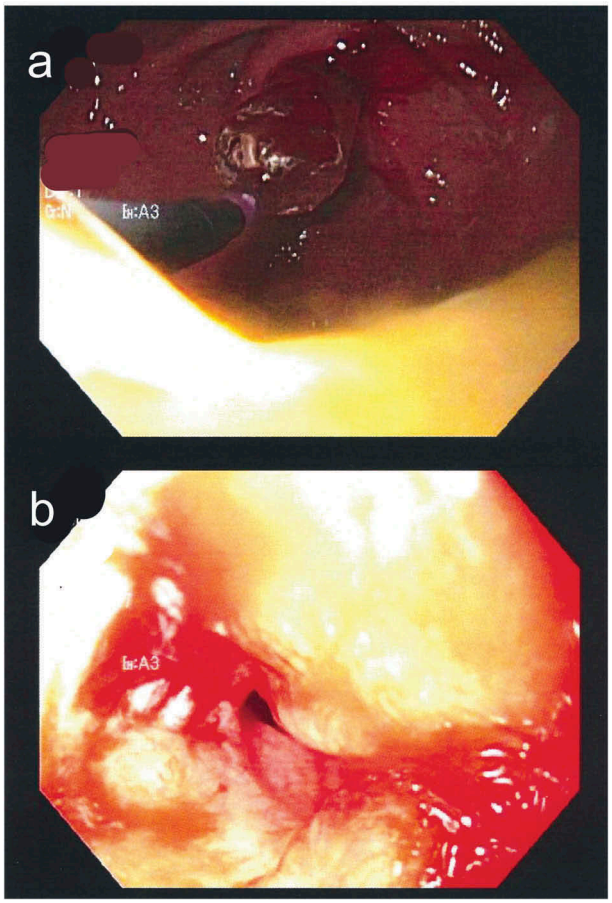

Figure 3. Endoscopy showing (a) Serial Balloon Dilator serially expanding the stenotic pyloric opening from 12 mm to 15 mm with bloody response; (b) on withdrawal after dilation a split defect of at least 5 mm is seen.

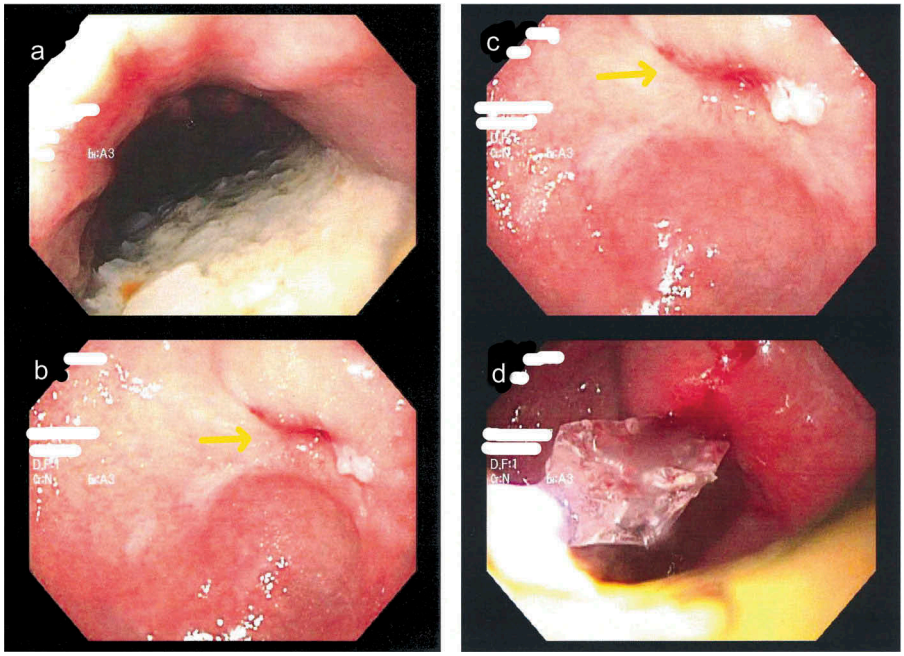

Figure 2. Endoscopy showing (a) 300 ml of residual oatmeal (b) & (c) Pylorus demonstrating 'cervix sign' (d) Serial Balloon Dilator being inserted into the small pylorus opening.

