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Case Report

Acute esophageal necrosis (black esophagus) associated with duodenal ulcer bleeding: a case report

Joon Hyun Cho*, Tae Nyeun Kim*

*Division of Gastroenterology and Hepatology, Department of Internal Medicine, Yeungnam University College of Medicine, Daegu, South Korea. *Equal contributors.*

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Abstract: Black esophagus is a rare multifactorial clinical condition with an impressive endoscopic appearance of black necrotic-appearing mucosa extending proximally in the esophagus and abruptly terminating at the gastroesophageal junction. The etiology and pathogenesis of black esophagus are unclear, but ischemia, impaired local defense barriers, and altered gastric motility with acid reflux may be inciting factors. Although black esophagus can develop from a duodenal ulcer or duodenal ulcer bleeding, this has been reported to be extremely rarely to date. Here, we present the case of an 88-year-old female who developed black esophagus associated with duodenal ulcer bleeding. The patient was referred to us for gastrointestinal bleeding, and the diagnosis was confirmed by esophagogastroduodenoscopy. She became stable and duodenal ulcer bleeding ceased after three therapeutic endoscopies with hemoclipping, high dose intravenous injections of proton-pump inhibitors, and supportive care by total parenteral nutrition and nil-per-os. Concomitant "non-bleeding" black esophagus was also improved by esophagogastroduodenoscopy.

Keywords: Black esophagus, acute esophageal necrosis, esophagus, duodenal ulcer, bleeding

Introduction

Acute esophageal necrosis (AEN), commonly referred to black esophagus or necrotizing esophagitis, is a rare disease characterized by typical endoscopic findings of diffuse involvement of the distal esophagus with various proximal extensions of black necrotic appearing mucosa that are abruptly interrupted at the gastroesophageal junction [1, 2]. A retrospective endoscopy series estimated that the prevalence of AEN ranges from 0.01 to 0.2% [3]. The etiology of AEN remains unclear, but is assumed to be multifactorial and to pivotally involve ischemia and impaired mucosal defense barriers resulting in topical injury ultimately leading to necrosis. The most frequent clinical presentation (90%) is acute upper gastrointestinal bleeding (UGIB), which is often preceded by hemodynamic instability [4]. Despite supportive care and treatment of underlying disease, overall mortality of AEN still nears 35% [5]. AEN may arise in the setting of cardiovascular compromise, shock, diabetic ketoacidosis, vascular disease, alcohol abuse, thromboembolic phenomena, malignancy, duodenal ulcer disease, gastric outlet obstruction, hiatal hernia, or malnutrition [6, 7], but although a number of case reports have been issued on AEN, only a few reports of AEN due to duodenal ulcer or duodenal ulcer bleeding have been described [8, 9]. Here, a rare case of AEN (black esophagus) associated with duodenal ulcer bleeding is reported and likely pathophysiology of the condition is discussed.

Case presentation

An 88-year-old woman was referred to our institution with acute onset melena during hospitalization of duration one week at another hospital for rib fracture. At the time of referral, she was also suffering from nausea and vomiting of 4 to 5 days duration and from poor oral intake of 2 weeks duration. Prior to transfer to our hospital, she had received intermittent analgesic injections and nonsteroidal anti-inflammatory drugs. In addition, she was taking medications regularly for type 2 diabetes mellitus and hyperten-

