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Adult Idiopathic Hypertrophic Pyloric Stenosis - Case Report -

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Abstract:

Background & Aim: Adult idiopathic hypertrophic pyloric stenosis (AIHPS) is a rare disease with undetermined pathogenesis which both clinically and grossly stimulates gastric malignancies. Methods:We present a rare case of AIHPS in a 28-year-old Syrian man with no abvious predisposing factor. Imaging studies demonstrated gastric distension and no evidence of extrinsic compression. The pylorus was visibly identified and was clearly thickened. Microscopically, prominent hypertrophy of muscularis mucosa with hypertrophic smooth muscle cells arranged in whorls and fascicles. Conclusion: It is important as it represents a diagnostic endoscopic and clinical challenge for gastroenterologists in general.

Key Words: Adult Idiopathic Hypertrophic Pyloric Stenosis (AIHPS)− Pyloric Stenosis− Gastric Outlet Obstruction−Cervix Sign.

تضيق البواب الضخامي مجهول السبب لدى البالغين – تقرير حالة–

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الملخص:

خلفية البحث وهدفه: تضيق البواب الضخامي مجهول السبب لدى البالغين هو داء نادر وآليته الإمراضية غير محددة والتي تقلد خباثات المعدة عيانياً وسريرياً.

طرائق البحث: نقدم حالة نادرة من تضيق البواب الضخامي مجهول السبب لدى البالغين لدى شاب سوري بالغ من العمر 28 سنة من دون عوامل مؤهبة واضحة. أظهرت الدراسات التصويرية توسع في المعدة من دون دليل على انضغاط خارجي، منطقة البواب كانت محددة بشكل واضح والتثخن مرئي. الفحص المجهري أظهر ضخامة في العضلية المخاطية مع عضلات ملساء تبدي فرط ضخامة تنتظم بشكل دوامات وحزم.

النتائج والخلاصة: هذه الحالة هامة كونها تمثل تحدي تشخيصي تنظيري وسريري بالنسبة للأطباء الهضم بشكل عام.

الكلمات المفتاحية: تضيق البواب الضخامي مجهول السبب لدى البالغين–تضيق البواب– انسداد مخرج المعدة–علامة العنق.

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1. Introduction:

Adult idiopathic hypertrophic pyloric stenosis (AIHPS) is a rare case of undetermined pathogenesis (Zarineh, Leon, Saad, & Silverman, 2010). The most common clinical symptom is abdominal distention relieved by vomiting. In addition, most patients with AIHPS suffering from weight loss (Du Plessis, 1966). An upper gastrointestinal (GI) barium series and upper endoscopy are used to confirm the diagnosis and to rule out other possible malignant disease (Dye, Vidals, Lockhart, & Snider, 1979; Zarineh et al., 2010). (Figure 1)>

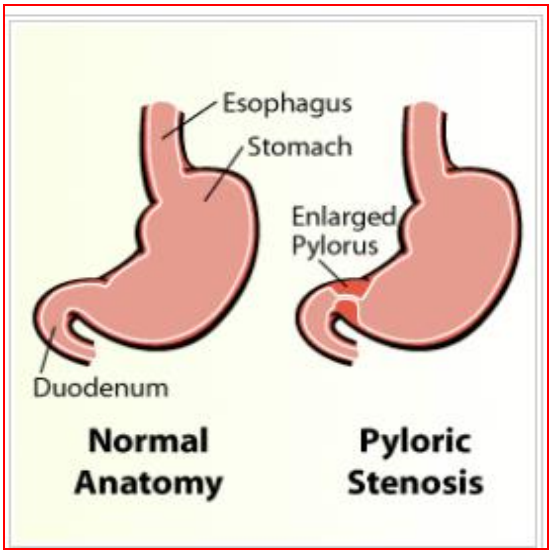

Figure 1: Graphic form shows anatomic site of enlarged pylorus.

