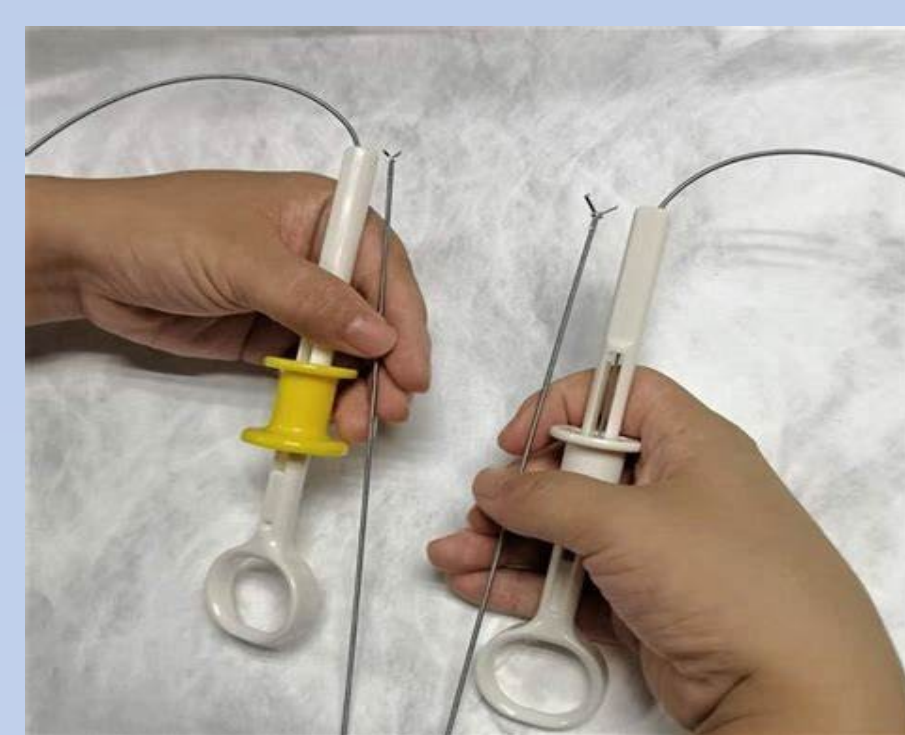


Black Esophagus (Gurvits syndrome): A Rare Case of Acute Esophageal Necrosis in a Young Adult

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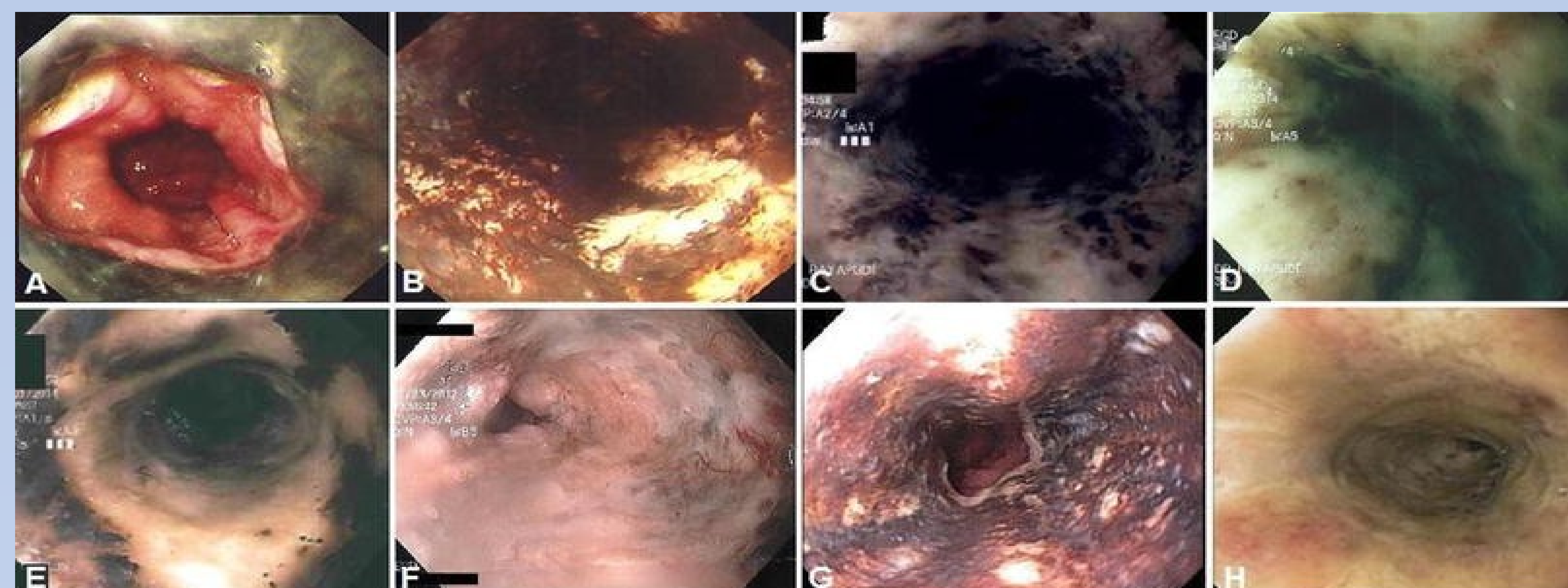
Introduction

