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Acute Esophageal Necrosis (Gurvits Syndrome) Presenting as Globus and Altered Phonation

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Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Female, 80-year-old
Final Diagnosis: Acute esophageal necrosis • acute pharyngitis • duodenal ulcerations • gastric erosions • gastritis
Symptoms: Dysphagia • emesis • globus • hoarseness • sore throat
Medication: —
Clinical Procedure: Esophagogastroduodenoscopy
Specialty: Gastroenterology and Hepatology

Objective: Rare co-existance of disease or pathology

Background: Acute esophageal necrosis (AEN), also known as black esophagus or Gurvits syndrome, is an infrequently seen clinical condition distinguishable by a visually striking endoscopic appearance of necrotic esophageal mucosa that involves the distal esophagus with proximal extensions ending at the gastroesophageal junction. Since its early recognition pathologically in the 1960s and endoscopically in the 1990s, AEN, despite its rarity, is being increasingly recognized as a demonstratable cause of upper gastrointestinal bleeding. Cases of pan-esophageal necrosis are sparsely reported, leaving management guidance to isolated case reports.

Case Report: An 80-year-old female smoker with advanced chronic obstructive pulmonary disease presented with signs and symptoms of acute pharyngitis and globus sensation that had been evolving over the preceding weeks. An esophagogastroduodenoscopy revealed circumferential necrotic mucosa encircling the entire lumen of the esophagus. The patient was made nil-per-os and started on high-dose anti-reflux therapy with adequate hemodynamic resuscitation.

Conclusions: AEN is multifactorial but primarily a combination of decreased tissue perfusion and a massive influx of gastric contents in settings of impaired local defense barriers. Despite its dramatic presentation, the majority of cases resolve with conservative medical management, foregoing surgical interventions.

MeSH Keywords: Endoscopy, Digestive System • Esophageal Diseases • Esophagitis • Pharyngitis

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/926019

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Background

Acute esophageal necrosis (AEN), also known as "black esophagus" or Gurvits syndrome, is a rare phenomenon with classic endoscopic findings. Dispersed circumferential black mucosal discoloration of the distal esophagus extending proximally to various lengths and ending abruptly at the gastroesophageal junction (GEJ) is most commonly noted. Its historic incidence varies from 0.01% to 0.28% according to earlier studies [1]; however, these numbers likely under-represent the true prevalence of this disease and can increase with the use of modern endoscopy. A recent study from Japan estimated that up to 6% of upper gastrointestinal bleeding (GIB) might be related to AEN [2]. Associated endoscopic findings can indicate duodenal ulcer disease in up to 50% of the cases [3]. AEN has a predilection for males and the elderly with a median age of presentation of 68 years, and the most common presenting symptoms are GIB, vomiting, melena, chest pain, abdominal pain, and dysphagia [4]. Its etiology is multifactorial, mainly comprising a combination of ischemic insult, the corrosive effect of a massive influx of gastric contents, and impaired local esophageal defense barriers. Systemic conditions such as diabetes mellitus, cardiovascular disease, chronic renal disease, alcohol abuse, malnourishment, and general debilitation place patients at the highest risk of developing AEN. The mainstay of management remains the correction of coexisting medical conditions, aggressive hemodynamic resuscitation, nil-per-os (NPO) restriction, and high-dose proton pump inhibitor (PPI) therapy. Repeat endoscopy may be performed to verify the normalization of the esophageal mucosa. The overall prognosis of AEN is poor, with 32% of patients succumbing to underlying critical illness, whereas AEN-related mortality is approximately 6% [1,3]. We present an unusual case of acute pan-esophageal necrosis in an 80-year-old woman with an atypical clinical presentation and nearly normal objective tests.

