

Acute Esophageal Necrosis. A Case Report.

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Abstract

Acute esophageal necrosis syndrome, is a rare cause of gastrointestinal bleeding. It is characterized by a particular black coloration, mostly of the distal esophagus, which ends abruptly in the gastroesophageal junction. The hemorrhagic manifestation may be hematemesis, melena or both of them. It is a life-threatening condition with significant difference between the two sexes. We report a case of a 67 years old male with schizophrenia, aspiration pneumonia and sepsis who presented with hematemesis. The endoscopic finding was a black, wide linear necrosis of the esophageal mucosa which extended from the middle esophagus to the gastroesophageal junction, compatible with acute esophageal necrosis. The esophageal lesion healed progressively at the same time with the improvement of the general health condition.

We must think of acute esophageal necrosis in critically ill male patients, in their sixth decade of life, with multiple comorbidities and gastrointestinal hemorrhage, because early recognition can help reducing the mortality of this rare syndrome.

In conclusion, we must think of acute esophageal necrosis in critically ill male patients, in their sixth decade of life, with multiple comorbidities and gastrointestinal hemorrhage. The diagnostic is made most of the time only by the characteristic endoscopic findings. Because of the gravity of this affection, the treatment must be quick and aggressive.

Keywords; esophageal necrosis, gastrointestinal bleeding, black esophagus

Introduction

Acute esophageal necrosis (AEN) syndrome, also known as “black esophagus”, necrotizing esophagitis or Gruvitis syndrome is a rare cause of gastrointestinal bleeding. The endoscopic finding is a particular black coloration mostly of the distal esophagus which ends abruptly in the gastroesophageal junction [1]. It is often found in severely ill patients. The hemorrhagic manifestation may be

hematemesis, melena or both of them. It is a life-threatening condition with a high mortality rate, approaching 32%, but the mortality specific to AEN is closer to 6% [2]. There is a significant difference between the two sexes, with a higher incidence in men. The physiopathology is still unclear and multiple events may be in cause.

We report a case with AEN in a schizophrenic patient with aspiration pneumonia and sepsis.

Case report

A 67 years old male with schizophrenia under treatment with Risperdal and Clozarem, living alone, is referred to the emergency endoscopy unit for upper endoscopy.

He has been taken to the hospital and presented to the emergency room for deterioration of the general condition, obnubilation, dyspnea and hematemesis.

At the admission he had: blood pressure 140/70 mmHg, heart rate 100 bpm, respiratory rate 25/min, oxygen saturation 89%. On the physical examination he was pale, agitated, uncooperative and the communication was difficult

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because of his mental disease. The laboratory findings were as follow: red blood cells 4.57×10^6 u/L, hemoglobin 12.8 g/dL, hematocrit 37,6%, white blood cells 24.4 K/uL, neutrophils 22.3 K/uL (91.5%), lymphocytes 0.9K/uL (3.7%), monocytes 1.1 K/uL (4.6%), eosinophils 0.0 k/uL (0%), platelets 413 K/uL, C-reactive protein 9.52 mg/dL, urea 100 mg/dL, creatinine 1.90 mg/dL.

The pulmonary CT scan showed fibrotic zones and images compatibles with aspiration pneumonia. The covid-19 rapid test was negative. HIV test was also negative.

The above findings were consistent with the diagnosis of sepsis and aspiration pneumonia.

The esophagogastroduodenoscopy (EGD), performed three hours after the admission, showed black, wide, linear necrosis of the esophageal mucosa which is extended from the middle esophagus to the gastroesophageal junction. The rest of the mucosa had active oozing (Fig. 1, 2). A deep duodenal ulcer was also found, with no signs of active hemorrhage. The patient was immediately transferred to the intensive care unit, because of the altered clinical condition and poor prognosis.

After 4 days of intravenous fluids, antibiotics, proton pump inhibitors and parenteral nutrition the patient's clinical condition improves distinctly and he underwent a second EGD, which demonstrates also an improvement of the black mucosal necrosis found in the first EGD. Most of them were covered by white exudate and there was no active bleeding (Fig. 3,4). The patient was discharged from the hospital after 16 days of hospitalization. He did not present to the follow-up endoscopy, planned one month later.

Discussion

Black esophagus is a relatively rare entity, firstly described in 1990 by Goldenberg [3]. The latest report of the cases diagnosed with necrotizing esophagitis encountered 118 patients [4]. According to different studies the incidence is 0,01-0,28% [4].

The etiology seems to be multifactorial, including ischemic events of the esophagus, the nonfunctional mucosal barrier and the chemical injury from gastric secretions. Most authors sustain the two-hit hypothesis, where there is a combination of ischemic event with increased metabolic requirement caused by the ischemia and the necrosis [5] [6].

