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Primary hypertrophic pyloric stenosis in the adult: A case report

Erişkinde primer hipertrofik pilor stenozu: Olgu sunumu

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A 66-year-old male was admitted with a two-month history of vomiting and weight loss. Endoscopy showed a pyloric obstruction and the patient underwent subtotal gastrectomy with gastro-jejunostomy. The histopathological study of the specimen revealed primary hypertrophic pyloric stenosis without any evidence of duodenal peptic disease. In the adult, this is a rare cause of gastric outlet obstruction of unknown etiology. It is usually recognized by histopathological examination of the specimen after a gastric resection performed to treat gastric outlet obstruction syndrome. However, some endoscopic and radiological signs, such as the cervix sign, or elongation of the pyloric channel, may give clues about the presence of the disease preoperatively. In symptomatic cases, surgery is the preferred treatment modality.

Key words: Pylorus, pyloric stenosis, primary hypertrophy, adult gastric outlet obstruction syndrome.

İki aydır devam eden kusma ve kilo kaybı yakınmalarıyla başvuran 66 yaşındaki erkek hastanın üst sindirim sistemi endoskopisinde, pilorda tam tıkanma saptandı. Subtotal gastrektomi ve gastrojejunostomi ameliyatı yapılan hastada, piyesin histopatolojik incelemesinde duodenal peptik ülser bulgusu göstermeyen primer hipertrofik pilor stenozu saptandı. Primer hipertrofik pilor stenozu mide çıkışı tıkanmasının etyolojisi bilinmeyen nadir bir nedenidir. Tanı genellikle mide çıkışı tıkanmasının tedavisi amacıyla yapılan mide rezeksiyonu sonrası piyesin histopatolojik incelemesi ile konulur. Ancak, serviks belirtisi ya da pilor kanalının elongasyonu gibi bazı endoskopik ve radyolojik bulgular ameliyat öncesi tanı için ipuçları olabilir. Semptomatik olgularda cerrahi tedavi tercih edilen yöntemdir.

Anahtar kelimeler: Pilor, pilor stenozu, primer hipertrofi, erişkinin mide çıkşı tıkanması sendromu.

INTRODUCTION

In children, congenital hypertrophic pyloric stenosis is a relatively common disease with an incidence of 0.25 to 0.50 % in all live births (1). However, hypertrophic pyloric stenosis is a rare cause of gastric outlet obstruction in adults (2,3). There are only about 200 reported cases of adult primary hypertrophic pyloric stenosis (PHPS) in the literature (4). Quigley et al. (2) reported only one case out of 100 adult cases of gastric outlet obstruction. It was first described by Cruveilhier (5) in 1835. In 1901, Robson and Moynihan (6) reported the first surgical correction by gastrojejunostomy in a man with the disease and Barling (7) described its histopathology in 1913. Although its etiology is still obscure, some cases may represent persistence of a mild form of the juvenile condition in adult life (1,8-10). Two studies with contradictory results reported that symptoms and radiological findings may regress with age (11,12). A primarily neurogenic etiology has also been suggested by some authors (13-15). Approximately 80% of the cases reported so far were male and the age of onset is extremely variable, ranging from 17 to 82 years (2,7,8,16-20).

In this report, a case with PHPS is presented, and the current diagnostic tools to establish the precise diagnosis and treatment modalities are discussed along with a brief review of the literature.

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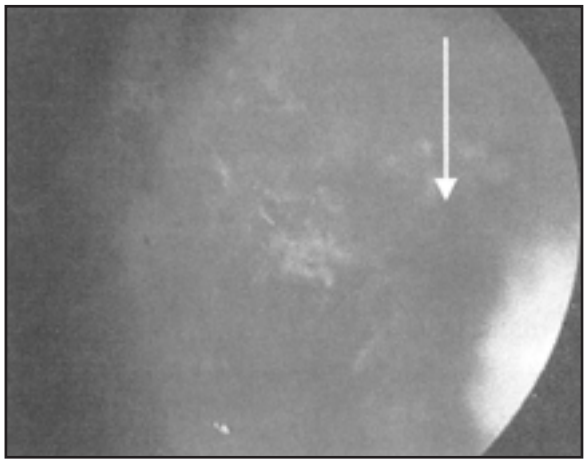

Figure 1. Endoscopic view of the prepyloric antrum, arrow shows the mucosal dimpling of the completely obstructed pylorus.