Case Report

An 80-year-old female smoker with a history of advanced chronic obstructive pulmonary disease (COPD) and major depression on 4 L of home oxygen presented to the hospital with a sore throat, globus sensation, voice hoarseness with a notable change in her speech sound, and dysphagia followed by repeated episodes of clear emesis which had been evolving over the preceding weeks. Her prescription medications included bupropion, albuterol, ipratropium, and fluticasone inhalers. On physical examination, she was visibly ill, although not toxic appearing, and the only notable vital sign was tachycardia with a heartrate up to 133 beats per minute. She displayed significant cachexia (Body Mass Index 13), dry mucous membranes, and an erythematous posterior pharynx that was unremarkable for exudate, mass effect, or lymphadenopathy.

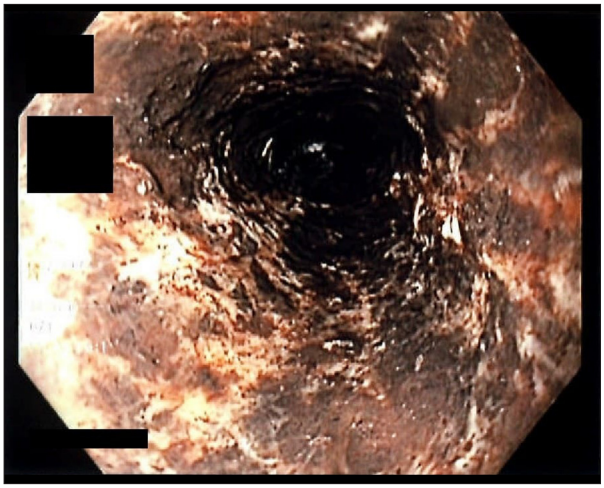

Figure 1. Esophagogastroduodenoscopy (EGD) exposing a necrotic esophageal mucosa.

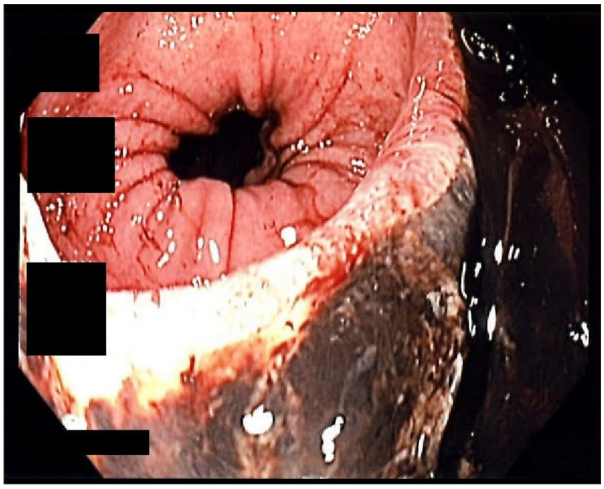

Figure 2. Esophagogastroduodenoscopy (EGD) showing a sharp demarcation at the gastroesophageal junction (GEJ).

Laboratory analysis demonstrated a white blood cell count of 16 100/mL with left shift and intact hemoglobin of 15.8 g/dL without any evidence of frank or occult GIB; her metabolic panel was unremarkable. A computed tomography scan of the neck revealed localized inflammatory changes of the pharynx, mucosal hyperemia, and submucosal edema along the upper thoracic esophagus. An esophagogastroduodenoscopy (EGD) revealed black necrotic-appearing mucosa (Figure 1) encompassing the entire esophagus and ending abruptly at the GEJ (Figure 2) with associated gastric erosions (Figure 3) and proximal duodenal ulcerations (Figure 4). A biopsy was obtained and the histochemical analysis revealed acute esophageal sterile gangrenous necrosis (Figure 5). The patient was diagnosed with AEN. Initially, there was a concern for mass effect, although the diffuse erythema and hyperemia compounded by the onset of symptoms were concerning for acute pharyngitis. Results of an antistreptococcal antigen test and upper