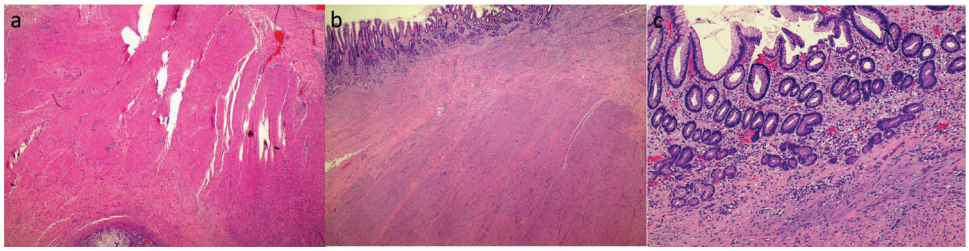

Figure 4. Cross section of the pylorus showing (a) increased thickness of the pylorus muscle, (b) focally hyperplastic gastric muscularis propria (c) 10× magnification and hyperplastic muscularis layer reaching up to the lamina propria.

a late stage; Type 2, which is the most common type, and which occurs during adult life and is likely secondary to underlying GI pathologies, such as peptic ulcer disease, malignancy, and certain inflammatory diseases; and Type 3, which is idiopathic HPS of adult onset [9]. Zarineh et al. divided HPS into primary, which has no underlying cause and under which AIHPS would be classified, and secondary, caused by an underlying disorder like the excessive healing of gastric or duodenal ulcers, malignancy, GISTs, postoperative intraabdominal adhesions, bezoars, and increased vagal activity causing pylorus hypertrophy [10]. The secondary type of HPS has no or mild hypertrophy of the pylorus muscle as compared to the primary type.

The exact etiology of AIHPS is unclear, with both genetic and environmental factors involved. The majority of researchers believe it to be the persistence of the mild juvenile form of HPS [11–13], which is further strengthened by the fact that both infantile and adult IHPS have similar histological and anatomical changes [14]. However, it still seems unclear as to the reason of the prolonged asymptomatic period until the age of 30–60 years. Some researchers postulate edema, spasm, or inflammation triggers pyloric occlusion in a predisposed individual [10]. Other possible etiologies include protracted pylorospasm, vagal hyperactivity, and changes in Auerbach's plexus. Our patient has had multiple symptoms since she was 13 years old which give credence to this theory.

The most common symptoms of AIHPS are postprandial nausea, vomiting, early satiety, and epigastric pain as seen in our patient. An abdominal mass is rarely felt as compared to the juvenile form of HPS [15].

The differential diagnosis includes malignancy and diabetic gastroparesis, both of which can present similarly to AIHPS. On pathology, GISTs can be hard to differentiate from AIHPS [10].

Diagnosing AIHPS on imaging is difficult as some cases may have completely normal studies with no pathognomonic signs [16]. 'Kirklin's sign' or 'mushroom sign' describes the protrusion of the pyloric muscle into the duodenal cap on the upper GI series. This sign has been extensively studied in juvenile HPS and has been found to occur in 50% of pediatric patients [17], but its frequency is unclear in AIHPS. The sign can also be elicited by manual pressure on the stomach [18–20]. Abdominal CT scan shows nonspecific distal gastric wall thickening in some cases, as seen in our patient. However, it is mainly used to rule out secondary causes of HPS including malignancy. Endoscopy usually shows a unique 'Cervix sign,' which is a fixed, markedly narrowed pylorus with a smooth border, and which persists after anticholinergic therapy. It can be differentiated from pylorospasm when pressure is applied through the endoscope. The cervix sign was present in our patient and was the main clue that helped us to diagnose AIHPS [21,22]. Up till now, there have been no studies describing the sensitivity and specificity of any radiological test or endoscopy findings in diagnosing AIHPS. Diagnosing AIHPS is based on a high index of clinical suspicion and suggestive radiological and endoscopic signs with the definitive diagnosis being made by the pathologist. Pathology shows grossly elongated and thickened pylorus. On microscopy, there is marked hypertrophy and hyperplasia of gastric muscularis propria with no marked inflammatory cells or malignancy seen [16].

Multiple treatments have been proposed for AIHPS, including endoscopic dilation, pyloromyotomy with or without pyloroplasty, gastrectomy with a Billroth 1 gastroduodenostomy. Laparoscopic pyloroplasty is a less invasive option. Endoscopic dilation has a high rate of recurrence and provides only temporary relief of symptoms. It is an option in high-risk surgical patients. Currently, there is no evidence of one surgical technique being superior to another. Further research on AIHPS is warranted before a method can be finalized as the standard of care [10,13,21].

Disclosure statement

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