Idiopathic Hypertrophic Pyloric Stenosis Combined with Left Paraduodenal Hernia in an Adult

Basilios Papaziogas Charalambos Lazaridis Anastasios Souparis

Ioannis Koutelidakis Marios Grigoriou Dimitris Dragoumis

Konstantinos Atmatzidis

2nd Surgical Clinic, Aristotle University Hospital 'G. Gennimatas', Thessaloniki, Greece

Key Words

Hypertrophic pyloric stenosis, adult · Paraduodenal hernia

Abstract

Objective: We report a case of primary hypertrophic pyloric stenosis combined with a paraduodenal hernia in a 35-year-old woman. **Clinical Presentation and Intervention:** The patient presented with signs of obstructive ileus. CT of the abdomen revealed a marked dilatation of the stomach and the proximal jejunum as well as a circumferential thickening of the antral-pyloric region with characteristics indicating hypertrophic pyloric stenosis. Exploratory laparotomy revealed the presence of a paraduodenal hernia containing jejunal loops and marked thickening of the pyloric region. The jejunum was reduced to its normal place and the ostium of the paraduodenal hernia closed with a running suture. The hypertrophic pyloric stenosis was treated with pyloromyotomy. Since the patient had no predisposing factors for the development of secondary pyloric stenosis, we considered the pyloric stenosis as congenital in origin. **Conclusion:** To our knowledge this is the first reported case of congenital pyloric stenosis combined with the presence of a paraduodenal hernia in an adult.

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Introduction

Congenital hypertrophic pyloric stenosis is a benign disease caused by hypertrophy of the circular muscle fibers of the pylorus. Its incidence is estimated at about 0.25–0.5% of all births. Its clinical manifestations usually become obvious during the first 2 months of life [1]. The presence of pyloric stenosis in adults with characteristics of hypertrophy of the circular muscle layer of the pyloric canal is classified as primary hypertrophic pyloric stenosis and it is considered a mild form of congenital pyloric stenosis revealed in adult life [2].

Paraduodenal hernias, also known as mesocolic or mesentericoparietal hernias, are the most common types of internal hernias (>50% of all internal hernias). They are rare congenital anomalies in which the small intestine is partially or completely trapped beneath the mesentery of the developing colon [3, 4].

We present the case of a 35-year-old woman with primary hypertrophic pyloric stenosis combined with a paraduodenal hernia.

Clinical Presentation

A 35-year-old woman was examined at the emergency room of our department complaining of diffuse, colicky abdominal pain and vomiting of several hours duration. The patient's medical history was not contributory to her problems. However, she

had experienced repeated episodes of dyspepsia after food intake, since her childhood. The physical examination revealed a distended abdomen and mild tenderness on palpation. Blood examination showed leukocytosis.

The X-ray images of the abdomen revealed the presence of dilated small intestine with air-fluid levels and marked distension of the stomach. Computed tomography (CT) of the abdomen also showed dilatation of the small intestine up to the middle of the jejunum. It also revealed a circumferential thickening of the antral-pyloric region with characteristics indicating hypertrophic pyloric stenosis (fig. 1). There was a significant delay (>10 min) in the emptying of the gastric content and no evidence of an external lesion or enlarged gastric lymph nodes.

Emergency laparotomy was performed. The abdominal cavity was reached through a midline incision. The exploration of the abdomen revealed the presence of a left paraduodenal hernia containing a portion of jejunum and obstructing its lumen (fig. 2). The ostium of the hernia was formed by the duodenum (right), the transverse mesocolon (cranially), the inferior mesenteric vein (left) and the left colic artery (dorsally).

Fig. 1. CT of circumferential thickening of the pylorus.

Fig. 2. Left paraduodenal hernia containing small bowel. Operative findings.

Fig. 3. Circumferential hypertrophic pyloric mass. Operative finding.

The jejunal loops were reduced. No signs of ischemia or necrosis were evident. The ostium of the paraduodenal hernia was closed with a running 2-0 Vicryl suture between the transverse mesocolon, the duodenum and the peritoneal folds covering the inferior mesenteric vein and left colic artery.

The stomach appeared markedly dilated. The circumferential hypertrophic antropylic canal was palpable and easily recognized by its pale, glistening color, as compared to the soft and pliable gastric wall. Since no signs of a secondary pyloric stenosis were present, we considered this to be a case of primary hypertrophic pyloric stenosis. The gastric outlet obstruction was relieved with a 5-cm-long pyloromyotomy (fig. 3). The postoperative course of the patient was uneventful and she was discharged on the 5th postoperative day.

