

Complete Esophageal Obstruction: A Rare Complication of Zollinger-Ellison Syndrome

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INTRODUCTION

- Complete esophageal obstruction (CEO) is a rare phenomenon due to severe stenosis that obliterates the esophageal lumen.
- Occurs in 5% of patient receiving radiation for head and neck cancer. Though given its rarity, there are no reports of incidence in other etiologies.¹
- Zollinger-Ellison Syndrome (ZES) is due to gastric acid hypersecretion via gastrin-secreting neuroendocrine tumors (NET) called gastrinomas.
- Prevalence of ZES in the United States 1.7 per 100,000 people. It occurs sporadically in ~ 80% of patients while ~20% occurs in associated with MEN1.^{2,3}
- Patients diagnosed with ZES typically have minimal esophageal morbidity given advancements made in anti-secretory medications.⁴
- This case presents a rarely documented sequela of modern-day ZES patients.

CASE PRESENTATION

- A 73-year-old man with history of HIV on HAART, GERD, and ZES presented with worsening dysphagia to both solids and liquids.
- Diagnosed with ZES 2 years prior. Labs significant for gastrin level of 927 pg/ml, gastric pH 7, and serum chromogranin A of 1836.
- Gastrinoma located with PET scan demonstrating a DOTATE avid mass in the first portion of the duodenum. Screening for MEN1 was negative.
- Medical therapy was maximized to omeprazole 180 mg/day and famotidine 80 mg/day.
- Underwent tumor resection with distal gastrectomy, resection of D1, roux-en-Y hepaticojejunostomy, and truncal vagotomy.
- Underwent a total of 21 EGDs and 7 esophageal stents for dysphagia secondary to refractory esophageal strictures.
- He lost 48 lbs over 2 years despite enteral feeds and TPN.
- In January 2022, he presented with inability to swallow his own secretions and profound weakness from malnutrition.

DIAGNOSTIC ASSESSMENT

Barium
Esophagram

- Significant narrowing in mid thoracic esophagus followed by second severe narrowing without passage of contrast (Figure 1).

EGD

- Two high-grade strictures at 25 cm and 35 cm. Second stricture resulted in blind pouch and CEO (Figure 2,3)

Figure 1. Barium Esophagram revealing a high-grade stricture in mid-thoracic esophagus followed by CEO.



Figure 2. Post-dilation of first severe stricture located ~25 cm from incisors.

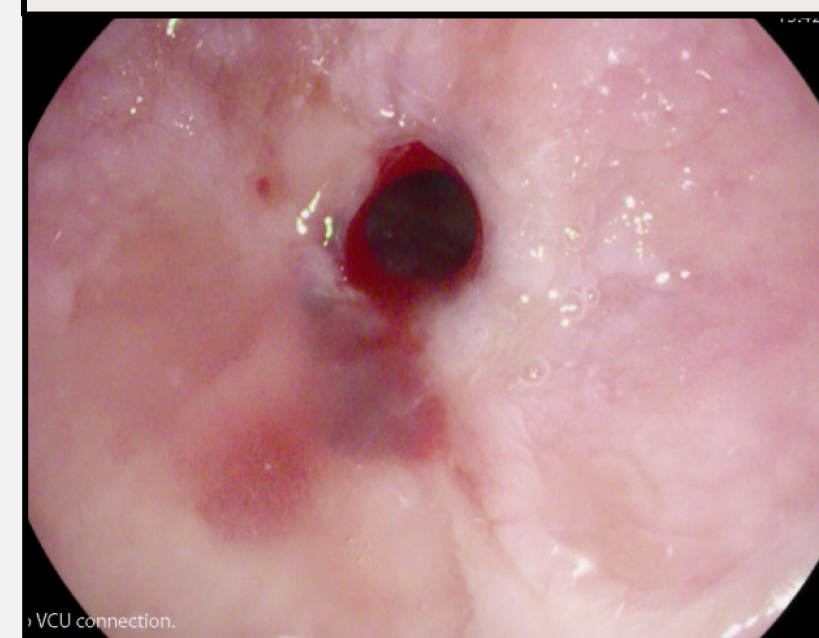
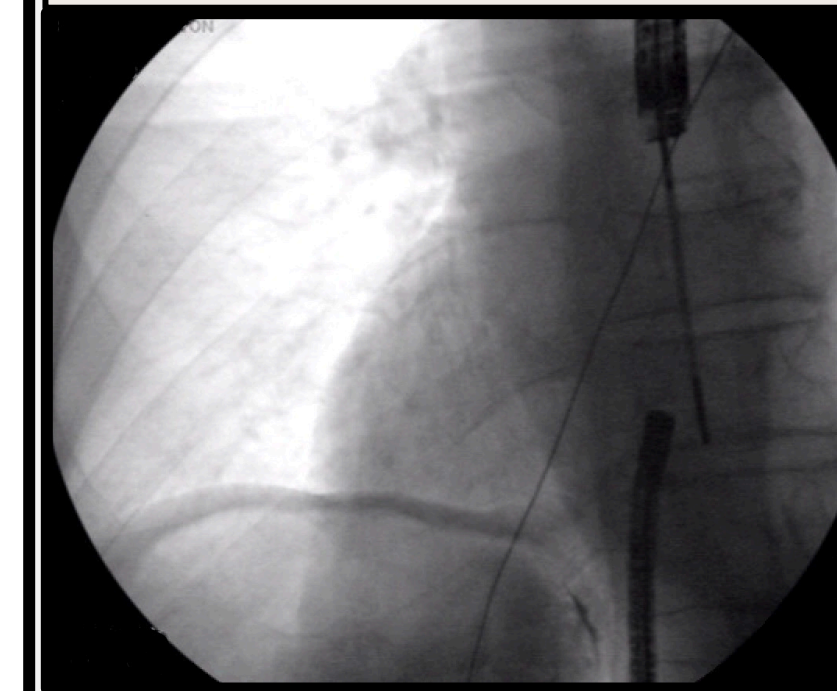


Figure 3. Second stricture resulted in blind pouch at ~35 cm from incisors.



INTERVENTION AND OUTCOME

Figure 4. Fluoroscopic imaging displaying attempt at CARD.



- Lumen restoration was attempted using combined antegrade and retrograde dilation (CARD) (Figure 4).
- This involved 2 operators, 2 scopes, and an EUS needle.
- Unfortunately, the two blind loops appeared to be ~6 cm apart in different alignments making lumen restoration unfeasible.
- Not a candidate for esophageal reconstruction with jejunal interposition due to poor nutritional status.

DISCUSSION

- This patient with ZES developed CEO despite maximal acid suppression, surgical resection, balloon dilations, and numerous esophageal stents.
- Complications of hypergastrinemia was once number one cause of morbidity in ZES but effective acid suppressive therapy has diminished rates of gastrectomy.⁵
- While there are reports of severe stenotic strictures, extensive literature review did not reveal any reports of CEO in ZES.
- Endoscopic lumen restoration such as CARD and per-oral endoscopic tunneling for restoration of the esophagus (POETRE) have been attempted.^{6,7}
- This case demonstrates that CEO is a possible sequela of ZES despite maximal medical management and endoscopic lumen restoration in non-surgical candidates can be difficult.

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