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CASE SERIES

Acute esophageal necrosis: a case series of five patients presenting with "Black esophagus"

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Abstract Acute esophageal necrosis (AEN), also known as "Black esophagus", is a rare condition that typically presents as upper gastrointestinal hemorrhage. A retrospective chart analysis was conducted at two tertiary care hospitals over a three-year period (2005–2007) using a computerized inpatient database. Out of 9,179 upper endoscopies performed, five patients (0.05% prevalence) were found to have black esophagus. Their mean age was 44 years and the most common presentation was upper gastrointestinal bleeding. All five patients had comorbid conditions, most commonly coronary artery disease, diabetes mellitus, and renal insufficiency. Two patients died, but the cause of death was not related to AEN in either. In conclusion, AEN is usually seen in critically ill elderly patients with multiple comorbid conditions, particularly vascular disease, diabetes mellitus and azotemia.

Keywords Acute esophageal necrosis · Black esophagus · Esophagitis · Necrotic esophagus · Upper gastrointestinal hemorrhage

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Introduction

Acute esophageal necrosis (AEN) is a rare entity described as a dark lesion distributed in a circumferential manner in the distal one-third of the esophagus with or without exudates [1]. Histologically, necrosis involves the mucosal and submucosal layers of the esophagus [1] and it usually occurs in the absence of caustic or other injurious agents. The exact prevalence is unknown, but in one prospective study [2] it was estimated to be 0.2%. Also known as "Black esophagus", the first endoscopic description was reported in 1990 by Goldenberg et al., although it had been reported earlier in postmortem reports [3]. Eighty-eight patients with AEN have been reported in literature between 1965 and 2006 [4].

AEN is becoming an increasingly recognized entity in gastroenterology, probably due to increased utilization of endoscopy. The pathogenesis of AEN is not fully understood, but tissue injury secondary to a hypoperfusion state or vascular thrombosis plays a dominant role. Clinically, patients may present with symptoms of upper gastrointestinal bleeding, and conservative treatment is usually an acceptable modality [4]. Complications include perforation, mediastinitis, and stricture formation. Mortality in such patients frequently depends on the underlying condition [4].

Methods

A retrospective audit of both inpatient and outpatient endoscopies done over a three-year period (January 1, 2005–December 31, 2007) at two facilities (Truman Medical Center and Saint Luke's Hospital), affiliated with the University of Missouri Kansas City, was done. These hospitals are tertiary referral centers and based in an urban

setting with a high procedure burden of more than 3,000 procedures at each hospital per year.

Results

A total of 9,179 upper endoscopies were performed in three years; five patients (0.05% prevalence) were found to have black esophagus on upper endoscopy. The clinical presentation, comorbid medical conditions, laboratory findings, and the presumed cause of AEN for all these patients are outlined in Table 1.

Case 1

A friend brought a 25-year-old Caucasian female to a hospital because of abnormal behaviour. The patient was lethargic and responded only to painful stimuli. Initial laboratory evaluation revealed a white count of 50,000 per cubic millimeter (91% granulocytes). Blood cultures grew *Candida glabrata*. Chest radiograph and computed tomography (CT) scan of the chest showed subcutaneous emphysema in the neck and chest regions. Gastroscopy showed extensive circumferential esophageal necrosis from 20 cm to 36 cm from the mouth extending to the esophageal junction. Biopsies revealed necrotizing esophagitis, and the giemsa stain was negative for fungi. Immuno-peroxidase stain for cytomegalovirus and herpes simplex virus were negative. After 2 weeks of supportive management, a repeat gastroscopy revealed esophageal bruising and a single nonbleeding ulcer in the distal esophagus.

Case 2

A 49-year-old gentleman was admitted with dysphagia and recurrent episodes of emesis. Previous gastroscopy showed an esophageal stricture, which was dilated. As a result of repeated vomiting, he developed hematemesis and underwent gastroscopy, which showed that the distal esophageal mucosa was markedly friable and necrotic, and bleeding in most areas. The gastroesophageal junction was obstructed and could not be negotiated with the endoscope; therefore dilation was not performed. He was kept nil per oral and on supportive management. Repeat gastroscopy 2 weeks later, showed grade 2 esophagitis, edematous mucosa, dilated esophagus with pooling of secretions and a "pop" was heard on passage through the lower esophageal sphincter. Botulinum toxin was administered in the lower esophageal sphincter resulting in resolution of patient's symptoms. He was discharged in stable condition. There was no identifiable source of infection in this patient.

