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EC GASTROENTEROLOGY AND DIGESTIVE SYSTEM

Case Study

Black Esophagus: Acute Esophageal Necrosis. Number of Cases

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Abstract

Acute esophageal necrosis, also known as the black esophagus, is a rare pathology diagnosed by endoscopy showing a black-looking esophageal mucosa. We present three cases of acute esophageal necrosis, the multifactorial pathogenesis was in our patients, especially hypoperfusion being probably the key factor for esophageal lesions. The patients presented the same endoscopic characteristics and the esophageal biopsy samples of two patients had a histological pattern of severe inflammation and necrosis, one of them associated with fungal infection. All had a favorable evolution, and esophageal stenosis was presented as a complication in one of the cases presented.

Keywords: *Black Esophagus; Acute Esophageal Necrosis*

Introduction

The black esophagus or acute esophageal necrosis (NEA) is a rare medical condition characterized by a black-looking circumferential esophageal mucosa that almost universally affects the distal third of the esophagus and there is a sudden transition in the gastroesophageal junction [1-3]. This syndrome is gaining acceptance as an important cause of upper gastrointestinal bleeding in hospitalized patients [4].

The exact prevalence is unknown [5], was described by Brennan in 1967 at an autopsy and the first endoscopic description was in 1990 by Goldenberg., *et al* [6]. Two large series of autopsies from the United States and France have reported zero cases from a series of 1000 adult autopsies and 0.2% at 3000 autopsies, respectively.

Two retrospective series that have reviewed the results of > 100,000 endoscopies have estimated the incidence of approximately 0.01% (12 patients), and another retrospective analysis of 10,295 endoscopies has shown an incidence of 0.28% (29 patients) [7].

The pathogenesis of NEA is not well known, but tissue injury secondary to a state of hypoperfusion plays a dominant role [8]. Approximately 70 percent of patients present with upper gastrointestinal bleeding with hematemesis and melena. Symptoms can develop rapidly after a triggering event [1,9]. Endoscopically, the esophageal mucosa is diffusely necrotic, "black esophagus", which preferentially affects the distal esophagus, is variable in length, and abruptly stops in the UGE [2,10].

The differential diagnosis includes malignant melanoma, acanthosis nigricans, pseudo melanosis, and melanocytosis of the esophagus [11]. Management is based on aggressive resuscitation, correction of underlying medical conditions, institution of therapy with proton pump inhibitors, sucralfate and monitoring for signs of infection or perforation [7].

Complications include perforation, mediastinitis, and stricture formation. Overall mortality is related in most cases to the underlying disease [8].

Here we present the clinical and endoscopic characteristics of three patients who underwent acute necrotizing esophagitis, we reviewed the possible causes that originated its management and follow-up.

Clinical Case No 1

Female patient of 56 years of age, illiterate, with a history of pott mal disease diagnosed 3 months ago, she underwent spinal decompression with arthrodesis and was receiving antitumor treatment.

Patient with a week of diarrheal stools in moderate amount several times a day, is accompanied by vomiting of food content and two days before his admission begins with manes 3 times a day and hematemesis in coffee well twice. Clinical picture is accompanied by general malaise, weakness so she is taken to the hospital by her son.

On admission, dehydrated, TA hypotension: 84/60 FC: 92X 'afebrile, with dry oral mucosa, Qx erythematous wound in the posterior thoracic region. Painful abdomen in the epigastrium.

Laboratory: Leukocytes: 3170, Hb: 7.3, Ht: 21.5%, PLT: 39,000 Creatinine: 0.9, Urea: 116, Na: 124 K: 2.5 TP: 23.8 INR: 1.89.

In the endoscopy, the black esophageal mucosa was observed in the middle and distal third (Figure 1). The biopsy revealed areas of necrosis with inflammatory infiltrate, cellular debris, as well as fungal hyphae and spores compatible with candida (Figure 2).