- Estimated prevalence of Acute Esophageal Necrosis (AEN), also known as "black esophagus," is 0.01-0.28% (1)
- AEN affects men four times more than women
 - Observed in every age group, but peak incidence is in the sixth decade of life (1)
- Clinical presentation
 - Upper gastrointestinal bleeding, epigastric pain, retro-sternal chest discomfort, dysphagia (2)
- Characteristic features
 - Diffuse circumferential black mucosal discoloration in the distal esophagus that abruptly stops at the gastroesophageal junction
- Treatment
 - Correct coexisting co-morbidities, restore hemodynamic stability, intravenous gastric acid suppression with proton pump inhibitors (3)



Case Report

- A 38-year-old-male with poorly controlled diabetes mellitus (Hb A1c 14.3) was admitted with diabetic ketoacidosis and sepsis to the medical intensive care unit. The patient had been found obtunded at home. On admission, laboratory findings revealed a β hydroxybutyrate level of 12.46, fingerstick glucose of 1159 and arterial PH of 7.0.
- The patient was intubated and sedated, given fluid boluses and started on an insulin drip. He was noted to have melena four (4) days after admission.
- The gastroenterology team performed Upper GI endoscopy, which revealed circumferential inflammation with black necrotic appearing esophageal mucosa present in the distal two-thirds of the esophagus ending at the level of the esophageal-gastric junction.
- Additional findings included a small hiatal hernia, a small erythematous area of trauma within the gastric antrum (likely secondary to nasogastric tube insertion), and an unremarkable appearing duodenum. The patient was started on high dose proton pump inhibitors, antimicrobials, and was given supportive measures including good glycemic control, fluid resuscitation and enteral diet as tolerated.
- The patient's overall condition improved, and he was discharged home with follow-up in the gastroenterology clinic.



Endoscopic images of individuals with AEN

Discussion

- The pathogenesis of AEN is not fully understood, but many investigators believe it to be caused by hypo-perfusion, reflux of gastrointestinal liquids, impaired mucosal defenses, or some combination of these factors (6)
- Patients found with AEN commonly present with chief complaint of hematemesis, coffee ground emesis, and melena (6)
- The average age for presentation for AEN is reported to be around 60-70 years old (1, 6, 7)
- Prognosis for patients with AEN is also poor when paired with other co-morbidities.
- Around 32% of patients die, primarily from underlying conditions (6, 7)
- Prognosis also seems to be related to the overall size of necrosis, with one review finding that 29 out of a group of 32 patients who died with AEN had 2/3 or more of necrotic tissue in the esophagus. (7)

Conclusion

- AEN or "black esophagus" is a rare yet intriguing gastroenterology disease syndrome. The etiology is variable and multifactorial dictating the consequent management plan. The diagnosis should be entertained mostly in old patients with multiple co-morbidities like hypertension, heart disease, stroke and diabetes who present with an upper gastrointestinal hemorrhage.
- Early upper endoscopy is essential for the diagnosis as well as fast and effective management. Patients who present without complications can be treated conservatively with proton pump inhibitors and management of other co-morbid conditions. However, the majority of patients present with anemia and other complications like esophageal perforation which are associated with increased mortality.
- Hence, patients' hemodynamic status needs to be closely monitored and managed effectively including surgical intervention where necessary.

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