Acute Esophageal Necrosis: A Case Report and Review of the Literature

by Jacob Lewey, Douglas G. Adler

CASE REPORT

A 45-year-old man with longstanding diabetes and alcoholism undergoes an elective outpatient laparoscopic cholecystectomy which is complicated by transient hypotension. The patient was rapidly resuscitated by the anesthesiologist but in recovery, after extubating, complained of severe chest pain. An EKG showed tachycardia but was otherwise normal. The patient then had witnessed hematemesis and a GI consult was obtained. A stat esophagogastroduodenoscopy was performed which showed severe mucosal necrosis of the mid to distal esophagus terminating at the gastroesophageal junction. (Figure 1a and 1b) The patient was admitted to the ICU with a diagnosis of acute esophageal necrosis.

I. Introduction

Acute esophageal necrosis (AEN), also known as "black esophagus" and rarely as "Gurvits Syndrome," is characterized by necrosis of the esophageal mucosa.[1] AEN generally manifests endoscopically as circumferential black tissue in the mid to distal esophagus with an abrupt transition to healthy mucosa occurring at the gastroesophageal junction (GEJ), although in some cases the entire esophagus can be involved.[2] AEN is rare with an occurrence rate of 0.01%-0.02%.[1] AEN carries a high mortality rate, variously reported as 32%-38%.[3] The mortality rate specific to complications secondary to AEN has been estimated to be 6%.[4] AEN has been shown to be most common in elderly men in their sixth decade of life.[2] Possible risk factors for AEN include coronary artery disease, diabetes mellitus, hypertension, malignancy, and alcohol abuse.[1]

Jacob Lewey, Biomedical Sciences, Colorado State University, Fort Collins, CO Douglas G. Adler MD, FACG, AGAF, FASGE Director, Center for Advanced Therapeutic Endoscopy (CATE) Porter Adventist Hospital, Centura Health, Denver CO

II. Pathophysiology and Presentation

The distal portion of the esophagus receives its blood supply from the left gastric and left phrenic arteries and is less densely vascularized than the proximal esophagus. As such, the distal esophagus is properly described as a "watershed" area.[5,6] Ischemic insult is one of the major factors in the pathophysiology of AEN.[7] A "two-hit" hypothesis has been proposed to explain the pathophysiology of AEN, consisting of hypoperfusion predisposing the mucosa and possibly the submucosa to chemical insult from gastric reflux resulting in necrosis of esophageal mucosa with a neutrophilic response.[8] Additionally, weakening of mucosal defense mechanisms, such as acid buffering, may contribute to the development of AEN.[9] AEN typically presents with acute upper gastrointestinal bleeding (including, hematemesis, melena, or both), odynophagia, dysphagia, epigastric pain, and chest pain.[2,4] Hyperglycemia is present in approximately 90% of cases, likely due to DKA being a common etiology.[8,21] Hematemesis, melena, and coffee ground emesis are present in 70% of cases.[6,7]

III. Etiology

A. Cardiovascular Compromise

Hypoperfusion may be caused by embolism, thoracic aortic aneurysm (TAA), or a drop in blood pressure. Pulmonary embolism may result in hypotension causing a low flow state.[10] Low flow states may also be caused by TAA.[10] A ruptured TAA can cause a mediastinal hematoma compressing the esophagus and reducing blood flow to the area.[10] An unruptured TAA may also compress the esophagus.[10] In both cases, the reduced blood flow to the esophagus may result in AEN.[10] In one report, a patient with multiple embolism following elective abdominal aortic aneurysm repair developed AEN.[11] In another case, pulmonary embolism and TAA caused hypoperfusion which led to AEN.[10]

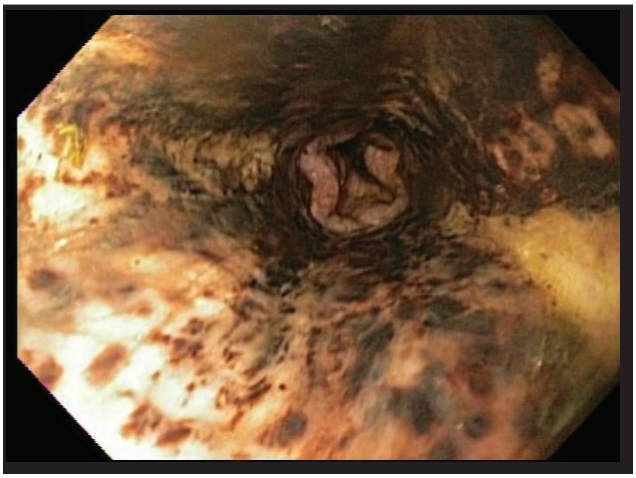

Figure 1a. Endoscopic photo showing mid to distal esophageal necrosis with large patches of black, necrotic tissue.