Case 3

A 50-year-old gentleman was admitted with worsening renal function. The patient had a renal transplant approx-

Table 1 Clinical features and laboratory investigations in patients with black esophagus

No	Age (y)	Sex	Presentation	Comorbid conditions	Laboratory results				
					Hemoglobin (g/dL)	Total leukocyte count ($X10^9$/L)	Platelet count ($X10^9$/L)	INR	Creatinine (mg/dL)
1	25	F	Altered mental status, pneumo-mediastinum, acute renal failure	DKA, seizure disorder, amphetamine abuse, Candidemia	14.5	50	420	1.25	2.5
2	49	M	Dysphagia, hematemesis, acute renal failure, severe GERD	Achalasia, CHF, ejection fraction 10%, anemia, heavy alcohol, tobacco, cocaine use	8.4	7.5	297	1.70	1.8
3	50	M	Epigastric pain, nausea	Renal transplant in rejection, DM, PVD, HD, ischemia, immunodeficiency	12.3	7.5	84	1.3	3.3
4	60	F	Syncope, hypotension, nausea, vomiting, GI bleed	Resected pancreatic cancer, nephrectomy for renal cancer, resected hepatic adenoma, DM, HTN, CHF, ESRD on HD	11.4	20.5	777	1.2	4.8
5	36	M	Nausea, vomiting, GI bleed	DKA, GERD, heavy smoking	7.5	14.2	211	NA	1.0

CHF congestive heart failure; *DM* diabetes mellitus; *DKA* diabetic ketoacidosis; *EF* ejection fraction; *ESRD* end stage renal disease; *GERD* gastroesophageal reflux disease; *HD* hemodialysis; *HTN* hypertension; *PVD* peripheral vascular disease

imately 3 months earlier and had a baseline creatinine of 1.4 mg/dL. On admission, his creatinine was 1.8 mg/dL and increased to 3.6 mg/dL during admission. The patient was diagnosed as acute graft versus host rejection and started on thymocyte protocol. After 3 days of hospitalization, he had epigastric pain, nausea and vomiting. Gastroscopy revealed dusky appearing distal 10 cm of esophagus, but biopsies were not done due to severe thrombocytopenia. The patient was continued on proton pump inhibitors (PPI) and was started on parenteral nutrition. Repeat gastroscopy 3 days later, showed resolution of the black esophagus. The patient was discharged but presented a few days later with hypoxic encephalopathy and died.

Case 4

A 60-year-old Caucasian female was admitted to the intensive care unit with syncope and hypotension. She had a prior diagnosis of pancreatic adenocarcinoma and underwent total pancreatectomy, splenectomy, antrectomy, and a Billroth II anastomosis. Non-invasive imaging studies, performed for persistent nausea and vomiting, were unremarkable. Gastroscopy showed diffuse blackened esophageal mucosa and severe, friable esophagitis. No biopsies were taken due to high risk of bleeding from the severe esophagitis. The patient was provided aggressive management, but her hospital course was complicated by gram-negative septicemia, non-ST elevation myocardial infarction with supraventricular tachycardia, and refractory *Clostridium difficile* diarrhea. The patient eventually succumbed to these complications.

Case 5

A 36-year-old Caucasian male with long standing history of type 1 diabetes was admitted with ketoacidosis, secondary to inadequate insulin intake. The patient had a history of reflux disease controlled with ranitidine; he was smoking one pack of cigarettes per day, and used cocaine for recreational purposes along with alcohol. He was admitted with persistent nausea and vomiting, and gastroscopy was done as the patient also reported coffee-ground blood in his emesis. The endoscopy revealed necrotic esophagitis in the middle and distal parts of the esophagus. The patient was kept nil per oral and started on pantoprazole twice daily. CT scan of the chest next day showed no perforation, and the patient's diet was cautiously advanced. He tolerated his diet well and was discharged. There was no source of infection identified in this patient. He followed up with his primary care physician 2 months later and was asymptomatic.

Table 1 shows the clinical data in these patients. The mean age of the patients was 44 years. The most common symptoms were upper GI bleeding with nausea and vomiting. Comorbid conditions included coronary artery disease, diabetes mellitus, and gastroesophageal reflux disease.

All the five patients were critically ill with hypotension, requiring vasopressor support and close monitoring in the critical care unit. Two patients had reflux esophagitis seen on endoscopy. Except for the first patient (who had candidemia), no infection was identified.