Men are more affected with a sex ratio 1:4 and the average age of diagnostic is 67 years [7].

In 70% of the patients the clinical presentation of AEN is hematemesis [5] [8], which never appears alone. Other symptoms are: dysphagia, epigastric pain, chest pain and signs of underlying comorbidities.

Laboratory findings in our patient were compatible with infectious syndrome, sepsis and altered renal function. The CT scan didn't reveal changes in the esophagus or stomach. In patients with AEN, leukocytosis and anemia are common findings. The computed tomography may find thickened distal esophagus, hiatal hernia or gastric outlet obstruction [9].

AEN may be underdiagnosed, because of the gravity of the patients at the presentation, who are severely ill, with alternated hemodynamics, multiple pathologies and the endoscopic procedure in this patient is often difficult to perform in the first hours. It also exists subclinical forms of the disease and a relatively early healing of the mucosal lesions in case of transitory ischemia of the esophagus [7] [10]. The diagnostic is histological and the samples show necrotic mucosal and submucosal lesions, but the biopsy is not always necessary. There is high risk of perforation during the biopsy and most of the time the diagnosis is based on endoscopic examination and clinical features.

The differential diagnosis must be done with melanocytosis, pseudo-melanosis, malignant melanoma, acanthosis nigricans, coal dust deposition, exogenous dye ingestion, lye ingestion [11].

In our case, referring to the mental disease of the patient, we had to do the differential diagnosis with esophagitis from voluntary exogenous corrosive substance ingestion. This diagnosis was quickly dismissed, based on the second EGD, performed 4 days after the first one.

In the first endoscopy we found stage I AEN, referring to a staging system proposed by Gurvitis et al [5], while in the second endoscopy the coexistence of white exudates with black mucosa is compatible with the healing phase or stage 2 AEN.

The relatively quick improvement of the mucosal lesions in parallel with the improvement of the general condition, their topography, the end of the lesions at the gastroesophageal junction are arguments in favor of acute esophageal necrosis. The lower third esophagus is more affected, because it is less vascularized comparing to the proximal esophagus and it has poor collateral blood supply [9] [12]. It is also the most exposed to gastric reflux and gastric acid. In 36% of the cases, the entire esophagus is involved [13].

In our patient we also found a duodenal bulbar ulcer (Fig. 5), which seems to be related to AEN. Pathologies of duodenum, as ulcers and severe duodenitis, and pyloric region (deformed pyloric region) are found in 2/3 of the patients with black esophagus [14]. The pseudo-obstruction favors gastro-esophageal reflux and exposes the distal esophagus to gastric acid, biliary acid and pepsin. This process potentiates hypoperfusion and cause mucosal injuries and impairment of the mucosal healing [14].

The etiology of AEN in our patient is probably ischemic, related to the hemodynamic compromise following the sepsis and the alteration of protective barriers due to the poor nutritional status because of the mental disease. The coexistence of a poor nutritional status, with the vascular insufficiency and the hemodynamic instability can cause hypoperfusion of the esophageal tissue, mucosal buffering, diminishing of protective barriers and impairment of mucosal repairing mechanisms.

The risk of developing AEN is higher in patient with diabetes mellitus, hypertension, coronary artery disease, alcohol abuse, chronic kidney disease, chronic liver disease,

malignancy, peripheral vascular disease, chronic obstructive pulmonary disease and poor nutritional status [5] [6] [15] [13] [8] [16] [17]. These conditions are associated with severe vasculopathy and low flow which lead to hypoperfusion of the esophagus and ischemic events of the organ, especially of its distal part.

The treatment of AEN consist in treating the underlying severe comorbidities, which always accompanied this condition. The patients must be placed NPO (nil-per-os) and parenteral nutrition must be initiated if a long time NPO is planned. Correction of fluid imbalance is very important. Proton pump inhibitors are systematically used to protect the damaged esophagus. Oral sucralfate is indicated in addition to anti-acid systemic therapy, if renal failure is not present, which was not our case.

Our patient was treated with broad spectrum antibiotics for the sepsis, but the use of antibiotics, antivirals and antifungal medicaments might be precautions, because they are often accused as a cause of AEN [6]. At the discharge he was given proton pump inhibitors until next consultation and endoscopy.

AEN early complications are bleeding (85%) and esophageal perforation (<7%) [13] [18]. Endoscopic examination must be performed with caution and interrupted if necessary, in case of suspicion of esophageal perforation. The esophageal stricture (70%) and trachea-esophageal fistulas (30%) are late complications [8].

