Black Esophagus or Acute Esophageal Necrosis: A Case Series and Single-Institution Experience

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INTRODUCTION: Acute esophageal necrosis (AEN) or ‘black esophagus’ is a rare disease of the esophagus with an endoscopic prevalence of 0.02%. It generally presents with signs of upper GI bleeding including melena and hematemesis. Its pathophysiology is largely attributed to severe illness states that affect the perfusion of the esophagus. The distal esophagus being least vascular is most commonly affected. Upper endoscopy reveals circumferential black discoloration. Biopsies are taken to confirm necrosis, as well as rule out infection or pigmentation. Management is generally supportive, with endoscopic resolution seen in many cases with correction of the underlying insult.

CASE DESCRIPTION/METHODS: Three patients with AEN were identified over the last 2 years at our institution. Two females aged 67 and 78 were admitted within the same month in the fifth and sixth decades of life. Existing literature reports DKA as a relatively common precipitating factor. All of our patients presented with upper GI bleed secondary to AEN and were in DKA superimposed on low perfusion states due to CHF, atherosclerotic disease and sepsis respectively. Two out of three patients were females. Based on our case series, it appears that DKA, a critical illness in itself, further worsens existing low perfusion states. This can manifest as upper GI bleeding. DKA demands prompt correction, especially in patients with concomitant vasculopathy.

CASE 1: A 35-year-old male with poorly-controlled T1DM (A1c 9.1%) who noticed intermittent odynophagia for a week. Family history of diabetes. Physical exam: Temperature 98.6°F, blood pressure 130/80 mmHg, heart rate 80 bpm, respiratory rate 18 breaths/min. Skin examination revealed tense bullae on upper extremities (Figure 2). Abdominal exam was notable for epigastric tenderness. Laboratory exams: Hgb 8.1 g/dL, WBC 13.5, Neutrophils 10.5. Blood cultures negative. Upper endoscopy showed a 3 cm esophageal obstructive mass-like lesion in the lower third of the esophagus with oozing at the base (Figure 2A). Biopsies showed non-specific findings including necrosis and foreign material. The following day, the patient tolerated solid food without dysphagia. A second upper endoscopy showed a ~3 cm esophageal tear with a small adherent clot at the site of the previously noted mass-like lesion (Figure 2B). He continued to tolerate a regular diet without further symptoms.

CASE 2: A 67-year-old female with T2DM and hypertension. She was diagnosed with CHF with reduced ejection fraction (40%). Both patients had documented T2DM and atherosclerotic disease. Laboratory findings showed elevated serum glucose and anion-gap metabolic acidosis with elevated beta-hydroxybutyrate. Both underwent EGD which was notable for a circumferential black esophagus that abruptly terminated at the gastro-esophageal junction (Figures 1 and 2). The third patient was a 35-year-old male with poorly-controlled T1DM (A1c 9.1%) who noticed melena stools 2 days prior and presented with hypovolemic shock. EGD was consistent with black esophagus. Biopsy showed acute inflammatory exudates with necrotic cells and numerous fungal hyphae. He was started on micafungin for two weeks and twice-daily PPI. EGD 8 weeks later showed complete resolution of initial findings (Figure 3). 

DISCUSSION: AEN is a rare cause of upper GI bleed, more commonly reported in men and in patients with underlying vasculopathies who present with severe illness. Peak incidence is in the sixth decade of life. Existing literature reports DKA as a relatively common precipitating factor. All of our patients presented with upper GI bleed secondary to AEN and were in DKA superimposed on low perfusion states due to CHF, atherosclerotic disease and sepsis respectively. Two out of three patients were females. Based on our case series, it appears that DKA, a critical illness in itself, further worsens existing low perfusion states. This can manifest as upper GI bleeding. DKA demands prompt correction, especially in patients with concomitant vasculopathy.

Black Esophagus: A Rare Finding in a Patient With Bullous Pemphigoid

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INTRODUCTION: Bullous pemphigoid (BP) is a chronic relapsing autoimmune blistering disease characterized by circulating antibodies directed against specific epitopes of hemidesmosomes involved in anchoring the epidermis to the dermal layer. We report a case of acute onset esophageal bullae identified in a patient with active skin BP seen only upon withdrawal of the upper endoscope, not present on insertion. This sign is analogous to Nikolsky’s sign where pressure or shearing results in formation of new bullae.

CASE DESCRIPTION/METHODS: A 57-year-old Caucasian female with Type II DM, BP, CKD stage 3a presented to hospital for 5-6 episodes of melanic stools for two days. She was diagnosed in September 2017 with BP by skin biopsy and direct immunofluorescence (Figure 1). At the time of presentation, she was normotensive, afebrile, heart rate 102 beats/min and respiratory rate of 14 breaths/min. Skin examination revealed tense bullae on upper extremities (Figure 2). Abdominal exam was notable for tenderness to palpate in the epigastric region. Digital rectal exam revealed subepidermal blister formation with numerous eosinophils within the cleft (black arrow).

DISCUSSION: Bullous pemphigoid (BP) is a chronic relapsing autoimmune blistering disease characterized by circulating antibodies directed against specific epitopes of hemidesmosomes involved in anchoring the epidermis to the dermal layer. We report a case of acute onset esophageal bullae identified in a patient with active skin BP seen only upon withdrawal of the upper endoscope, not present on insertion. This sign is analogous to Nikolsky’s sign where pressure or shearing results in formation of new bullae.
Esophageal diverticula (ED) are a rare structure abnormality with prevalence less than 10% of all esophageal diverticula, with an incidence of 1:500,000/year. Here we report a case of a true esophageal epiphrenic diverticula in the setting of heterotopic gastric mucosa patches (HGMPE) found after presentation with epigastric symptoms.

**CASE DESCRIPTION/METHODS:** A 35-year-old female with a past medical history of endometriosis presented with a few months of epigastric pain, nausea, and vomiting with no response to treatment for gastroesophageal reflux disease (GERD) prompting investigation with esophagogastroduodenoscopy (EGD) which revealed a diverticulum 28cm from the incisors suggestive of a true diverticulum at the posterolateral aspect of the esophagus. Z line was at 38 cm from the incisors. EUS showed a single esophageal diverticulum from 28 to 30 cm with all four wall layers indicating a true diverticulum. She returned for endoscopic ultrasound (EUS) for further evaluation with development of dysphagia with no regurgitation or halitosis. Repeat EGD prior to EUS revealed areas suggestive of ectopic gastric mucosal patches 28 cm from the incisors with a single diverticulum at the posterolateral aspect of the esophagus. Pathology from biopsy at the 29 cm showed cardiac type mucosa with chronic inflammation without evidence of metaplasia, dysplasia, or intramucosal eosinophils significant for HGMPE.

**DISCUSSION:** ED, particularly EED are a rare entity with varying clinical manifestations including dysphagia, regurgitation, weight loss, chest pain, halitosis, aspiration, and heartburn with the majority of patients being asymptomatic until the diverticula are larger than 5cm. Moreover, EED typically are false diverticula with the involvement of the mucosa and submucosa as a result of an esophageal motility disorder. Within ED, EED constitutes less than 10% of all esophageal diverticula, with an incidence of 1:500,000/year. Here we report a case.