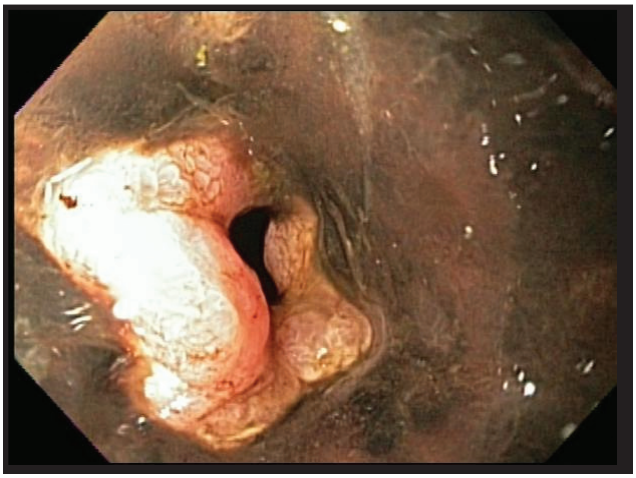

Figure 1b. Note the sharply demarcated end of the acute esophageal necrosis at the gastroesophageal junction.

Multiple instances of acute hypotension resulting in AEN have been reported in patients suffering from vascular disease.[3,12] These acute drops in blood pressure may occur during coronary artery interventions or as isolated events.[3,12] Henoch-Schönlein Purpura (HSP) has been proposed as a relevant factor of the etiology of AEN in some patients, as the inflammation and bleeding within small blood vessels may contribute to development of AEN.[13]

B. Alcohol and Cocaine Abuse

Chronic alcohol abuse can cause GI bleeding or alcohol lactic acidosis leading to a low flow state, and it can cause impairment of gastric defenses, increasing the risk of AEN.[14,15]

Cocaine may cause non-occlusive mesenteric ischemia (NOMI) which may cause lactic acidosis. Both NOMI and lactic acidosis may promote the development of AEN.[16] In patients with both alcohol and cocaine abuse, these two agents can react to form the more toxic compound, cocaethylene, potentially increasing the risk of AEN.[16]

C. Infection

Various infections, including methicillin resistant *Staphylococcus aureus* (MRSA), *Klebsiella pneumonia*, *Penicillium chrysogenum*, herpes simplex virus (HSV) cytomegalovirus (CMV), SARS-CoV-2, *Strongyloides stercoralis* and *Candida sp.* and other fungal infections have been attributed to the cause of AEN.[17,18,19] SARS-CoV-2 has been noted to cause tissue hypoperfusion, increasing the risk of AEN.[18] In one case, MRSA infection caused septicemia and led to the development of AEN.[17] Another case credited *Strongyloides stercoralis* as the cause of intestinal damage and AEN.[20]

D. Diabetic Ketoacidosis

Diabetic ketoacidosis (DKA) predisposes some patients to AEN, and may be in part due to hypovolemia.[21] Hyperglycemia secondary to DKA may decrease gastric motility and increase acid reflux.[21] A correlation between DKA and AEN has been noted in the presence of GI bleeding.[22] On a related note, Type I diabetes could be a potential risk factor for esophageal ischemia.[23] Additionally, it has been proposed that hyperglycemic states, such as DKA, could be complicated by AEN.[24]

E. Renal Transplantation

Patients receiving renal transplantation are, by definition, taking immunosuppressive medications.[25] This immunosuppressed state makes them more susceptible to infections known to cause AEN, such as CMV and *Candida* species.[25] Development of AEN has also been associated with graft versus host disease in kidney transplant patients.[25] AEN has also been associated with renal transplant surgeries in the presence or absence of acute hypotension.[26,27]