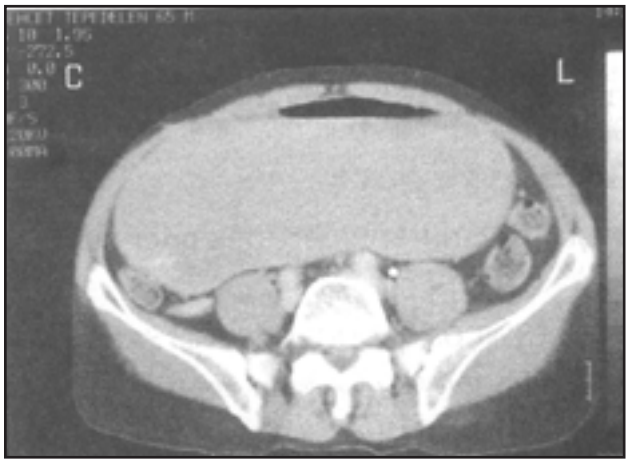

Figure 2. The extreme gastric dilatation due to the complete pyloric obstruction is shown in the abdominal CT scanning.

CASE REPORT

A 66-year-old male was admitted to our surgical unit in January 2000 with a two-month history of nausea, vomiting after meals and weight loss of 10 kg. Previous medical history revealed only mild dyspeptic complaints during the previous 30 years. There was a succussion splash on physical examination. Endoscopic examination showed complete pyloric obstruction with gastritis and grade III distal esophagitis (Figure 1). On abdominal computed tomography (CT), there was no evidence of gastric neoplasia but extreme gastric dilatation (Figure 2). A subtotal gastrectomy with a gastrojejunostomy (Billroth II) was performed and the postoperative course was uneventful.

The subtotal resected stomach was 28x13x15cm in size. The thickness of the gastric wall exceeded 10 mm at the distal surgical border. Serial sections were cut from the specimen but macroscopic inspection did not reveal any ulcer or neoplasm. Histopathologic examination of the specimen

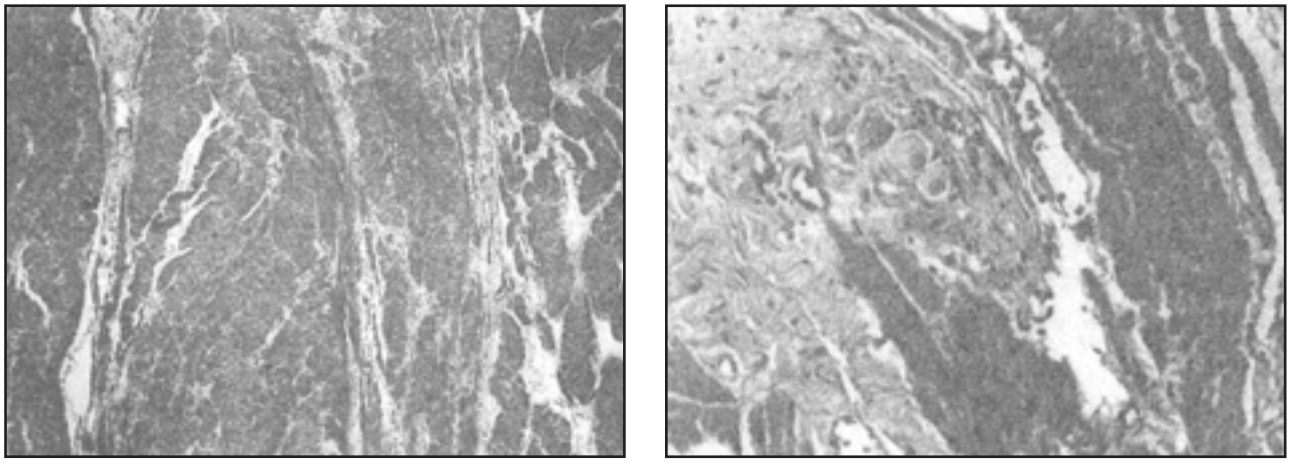

Figure 3. a) X40, H+E, hypertrophic muscle fibres of the muscularis propria at the distal stomach (pylorus), b) X200, H+E, hypertrophic nerve bundle with mild hypertrophic changes of the ganglion cells among the disorganized muscle fibers at the pyloric region.