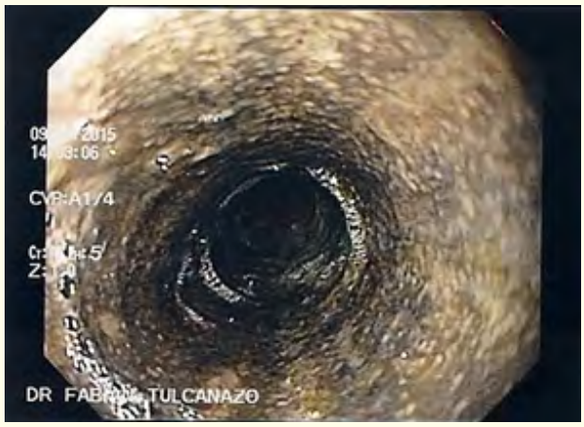

Figure 1

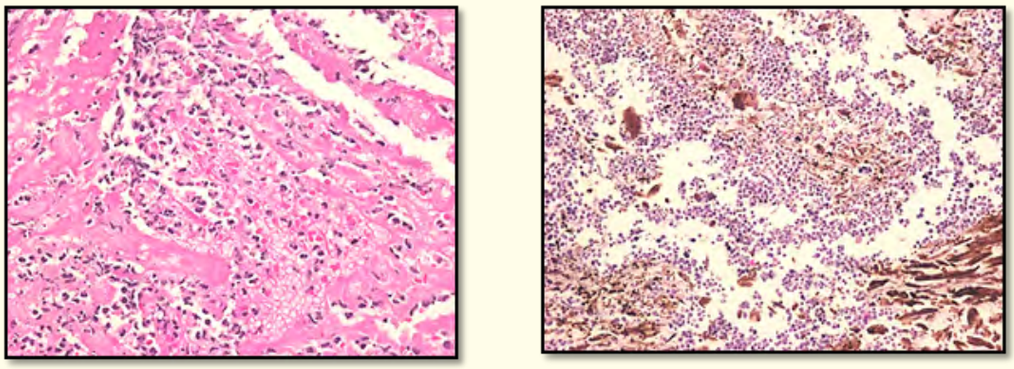

Figure 2

Patient was managed in intensive care unit with satisfactory evolution, was discharged. Endoscopic control at 5 weeks revealed esophageal stenosis (Figure 3), which was managed with dilatation.

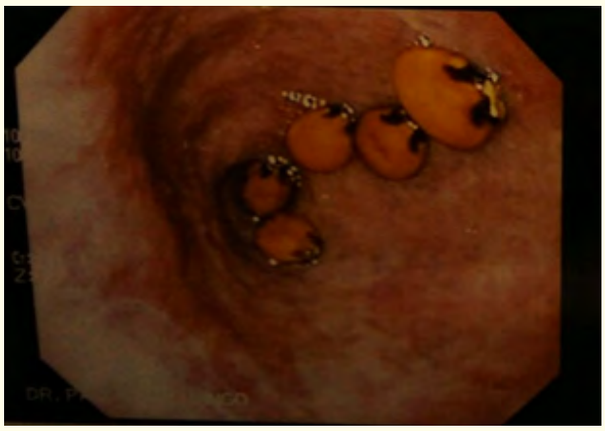

Figure 3

Clinical Case No 2

A 71-year-old male patient with a history of hypertension treated with telmisartan.

Approximately 30 hours ago she presented retrosternal and epigastric chest pain, without irradiation, of slight intensity and 24 hours later she was accompanied by manes for 3 times, asthenia and hyporexia. Patient goes to emergency with vital signs within normal parameters, normal cardiopulmonary, little painful abdomen in epigastrium.

Laboratory: Leukocytes: 16,480 Hb: 13.3 Ht: 38.3% PLT: 201,000 Creatinine: 1.5 Urea: 140 Na: 138 K: 4.09 INR: 1 EKG: normal.

The endoscopy showed a necrotic esophageal mucosa, friable in the middle and distal third (Figure 4).

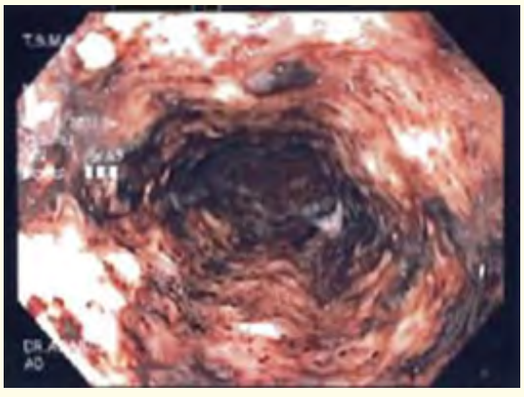

Figure 4

A chest CT was performed, evidencing a concentric thickening of the walls of the esophagus with thickness up to 1 cm (Figure 5).

The patient received treatment with proton pump inhibitor with good response, after 5 days an endoscopic control was performed that showed a significant improvement of the esophageal mucosa, stable patient was discharged (Figure 6).