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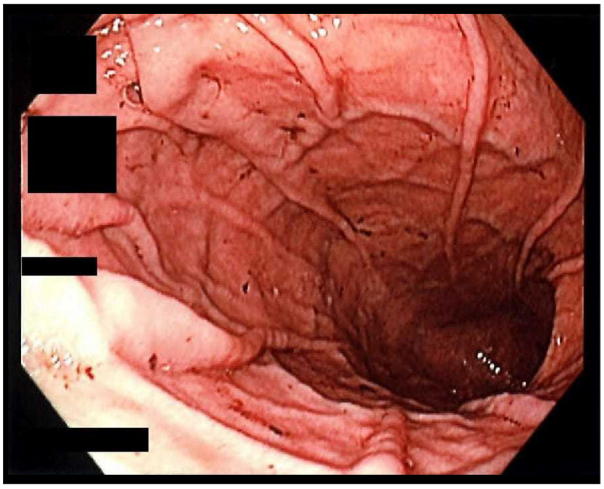

Figure 3. Esophagogastroduodenoscopy (EGD) revealing the gastric mucosa with erosions.

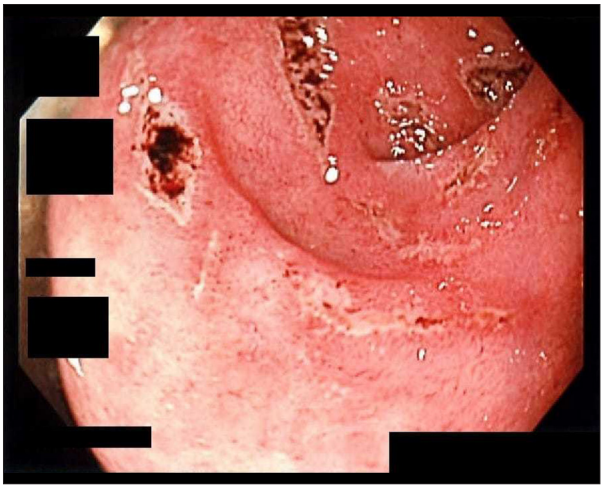

Figure 4. Esophagogastroduodenoscopy (EGD) revealing proximal duodenal ulcerations.

respiratory viral pathogen panel were negative. Upon visualization of the necrotic surface on EGD, other possible etiologies such as malignant melanoma, pseudomelanosis, caustic injuries, iron supplement usage, coal dust deposition, acanthosis nigricans, and melanocytosis of the esophagus were considered. However, a detailed history and physical examination, along with endoscopic findings in this clinical setting, established the diagnosis of AEN. Shortly after the patient arrived in the emergency department, intravenous fluids were administered because she appeared dehydrated. Intravenous dexamethasone and ceftriaxone were administered for pharyngitis, and nausea was corrected with ondansetron. Prophylactic doses of enoxaparin for deep vein thrombosis prevention were initiated. Her oxygen requirements remained unchanged during the admission, and she was maintained at her home level of 4 L per minute. Intravenous esomeprazole 40 mg twice daily was started in the emergency room, and liquid sucralfate

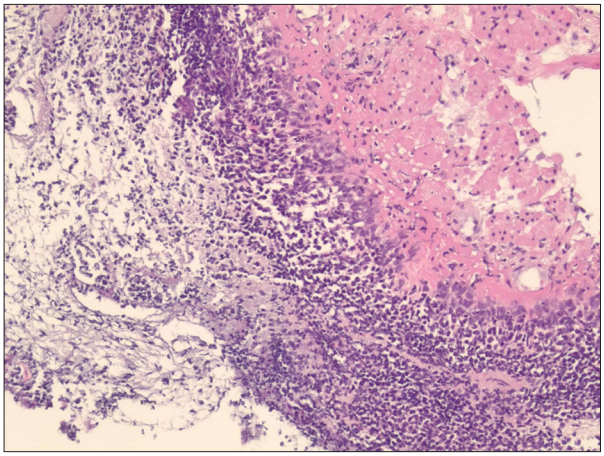

Figure 5. Histologic appearance of acute esophageal necrosis or black esophagus: hematoxylin and eosin (H&E) staining showing necrotic debris, absence of epithelium, granulation tissue, and robust leukocyte infiltrates at 100× magnification.

suspension was administered after the EGD. Our hospital dietician regulated nutritional support with bowel rest emphasized in the immediate post-endoscopy period, and a speech therapist initiated phonation exercises. The patient was admitted for 4 days, and toward the end of her admission, feeds were restarted and advanced sequentially based on tolerance. Before discharge, the patient was scheduled for a follow-up EGD in 3 months. Unfortunately, however, because of the COVID-19 epidemic, our endoscopy center services were limited, and the patient was lost to follow-up despite our attempts to re-establish care.

