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Is Laparoscopic Distal Gastrectomy A Justified Approach for Adult Hypertrophic Pyloric Stenosis, a Rare Cause of Gastric Outlet Obstruction? (with video)

Nadir Bir Mide Çıkış Obstrüksiyonu Nedeni Olan Erişkin Hipertrofik Pilor Stenozunda Laparoskopik Distal Gastrektomi Yaklaşımı Uygun Bir Tedavi Seçeneği Midir? (video ile)

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ABSTRACT

Idiopathic hypertrophic pyloric stenosis (IHPS), which affects 0.2% to 0.4% of children, is caused by the hypertrophy of the pylorus muscle layer. This condition leads to stenosis and non-bilious vomiting. Adult IHPS (AIHPS) is rare and typically occurs in males aged 30-60 years. This report discusses the case of a 39-year-old male diagnosed with IHPS following surgery. The patient, who complained of abdominal swelling, nausea, and vomiting, was diagnosed with gastric outlet obstruction on computed tomography (CT) and upper gastrointestinal endoscopy (UGE). Laparoscopic distal gastrectomy with Roux-en-Y reconstruction was performed due to unsuccessful endoscopic intervention and ongoing symptoms. The pathological diagnosis was consistent with that of IHPS. Laparoscopic distal gastrectomy with Roux-en-Y reconstruction is an effective treatment for AIHPS. It minimizes discomfort, reduces hospital stay and recovery time, and provides definitive diagnoses with minimal invasiveness. Laparoscopic skills are crucial. Further research and extended follow-up are required to confirm standard care for AIHPS.

Keywords: Gastric outlet obstruction, adult idiopathic hypertrophic pyloric stenosis, laparoscopic distal gastrectomy, Roux-en-Y reconstruction

ÖZ

İdiyopatik hipertrofik pilor stenozu (IHPS), çocukların %0,2 ila %0,4'ünü etkileyen ve pilorun muskular tabakasının hipertrofisinden kaynaklanan bir hastalıktır. Bu durum stenoza ve safrasız kusmaya neden olur. Eriskin idiyopatik hipertrofik pilor stenozu (AIHPS) ise nadir görülen ve genellikle 30-60 yaş arası erkekleri etkileyen bir rahatsızlıktır. Bu takdimde, ameliyat sonrası IHPS tanısı alan 39 yaşındaki bir erkek hasta ele alınmaktadır. Karın şişkinliği, bulantı ve kusma şikayetleriyle başvuran hastaya, bilgisayarlı tomografi ve üst gastrointestinal endoskopi sonucunda gastrik cıkış obstrüksiyonu tanısı konuldu. Endoskopik müdahalenin başarısız olması ve semptomların devam etmesi üzerine laparoskopik distal gastrektomi ve Rouxen-Y rekonstrüksiyonu uygulandı. Patolojik tanı IHPS ile uyumluydu. AIHPS tedavisinde Roux-en-Y rekonstrüksiyonlu laparoskopik distal gastrektomi etkili bir yöntemdir. Bu yöntem hasta rahatsızlığını, hastanede kalış süresini ve iyileşme süresini minimize eder, ayrıca minimal invaziv şekilde kesin tanı sağlar. Bu prosedürde laparoskopik beceriler kritik önem taşır. AIHPS için standart tedavi protokolünün doğrulanması amacıyla daha kapsamlı araştırma ve uzun süreli takip gerekmektedir.

Anahtar Sözcükler: Mide çıkış obstrüksiyonu, erişkin idiyopatik hipertrofik pilor stenozu, laparoskopik distal gastrektomi, Roux-en-Y rekonstrüksiyon

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INTRODUCTION

Idiopathic hypertrophic pyloric stenosis (IHPS) affects children at an incidence of 0.2% to 0.4% (1). IHPS is caused by hypertrophy of the muscle layer of the pylorus, leading to stenosis. This condition is characterized by vomiting of non-bilious gastric contents and partial gastric outlet obstruction. There is no congenital component to it. Adult-type IHPS (AIHPS) is extremely rare; most physicians are unaware of it. Most patients with AIHPS present with symptoms and signs between the ages of 30 and 60 years (2). Males are three times more likely to experience this condition than females (2). In this case report, we discuss a 39-year-old male patient who was diagnosed with IHPS stenosis postoperatively.

CASE REPORT

A 39-year-old male patient was recently admitted to our emergency department complaining of abdominal swelling, nausea, and vomiting. The patient also experienced weight loss of approximately 6 kg in the previous few months. Upon admission, an upright abdominal radiograph revealed a nonspecific small-bowel gas pattern in the upper right and lower quadrants (Figure 1). Laboratory results showed elevated blood urea nitrogen levels at 48 mg/dL; creatinine levels at 2.43 mg/dL; low serum potassium at 2.6 mEg/L; low chloride at 69 mEq/L; and a high pH of 7.57. These findings indicated hypokalemic-hypochloremic metabolic alkalosis and the development of acute renal failure. Following these diagnoses, the patient was hospitalized at a nephrology clinic for further evaluation and treatment. A multidetector CT was performed upon renal function restoration, revealing only gastric distension with no masses or lymphadenopathy (Figure 2). UGE revealed a distended stomach with food residue and stenotic pylorus (Figure 3). Despite nasogastric decompression and fluid resuscitation, a subsequent UGE intended for pyloric sphincter dilation failed, as the scope could not pass through the stenotic pylorus. The patient whose symptoms did not improve was referred to our clinic. The patient, who had no other known health issues, had experienced occasional bouts of nausea and vomiting over the past decade, typically involving the patient's ingested food. Since we could not rule out gastric malignancy from the differential diagnosis, we planned distal gastrectomy.