The management options for relief of pyloric stenosis or obstruction include endoscopic dilation, pyloromyotomy, pyloroplasty, gastrojejunostomy, and gastrectomy (Simson, Thomas, & Stoker, 1986). We report a case of adult IHPS to remind young recent physicians of this rare entity and to highlight this difficult to diagnose but potentially curable disease.

2.Case Report:

A 28-year-old man presented with a long-standing history of epigastric pain, nausea, vomiting, and epigastric fullness, but there is no history of vomiting during infancy. The patient's medical history was not typical for peptic ulcer disease (PUD), gastroesophageal reflux disease (GERD) or diabetes mellitus and he had not undergone prior surgical interpositions. No history of taking any drugs such as NSAIDs. Abdominal films obtained before admission showed massive marked gastric distension. Physical exam revealed a soft, moderate distended abdomen. No tenderness, mass, and/or hernia were discovered .The remainder of the examination was unremarkable. He was admitted with diagnosis of gastric outlet obstruction. A CT scan did not show any evidence of extrinsic compression. An upper endoscopy demonstrated massive gastric distension and the pyloric channel did not appear to relax and dilate.

The endoscope passed the channel with slight resistance, which was interpreted as pyloric stenosis. The pylorus demonstrated a 'cervix sign'(figure 2),the duodenum was unremarkable.

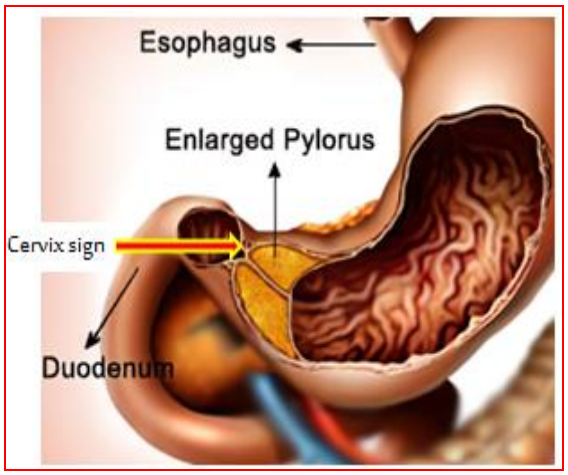

Figure 2: Graphic form shows cervix sign in AIHPS.

At this time, our main differential diagnosis included AIHPS, peptic ulcer disease, malignancy, and gastrointestinal stromal tumors (GISTs). Surgical resection was performed at Mouwasat University Hospital. The final diagnosis could only be made on the final pathology report after discussion with gastroenterology, general surgery, and the patient who wished for a more permanent treatment for her underlying condition, he underwent a distal partial gastrectomy with a Billroth 1 gastroduodenostomy.

3. Histopathologic Features:

The specimen consisted of a segment of stomach, measuring 33×11×5 cm with lymph node dissection . The wall of the stomach was uniformly thickened over the proximal portion of the specimen involving the entire circumference of the gastric

wall. A very prominent gastric fold measuring 1.2 cm in thickness and located at 1 cm from the distal margin was observed at the pylorus (Figure 3) with 16 reactive lymph nodes.

Figure 3: Panoramic image of the thickness of the pyloric stenosis.

There were focal areas of congestion and edema in the mucosa, but no masses or ulcerations were seen. Pathology was significant for marked muscularis propria hypertrophy with smooth muscle cells arranged in whorls and fascicles were seen in the pyloric region, while the remaining stomach also showed a thickened muscularis propria (Figures 4 A,B)