We could not evaluate the long-term evolution of AEN in our patient, because he did not present at the consultation planned after one month of the discharge from the hospital, but the endoscopic follow-up is recommended to evaluate the healing or the complications of AEN.

The mortality rate alone is approximately 6%, but it grows exponentially to 31.8-50% and has a poor prognosis, when in presence of other serious comorbidities [16] [19] .

In conclusion, we must think of AEN in critically ill male patients, in their sixth decade of life, with multiple comorbidities and gastrointestinal hemorrhage. The diagnostic is made most of the time only by the characteristic endoscopic findings. Because of the gravity of this affection, the treatment must be quick and aggressive.

Figures

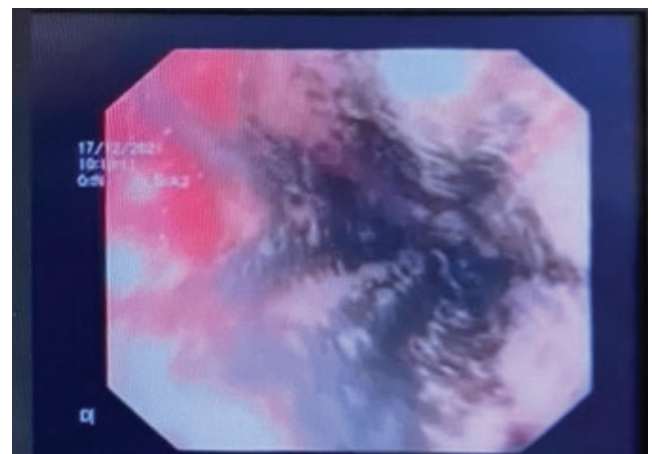


Fig. 1,2: Black linear necrosis of the esophageal mucosa from middle esophagus to the gastroesophageal junction (Day 0)

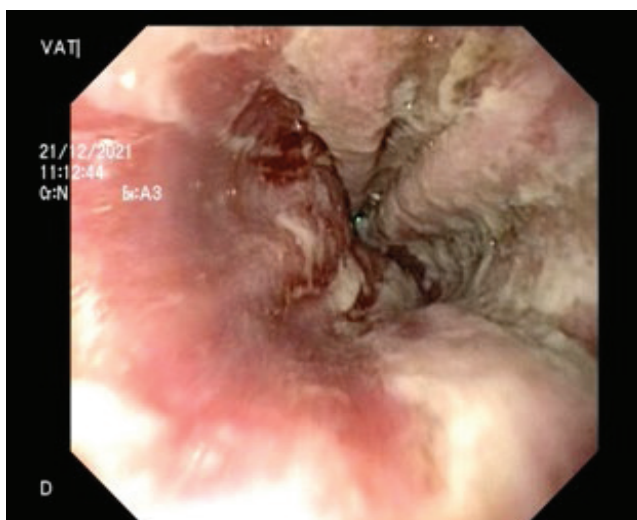


Fig. 3,4: Beginning of the mucosal healing, white exudate covering the linear necrosis (Day 4)

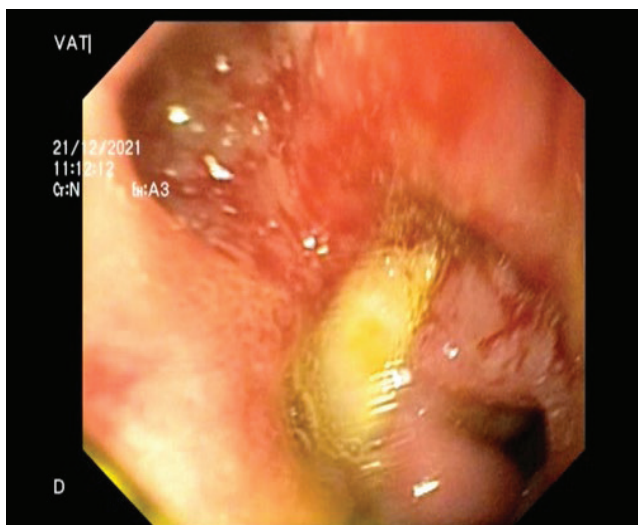


Fig. 5: Ulcer of the anterior face of the duodenal bulb

Declaration of any potential financial and non-financial conflicts of interest:

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Ethics approval and consent to participate – all the patients/their parents have signed informed consent.

Consent for publication - the patient/his parents have signed informed consent

Clinical trial registration information provided – not applicable

We confirm that the manuscript, including related data, figures and tables has not been previously published and that the manuscript is not under consideration elsewhere.

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