CASE REPORT

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Idiopathic Hypertrophic Pyloric Stenosis - A Rare Condition Mimicking Gastric Cancer in An Older Adult

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Abstract

Idiopathic hypertrophic pyloric stenosis is a very rare entity in older adults, unlike in childhood. Although the etiology of idiopathic hypertrophic pyloric stenosis is not clear, secondary causes such as excessive healing of gastric or duodenal ulcers, malignancies postoperative intraabdominal adhesions should be excluded. In this case presentation, we present an older adult admitted with symptoms mimicking gastric cancer and diagnosed with idiopathic hypertrophic pyloric stenosis.

Keywords: Geriatrics, hypertrophic, idiopathic, pyloric, stenosis

Introduction

Idiopathic hypertrophic pyloric stenosis (IHPS) is a predominantly infantile disease characterized by idiopathic thickening of gastric pyloric musculature. Infantile IHPS incidence is between 0.1% and 0.8% (1). and it is usually diagnosed during the first 2 months of life. Adult idiopathic hypertrophic pyloric stenosis (AIHPS) is a rare condition with very few cases reported in the literature (2). Secondary causes for pyloric obstruction appear to be far more common in middle-aged males (3,4) but still, AIHPS should be kept in mind. Although the etiology remains unclear, theories have been proposed regarding the persistence of the juvenile form into adulthood. In this report, we present a case of AIHPS, to remind physicians of this benign entity that rarely comes to mind in the differential diagnosis of an older adult presenting symptoms mimicking gastric cancer.

Case Report

A 66-year-old male patient was admitted to the geriatric outpatient clinic with complaints of abdominal pain, weight loss, nausea, postprandial vomiting, and early satiety. He had been experiencing these symptoms for two years. He had lost twenty kilos in a year. His symptoms worsened progressively over the previous three months. The patient's complaints of nausea and vomiting began after meals. The vomit content consisted mostly of undigested food and there was no bile. He had a medical history of diabetes mellitus and benign prostatic hypertrophy. He had no specific surgical history. His diabetes mellitus disease was under control, there was no gastrointestinal involvement. A physical exam revealed a soft, mildly distended abdomen. No mass or tenderness was detected. The rest of the examination was normal. He had iron deficiency anemia, hemoglobin level was 8.4 gr/dL, ferritin level was 59 nq/mL, and transferrin saturation was 5%. He had no vitamin B12 or folate deficiency. Hba1c was 6.3% and the fasting blood glucose level was 116 mg/dL. The potassium level (3.24 mmol/L) was detected as low due to vomiting. Chest X-ray demonstrated the "Kirklin's sign" defined as the deformity of the normal gastric bubble (Figure 1).

Due to the persistent symptoms and iron deficiency anemia, abdominal computed tomography (CT) and upper gastrointestinal endoscopy were performed to exclude gastric malignancy or peptic ulcer. CT's abdomen demonstrated a wide stomach with thickening of the wall in the antrum and a stenotic pylorus (Figure 2). Upper gastrointestinal endoscopy showed a distended stomach with a large volume collection of partly digested food. Due to this much collection, the distal stomach could not be seen clearly by endoscopic examination. After non-oral feeding and

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nasogastric decompression, upper endoscopy was performed again. The patient's stomach was still filled with food content and the endoscope couldn't pass the pyloric channel which was interpreted as pyloric stenosis. His symptoms did not improve, he underwent laparotomy for diagnostic and therapeutic purposes. A palliative gastrojejunostomy was performed and pathologic samples were taken from the paraesophageal lymph node. Pathology was reported as a benign reactive lymph node. In the postoperative period, the patient's recovery was slow, and symptoms completely improved at the 1-month follow-up visit. Upper gastrointestinal endoscopy was reperformed after 1 month of surgery. It showed an intact stomach and pylorus with a "cervix sign" (Figure 3). Endoscopic pathology was significant for foveolar epithelial hyperplasia of the stomach. There was no evidence of gastric cancer, ulcer, metaplasia, or helicobacter pylori infection. According to the results of these clinical and radiological findings, the patient was diagnosed with AIHPS.

Discussion

AIHPS is a rare disease and more common in middle-aged men (3,4). Hypertrophic pyloric stenosis (HPS) has been divided into several types by different authors. The most widely accepted etiologic classification includes the primary type with no obvious underlying disease and the secondary type caused by an underlying disorder such as overhealing of gastric or duodenal ulcers, malignancy, gastrointestinal stromal tumors (GISTs), postoperative intra-abdominal adhesions (5-7). In the secondary type, the muscle fibers are usually replaced by fibrous tissues, and there is little or no smooth muscle hypertrophy, as compared to the primary type. The etiology of AIHPS is unclear. Most researchers believe that AIHPS is the persistent form of mild juvenile HPS (6). Infantile and adult forms of IHPS have anatomical and histologically similar changes (8). However, it still seems unclear why most of the patients stay asymptomatic until middle age.