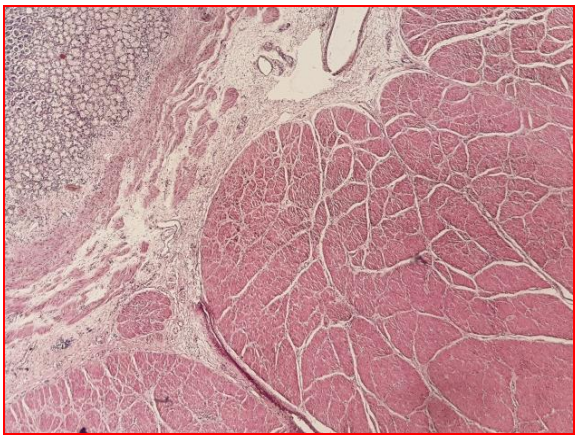

Figure 4 A: Smooth muscle hypertrophy

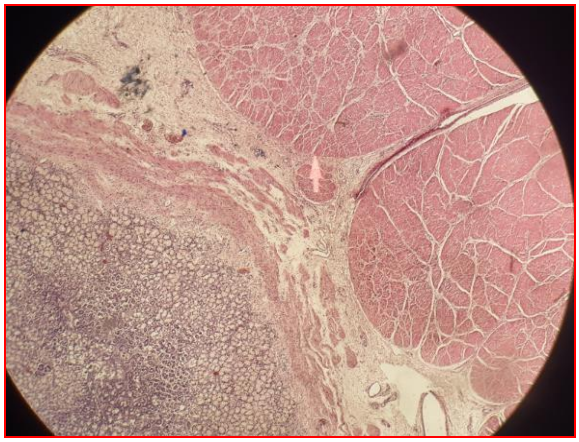

Figure 4 B: Smooth muscle hypertrophy microscopically.

Transition between thickened and normal areas was gradual. The maximum pylorus muscle thickness measured at 1.2 cm with edematous changes in the mucosa and eosinophils in the mucularis propria (Figure 5).

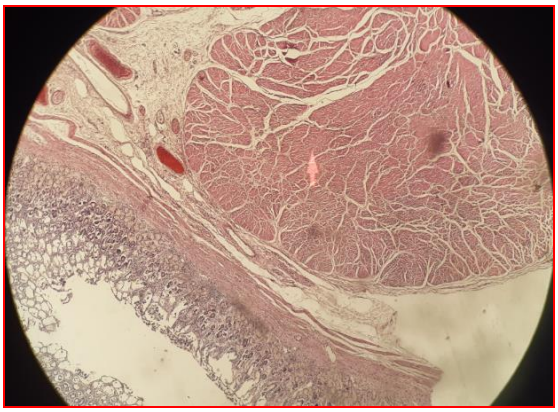

Figure 5: Smooth muscle hypertrophy with overlying gastric mucosa microscopically.

4. Discussion:

The etiology of AIHPS remains unclear, despite several theories advanced in the literature. In a case report of Zarineh et al. (Zarineh et al., 2010) stratified the etiological classification of adult idiopathic hypertrophic pyloric stenosis to two main types: primary, which has no underlying pathology, and secondary, which may encompass several causes, including healing of a previous gastric or duodenal ulcer, cancer, postoperative adhesions, and vagus nerve hyperactivity, ultimately leading to hypertrophy of the muscular anatomy (Ger, 1964; Zarineh et al., 2010). In another report, Danikas and

colleagues (Danikas, Geis, Ginalis, Gorcey, & Stratoulias, 2000) stratified AIHPS to three types. Type 1 was described as a late-stage, infantile, hypertrophic pyloric stenosis. Typically the patient has symptoms and a history that can be traced back to infancy. Type 2, the most common type, has an onset in adult life and is secondary to an underlying pathologic condition in the GI tract, such as duodenal and gastric ulcers, cancer, and other inflammatory diseases. Finally, idiopathic pyloric stenosis, which has an onset in adulthood without a known cause,was classified as type 3. In this type, there is no history of vomiting during infancy (Danikas et al., 2000; Du Plessis, 1966; Hadad & Mallick, 2014).

5.Conclusion:

AIHPS is a rare clinical condition that may mimic other more common GI lesions. Minimally invasive surgery may be an option for the management of this condition. Attention to the clinical and histologic examination should allow a right and accurate interpretation and prevent misinterpretation of a neoplasm, especially at the time of frozen section.

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