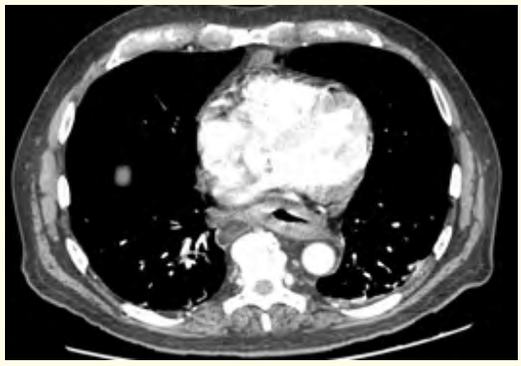

Figure 5

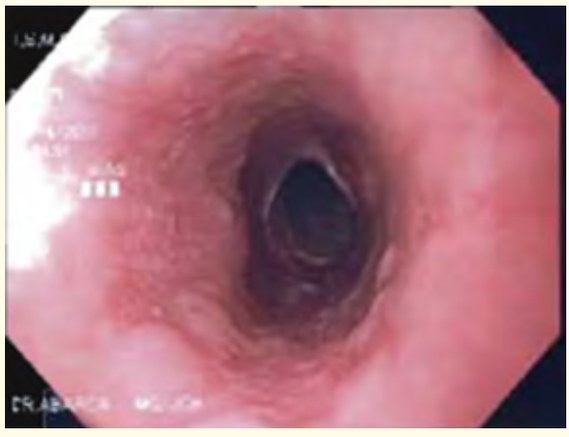

Figure 6

Clinical Case No 3

An 84-year-old male patient with a history of dyslipidemia, benign prostatic hypertrophy, atorvastatin, doxazosin. Two weeks ago he was hospitalized for food poisoning and received treatment with prednisone, epinastine, hydroxyzine, montelukast, ranitidine, he reported having maintained vomiting after discharge.

Patient who two hours prior to admission with apparent cause of vomiting of food content for several occasions, has lipothymia, receiving a blow in the left frontal region without loss of consciousness. Later he gets up by his means and presents pain in epigastrium of great intensity, is accompanied by anxiety so he comes for his assessment.

On admission, the patient presented with TA: 133/72 FC: 91X 'excoriation in the left frontal region, normal cardiopulmonary, non-painful abdomen. Laboratory: Leukocytes: 14,990 Hb: 15.6 Ht: 43.4% PLT: 182,000 Creatinine: 1.1 Urea: 28.3 Na: 129 K: 3.45 INR: 1 EKG: normal. Normal skull CT.

Patient presented pain in the epigastrium again, so an endoscopy was performed that showed a black esophageal mucosa throughout its extension to the gastro esophageal junction (Figure 7).

A chest CT angiography was performed where a concentric thickening of the walls of the esophagus was observed (Figure 8).

The patient evolved favorably to treatment with proton pump inhibitor, medical discharge was given and later an endoscopic control was performed at 6 weeks that showed an esophageal mucosa with normal characteristics.

