Acute Esophageal Necrosis, A Rare Entity: Case Report and Review

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Abstract

Acute esophageal necrosis (AEN), also known as "black esophagus" or "acute necrotizing esophagitis", is a rare condition with a multifaceted pathogenesis. Patients typically present with upper gastrointestinal bleeding and have associated epigastric pain, fever, nausea, and dysphagia. AEN is characterized by circumferential esophageal mucosal necrosis, appearing as black discoloration on esophagogastroduodenoscopy (EGD). AEN is usually seen in patients with chronic comorbidities (renal insufficiency, diabetes mellitus, malnutrition, hematologic/solid organ malignancies, etc.) and many cases arise in acute settings that involve either multiorgan dysfunction, vasculopathies, and/or low-flow states such as diabetic ketoacidosis (DKA) and sepsis. Although AEN is a rare phenomenon, mortality rates have been reported to be as high as 32% in this population. Here, we discuss a case of a 43-year-old patient that presented with non-bloody emesis and abdominal pain, who was found to have DKA and subsequently AEN.

Introduction

Esophagitis has many etiologies including gastric reflux, infection such as CMV or HIV, radiation or drug induced, and caustic ingestion (5). Findings on CT for esophagitis may be similar to that seen in AEN, therefore prompt EGD and biopsy are needed to rule out causes of esophagitis and to confirm AEN. The pathogenesis of AEN is multifaceted and likely caused by ischemia due to low flow states, shock, DKA, gastric outlet obstruction with gastroesophageal reflux, and even reported after certain medication use (3). Patients with AEN are usually elderly males with multiple comorbidities (cardiovascular disease, diabetes, malignancy, drug abuse, renal insufficiency) and commonly but not always present with signs of upper gastrointestinal bleeding. Other associated symptoms include abdominal pain, dysphagia, nausea, vomiting, low-grade fever, and symptoms of low volume status. AEN is a rare condition, and the true prevalence is relatively unknown due subclinical presentations, potential delay in performing endoscopy, and early mucosal healing in the disease process with transient ischemic or chemical insult. AEN may have an incidence ranging from 0.01-0.2% according to certain endoscopic series. It is also more common among males with a peak incidence in the sixth decade of life (3).

Case Presentation/ Differential Diagnoses

HPI: 43-year-old male with a past medical history of uncontrolled insulin-dependent **diabetes mellitus type 1**, osteomyelitis s/p toe amputation and neuropathy presents following loss of consciousness and three days of **abdominal pain**, **persistent non-bloody vomiting**, nausea, and reports poor p.o. intake. The patient also had non-bloody diarrhea two days prior which had resolved. He reports epigastric pain with no radiation. Nothing provokes or relieves the pain.

Admitted to: ICU for severe DKA.

ED Vitals: HR 147, Temp 36.5, RR 18, BP 105/74, SpO2 100% on RA

Physical Exam: Tender to palpation Diffuse abdominal pain, mainly in LLQ

Pertinent Labs: VBG: pH 7.28, pCO2 29, pO2 22, Bicarb 12

CBC- WBC 40.5, Hgb 13.6, platelets 465

BMP: Na 118, K 3.8, Cl 86, Anion Gap 20, Cr 1.63, BUN 53, Glucose 775

Lactate 5.0, A1c 13.1

Differentials based on EGD:

-Melanosis
-Pseudomelanosis
-Acanthosis nigricans
-Coal dust
-Caustic ingestion
-Pseudomembranous Esophagitis