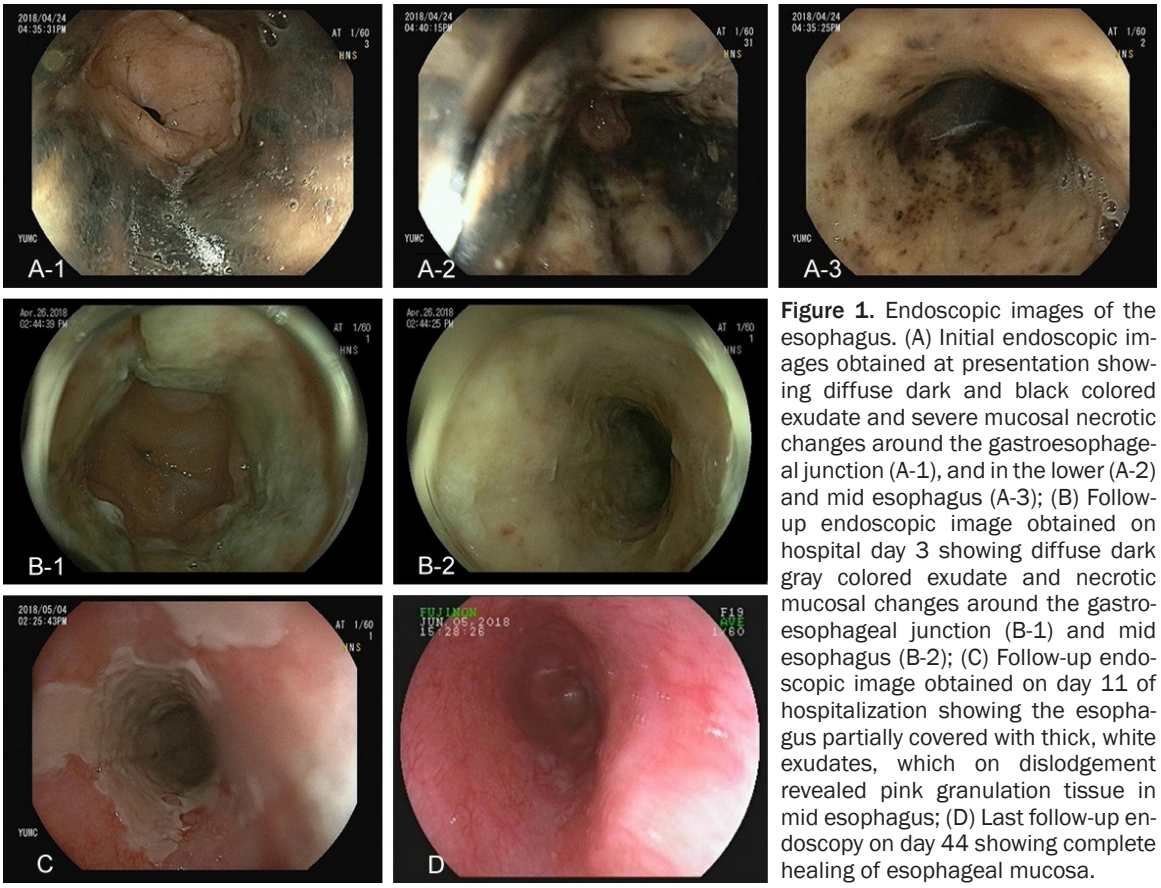

Figure 1. Endoscopic images of the esophagus. (A) Initial endoscopic images obtained at presentation showing diffuse dark and black colored exudate and severe mucosal necrotic changes around the gastroesophageal junction (A-1), and in the lower (A-2) and mid esophagus (A-3); (B) Follow-up endoscopic image obtained on hospital day 3 showing diffuse dark gray colored exudate and necrotic mucosal changes around the gastroesophageal junction (B-1) and mid esophagus (B-2); (C) Follow-up endoscopic image obtained on day 11 of hospitalization showing the esophagus partially covered with thick, white exudates, which on dislodgement revealed pink granulation tissue in mid esophagus; (D) Last follow-up endoscopy on day 44 showing complete healing of esophageal mucosa.

sion, which included glimepiride, telmisartan, and amlodipine. The patient had no history of prior GI bleeding.

On arrival at our emergency room, the patient appeared a little sick but relatively stable, excepting mild hypotension. Initial vital signs were temperature 37.1°C, blood pressure 90/60 mmHg, heart rate 84 beats/min, respiratory rate 16/min, and oxygen saturation 97% in room air. A laboratory study revealed an acute decline in hemoglobin to 4.3 g/dL from 11.3 g/dL (determined by routine testing two days earlier at the previous hospital), acute renal impairment (creatinine 2.9 mg/dl, ureic nitrogen 120 mg/dl), hyperkalemia (serum potassium 6.1 mmol/L), hypoalbuminemia (serum albumin 2.55 g/dL), and neutrophilic leukocytosis (WBC 27.4 k/mL, neutrophils 79.7%). Other laboratory findings included platelets 283 k/mL and INR (international normalized ratio) 1.4. After transfer, the patient was immediately administered intravenous fluids and packed RBCs without vasopressors. Emergency esophagogastroduodenoscopy (EGD) revealed diffuse black and dark exudate, severe mucosal changes characterized by necrosis at the middle and lower thirds of the esophagus (**Figure 1A**), which was consistent with AEN, and multiple huge ulcers with adherent clots at the duodenal bulb (**Figure 2A**). The patient was placed on nil-per-os (NPO), total parenteral nutrition (TPN), intravenous high dose proton-pump inhibitors (PPIs), and intravenous antibiotics.