Discussion

AEN, or black esophagus, is becoming an increasingly recognizable cause of upper GIB with an eye-catching appearance of black necrotic esophageal mucosa almost universally involving the distal esophagus, with various degrees of proximal extensions and an abrupt cessation at the GEJ [1,3]. Its etiology is triggered by a combination of tissue hypoperfusion (seen in cardiovascular compromise), prolonged and massive reflux of gastric contents (seen in alcohol intoxication, diabetic ketoacidosis, and duodenal ulcer disease), and curtailed mucosal defenses of the esophagus (seen in malnourished, oncologic, and diabetic patients) due to ineffective compensatory mechanisms [5,6]. Common blood supply (left gastric or left phrenic) off the celiac axis to the distal esophagus and duodenum may expound the frequently associated findings of duodenal ulcer disease, which can, in turn, parallel the extent of the esophageal injury from concomitant gastric outlet obstruction [3]. The middle and distal esophagus is the most prone to ischemic injury due to its watershed vascular characteristics [4,7].

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AEN has a predilection for the elderly, as this population often has comorbidities predisposing them to a low hemodynamic state and the inability to maintain a sufficient physiologic reserve [8,9]. Endoscopic appearance is usually diagnostic, with the majority of cases being managed conservatively and a small fraction by surgical intervention [1,3,5]. Tissue biopsy is not required but can confirm the diagnosis with supported histologic findings of necrotic debris, absence of a viable epithelium, and frequent submucosal involvement, potentially leading to full-thickness lysis [10,11]. In our patient, the pharyngitis which led to inflammation with decreased oral intake and dehydration from post-tussive emesis could explain her presentation.

Most AEN cases resolve spontaneously with reversal of the underlying medical illness, hemodynamic resuscitation, NPO restriction, and aggressive PPI therapy. Antimicrobial therapy can be warranted in cases of evolving sepsis, rapid decompensation or immunocompromise, or suspected esophageal perforation. In contrast, surgical intervention is reserved for rare cases of esophageal perforation complicated by mediastinitis or abscess formation. Stricture formation or stenosis can be observed in over 10% of the cases, and endoscopic dilatation is typically successful. The overall prognosis of AEN is poor, with a third of patients dying from associated critical illness, while mortality specific to AEN is nearly 6% [1,3,11]. To our knowledge, this is the first presentation of esophageal necrosis associated with pharyngitis-induced vomiting. Our patient's COPD-related vasculopathy, massive gastroesophageal reflux from repeated emesis, and generalized cachexia potentiated by pronounced dehydration and hypovolemia led to the development of the black esophagus. Prompt recognition and treatment of the AEN coupled with appropriate management of her coexisting medical conditions resulted in effective care for this elderly patient.

Conclusions

AEN is a medical condition with a striking endoscopic appearance commonly found in the elderly and individuals with multiple comorbidities and remains an unusual but significant cause of upper gastrointestinal bleeding. The distal esophageal mucosa is at particular risk from hypoperfusion because of its watershed architecture, and pan-esophageal necrosis has been reported in association with duodenal ulcer disease. Tissue biopsies are not required but can support the diagnosis. The overall prognosis of AEN is poor, with nearly a third of patients dying from associated critical illness, while mortality specific to AEN is nearly 6%. Stricture formation or narrowing can be observed in over 10% of the cases, and endoscopic dilatation is typically successful.

Conflict of interest

None.

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