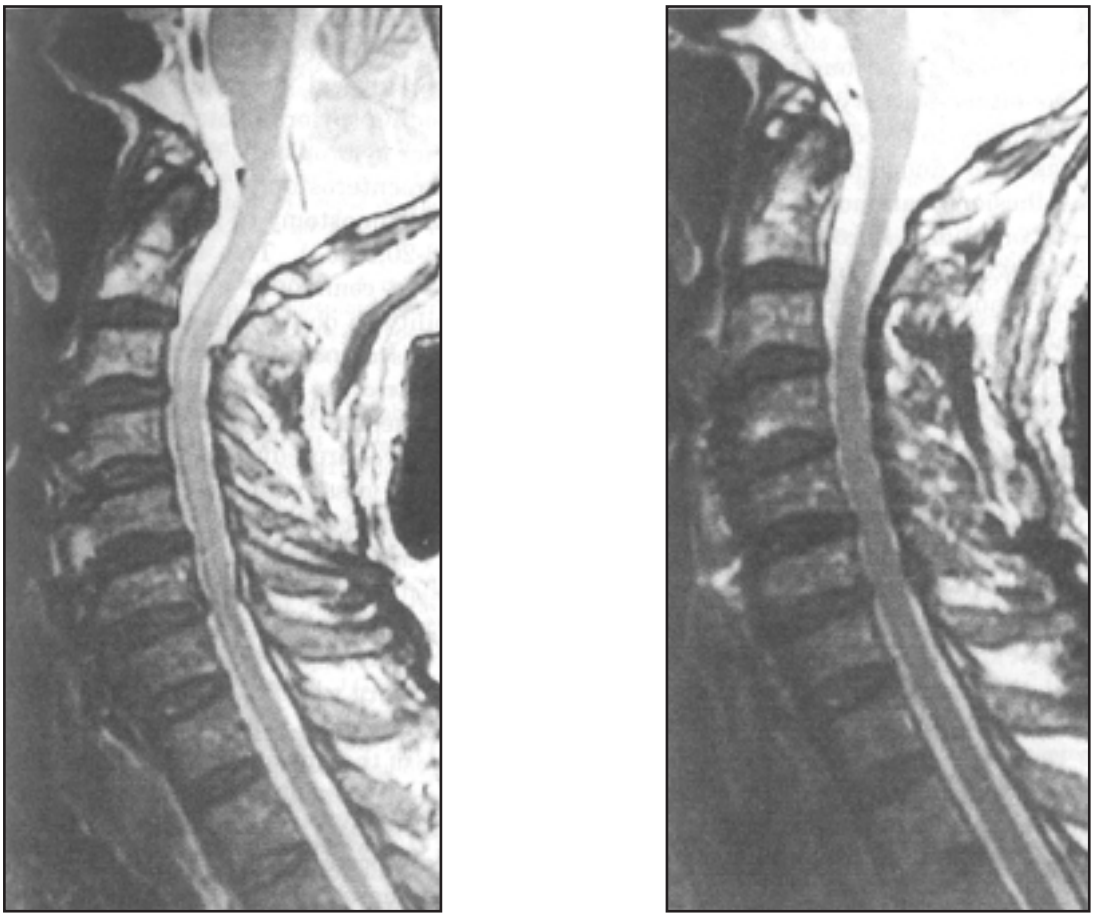

Figure 4. a) The preoperative T2 weighted cervical MRI section showed high-grade cervical degenerative and osteophytic changes from C3 to C7 anteriorly; compressing both esophagus and trachea, b) the postoperative T2 weighted cervical MRI section revealed that almost all degenerative changes that seen preoperatively were resected.

showed advanced hypertrophy of the muscularis propria (Figure 3a) and muscularis mucosa without ulceration, scarring or neoplasm at the pyloric region. Hypertrophied muscle fibers revealed architectural disorganization in haphazard fashion in some areas of the muscularis propria. Hyperplastic and hypertrophic nerve bundles were seen with mild hypertrophic ganglion cells (Figure 3b). The lamina propria revealed mild lymphoplasmacytic infiltration, which was considered as a chronic superficial nonspecific gastritis. Neither collagen deposition nor fibrosis was seen. Sections were analyzed for the presence of H. pylori by using Giemsa stain and were found to be negative. Using the standard streptavidin-biotin peroxidase method, a monoclonal antibody for desmin (clone D33, Neomarkers, CA) showed positive cytoplasmic immunoreactivity in hypertrophic muscle cells.

