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Case Report

Gastric Outlet Obstruction (GOO) Secondary to Adult Idiopathic Hypertrophic Pyloric Stenosis (AIHPS): A Rare Cause of GOO And Review of Literature

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Abstract

Background: An adult form of hypertrophic pyloric stenosis has been recognized since 1835 when Jean Cruveilhier first reported the case of a 72-year-old woman who presented with a lifelong history of dyspepsia and vomiting. Although this disease has received far less attention than its more common pediatric counterpart, reports of cases have continued to appear sporadically since that time.
The continuous debate on the fundamental issue of whether the disease entity is congenital or acquired. This report has been prepared not only to add one more case to the literature but also in the hope that a summation of available data may provide sufficient information reasonably to resolve this central problem of aetiology.

Case Summary: We report a 60-year-old Male with a lifelong History of recurrent Vomiting which was exercabated a month before presentation. Not a known peptic Ulcer disease patient, no clinical features of gastric or colonic malignancy.
He had an upper GI endoscopy which revealed stenosed pylorus with no gastric lesion with abdomino pelvic ultrasound revealing a dilated stomach and hypertrophy of the pylorus he was resuscitated and intraoperative findings revealed markedly dilated stomach stenosed and hypertrophied pylorus no mass lesion on the stomach or peritoneal cavity. Histology revealed hypertrophied pyloric muscles.

Objective: We aim to report this case to reawaken our consciousness that even though rare, with a high index of suspicion and availability of endoscopic services, the diagnosis of Adult idiopathic hypertrophic pyloric stenosis(AIHPS) is possible.

Conclusion: Idiopathic pyloric stenosis is an Uncommon cause of GOO with a rewarding and good prognosis when promptly managed

Keywords: Adult, Idiopathic, Hypertrophic, Pyloric, Steonis, Gastric, outlet, obstruction

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Introduction

Jean Cruveilhier is thought to be the first to describe Adult Idiopathic Hypertrophic pyloric stenosis (AIHPS in 1835). The most common clinical symptom is abdominal distention relieved by vomiting. In addition, most patients with AIHPS experience weight loss (1). Adult idiopathic hypertrophic pyloric stenosis (AIHPS) is a rare entity of uncertain pathogenesis, it is estimated to have an incidence of 0.2% to 0.5% of all live births (2-4). An upper gastrointestinal (GI) barium series and upper GI endoscopy is used to confirm the diagnosis and to rule out possible malignant disease, the treatment modalities for the relief of pyloric obstruction include endoscopic dilation, pyloromyotomy, pyloroplasty, gastrojejunostomy, and gastrectomy (5-8).
AIHPS- is a rare entity, requiring a high index of suspicion to diagnose. Cases need to be reported to add to the sparse body literature to understand the Natural history of the disease which will enhance better treatment.

Case Report

We report a 60-year-old peasant farmer, who presented at the Emergency unit with the complaint of persistent vomiting, epigastric pain and generalised body weakness. He has been having recurrent episodes of vomiting stale feeds eaten 3 to 5 days, over the last 4 decades, However no haematemesis, melena, yellowness of the eyes, abdominal distention or absolute constipation. No change in bowel habits no features suggestive of malignancy. He has no comorbidities. He has had several Hospital admissions on account of these symptoms. Examination revealed a chronically ill-looking man, pale, dehydrated, anicteric, and acyanosed. The abdomen was scaphoid with no palpable mass or organomegaly. Other systems were not remarkable.
He had fluid resuscitation and Potassium replacement. Upper GI endoscopy revealed a distended stomach with food debris and constricted pylorus that cannot be intubated

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showing the classical 'cervix sign ' as shown in Figure I. Had laparotomy with gastrojejunostomy with intraoperative findings of dense adhesion at the pylorus, markedly distended stomach. He had an uneventful post-operative recovery and symptoms improved significantly. Full thickness biopsy of the pylorus showed hypertrophy of the circular smooth muscle of the pylorus.

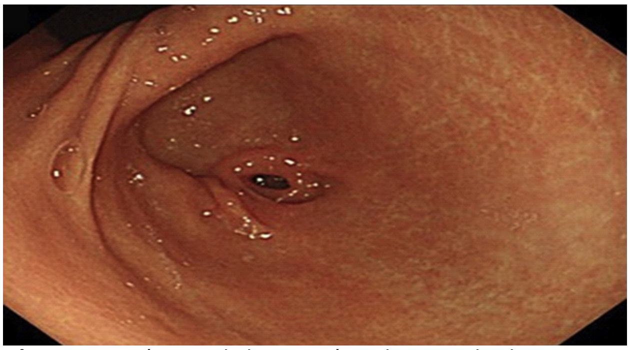

Figure I: Endoscopic image showing cervix sign.