Surgical Procedure

The procedure is also shown in detail in the video. The patient was positioned supine with their legs apart in the reverse Trendelenburg position under general anesthesia. Subsequently, the Veress needle was inserted. The abdomen was then inflated with CO_2 gas until a pressure of 12 cm H₂O was attained. A 30-degree camera was inserted through the 10 mm umbilical port. Under laparoscopic visualization, a 12 mm trocar was inserted 1 cm above the umbilicus along the left midclavicular line, serving as the operator's primary manipulation port. A 5 mm trocar was inserted at a symmetric location on the right side to serve as the operator-assisted manipulation port. A 5 mm trocar was inserted 2 cm below the subcostal margin at the left anterior axillary line to serve as the manipulation port for the first assistant. A Nathanson liver retractor was inserted through the subxiphoid area to elevate the left lateral segment of the liver (Figure 4).

The gastrocolic ligament was divided 3 cm away from the gastroepiploic vessels using a laparoscopic harmonic scalpel.

Specific branches of the left gastroepiploic artery and the short gastric artery were cut along the greater curvature of the stomach at a predetermined level for the division of the stomach. The lymph nodes around the origin of the right gastroepiploic vein and artery were dissected en bloc. Then, the vessels were divided at their origins. The duodenum was transected immediately distal to the pylorus using a 60 mm Endo-GIA stapler. The stomach was transected at a designated level using an endoscopic stapler.

The jejunum was grasped and transected using an Endo-GIA stapler, 20-25 cm away from the ligament of Treitz, and then marked with purple dye. The Roux-limb side of the jejunum was followed distally at a distance of 50 cm. With a harmonic scalpel, small enterotomies were performed on the antimesenteric side of the proximal loop of the jejunum and the Roux limb's distal segment to place the endoscopic stapler's fork. The stapler was introduced through the 12-mm trocar on the left abdomen. A side-to-side jejunojejunostomy was conducted using a 60-mm Endo-GIA stapler. The common entry hole was closed with intracorporeal running sutures.

The proximal segment of the Roux limb was carefully mobilized while preserving the mesenteric arch. The tension-free jejunal loop was then moved via an antecolic route. Small incisions were made at the end of the transected site of the antimesenteric surface of the Roux limb, and greater curvature of the remnant stomach. An isoperistaltic gastrojejunostomy was performed, creating a side-to-side anastomosis using an endoscopic stapler. The common entry hole for gastrojejunostomy was approached, suspended with Lambert sutures, and closed using a 60 mm endoscopic stapler. The dissected stomach was extracted through an extended umbilical incision measuring approximately 4 cm. Finally, a flat drainage tube was positioned close to the duodenal stump and behind the anastomotic site.

The patient began a clear liquid diet, gradually progressing to a regular diet on the third postoperative day. The drain was removed



Figure 1. The upright abdominal radiograph reveals a nonspecific gas pattern in the small bowel in the upper right and lower quadrants



Figure 2. A. The abdominal CT scan shows a thickened distal gastric antrum and a narrowed pylorus (indicated by the blue arrow). B. The coronal view of the abdominal CT scan shows marked gastric distension

CT: Computed tomography



Figure 3. During an upper gastrointestinal endoscopy, the stenotic pylorus is visible

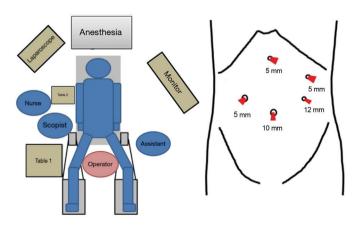


Figure 4. Positions of the operator, surgical equipment, and trocar sites for laparoscopic distal gastrectomy

on the sixth postoperative day, and the patient was discharged without complications. Pathological examination revealed that the distal gastrectomy material was characterized by increased pyloric muscle thickness and hypertrophy of the muscularis propria layer (Figure 5). The removed lymph nodes were also reactive.

DISCUSSION

AIHPS is a rare disease often underreported due to diagnostic challenges. It is most frequently reported in middle-aged men, as exemplified by our case (3). Hypertrophic pyloric stenosis (HPS) is classified into three types: type 1 HPS, diagnosed at a late stage in infants; type 2, the most common type, is occurring in adults and is often secondary to underlying gastrointestinal pathologies such as peptic ulcer disease, malignancy, and certain inflammatory diseases; and type 3, idiopathic HPS, is occurring in adults (4). Zarineh et al. (5) further divide it into primary (no underlying cause) and secondary (due to disorders like ulcers, malignancies, adhesions, bezoars, and increased vagal activity). Unlike Primary HPS, which shows muscle hypertrophy, the secondary type exhibits mild or no hypertrophy, with fibrous tissues replacing muscle fibers.