Follow-up examination of the patient, 6 months after the operation, showed complete remission of the patient's dyspeptic symptoms that could be attributed to the pyloric stenosis. Furthermore, endoscopic examination of the stomach and duodenum was negative for the presence of ulcer or other pathology of the stomach or duodenum.

Discussion

Pyloric stenosis in adults is usually associated with the presence of gastroduodenal ulcers, benign or malignant neoplasms, extrinsic postoperative or post-inflammatory adhesions, hypertrophic antral-pyloric folds and bezoars. This form of pyloric stenosis is considered secondary and results from localized replacement of muscle fibers by fibrous tissue, whereas hypertrophy of the smooth muscles is usually absent or minimal. The development of pyloric stenosis in adults caused by hypertrophy of the pyloric

circumferential smooth muscle layer, in the absence of the above-mentioned predisposing conditions, is called idiopathic or primary hypertrophic pyloric stenosis [5].

The exact occurrence of this form of hypertrophic pyloric stenosis cannot be estimated with accuracy, since the great majority of the cases remain asymptomatic. A certain percentage probably represent an incomplete form of congenital pyloric stenosis, which becomes symptomatic later in life. Zavala et al. [1] reported on a family with both congenital and adult type of hypertrophic pyloric stenosis. This assumption is further supported by the fact that almost 80% of the patients with primary adult hypertrophic pyloric stenosis are men, which is in accordance with the male predominance of congenital pyloric stenosis [1, 6, 7].

On the other hand, paraduodenal hernias are a rare, but surgically important malformation, which result from failure of fixation of either the right or left mesocolon to the posterior abdominal wall. Overall, they account for approximately 1% of all causes of intestinal obstruction. Incarcerated paraduodenal hernias are associated with 20% mortality.

A right paraduodenal hernia is associated with non-rotation of the pre-arterial midgut segment and is characterized by small bowel entrapment posterior to the right colon and cecum. A left paraduodenal hernia entraps the small intestine, which is contained within a hernia sac with a neck composed of the inferior mesenteric vein and peritoneal attachments extending to the posterior wall. It is associated with normal positioning of the colon and cecum [8, 9].

In our patient, the diagnosis of pyloric stenosis was suspected from the plain X-ray and CT findings. The stomach was markedly dilated on plain radiography, although a nasogastric tube was in situ indicating a chronic gastric outlet obstruction. Secondly, the CT of the abdomen also showed marked dilatation of the stomach along with the characteristic circumferential thickening of the antropylic region. However, the presence of dilated jejunal loops and air-fluid levels of the small intestine on plain radiography did not conform with the acceptance of this condition as the cause of the patient's symptoms. Thus, the paraduodenal hernia was probably the cause of her symptoms, while identification of the primary hypertrophic pyloric stenosis can be considered an incidental finding. Unfortunately, we have no histological studies to confirm the presence of hypertrophic pyloric stenosis because a pyloromyotomy was performed. Our diagnosis was based on the computed tomographic and intraoperative findings, which were convincing.

Partial gastrectomy, gastroenterostomy, pyloromyotomy, pyloroplasty, and endoscopic dilatation are proposed as treatments for primary hypertrophic pyloric stenosis by various authors [10, 11]. We decided to perform a pyloromyotomy, in order to avoid a gastrectomy for a benign disease. The ostium of the paraduodenal hernia was closed with a running Vicryl 2-0 suture.

To our knowledge this is the first reported case of congenital pyloric stenosis combined with left paraduodenal hernia in an adult. Probably there is no common etio-pathogenetic background that could connect these two congenital conditions. However, congenital hypertrophic pyloric stenosis has been described in association with other congenital anomalies, such as congenital mesoblas-

tic nephroma, nephrocalcinosis, recessive polycystic kidney disease, penile agenesis, jejunal atresia, congenital short bowel, congenital diaphragmatic hernia and congenital hypothyroidism [1, 7].

Conclusion

This is the first reported case of congenital pyloric stenosis combined with left paraduodenal hernia in an adult. Both conditions are extremely rare and may cause great difficulties in the differential diagnosis of acute abdomen, especially in cases with a high obstructive ileus.

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