A 73-year-old African-American male was transported to the emergency department due to what emergency personnel described as “coffee ground emesis.” He was pronounced dead shortly after arrival. An unlimited autopsy examination was conducted under authorization of the coroner’s office. Medical record review revealed that the decedent had been discharged from the hospital just one day prior to his death following a three-day admission for abdominal pain, bloody diarrhea, and a 22-lb unintentional weight loss. Medical history documented hypertension, chronic obstructive lung disease, and a 57-pack-year smoking history. Alcohol abuse was also endorsed, but cessation of use was reported six months prior. During that admit, he was treated for volume-depletion, a urinary tract infection, and suspected infective colitis with antibiotics. Symptoms had resolved on hospital day three, and the patient was discharged home with a two-week course of ciprofloxacin and metronidazole and a follow-up colonoscopy appointment in one month.

At the time of autopsy, the decedent was described as cachectic. Figure 1a shows the decedent’s esophagus, opened longitudinally. Figure 1b shows the corresponding histology from the esophagus. Other findings documented at autopsy included ischemic bowel disease in the descending colon with patchy superimposed pseudomembranous colitis, emphysematous change, papillary renal cell carcinoma of the right kidney, microscopic prostatic adenocarcinoma, hepatic fibrosis, and intact hepatic hemangiomas.
DIAGNOSIS: Acute esophageal necrosis, ischemic and pseudomembranous colitis

DISCUSSION

Acute esophageal necrosis (AEN) is a rare disorder with an unclear etiology that is also known as “black esophagus.”1,2 A relatively newly described diagnosis, AEN was first reported by Goldenberg et al. in 1990.1,3 Affected patients most often present with upper gastrointestinal bleeding, including hematemesis. Other symptoms may include nausea and vomiting, dysphagia, fever, lightheadedness, and syncope. AEN occurs more often in patients with multiple co-morbidities, particularly those with diabetes mellitus, malignancies, hypertension, alcohol abuse, coronary artery disease, chronic obstructive pulmonary disease (COPD), renal insufficiency, immunosuppression, sepsis, broad spectrum antibiotic use, and aortic dissection.1,2,4,5

AEN is a rare disorder. Two large autopsy studies have found zero cases in 1,000 consecutive autopsies and a 0.2% frequency in 3,000 total autopsies.1,6 Two retrospective endoscopic studies have estimated the incidence at 0.01% and 0.28%.1,8,10 Men are affected four times more commonly than women. The disease has been documented in all age groups, but the peak incidence occurs in the sixth decade with an average age of 67 years.1,4

The gross pathology of AEN is pathognomonic characterized by circumferential black discoloration of the esophageal mucosa, primarily in the distal segment (97%) but occasionally with proximal extension to involve the upper one-third.1 Notable is the sharp contrast and abrupt transition with normal appearing gastric epithelium at the gastroesophageal junction.1,4 Gross features are so remarkable that diagnosis is frequently made by esophagogastroduodenoscopy (EGD) without the requisite need for biopsy.1,2 Histology, however, is also typical and includes necrosis of the mucosa and submucosa without any remaining recognizable viable squamous epithelium. Necrosis into the muscularis layers and full thickness into the adventitia have also been described. Additional findings include a heavy leukocytic infiltrate, deranged muscle fibers, and vascular thrombi.1,2,4,5 Altenberger et al. also describe a black nonspecific granular pigment that stains positively with periodic acid Schiff (PAS) and negatively for iron.5

The etiology of AEN is unclear but is thought to be multi-factorial and result from a combination of ischemia, backflow reflux of acidic gastric contents, and impaired local defense barriers.1,2,4 Several studies have supported a role for ischemia in the pathogenesis of AEN, particularly since the distal esophageal segment is relatively hypovascular compared to the more proximal segments.1,2,4,5 The histo-
logical appearance of AEN also bears similarities to that which is seen in ischemic colitis, offering added support for the hypothesis that the two diseases may share common mechanistic etiologies.2

Other diagnoses that need to be entertained upon identifying a black esophagus include infectious agents and toxic ingestions. Infectious esophagitis is more likely when microscopy reveals inclusion bodies, microorganism colonies, and multinucleated giant cells. Caustic ingestions do not display the typical gross pattern of AEN such that there is distal segment involvement with relative sparing of the proximal esophagus and sharp delineation from the gastric mucosa.1,2,4

The mortality rate for AEN is reportedly as high as 32% and is thought primarily to be due to patients’ existing co-morbidities.1,5 Uncomplicated AEN in a patient without co-morbidities follows a far more indolent clinical course and is potentially reversible with a far lower mortality rate at approximately 6%.1 The goal for treatment in AEN mainly targets treating the patient’s underlying conditions but also includes parenteral nutrition, proton pump inhibitors, H2 blockers, and sucralfate. Healing and resolution of the esophageal mucosa have been observed over a varied time course, ranging from one week to one month following diagnosis. Perforation of the esophagus is the most serious primary complication in AEN, reported to occur in 7% of cases; but stricture and stenosis are also described in another 10% of cases.1,4

In conclusion, acute esophageal necrosis or AEN is a rare condition with a high mortality rate that occurs most often in patients with multiple co-morbidities. The characteristic black stained esophageal mucosa, circumferentially favoring the distal segment, is easily seen on endoscopic visualization. The discoloration abruptly terminates at the gastroesophageal junction but may extend proximally into the mid and even proximal one-third. Histology of the involved segment shows no viable squamous mucosa, prominent necrosis, or overlying black granular pigment and small caliber thromboemboli. The etiology is unclear but is hypothesized to be a combination of ischemia, reflux of gastric acids, and impaired barrier defenses. The high mortality rate and poor prognosis are due in large part to the patient’s co-morbidities, and treatment is aimed at treating the coexistent conditions and maintaining hemodynamic stability in the patient.

The case illustrated here demonstrates the classic clinical presentation and typical gross and microscopic pathology of AEN. Even though retrospective historical review offers the possibility that the decedent’s complaints upon his prior admission may have suggested AEN, a remarkably high index of suspicion, and EGD would have been required in order to have diagnosed this exceedingly rare and underappreciated pathologic entity. Still further, this patient’s multiple significant comorbidities, which included two primary incidental carcinomas, chronic lung disease and prior alcohol abuse as well as the more recent evolution of an ischemic bowel and overlying pseudomembranous colitis clearly had a substantial contribution to his fatal outcome.

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REFERENCES


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