

Black Esophagus (Acute Esophageal Necrosis) Caused by Liver Cirrhosis

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ABSTRACT

Acute necrotizing esophagitis (black esophagus) is a rare clinical entity that is characterized by partial or total loss of the epithelium, ulceration up to the circumferential slough of all layers of the mucosa and submucosal, frequent involvement of deep muscle layers, and frequent perforations.

Its blackish appearance in endoscopic examinations has given it the name of black esophagus. We describe a rare case of black esophagus caused by hepatic cirrhosis associated with gastric ulcer.

Keywords

Necrotizing esophagitis, Black esophagus, Acute esophageal necrosis.

Introduction

Acute esophageal necrosis (AEN) is also known as black esophagus or Gurgits Syndrome. It was first described in the medical literature in 1990 by Goldenberg and colleagues but became a distinct clinical syndrome in 2007 by Gurgits [1-3]. The incidence of the disorder is low, and the diagnosis requires a high degree of clinical suspicion and visual inspection of esophagogastroduodenoscopy.

It is virtually always associated with comorbidities including diabetes mellitus, malignancy, thromboembolic disorders, alcohol abuse, renal insufficiency, and cardiovascular disease [2]. In fact, the degree of collective concurrent illness will, in large part, determine the prognosis. The mortality rate has been reported to be as high as 32%-36%, which reflects the underlying illnesses [3,4].

Treatment includes active management of the comorbidities, intravenous administration of a proton pump inhibitor, and possibly short-term parenteral nutrition. The main aim of treatment is to

avoid extension of the injury and allow spontaneous healing. We present an unusual case of a young diabetic male with documented black esophagus.

Case Report

A 41-year-old man visited the emergency room with a 12 hours history of epigastric pain, violent blood vomiting and hematemesis. His past medical history included alcoholic liver injury with diagnosis of cirrhosis.

The previous day, he had consumed distilled drinks containing 40% alcohol. He denied the ingestion of any caustic agents, antibiotics, and Non-steroidal anti-inflammatory drugs (NSAIDs). He had a regular pulse of 120 bpm, a blood pressure of 84/56 mmHg, a respiratory rate of 19 breaths/min and a core temperature of 36,7°C.

Physical examination revealed a mild distended abdomen (ascitis) and epigastric tenderness. The liver edge was not measured below the right costal margin. Arterial blood gas measurements using with O₂ flow at 3 L/min was: pH 7-12, PaCO₂ 26.4 mmHg, PaO₂ 112.4 mmHg, AG 55.7 mmol/L, HCO₃⁻ 19.9 mmol/L, lactate 56.4

mmol/L. Laboratory data were: WBC 20.630/mm³, hemoglobin 89 g/L, platelets 134 K/mm³, TP 385 g/L (normal 67-83 g/L), SGOT 335 U/L (8-38 U/L), SGPT 136 U/L (4-44 U/L), BUN 320 mg/L (80-200 mg/L), creatinine 11 mg/L (6-10 mg/L), Na 139 mmol/L (138-149 mmol/L), K 5.1 mmol/L (3.5- 5.0 mmol/L), Cl 91 mmol/L (99-110 mmol/L), amylase 48 U/L (43-116 U/L).

Abdominal US demonstrated the presence of signs of portal hypertension with no flow in the portal vein with high influx and presence of esophageal collateral circulation, besides signs of liver cirrhosis.

The patient was admitted and received intravenous fluids, and he was rapidly rehydrated and urinalysis did not reveal ketones. Arterial blood gas, 5 h after his admission, showed pH 7.505, lactate 4.1 mmol/L.

Esophago-gastro-duodenoscopy (EGD) on the same day, demonstrated black and friable mucosa from 5 cm up to the cardia. Biopsies of the esophagus were not taken at this time due to concern over possible perforation.

The stomach had a few linear ulcers and the mucosa was edematous, and the stomach had a gastric ulcer in the antrum in the posterior wall with a diameter of 2.5 cm with recent signs of bleeding and adhered clots.

Treatment was conservative management with oral intake restriction, intermittent nasogastric tube suction, total parenteral nutrition, intravenous omeprazole, antibiotics and clinical support.

