A Case of Acute Esophageal Necrosis

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ABSTRACT

Acute esophageal necrosis (AEN) is a rare condition that impacts the distal esophagus after a vascular insult. In this case report, we describe a patient with multiple co-morbid conditions presenting with shock from gastroenteritis who subsequently developed AEN complicated by refractory strictures.

KEYWORDS: acute esophageal necrosis; black esophagus; esophageal ischemia; gastroenteritis; upper gastrointestinal bleeding

INTRODUCTION

Acute esophageal necrosis (AEN), also known as black esophagus or necrotizing esophagitis, is a rare but serious condition found in approximately 0.2% of upper gastrointestinal (UGI) endoscopy cases.[1-3] Patients typically present with dysphagia, chest pain, abdominal discomfort or signs of upper gastrointestinal bleeding.[4] The etiology for AEN is unknown but is theorized to result from microvascular thrombosis to the esophagus due to ischemic, infectious or chemical mucosal injury from vomiting, severe reflux, acid buildup or gastric outlet obstruction caused by gastric volvulus or peripyloric ulcer.[3-5] While AEN can resolve with supportive care, the mortality rate can be as high as 35%, largely due to impaired esophageal function in combination with underlying medical co-morbities.[4,6] Potential complications include bacterial superinfections, esophageal perforation and esophageal stricture formation.[6] In this case report, we present a patient diagnosed with AEN in the setting of gastroenteritis complicated by refractory esophageal stricture.

CASE PRESENTATION

A 78-year-old female presented to the emergency department with several weeks of watery diarrhea, abdominal pain and intermittent chest pain provoked by eating, as well as decreased oral intake due to dysphagia. She had a history of ulcerative colitis, Schatzki's ring, atrial fibrillation, hypertension, hyperlipidemia, and cerebrovascular accidents with residual left-sided weakness, At admission, medications included allopurinol, amlodipine, aspirin, atorvastatin, clopidogrel, diazepam, doxazosin, labetalol, and mesalamine. Upon presentation, she was hypotensive with a blood pressure of 70/40 mmHg, pulse of 100 beats/min, and a temperature of 95.8 F. Physical examination was notable for diffuse abdominal tenderness without signs of peritoneal irritation. Pertinent laboratory studies included a white blood cell count of 26,000 leukocytes/mcL, hemoglobin of 7.7 g/dL, and platelet count of 234,000. She had several metabolic abnormalities including a sodium 133 meq/L, chloride 97 meq/L, bicarbonate of 13 meq/L, blood urea nitrogen of 168 mg/dL, and serum creatinine of 8.96 mg/dL. After hemodynamic stabilization, a computer tomography (CT) scan of the abdomen and pelvis revealed multiple fluid-filled, nondilated loops of small and large bowel as well as circumferential thickening at the gastroesophageal (GE) junction. The patient was diagnosed with gastroenteritis. She continued to have episodes of hypotension and had an acute drop in hemoglobin to 6.4 g/dL, but was responsive to intravenous fluids, packed red blood cells, and broad-spectrum antibiotics.

Several days following presentation, the patient clinically improved but continued to complain of intermittent odynophagia with vague substernal chest pain. A barium swallow study revealed partial obstruction at the gastroesophageal junction concerning for a thickened gastric fold or polypoid mass. An UGI endoscopy showed black, necrotic mucosa circumferentially throughout the mid and lower third of the esophagus (**Figure 1**). Biopsies revealed necrotic tissue fragments colonized by coccoid organisms. Although cultures failed to grow any organisms, she was managed empirically with Fluconazole and Amoxicillin/Clavulanate.

Repeat endoscopic evaluation six weeks later revealed a significant stenosis with stricture formation in the area overlying the initial insult (**Figure 2**). Multiple endoscopic sessions with balloon dilatation and stenting were attempted, but the patient continued to suffer from dysphagia and refractory strictures. A percutaneous endoscopic gastrostomy (PEG) tube was ultimately placed.

DISCUSSION

Acute esophageal necrosis is a rare phenomenon characterized by diffuse, circumferential, black-appearing, distal esophageal mucosa that stops abruptly at the gastroesophageal junction (GEJ).[8] Exceptionally rare cases of proximal

Figure 1. Initial endoscopic appearance revealing diffuse, circumferential esophageal necrosis involving the mid and distal esophagus.