F. Medications

Various medications including sodium polystyrene sulfonate (SPS; Kayexalate), tacrolimus, and the combination of clozapine and olanzapine have been associated with development of AEN.[28,29,30] SPS is thought to cause acute inflammation of the gastrointestinal mucosa due to decreased prostaglandin levels causing vasospasm and necrosis with the possibility of AEN developing.[28] In one case report, AEN developed after a patient accidentally took clozapine and olanzapine.[29] In another case, AEN developed after a patient took tacrolimus, and rapid resolution was seen after discontinuing the medication.[30]

IV. Sequelae of AEN

Potential sequelae of AEN include esophageal perforation, esophageal stenosis/stricture formation (which can be acute or delayed), mediastinitis, and/or the formation of mediastinal abscesses.[31,32,33] Perforation of the esophagus is the most severe sequelae of AEN and is most commonly treated surgically, although endoscopic options can be considered if the injury is localized.[31] Esophageal strictures are seen in approximately 10% of cases.[31] Endoscopic balloon dilation (EBD) or esophageal stents may be used as treatments for esophageal strictures.[32] If endoscopic approaches are unsuccessful, esophagectomy or colonic interposition may, rarely, be considered.[31,34]

V. Management and Treatment Options

The gold standard for the diagnosis of AEN is upper endoscopic examination.[10] Treatment of AEN should be focused on the underlying etiologies.[1] Initially patients should be made NPO and treated with IV fluid, potentially with parenteral nutrition if felt to be warranted.[1,35] Proton pump inhibitors and sucralfate can be used for gastric acid suppression and mucosal protection, respectively.[1] Packed red blood cell infusions may be used as needed in patients with hemorrhage.[35] Antimicrobial therapy may be used if concerns about bacterial translocation or sepsis exist.[1] Surgical intervention should, in most cases, be avoided unless a severe adverse event such as perforation has occurred, and even some of these cases can be managed endoscopically.[1]

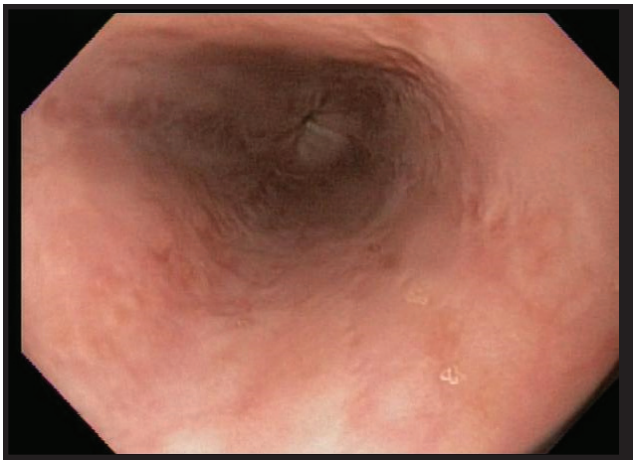

Figure 2. Findings at EGD in the same patient 6 weeks after the initial event. Note the healed esophageal mucosa with diffuse scar formation. There is no stricture formation.

VI. Outcomes

The prognosis of AEN is usually poor. Sequelae of AEN include perforation, stricture/stenosis of the esophagus, mediastinitis and/or the formation of mediastinal abscesses.[31,34] Recovery may be achieved when treatment is focused on the underlying pathologies and restoration of proper hemodynamics.[32]

Outcome of Case

The patient was treated supportively with fluids and intravenous proton pump inhibitors and parenteral nutrition and made a good recovery. An esophagogastroduodenoscopy performed 6 weeks later showed good healing of the esophageal mucosa without stricture formation but with diffuse, superficial, scar formation. (Figure 2)

VII. CONCLUSION

AEN usually has a circumferential black appearance affecting the mid to lower esophagus and sometimes the upper esophagus**.** It is important for treatment to be focused on the underlying etiologies, suppression of gastric acid, mucosal protection, and

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restoration of hemodynamics. Sequelae of AEN may require surgical or endoscopic intervention. AEN is a rare disease and patients may have a poor outcome with a high mortality rate despite aggressive treatment. ■

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