Three days after admission, EGD revealed the presence of a thick, white exudate on the pink esophageal mucosa. Oral feeding started on the seventh day. The patient was discharged without complaints on the thirteenth day, taking 20 mg of omeprazole once a day, and his alcoholic hepatitis was sustained.

Discussion

Epidemiology

Literature analysis shows that the estimated prevalence of AEN is low. Two large autopsy series from the United States and France have reported zero cases in a series of 1,000 adult autopsies and 0.2% in 3,000 autopsies, respectively [5].

Two large retrospective series that have reviewed the findings in > 100,000 endoscopies have estimated the incidence at approximately 0.01% (12 patients), and another retrospective analysis of 10,295 endoscopies has shown an incidence of 0.28% (29 patients) [6].

However, these numbers are likely underestimated and the true prevalence of AEN remains unknown, largely due to the potential of subclinical presentation of the disease and the early healing of the mucosa that can be seen with transient ischemic or chemical injury. Besides, it's observed a tendency of AEN occurring more frequently in patients in critical states, such as refractory shock or systemic illness [7].

Ramos R et al. 2008 demonstrated a mortality rate of 63.6% of the patients with AEN, although most probably that rate reflects the worsening of underlying illness and thus not wholly attributable to AEN. It's also reported that 18.2% of the patients remained asymptomatic and had no residual effects [8].

An intriguing paper by Japanese authors has reported AEN as the fourth leading cause of coffee ground emesis, hematemesis, or melena in a series of 239 hospital admissions for upper gastrointestinal bleeding, which accounted for approximately 6% of the cases [9].

AEN clearly shows sex and age predilection. Men are four times more commonly affected than women, represent approximately 80% of cases [1], and although the disease has been documented in every age group, the peak incidence occurs in the sixth decade of life with an average age of 67 years [10].

Clinical presentation

General debilitation, poor clinical condition and advanced age with multiple medical comorbidities are associated with increased likelihood of development of AEN. Personal history of diabetes mellitus (24%), malignancy (20%), hypertension (20%), alcohol abuse (10%), and coronary artery disease (9%) places a patient at risk of developing AEN [6,10].

Other comorbidities associated by literature are alcoholic cirrhosis, malnutrition, diabetes mellitus, diabetic ketoacidosis, malignant neoplasia, hypotension, chronic kidney disease, cardiovascular diseases, duodenal or gastric ulcer, gastric obstruction, hypersensitivity to antibiotics, certain viral infections, alcoholic hepatitis, severe acid reflux, acute pancreatitis, severe vasculopathies, sepsis, septic shock, aortic post-traumatic injuries, aortic dissection, and thromboembolism. Diagnosis requires that other etiologies that produce similar endoscopic alterations be discarded, including melanosis, malignant melanoma, pseudomelanosis, acanthosis nigricans, adverse drug effects (quinidine and tetracycline), and infection (Candida and herpes) [11,12].

The most frequent clinical manifestations of AEN are gastrointestinal bleeding, hematemesis or melena, or nausea and vomiting, thoracic or abdominal pain, dysphagia, syncope or sudden death. It is often preceded by hemodynamic instability and symptoms of gastroesophageal reflux. Patients with black esophagus usually present initially with GI hemorrhage in 65% to 90% of cases [1,10,12,13].

This condition is more common in the elderly and the Diagnosis of AEN is essentially endoscopic, although it is occasionally made during emergency surgery or autopsies [14].

Histopathology and Etiology

The etiology is believed to be associated with hypoperfusion of the mucosa, caused by decreased blood flow of arterial branches, secondary to hypovolemia from either third spacing or fluid loss.

Another theory includes reflux of gastric contents, which can also lead to necrosis. This corroborates the associated comorbidities such as multiorgan and hemodynamic compromise, digestive bleeding and electrolyte imbalances [14,15].

Black esophagus is defined by diffuse, circumferential necrosis of the esophagus with preferential involvement of the distal third of the esophagus that abruptly stops at the gastroesophageal junction, and in the absence of caustic ingestion [16].