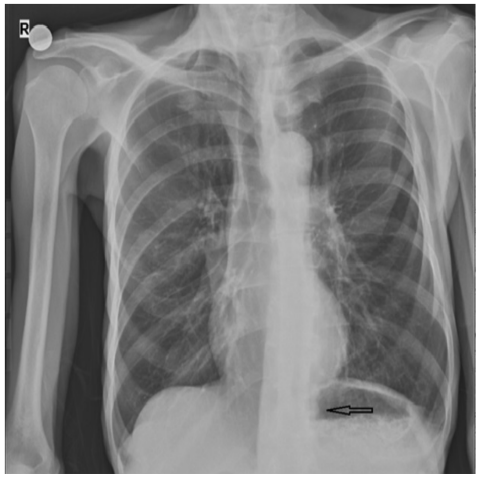

Figure 1. Deformity of the normal gastric bubble on an upright chest X-ray: "Kirklin's sign"

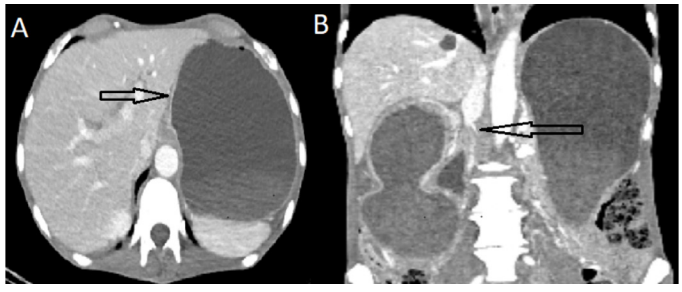

Figure 2. A. Wide stomach, B. Thickening of the wall in antrum, stenotic pylorus

The clinical features of adult IHPS are similar to other conditions that cause gastric outlet obstruction. As in our case, postprandial nausea, vomiting, and early satiety are the most common symptoms of AIHPS. Unlike infantile IHPS, an abdominal mass is rarely felt (9).

In the differential diagnosis, more common malignancies and diabetic gastropathy should be considered. Adult IHPS is not easy to diagnose without surgery. The image of the protrusion of the pylorus into the duodenal valve is called Kirklin's (10) sign or "fungus sign" in radiological examination. Kirklin (10) were the first investigators to describe this finding radiologically in 1993. This sign is common in 50% of pediatric patients (11). Its frequency in AIHPS is uncertain, as it is also can find in the normal population. Abdominal CT helps to exclude secondary causes of HPS such as malignancy, and distal gastric wall thickening can be detected as a unique nonspecific sign of İHPS as in our case. Upper gastrointestinal endoscopy is used to diagnose HPS. The endoscopic sign, which is called the "cervix sign" indicating the narrowing of the pylorus, was first described by Schuster and Smith (12). The "cervix sign", which was also found in our patient, was one of the clues that helped us to diagnose IHPS.

Multiple treatments have been suggested for AIHPS, such as endoscopic dilatation, pyloromyotomy, or gastrectomy with Billroth I gastroduodenostomy. The treatment of choice is usually

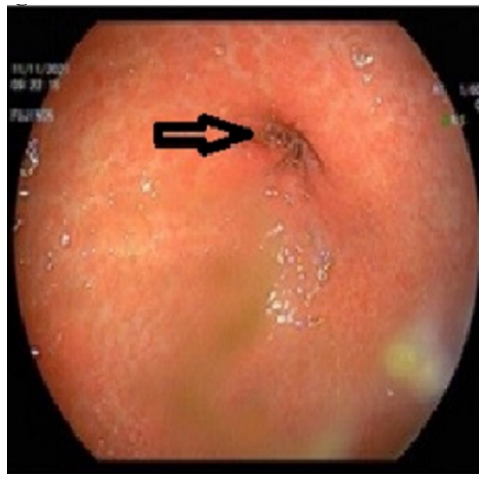

Figure 3. Pylorus demonstrating "cervix sign"

surgery with gastric resection and Billroth I anastomosis (3). Although pyloroplasty and vagotomy also showed successful results (6). Currently, there is no evidence of which surgical technique is superior, but some authors recommend partial gastrectomy as long-standing pyloric hypertrophy may increase the risk of gastric carcinoma (9,13,14). A definitive diagnosis of IHPS is made by demonstrating smooth muscle hypertrophy in the histopathology of the pylorus. In our patient, pyloric histopathology was not available because a palliative gastrojejunostomy was performed by preserving the pylorus due to the patient's advanced age and frail nature. However, our patient has not had a history of peptic ulcer, surgery, and previous malignancy. In addition, preoperative and postoperative endoscopy and computed tomography have not revealed any secondary causes. After 1 month of surgical treatment symptoms were completely improved.

Conclusion

Adult IHPS is a long-standing disease of uncertain etiology. AIHPS is very rarely reported in older adults so this report can remind clinicians this benign entity should be kept in mind in an older patient presenting with persistent vomiting and weight loss. Partial gastrectomy with Billroth I reconstruction is the preferred treatment method by most physicians but in older frail adults gastrojejunostomy could be an alternative surgical option due to the low risk of complications and faster recovery time, as in our case.

Ethics

Informed Consent: Informed consent was obtained.

Peer-review: Internally and externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: E.Ç., E.Çe., F.G., H.D.V., Concept: E.Ç., H.D.V., Design: H.D.V., Data Collection or Processing: E.Ç., E.Çe., Analysis or Interpretation: E.Ç., E.Çe., Literature Search: E.Ç., H.D.V., Writing: E.Ç., H.D.V.

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