Diagnosis is based on medical history, clinical and radiological evidence, and endoscopic appearance. However, pathologists typically make the diagnoses. Patients mainly experience postmeal abdominal distension, relieved by vomiting. These symptoms include nausea and minimal pain. Symptoms, such as anorexia and early satiety, can also occur. Weight loss is a common experience among all patients. The absence of pain at symptom onset is a key diagnostic indicator. The duration of the symptoms varies from 5 weeks to 16 years (4). Our patient's intermittent symptoms of bloating, nausea, and vomiting for ten years align with the clinical findings of AIHPS.

Specific radiological findings may indicate AIHPS. A convex indentation at the base of the duodenal bulb, known as Kirklin's sign, indicates AIHPS (6,7). The "string sign" refers to an eccentric or concentric narrowing of the pyloric region (8). However, diagnosing AIHPS through imaging can be challenging because some cases

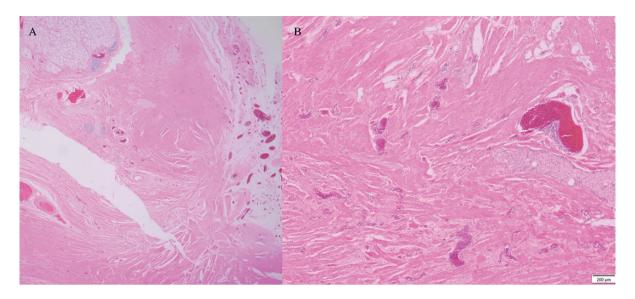


Figure 5. A. Increased thickness of the muscularis propria layer in the gastroduodenal junction (H&E x12,5) B. Hypertrophic muscle fibers in the muscularis propria layer (H&E x100)

may show average results with no distinctive signs. Abdominal CT scans help rule out secondary causes of HPS, such as malignancy. Additionally, thickening of the distal gastric wall can be identified as a unique, though nonspecific, sign of IHPS, as demonstrated in our case. Endoscopically, the pylorus is fixed, significantly narrowed, and has a smooth border. Its appearance is often described as the "cervix sign". Although peristaltic waves appear to result in some contractions, the pylorus fails to close entirely in the pyloric area. In our patient, the endoscopic observation of the "cervix sign" further validates the diagnosis of AIHPS.

Various treatments for AIHPS have been suggested, including endoscopic dilation, gastrectomy, gastrojejunostomy, pyloromyotomy, and pyloroplasty (4,9). Surgical exploration is recommended for the diagnosis and treatment of AIHPS. Full-thickness biopsy is often proposed to rule out malignancy. Most studies advocate subtotal distal gastric resection (4). Laparoscopic pyloroplasty is less invasive, but technically challenging with a thickened pylorus, typically reserved for patients in a debilitated condition (10). Despite high recurrence, endoscopic dilation is an option for high-risk patients (7). No current evidence favors one technique over another. Our patient underwent laparoscopic distal gastrectomy with Rouxen-Y reconstruction due to the consideration of malignancy in the differential diagnosis, in addition to pyloric stenosis secondary to a peptic ulcer as suggested in the literature.

Several factors led us to the decision of performing vagotomy. First, vagotomy, typically combined with other stomach procedures such as drainage, resection, or diversion, is commonly used in the management of complicated peptic ulcer disease. However, vagotomy might exacerbate gastroparesis in a chronically obstructed stomach. Therefore, the patient may be at risk of delayed gastric emptying. Second, the right vagus nerve typically appears as a single structure crossing the crura and deviating from the posterior surface of the esophagus. Conversely, laparoscopic identification of the left vagus nerve is more challenging. It often splits into smaller branches once it reaches the abdomen (11). Finally, vagotomy is less frequent today due to the application of postoperative proton pump inhibitors (PPIs) to reduce acid secretion. Furthermore, the success of primary procedures, rather than vagotomy, mainly determines patient outcomes after peptic ulcer surgery (12).

CONSLUSION

In conclusion, laparoscopic distal gastrectomy with Roux-en-Y reconstruction is not only technically feasible but also a safe operation for AIHPS. It provides definitive diagnoses with minimal invasiveness. Also, this minimally invasive surgical technique offers patients benefits, such as decreased pain, shorter hospital stays, and reduced postoperative disability. Nevertheless, proficient laparoscopic techniques are vital for successful implementation. Additional studies and prolonged follow-up are necessary to establish a clear standard of care for patients with AIHPS.

Ethics

Informed Consent: It was obtained.

Footnotes

Authorship Contributions

Surgical and Medical Practices: M.A.T., M.E., Concept: M.A.T., M.E., Design: M.A.T., Supervision: M.A.T., Material: M.A.T., M.E., F.P.U.G., Data Collection or Processing: M.A.T., M.E., F.P.U.G., A.S., B.E.A., Analysis or Interpretation: M.A.T., Literature Search: M.A.T., Writing: M.A.T., M.E., F.P.U.G., Critical Review: M.A.T.

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