The predilection toward involvement of the distal esophagus is thought to be due to its relatively poor perfusion. Blood flow to the distal esophagus is highly variable, but typically occurs through the left gastric and left inferior phrenic arteries. This is believed to result in a “watershed region” that creates a susceptibility to insult [17].

The main pathogenesis in alcoholics can be accounted by low systemic perfusion caused by severe lactic acidosis [18-20]. It is caused by an elevation of the NADH/NAD⁺ ratio with increased formation of lactate from pyruvate, diminished gluconeogenesis and decreased lactate uptake into the liver [21,22]. Thiamine deficiencies due to malnutrition and the direct action of alcohol are responsible for the occurrence of lactic acidosis. Moreover, malnutrition may compromise the mucosal defense system and healing capacity after injury [8,23,24]. Five typical symptoms of alcoholic lactic acidosis are vomiting, nausea, abdominal pain, tachycardia and the Kussmaul form of respiration.

Diabetic decompensation could be an important factor, particularly ketoacidosis, suggesting a relationship between the degree of proximal damage and of hyperglycemia [9]. The fall in blood pH had profound effects on circulation systems. Intrinsic cardiac contractility may be depressed, but inotropic function can be normal because of catecholamine release. Both peripheral arterial vasodilation and central venoconstriction can be present. Several reports described cases of stroke [25], intestinal necrosis [26], heart failure, and pancreatitis [27]. In severe metabolic acidosis and those may be associated with physiologic response to hypovolemia and subsequent poor organ perfusion caused by severe acidosis. Low esophageal perfusion pressure, a presumed cause, is suggested by hypotension. In addition, coagulation abnormality by acute pancreatitis can aggravate esophageal mucosal perfusion. Severe peripheral circulatory failure also makes massive gastric distension and renal insufficiency. Moreover, alcohol overindulgence induces irritation of the gastric mucosa and the accumulation of large volume of gastric secretions. This can overwhelm the normal defense of the esophagus, and could be another cause of necrosis [18].

Histologically, there is necrosis of the mucosa and submucosal, inflammation of the muscle fibers, and occasional thrombosis of blood vessels, not only of the esophagus, but also of the stomach and duodenum. However, gross findings alone are sufficient for diagnosis, and biopsy is not mandatory [14,21].

The vascularization of esophagus can be represented by three

different vascular zones: a proximal one, depending on thyroidal inferior arteries, with support from subclavian and carotid branches; a median zone referring to bronchial and intercostal arteries of direct aortic origin; a distal zone supported by branches of gastric and phrenic arteries, with possible contribution from hepatic and splenic vessels. This last zone seems particularly vulnerable to impairment of vascular support, so justifying the almost invariable implication of distal esophagus in the cases described in the literature [12].

Treatment

The mainstay of managing black esophagus is aggressive fluid resuscitation, bowel rest, and treatment with IV proton pumps inhibitors. NG tube insertion is contraindicated due to fear of perforation. Patients with an upper GI hemorrhage often respond to resuscitation with intravenous fluids and blood products. The management with IV broad-spectrum antibiotics and surgical consultation should be considered in cases wherein patients fail to respond to appropriate resuscitation, subsequently decompensate despite resuscitation or appear septic [1,14,15].

However, antibiotics, antiviral or antifungal are not indicated unless the patient has an infection, is immunocompromised, continues to decompensate despite adequate IV fluid resuscitation, critically ill, septic patients or an esophageal perforation [16].

In practice, the differential diagnosis is made with melanosis, pseudomelanosis, malignant melanoma, acantosis nigricans, charcoal deposits, and caustic poisoning [28].

The necessity of early antibiotic therapy may be unclear in the ED due to other considerations in the differential diagnosis; therefore, it is prudent to treat the patient empirically until these etiologies can be ruled out. Some clinicians recommend sucralfate due to its ability to bind pepsin and stimulate mucus secretion, which theoretically prevents further esophageal injury [6,15].

Prognosis

It is typically a disease of the elderly male, who may have long-standing general malnutrition. Additionally, this group already is at risk for ischemic insults. If the ischemia lasts long enough the damage becomes progressive and complete healing is less likely, a rapid diagnosis and medical remedy of the hypoperfusion by hemodynamic stabilization and restoring normal fluid balance is critical [29].