Endoscopic course and treatment of the five patients is noted in Table 2. All patients had black esophagus on endoscopic exam (Figs. 1a, 2a) that eventually resolved on

Table 2 Endoscopic course and treatment of the five patients

No.	Gastroscopy finding	Repeat gastroscopy (day)	Treatment	Outcome
1	Blackened esophagus, necrotic appearing pale esophageal mucosa, and normal GE junction	Ulcer in distal esophagus. Healed esophagus without blackened mucosa (day 14)	Nasogastric tube, PPI IV daily, broad-spectrum antibiotics, antifungal agent	Survived
2	Necrotic appearing friable areas of spontaneous bleeding	Grade 2 esophagitis, dilated esophagus, pooling of secretions, "pop" on passage through the LES (day 15)	NPO, PPI IV twice daily, 100 units of botulinim toxin injected at LES.	Survived
3	Dusky appearance in distal 10 cm of esophagus with areas of superficial ulceration suggestive of ischemia	Diffuse whitish appearing exudate with areas of dusky mucosa, at the distal 18 cm of esophagus, markedly improved from initial EGD (day 3)	NPO, PPI IV twice daily, total parenteral nutrition	Died
4	Diffuse blackened esophageal mucosa, friable mucosa	Not attempted	PPI IV twice daily, broad spectrum antibiotics	Died
5	Middle to distal esophagus appeared necrotic	Not done	NPO, PPI IV twice daily	Survived

GMS giemsa silver stain; *IV* intravenous; *LES* lower esophageal sphincter; *NPO* nil per oral; *PPI* proton pump inhibitor

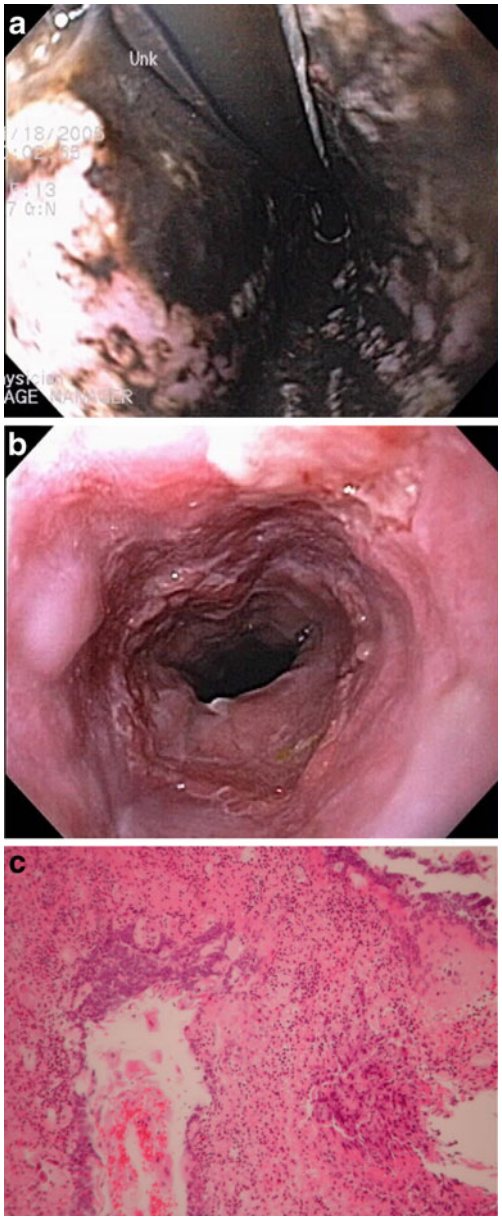

Fig. 1 Twenty-five-year-old female had an upper endoscopy showing black esophagus (**a**) followed by a repeat endoscopy 14 days later showing resolution of the lesions (**b**). Esophageal biopsy (**c**) with hematoxylin and eosin staining showing necrotic debris, absence of epithelium, granulation tissue, and heavy leukocytic infiltrates

repeat endoscopy in three patients (Figs. 1b, 2b). Two patients did not have a repeat endoscopy. All the patients required intensive care admission and monitoring, and were treated with aggressive fluid resuscitation and intravenous PPI. Four out of five patients had oral nutritional rest, whereas one patient required parenteral nutrition. Empirical supportive therapy, including oral nutritional rest, and PPI in the intensive care unit (five patients) and broad-spectrum antibiotics (two patients) was provided. One patient was lost to follow up. Of the remaining four patients, two died.

