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Sporadic gastrinoma with refractory benign esophageal stricture: A case report

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Abstract

BACKGROUND

Gastrinoma is characterized by an excessive release of gastrin, leading to hypersecretion of gastric acid, subsequently resulting in recurrent peptic ulcers, chronic diarrhea, and even esophageal strictures. This case report aims to improve awareness and facilitate early diagnosis and treatment of gastrinoma by presenting a rare case of gastrinoma with refractory benign esophageal stricture (RBES). Additionally, it highlights the persistent challenges that gastroenterologists encounter in managing RBES.

CASE SUMMARY

This case demonstrates a patient with gastrinoma who developed RBES and complete esophageal obstruction despite management with maximal acid suppressive therapy, multiple endoscopic bougie dilations (EBD) and endoscopic incisional therapy (EIT).

CONCLUSION

It is essential to diagnose gastrinoma as early as possible, as inadequately controlled acid secretion over an extended period increases the risk of developing severe esophageal strictures. In patients with esophageal strictures causing complete luminal obstruction, the blind reopening endoscopic incisional therapy (EIT) presents challenges and carries a high risk of perforation.

INTRODUCTION

Gastrinoma, also known as Zollinger-Ellison syndrome (ZES), is a rare pancreatic neuroendocrine neoplasm characterized by excessive gastrin release, leading to hypersecretion of gastric acid and subsequent manifestations of recurrent peptic ulcers, vomiting, abdominal pain, and chronic diarrhea[1]. Many of these symptoms can be significantly alleviated through treatment with acid inhibitors, particularly proton pump inhibitors.

The diagnosis of ZES is challenging as its symptoms are nonspecific and often overlap with those of other gastrointestinal disorders, leading to an initial correct diagnosis by the referring physician in only 3% of patients. Moreover, its rarity, with an incidence of 0.5 to 15 cases per million population, makes it extremely difficult to diagnose accurately in an acute setting[2]. Consequently, patients are typically diagnosed only after years of presenting symptoms.

In this report, we present a rare case of sporadic ZES in a young woman who presented with chronic diarrhea, abdominal pain, vomiting and later developed dysphagia due to severe reflux esophagitis with esophageal stricture. We will discuss in detail the diagnostic process and the treatments administered for her refractory benign esophageal stricture (RBES). This case report aims to improve awareness and facilitate early diagnosis and treatment of gastrinoma by presenting a rare case of gastrinoma with RBES. Additionally, it highlights that blind reopening endoscopic incisional therapy (EIT) presents challenges and carries a high risk of perforation.

CASE PRESENTATION

Chief complaints

A 33-year-old female patient was admitted with intermittent chronic watery diarrhea, abdominal pain, vomiting, and progressive dysphagia.

History of present illness

Over the past 2 years, she sought medical attention at a local hospital and was initially diagnosed with chronic idiopathic diarrhea. Laboratory and radiological investigations including stool for culture, parasites, fat analysis, computed tomography (CT) scan of the abdomen, colonoscopy, and enteroscopy showed no abnormal findings. However, her condition worsened with more frequent watery diarrhea combined with abdominal pain, vomiting, progressive dysphagia, and weight loss (10 kg over 3 months).

History of past illness

The patient also had a history of multiple peptic ulcers and had undergone an exploratory laparotomy with gastric perforation repair three months ago.

Personal and family history

She denied a family history of hereditary diseases or cancer.

Physical examination

Upon physical examination, mild tenderness in the epigastrium was noted.

Laboratory examinations

Fasting serum gastrin (FSG) and chromogranin A (CgA) levels were measured, showing elevated values of FSG (845.00 pg/mL; normal range 30-100 pg/mL) and CgA (500 ng/mL, normal range 10-110 ng/mL).

The serum calcium and parathyroid hormone levels were normal .

Imaging examinations

The esophagus-gastro-duodenoscopy (EGD) revealed severe reflux esophagitis (grade D), esophageal ulcers, and strictures.

The abdominal contrast-enhanced magnetic resonance imaging (MRI) unveiled a mass of approximately 2.6 * 3.2 cm at the head of the pancreas without liver metastasis

PATHOLOGY AND IMMUNOHISTOCHEMISTRY

The tumor cells, arranged into solid nests, exhibited abundant cytoplasm, fine and granulated chromatin, and inconspicuous nucleoli; Tumor cells showed positivity for Synaptophysin , Neuron-Specific Enolase, and gastrin.