On the 3rd day of hospitalization, despite vigorous intravenous fluid resuscitation and packed RBCs transfusion, anuria developed and azotemia worsened, and emergency hemodialysis was initiated via a central venous cannula. Follow-up EGD showed a diffuse dark gray exudate and necrotic mucosal change at the middle and lower thirds of the esophagus (**Figure 1B**). Endoscopic biopsies at the esophagus were deferred due to concerns about bleeding. EGD also showed an exposed vessel with active oozing on the huge ulcer at the duodenal bulb, which was successfully prevented using two pieces of hemoclip (**Figure 2B**). On day 4, repeat endoscopy showed no bleeding from the

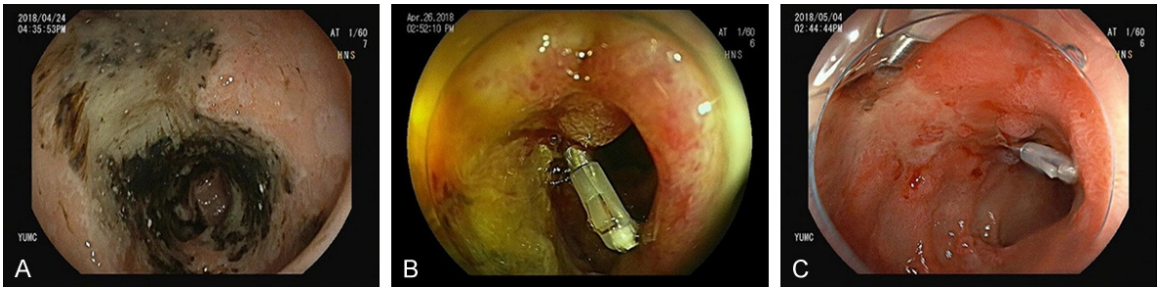

Figure 2. Endoscopic images of duodenum. A. Initial endoscopic image obtained at presentation showing multiple huge ulcers and adherent clots at the duodenal bulb; B. Follow-up endoscopic image obtained on day 3 of hospitalization showing an exposed vessel with active oozing on the huge ulcer at the duodenal bulb that was treated successfully by endoscopic hemoclipping; C. Follow-up endoscopic image obtained on day 11 after hemoclipping of the oozing site at the duodenal bulb.

multiple duodenal ulcer including the hemoclipping site and no specific interval change of AEN. Follow-up laboratory testing revealed thrombocytopenia (platelets 48 × 10^3/uL), and thus, an esophageal biopsy (due to bleeding concerns) was performed in order to exclude infectious or other diseases. However, the biopsy specimen obtained at the esophageal lesion indicated only inflammatory exudates. The rapid urease test was negative. Thereafter, the patient presented with intermittent, slight melena, and was supportively managed by TPN, packed RBC and platelet transfusions, and ongoing high dose intravenous PPI.

On day 9, the patient presented a large amount of melena and hematochezia. Emergent EGD showed an exposed vessel with active oozing on the duodenal ulcer, which was treated endoscopically with two pieces of hemoclip. At this time, esophageal lesions exhibited no specific interval change. On day 11, she presented ongoing intermittent slight melena and follow-up EGD with hemoclipping on the oozing site at the duodenal bulb was performed (**Figure 2C**). On this occasion, EGD showed an improvement in the appearance of the esophagus, which was partially covered by thick, white exudates that on removal revealed pink granulation tissue (**Figures 1C**). Thereafter, GI bleeding ceased and the patient remained stable. In addition, urine volume returned to near normal and serum creatinine decreased. After consulting a nephrologist, tapering the number of hemodialysis sessions was planned.

On day 21, follow-up EGD showed a remarkable improvement in esophagus appearance and the esophagus was only partially covered with white exudates. Oral feeding was resumed, and subsequently, the patient continued to improve. She was discharged from the hospital on day 30. Last follow-up EGD was performed at an outpatient department 44 days after transfer, and showed complete healing of the necrotic-appearing esophageal mucosal changes (**Figure 1D**).