Consequently, patients' prognosis is poor [30]. While in some cases AEN is associated with a fatal course, in other cases it is not necessarily a terminal event [8]. Depends on the severity of compromise of the patient's general health status and on the patient's age. Risk factors seem to be related to fatal outcomes with a mortality nearly 6% [30], including advanced age, male gender, extension of necrosis and associated comorbidities including hemorrhages, shock, sepsis, hypertension, hypotension, heart disease, liver failure, kidney failure, diabetic ketoacidosis and alcoholism [12,14].

Complications

Long-term complications of the AEN include esophageal stenosis, stricture formation, and esophageal perforation [2,3]. Stricture formation occurs with an incidence of 25% and may require endoscopic balloon dilation [2,4,31,32]. The most serious complication of AEN is esophageal perforation which can occur in less than 7% of patients, perforation can be fatal if not recognized and treated immediately with surgical intervention [2,33].

The management of relevant medical and surgical complications such as super infection, perforations, mediastinitis and fistulas is important, some interventions considered are dilating stenosis, stenting even esophagectomies in cases of poor response or serious complications [14,34].

Conclusions

Necrotizing esophagitis is a rare and severe esophageal disorder with high mortality and complication rates. The high comorbidity that accompanies it is, apparently, the trigger and cause of mortality for a large group. Comorbidity, together with advanced age, male gender, liver cirrhosis and the extent of necrosis are important risk factors.

The therapeutic approach must therefore be interdisciplinary and must aim at restoring the hemodynamic, acid-base, metabolic and acid-peptic equilibrium while managing infectious complications. Serious complications occur frequently and require close evaluation and timely management due to the risk to the patient's life.