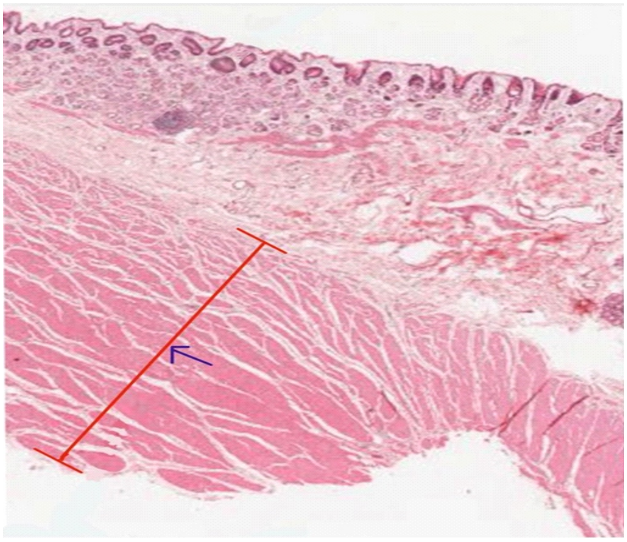

Figure II: Pyloric Biopsy showing Muscular Hypertrophy.

Discussion

The clinical symptoms of adult IHPS are similar to those of gastric outlet obstruction induced by other causes, including epigastric pain, easy satiety, and postprandial nausea or vomiting (2-5). The Presentation of our patient is similar to the above-reported symptoms in addition to weight loss and Malnutrition. Unlike infantile IHPS, an abdominal mass is seldom felt in patients afflicted with adult IHPS as in our index report the patient has no palpable intra-abdominal mass (1,2,14).

Radiologic imaging can be normal and upper GI endoscopy is often needed to make the diagnosis of IHPS and exclude other causes (3-12). However, several radiographic findings in patients with AIHPS may aid in establishing diagnosis. Kirklin's sign, which is described as a convex indentation at the base of the duodenal bulb, is a strong indicator of AIHPS. Other findings include the string sign, which can be visualized as an eccentric or concentric narrowing of the pylorus (4). the abdominopelvic Ultrasound of this index patient revealed a markedly dilated stomach with residual volume and intra-peritoneal lesion. In the instances where radiographic findings are normal, an endoscopy is needed to establish the diagnosis of AIHPS (1-7). Schuster defined a unique endoscopic sign called the “cervix sign” to describe the narrowing of the pylorus in our index patient also as shown in Figure I below (3).

The etiology of AIHPS remains unclear however It seems that both genetic and environmental factors are involved (1-6). Most physicians believe that adult IHPS is due to the persistence of the mild infantile form that persists into adult life (1,3,9,12,14).

The most widely accepted etiologic classification includes a primary type with no apparent underlying disease and a secondary type (1-14). The secondary type is more common and may be induced by many causes, including the healing of previous gastric or duodenal ulcers, hypertrophic gastritis, carcinomas, gastrointestinal stromal tumours, bezoars, vagal hyperactivity, and postoperative extrinsic adhesions (3-7). This type of HPS has no or only mild hypertrophy of the pyloric muscle (5,9,13). The muscle fibres are usually replaced by fibrous tissues. Microscopic examination demonstrates marked hypertrophy of pylorus muscles which may be associated with reactive mucosal gastropathy (3-7). However, no other significant pathologic process such as inflammation or neoplasm should be seen in the muscularis propria (4). The transition of the hypertrophied section is usually gradual from the normal areas, this is in agreement with the Histologic findings of this patient as shown in Figure II (5).

The primary or idiopathic type is characterized by hypertrophy and hyperplasia of the pyloric muscle. The only explanation proposed for the primary type is the persistence of the juvenile form that later presents in adult life, similarly the index case revealed hypertrophy of circular Muscles of the pylorus with no identifiable secondary cause, hence signifying this case to be likely Primary AIHPS (1,3,5,10).

The exact occurrence of adult IHPS cannot be estimated accurately, since some of the cases may remain asymptomatic (1). It is unclear why adult IHPS occurs more commonly among middle-aged Males, it is unclear why the majority of patients remain asymptomatic until middle age, as is the case with the index case report (3-11).

There are several means of treatment options including endoscopic dilation, gastrojejunostomy, partial gastrectomy, and pyloromyotomy with or without pyloroplasty (1-12). Studies have shown that endoscopic dilation may serve for temporary relief of symptoms, but it has a high rate of recurrence. Thus surgical treatment is the mainstay of treatment as is in this case who had gastro jejunostomy with significant symptom relief (3-14).

Conclusion

Although the true incidence of adult IHPS is unknown, there is a need to have a high index of suspicion when an adult presents with persistent or recurrent episodes of vomiting without apparent secondary causes of GOO. In these patients, a careful history of vomiting in infancy may be able to provide information about the illness's aetiology. While the majority of physicians prefer partial gastrectomy with Billroth I reconstruction, gastrojejunostomy may be a more effective surgical procedure given its lower risk of complications and quicker recovery period, as in the case of our index patient.

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