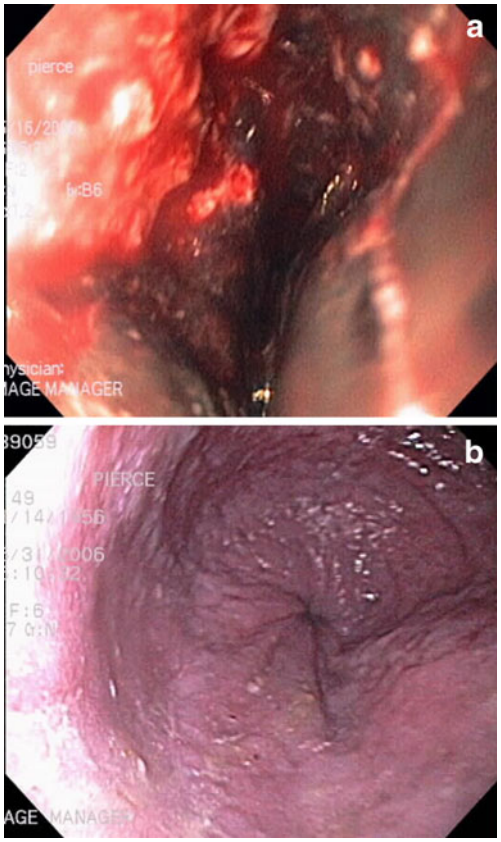

Fig. 2 Forty-nine-year-old male had an upper endoscopy showing black esophagus (**a**) followed by a repeat endoscopy 15 days later showing resolution of the lesions (**b**)

The cause of death of the two patients was not related to AEN; one patient died of anoxic encephalopathy secondary to cardiorespiratory arrest and the other died due to acute myocardial infarction.

Discussion

Moreta et al. [5] described the following criteria for the diagnosis of acute esophageal necrosis: circumferential black esophagus with or without exudates; distal esophageal involvement, ending sharply at the gastroesophageal junction; and absence of caustic ingestion. Biopsy material may be obtained for definitive histologic confirmation, but is not required. Histologic findings include the presence of necrotic debris on esophageal biopsy, necrotic mucosa without viable epithelium, and frequent submucosal involvement, occasionally extending into the muscularis propria, potentially leading to a full-thickness lysis [4]. The first patient had classic histologic changes on biopsy (Fig. 1c) suggestive of AEN. Biopsy could not be obtained in the other patients either due to risk of bleeding or thrombocytopenia.

Although the pathogenesis of AEN appears to be multifactorial, the precise cause is still unknown. Ischemia is the etiological mechanism most frequently referred to [3, 6], but the condition has been associated with a wide variety of conditions and risk factors, mainly advanced age, malnutrition, vascular disease, diabetes, infections especially in immunocompromised population, medications, alcohol intoxication, trauma, burns, and shock. A low-flow state with mucosal injury and underlying impaired defense mechanism at the cellular level might provide the ideal environment for the development of AEN. The blood supply to the proximal esophagus is derived from the superior and inferior thyroid arteries, while the rest of the esophagus receives its blood supply from the esophageal branches of the descending aorta and splenic and left gastric arteries more distally [7]. The latter is usually less vascularized and thus is more prone to ischemic injury [4]. This condition is generally seen in the elderly and those having comorbid conditions; two of our patients were 60 years or older.

The diagnosis of AEN is based on direct visualization by endoscopic exam. The role of other modalities for diagnosis of this condition, such as arterial Doppler study has not been studied. However, it is likely that AEN is more due to generalized low-flow state rather than one specific vessel as a cause of the condition.

Treatment is mainly supportive and consists of maintaining hemodynamic stability through adequate volemic resuscitation and minimizing acid exposure with intravenous proton pump inhibitors [1]. Bowel rest is advised, and consideration can be made for short-term parenteral alimentation. Nasogastric decompression may be required where persistent vomiting, gastric outlet obstruction, and bleeding are present [8]. Antimicrobials are indicated in select patients with signs of sepsis or immunocompromised states [8]. Endoscopy is diagnostic, and timing of follow up endoscopies needs to be dictated by patient's clinical status [9].

Early and late complications of AEN include esophageal stenosis (10.8%), esophageal perforation (6.8%), and mediastinitis and abscess formation (5.7%). Surgical intervention is required in cases where there is massive necrosis with esophageal perforation, and herniated gastric volvulus. In cases of mediastinal collection or abscess, prompt esophagectomy with surgical drainage and antibiotic therapy is needed [4]. One patient in our series had pneumomediastinum on presentation. There has only been once previously described case in the literature with such a presentation [10].

Natural history of this disease is a spontaneous resolution [6]. The overall mortality reported in the literature is 31.8%, and is probably related to underlying conditions, but death secondary to esophageal necrosis occurs in fewer than 6% of the cases [4]. The high mortality reported is due to the comorbid illnesses rather than to AEN.

In conclusion, black esophagus is an uncommon endoscopic finding, usually present with upper gastrointestinal tract bleeding. In our analysis, all patients with black esophagus had comorbid conditions, most common being coronary artery disease, diabetes mellitus, and renal insufficiency. The treatment of this condition should focus on supportive therapy. Compared with other causes of upper gastrointestinal tract bleeding, the mortality rate among patients with this condition is high.

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