Diagnostic Imaging/Test Findings

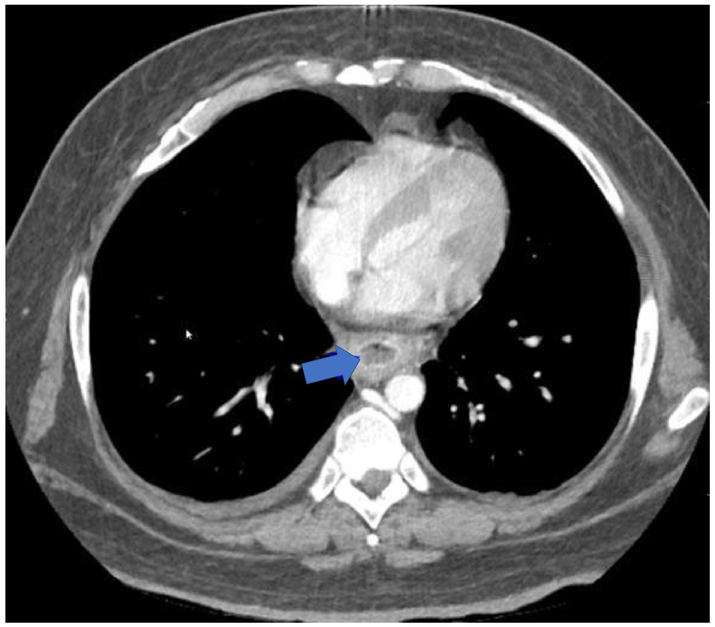

Figure 1. Axial IV contrast-enhanced CT of the thorax in the mid-lower esophagus and abdomen. Figure 1. shows circumferential wall thickening with external wall enhancement with lack of mucosal enhancement (blue arrow). This is a non-specific finding.

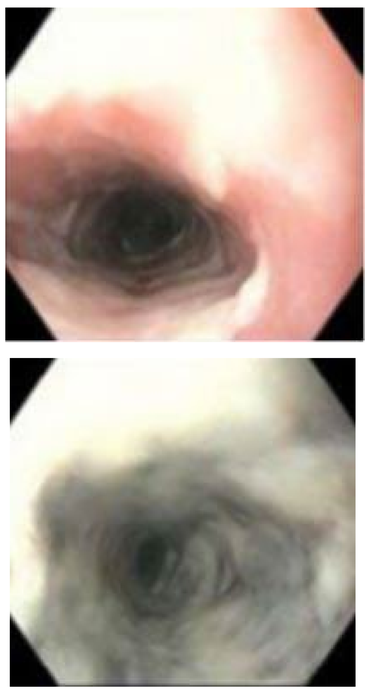

Figure 2. EGD showed circumferential white exudative esophagus with underlying black pigmentation from 25cm to 35cm and scattered erythema/ulceration and white exudates in upper esophagus.

Biopsy Results

-Small intestines with ulceration and focal necrosis, negative for H. pylori

-Stomach with antral gland mucosa showed chronic gastritis and mild reactive changes, Negative for H. pylori.

-Esophagus with extensively necrotic tissue with abscess formation

Treatment Options

There have been no established guidelines on management of AEN, largely due to its low incidence and prevalence along with unclear pathophysiology. Mortality related specifically to AEN may only be around 6% but the overall prognosis depends on comorbidities and the etiology of AEN (3). Management usually consists of intravenous fluids, correcting anemia, gastric acid suppression with proton pump inhibitors, sucralfate, and nil per os for at least 24 hours. It is vital to also treat underlying conditions that potentially incited the event, such as DKA, sepsis, anemia, gastric outlet obstruction etc., to prevent further complications such as esophageal strictures and less commonly esophageal perforation (3). Surgical intervention is only necessary if there is esophageal perforation resulting in mediastinitis and/or abscess formation (3).

Summary/Conclusion

-Comorbidities such as CKD and diabetes mellitus are risk factors, and low flow states may incite the event.

-Findings on CT are nonspecific and are suggestive of esophagitis which has an extensive differential diagnosis. Clinical presentation and history are key.

-Almost always affects the distal esophagus and stops at GE junction.

-EGD is required for diagnosis.

-Treatment is managing the underlying acute conditions and rarely surgery if there is esophageal perforation/mediastinitis.

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