The patient was discharged on the seventh postoperative day. He had no complaints during the following five months, but in May 2000 he was admitted to our clinic with progressive dysphagia, cough and dyspnea. Endoscopy was attempted but it was not possible to insert the endoscope into the cervical esophagus due to an extrinsic pressure at the level of cricopharyngeal muscle. Computed tomography and magnetic resonance imaging (MRI) revealed high-grade cervical degenerative and osteophytic changes from C3 to C7 anteriorly; compressing both the esophagus and trachea (Figure 4a). The patient underwent an anterior corpectomy with tracheostomy. Follow-up MRI revealed that almost all degenerative changes that had been seen preoperatively were resected (Figure 4b). The patient was able to swallow in the early postoperative period, but then died from a cause unrelated to the primary disease.

DISCUSSION

Adult pyloric obstruction has various causes that can be classified as either primary or secondary. Most cases are secondary to a local disease such as scarring of a gastric or duodenal ulcer, extrinsic postoperative adhesions, carcinoma, or bezoars (8,21,22). In these conditions, fibrous tissue predominantly replaces with no or very little hypertrophy of the smooth muscle in the pylorus (8). In the primary form, which our case had, there is a diffuse or focal hypertrophy of the smooth muscle without any underlying identifiable disease (8,23).

Preoperative diagnosis of PHPS in the adult may be difficult. Upper gastrointestinal radiologic studies may demonstrate an elongation of the pyloric channel up to four cm (normally 1cm) (2,24), while endoscopy may reveal a fixed narrow pylorus with an intact smooth border analogous to a doughnut, called the "cervix sign" (25). It can also be successfully diagnosed by ultrasound (26). In a recent case report, endoluminal ultrasonography showed thickening of the different layers of gastric wall with the suspicion of stromal neoplasia or lymphoma (13). A differential diagnosis with cancer or peptic ulcer should be established and a definitive diagnosis requires histopathological study of the entire wall (24). The only histopathological finding is the hypertrophied pyloric muscle fibers that end abruptly at the duodenum, sometimes accompanied by a mild degree of fibrosis. Mild hypertrophic changes of the ganglion cells may be seen (1) and chronic gastritis is usually present (24,27).

Although pyloric dilatation, which is associated with a high rate of recurrence, can be performed, the treatment of choice for adult PHPS is surgical (2,28). Various surgical procedures have been reported, including pyloromyotomy, Finney or Heineke-Mikulicz pyloroplasty, double pyloroplasty, simple gastroenterostomy, and gastric resection with gastrojejunostomy or gastroduodenostomy (1,13,16,18-20,29-34). The Freted-Ramstedt pyloromyotomy is commonly performed in children, but inability to observe the mucosa in this procedure and its association with diverticulosis and scarring restrict its use in adults (29,31,35). Finney and Heineke-Mikulicz pyloroplasties are short and curative procedures, but these techniques have some disadvantages including difficulty of closure under considerable tension due to a notably hypertrophied pylorus and the tendency to recurrent obstruction (16,29,30,33). Therefore, a double pyloroplasty technique has been advocated. In this technique, a posterior myotomy is combined with Weinberg modification of the Heineke-Mikulicz procedure and it is particularly useful for the safe closure of the initial anterior longitudinal myotomy incision without excessive tension and compromise of the lumen (2,29). Gastric resection is the preferred procedure in patients who develop persistent high-degree obstruction or when a neoplastic origin cannot be totally eliminated. It has the advantage of completely removing the lesion and provides a reliable specimen for histopathological examination (1,13,19,24,36).

In conclusion, primary hypertrophic pyloric stenosis in the adult is a rare condition. It necessitates surgical treatment in symptomatic cases and the elimination of a neoplasia in the etiology is mandatory.

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