Discussion

AEN, also known as “black esophagus”, is a rare clinical disorder first defined by Goldenberg et al. in 1990 [10], and later further characterized by Gurvits [4]. Although its incidence has been estimated to lie between 0.01% and 0.2% [3], it may be underestimated due to sometimes subtle clinical presentation of the disease and the spontaneous healing capacity of esophageal mucosa. Actually, AEN has received increased recognition over the last decade presumably due to more frequent endoscopic evaluations of patients presenting with UGIB. The most common clinical manifestation of AEN is UGIB with hematemesis or melena. Our patient visited with duodenal ulcer bleeding, and “non-bleeding and asymptomatic” black esophagus was incidentally diagnosed during therapeutic EGD, although the pathologic examination conducted failed to detect its presence.

Typical endoscopic findings of AEN are the presence of ulcero-necrotic circumferential black lesions of esophageal mucosa that show variable proximal extension to but abruptly stop at the gastroesophageal junction. AEN tends to occur in the distal third of the esophagus (97%) due to its relative hypovascularity as compared with the proximal esophagus [1].

The etiology of AEN is unclear and probably multifactorial. Several factors may act in concert to determine its clinical presentation. The important inciting events are tissue hypoperfusion (e.g., cardiovascular compromise or shock), impaired local defense barriers due to a debilitated condition (e.g., cancer or malnutrition), and altered gastric motility with acid reflux due to, for example, alcohol intoxication, or duodenal ulcer disease that overwhelm vulnerable esophageal mucosa [1, 4]. In other words, this syndrome may be an "ischemic condition" of the esophageal wall, wherein reduced blood flow is sufficient to reduce the ability of mucosal defense to withstand gastric acidity. Our patient had been suffering from nausea and vomiting for 4 to 5 days and poor oral intake for 2 weeks prior to presentation, which were suspected to have been caused by duodenal ulcer. The malnutrition thus derived might have impaired esophageal mucosal defenses and the reflux of gastric contents might have further damaged already vulnerable esophageal mucosa. In this harmful situation especially to the esophagus, hypovolemia due to duodenal ulcer bleeding and acute renal failure might have provoked tissue hypo-perfusion in esophageal mucosa and triggered AEN. The present case was incidentally diagnosed AEN related to duodenal ulcer bleeding. In absence of significant hemodynamic impairment or shock, this case is important to be reported.

Laboratory tests may show anemia and leukocytosis. Endoscopic findings are diagnostic and tissue histology is not required but may be supportive. As necrosis resolves, the esophagus may become partially covered with thick, white exudates, which probably represent mucosal sloughing, that can be easily dislodged to reveal pink granulation tissue [4]. The differential diagnosis of AEN includes malignant melanoma, acanthosis nigricans, corrosive agent ingestion, and infectious diseases [4].

Therapeutic modalities have not been standardized and the development of AEN generally carries a poor prognosis. The goal of therapy should be directed toward restoring hemodynamic stability and correcting underlying conditions, and includes fluid resuscitation, blood transfusion, NPO, TPN, and high doses of intravenous PPIs. Decision regarding antimicrobial, antiviral, and antifungal use should be made on an individual basis, especially in critically ill or septic patients, as antibiotic use per se has been reported to cause AEN [4, 11].

Prognosis is dependent on associated conditions, and mortality varies from 15 to 36% [7]. AEN associated mortality directly related to complications of esophageal necrosis, such as, perforation, hemorrhage, and sepsis, is low at around 6% [12]. The most common long-term complication is stenosis or stricture formation, which may occur in up to 10 to 25% of cases as early as 2 weeks after diagnosis [4, 12], and requires endoscopic or surgical treatment.

Conclusions

Although a number of case reports have been issued on AEN, little is known about the etiology and pathogenesis of acute esophageal necrosis. In some reports it has been suggested AEN might develop from duodenal ulcer or duodenal ulcer bleeding, however, after extensive literature search, only two reports were found on AEN due to duodenal ulcer in the English literature [8, 9]. One case of non-bleeding black esophagus was incidentally diagnosed and associated with duodenal ulcer bleeding. The case report described here supports the notion that, owing to several coordinated mechanisms and complicated causes, duodenal ulcer and hypovolemia resulting from ulcer bleeding may be associated with the development of AEN. Furthermore, AEN may present without esophageal bleeding and develop in the absence of serious comorbidity, significant hemodynamic impairment, or shock.

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Disclosure of conflict of interest

None.

Address correspondence to: Dr. Tae Nyeun Kim, Division of Gastroenterology and Hepatology, Department of Internal Medicine, Yeungnam University College of Medicine, 170 Hyeonchung-ro, Nam-gu, Daegu 42415, South Korea. Tel: +82-53-620-3842; Fax: +82-53-654-8386; E-mail: tnkim@yu.ac.kr

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