References

1. Choksi V, Dave K, Cantave R, et al. Black Esophagus or Gurgits Syndrome A Rare Complication of Diabetic Ketoacidosis. *Case Reports in Gastrointestinal Medicine*. 2017; 2017: 3.
2. Gurvits GE. Black esophagus acute esophageal necrosis syndrome. *World Journal of Gastroenterology*. 2014; 16: 3219-3225.
3. Worrell SG, Oh DS, Greene CL, et al. Acute Esophageal Necrosis A Case Series and Long- Term Follow-Up. *The Annals of Thoracic Surgery*. 2014; 98: 341-342.
4. Rigolon R, Fossa I, Rodella L, et al. Black esophagus syndrome associated with diabetic ketoacidosis. *World Journal of Clinical Cases*. 2016; 4: 56.
5. Lacy BE, Toor A, Bensen SP, et al. Acute esophageal necrosis report of two cases and a review of the literature. *Gastrointest Endosc*. 1999; 49: 527-532.
6. Augusto F, Fernandes V, Cremers MI, et al. Acute necrotizing esophagitis a large retrospective case series. *Endoscopy*. 2004; 36: 411-415.
7. Monteiro JMC, Castelo LF, Fischer WGG, et al. Black esophagus. *Autops Case Rep*. 2019; 9: e20180077.
8. Ramos R, Mascarenhas J, Duarte P, et al. Esofagitis necrosante aguda análisis retrospectivo. *Rev Esp Enferm Dig*. 2008; 100: 583-585.
9. Yasuda H, Yamada M, Endo Y, et al. Acute necrotizing esophagitis role of nonsteroidal anti-inflammatory drugs. *J Gastroenterol*. 2006; 41: 193-197.
10. Gurvits GE, Shapsis A, Lau N, et al. Acute esophageal necrosis a rare syndrome. *J Gastroenterol*. 2007; 42: 29-38.
11. Akkinepally S, Poreddy V, Moreno A, et al. The clinical picture Black esophagus. *Cleve Clin J Med*. 2009; 76: 400.
12. Manno V, Lentini N, Chirico A, et al. Acute esophageal necrosis a case report and literature review. *Acta Diabetol*. 2017; 54: 1061-1063.
13. Zacharia GS, Sandesh K, Ramachandran T, et al. Acute esophageal necrosis an uncommon cause of hematemesis. *Oman Med J*. 2014; 29: 302-304.
14. Ángel Arango LA, Cortés Mora EJ, Forero Piñeros EA, et al. Necrotizing esophagitis Presentation of cases and literature review. *Rev Colomb Gastroenterol*. 2018; 33: 249-268.
15. Arthur J, Caro D, Lester D, et al. Black esophagus A rare cause of gastrointestinal hemorrhage in the emergency department. *Emerg Med*. 2018; 50: 155-160.
16. Gurvits GE, Cherian K, Shami MN, et al. Black esophagus new insights and multicenter international experience in 2014. *Dig Dis Sci*. 2015; 60: 444-453.
17. Burtally A, Gregoire P. Acute esophageal necrosis and low-flow state. *Can J Gastroenterol*. 2007; 21: 245-247.
18. Endo T, Sakamoto J, Sato K, et al. Acute esophageal necrosis caused by alcohol abuse. *World J Gastroenterol*. 2005; 11: 5568-5570.
19. Bear BC, Mathew J, Parker CW III, et al. Acute esophageal necrosis black esophagus in setting of diabetic ketoacidosis. *J Case Rep Images Med*. 2015; 1: 18-21.
20. Doehn M, Schwartau M. Alcohol induced lactic acidosis in thiamine deficiency. *Z Rechtsmed*. 1982; 89: 125-130.
21. Duffens K, Marx Ja. Alcoholic ketoacidosis a review. *J Emerg Med*. 1987; 5: 399-406.
22. Fulop M. Alcoholism ketoacidosis. *Diabet Metab Rev*. 1989; 5: 365-378.
23. Hoyumpa AM. Mechanism of thiamin deficiency in chronic alcoholism. *Am J Clin Nutr*. 1980; 33: 2750-2761.
24. Klein M, Weksler N, Gurman GM, et al. Fatal metabolic acidosis caused by thiamine deficiency. *J Emerg Med*. 2004; 26: 301-303.
25. Keane S, Gallagher A, Ackroyd S, et al. Cerebral venous thrombus during diabetic ketoacidosis. *Arch Dis Child*. 2002; 86: 204-206.
26. DiMeglio LA, Chaet MS, Quigley CA, et al. Massive ischemic intestinal necrosis at the onset of diabetes mellitus with ketoacidosis in a three year old girl. *J Pediatr Surg*. 2003; 38: 1537-1539.
27. Tanaka M, Miyazaki Y, Ishikawa S, et al. Alcoholic ketoacidosis associated with multiple complications report of 3 cases. *Intern Med*. 2004; 43: 955-959.
28. Julián Gómez L, Barrio J, Atienza R, et al. Esofagitis necrotizante aguda. Una patología poco conocida. *Rev Esp Enferm Dig*. 2008; 11: 701-705.
29. Field Z, Kropf J, Lytle M, et al. Black Esophagus A Rare Case of Acute Esophageal Necrosis Induced by Diabetic Ketoacidosis in a Young Adult Female. *Case Rep Gastrointest Med*. 2018; 2018: 1-3.

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30. Deliwala S.S, Areeg Bala, Tarek Haykal, et al. AEN and acute pharyngitis. *Am J Case Rep.* 2020; 21: e926019.
 31. Inayat F, Hurairah A, Ul Hassan Virk H, et al. Acute Esophageal Necrosis An Update. *North American Journal of Medical Sciences.* 2014; 8: 320-322.
 32. Jeyalingam T, Shimamura Y, Teshima C, et al. Endoscopic Resolution of Black Esophagus With Treatment of Diabetic Ketoacidosis. *Clinical Gastroenterology and Hepatology.* 2018; 2018: 68-69.
 33. Talebi Bakhshayesh M, Samiee Rad F, Zohrenia H, et al. Acute esophageal necrosis a case of black esophagus with DKA. *Archives of Iranian Medicine.* 2015; 18: 384-385.
 34. Siddiqi A, Chaudhary FS, Naqvi HA, et al. Black esophagus a syndrome of acute esophageal necrosis associated with active alcohol drinking. *BMJ Open Gastro.* 2020; 7: e000466.