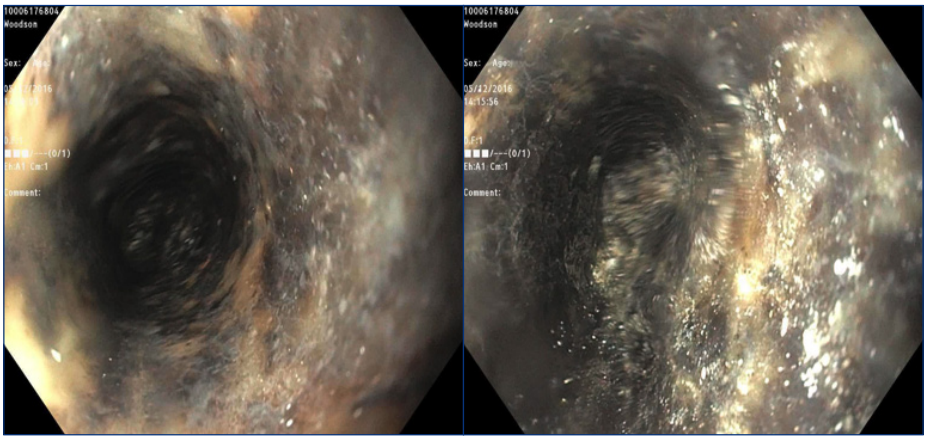
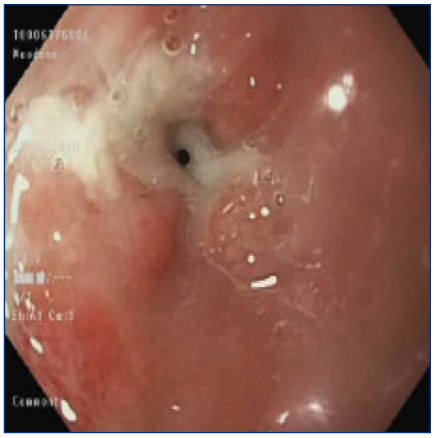


Figure 2. Severe esophageal stricture persisting after healing and requiring serial dilatation



"black esophagus" have been reported.[9] While the exact mechanism resulting in AEN is unknown, it is suspected to be multifactorial. Current hypothesis include tissue hypoperfusion, massive esophageal influx of gastric content, and impaired local defense barriers overwhelming the esophageal mucosa, resulting in profound ischemic changes.[10] Comorbid conditions, including diabetes, atherosclerotic disease, immunosuppressive states, chronic kidney disease, alcohol or cocaine abuse and advanced age, may act as predisposing factors in susceptible individuals.[7,11] Low flow states, as seen in sepsis, shock, congestive heart failure, cardiac arrhythmia, pancreatitis, acute blood loss, and hyperthermia, may also compromise the esophagus. Critical illness, poor nutritional status, and general deconditioning may further compromise local protective barriers, impair defense mechanisms and potentiate ischemic and chemical injury.

Clinically, the majority of patients present with hematemesis or melena after an UGI bleed.[1-9] Other associated symptoms can include dysphagia, epigastric pain, atypical chest pain, and low-grade fever. Symptoms may also be related to the underlying process including signs of sepsis, tachycardia, hypotension or altered mental status, usually presenting within 24 hours after an inciting event. Anemia and leukocytosis are often present; if CT is performed, it may reveal distal esophagus thickening.

The definitive diagnosis for AEN is made with endoscopy, which, in this patient, revealed a grossly necrotic, friable and ulcerated esophagus in the mid to distal portions with an abrupt transition point into a normal appearing stomach. The distal third of the esophagus is commonly affected due to its hypovascular anatomy, rendering it more vulnerable to ischemic insult. Histologically, necrotic debris extends into the submucosa with local inflammatory response, which, as demonstrated in this case, should always be sent for culture to exclude an underlying bacterial or fungal infection.[10] Rarely, necrosis is mild or healing at endoscopy, increasing the risk of delayed or missed diagnosis.

AEN carries a very poor prognosis. As a result, the initial management requires hemodynamic stabilization with intravenous fluids, aggressive acid suppressive medications with a PPI, and NPO status.[12] Nasogastric tubes are usually contraindicated as they may increase the risk of perforation.

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