MULTIDISCIPLINARY EXPERT CONSULTATION

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FINAL DIAGNOSIS

1. sporadic gastrinoma

2.refractory benign esophageal stricture

TREATMENT

1.The patient underwent pancreaticoduodenectomy for gasrinoma.

2.The patient underwent five sessions endoscopic bougie dilations and four sessions of endoscopic incisional therapy for the esophageal stricture

OUTCOME AND FOLLOW-UP

During the 2-years follow-up,the patient recovered from watery diarrhea after surgery. Unfortunately, during the patient's fourth EIT session, when the esophageal lumen was completely obstructed, she unexpectedly developed esophageal perforation. She was managed with conservative treatment because she declined further surgery. As a result, she could only receive long-term enteral nutrition through a nasogastric tube, significantly reducing her quality of life.

DISCUSSION

The reported case exhibits several distinctive features that warrant discussion. Firstly, the patient was characterized by chronic diarrhea and multiple peptic ulcers, which were initially misdiagnosed as chronic idiopathic diarrhea and sporadic peptic ulcers. However, it is important to note that ZES-related ulcers are usually multiple, located in unusual sites, and usually complicated by bleeding, perforation, or esophageal strictures in contrast to sporadic peptic ulcers. Moreover, approximately 75% of patients with gastrinoma present with watery diarrhea, which can easily be misdiagnosed as chronic idiopathic diarrhea, as was the case in our patient[4].

Interestingly, the case presented with an uncommon symptom of refractory esophageal stricture. ZES is typically associated with minimal esophageal morbidity because of advancements in antisecretory medications. A prospective study investigating esophageal involvement and its complications in ZES found that sporadic ZES rarely presents with a benign esophageal stricture, with a frequency as low as 0.4%[5]. After excluding esophageal cancer and drug or chemical-induced esophageal stricture, we considered that the prolonged acid reflux, attributed to gastrinoma, led to the formation of esophageal ulcers and scars, consequently resulting in refractory esophageal stricture. It is also noteworthy that the abdominal CT of the patient did not reveal any abnormalities, while the contrast-enhanced MRI showed a mass in the head of the pancreas. It consisted with the research from Peking Union Medical College Hospital indicates that, compared to conventional contrast-enhanced CT, MRI exhibits higher sensitivity and specificity in diagnosing pancreatic gastrinoma[6]. Additionally, MRI aids in determining the proximity of the tumor to the pancreatic duct.

The patient experienced severe esophageal stricture and showed no effects of proton pump inhibitor therapy. Therefore, endoscopic treatment was considered to improve esophageal stricture. Alessandro *et al* reported that endoscopic dilation employing bougies or balloon has proven successful in the majority of cases[7]. However, it was difficult to maintain a satisfactory luminal diameter despite five sessions of EBD in our case, indicating the presence of RBES. RBES is not only difficult to dilate but also tends to recur within weeks. In such instances, alternative endoscopic treatments like stent

placement or EIT combined with steroid injections should be considered. Michael *et al* conducted a study to evaluate the safety and efficacy of EIT in a pediatric population with refractory esophageal strictures and showed that in the refractory group, 61% of the patients met the criteria for treatment success and 2.3% with adverse events[8]. The patient in our case undered ultrasonography-guided and wire-guided EIT combined with triamcinolone acetonide injection and showed great improvements initially. Unfortunately, the patient experienced esophageal perforation following the fourth session of EIT. Therefore, even though EIT shows promise as an adjunct treatment option for RBES and may be considered before surgical resection even in severe cases, the complication rate, albeit low, is significant, and EIT should only be considered by experienced endoscopists in close consultation with surgery. Further prospective longitudinal studies are needed to validate this.

CONCLUSION

In conclusion, it is essential to diagnose gastrinoma as early as possible, as inadequately controlled acid secretion over an extended period increases the risk of developing severe esophageal strictures. In patients with esophageal strictures causing complete luminal obstruction, the blind reopening endoscopic incisional therapy (EIT) presents challenges and carries a high risk of perforation. Additionally, it highlights the persistent challenges that gastroenterologists encounter in managing RBES despite the availability of various treatment options.

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