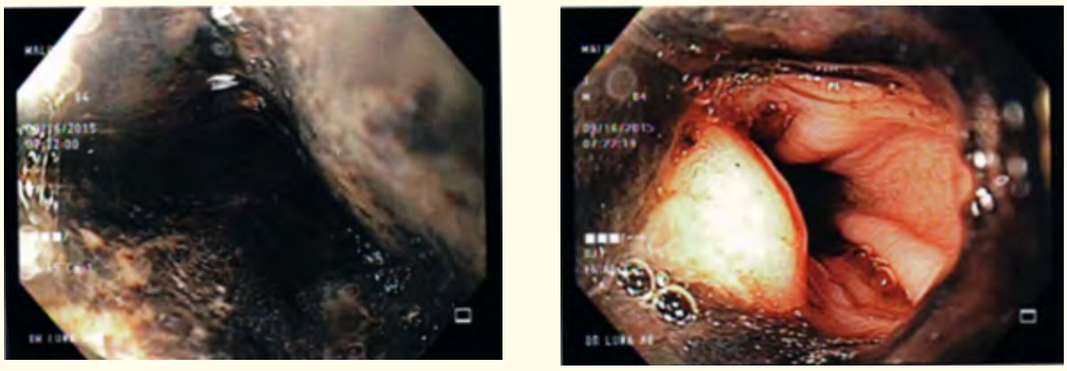

Figure 7

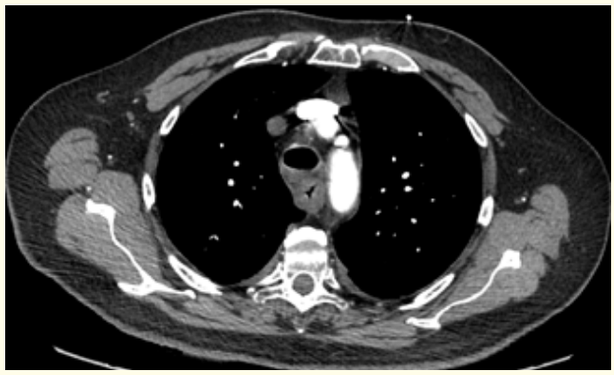

Figure 8

Discussion

The clinical presentation of NEA is with manifestations of high digestive bleeding, hematemesis and melena. In the review of systems, epigastric abdominal pain, nausea, vomiting, dysphagia, low-grade thermal rise, dizziness and syncope can be noted [12]. Other associated clinical conditions may include multiple organ failure, cardiac, pulmonary, and kidney disease, diabetes mellitus and ketoacidosis, vasculopathy, coagulopathy, peptic ulcer disease, generalized weakness and malnutrition, cirrhosis, acute alcoholic hepatitis, acute fatty liver of pregnancy, acute pancreatitis, sepsis, ischemic processes (cerebral infarction, intestinal ischemia), and trauma. The physical findings in a patient with NEA are usually confused by the underlying medical conditions, but may be notable for cachexia, fever, hypoxia, hemodynamic instability, including arrhythmias and hypotension, pallor, abdominal pain and positive guaiac in feces [8,14,15].

Morita., et al. describe the following criteria for the diagnosis of acute esophageal necrosis: circumferential black esophagus with or without exudates; involvement of the distal esophagus, which ends abruptly in the gastroesophageal junction; and the absence of caustic ingestion. The biopsy material can be obtained for definitive histological confirmation, but it is not required. Histological findings include the presence of necrotic debris in the esophagus biopsy, necrotic mucosa without viable epithelium, and there may be involvement of the submucosa, sometimes extending within the muscularis propria layer, which can lead to full-thickness lysis [8,13].

Most investigators have suggested that NEA has an ischemic origin based on histopathological study and clinical data [16]. The association of esophageal lesions with a low flow state, and the rapid resolution of the esophageal lesion after hemodynamic stabilization suggest that a temporary reduction of the blood perfusion of the esophagus can lead to extensive esophageal necrosis. The frequent involvement of the distal third of the esophagus, which is less vascularized, represents another clue; In addition, malnutrition can compromise the mucosal defense system and healing capacity after an injury, either by ischemia or another cause, resulting in NEA [17].

The initial management consists of the volume expansion with intravenous fluids and the treatment of the underlying disease. The suppression of gastric acid with intravenous proton pump inhibitors should be used to reduce additional peptic acid injury to the esophagus. Probably, oral therapy should be continued for a few months after resolution of symptoms to help prevent the formation of strictures. Reports of esophageal manometry and pH monitoring in patients with NEA have shown a lack of significant acid reflux and normal motility in 5 - 7 months after recovery [18].

Oral intake should be avoided for at least 24 hours, after which administration of sucralfate should be considered due to its cytoprotective effects and its ability to bind to pepsin and stimulate mucus secretion [19]. The nasogastric tube should not be used to prevent perforation. A decision regarding antimicrobials and antifungal use should be made on an individual basis, especially in the setting of patients who are critically ill or septic [19]. Empirical antibiotics should be initiated in cases of suspected esophageal perforation, rapid clinical decompensation, unexplained fever, in patients with AIDS, cirrhosis, transplant recipients, and patients on dialysis. Surgical intervention is reserved for the esophagus perforated with mediastinitis and the formation of abscesses [8].

The most serious complication of the NEA is perforation, which can occur in severe cases resulting from necrosis of the total thickness of the esophageal tissue. Its incidence is less than 7%. Other possible sequelae of the NEA is the formation of stenotic areas, which can be seen in > 10% of patients, the suppression of acid followed by an endoscopic dilation if necessary, usually produces a resolution [20].

The prognosis in general is poor, with mortality of almost a third of the patients due to the underlying critical illness. Mortality is variable as reported in the literature from 15% to as high as 36% reported in some series [21]. However, the specific mortality of the ENA is much lower, around 6%. Important risk factors include esophageal perforation, diabetic ketoacidosis, and compromised immune system [1].

	Case 1	Case 2	Case 3
Etiopathogenesis	Sepsis, hipovolemia	Hyperperfusion	Hypovolemia
Clinical Symptoms	Melena and hematemesis	Thoracic pain in the epigastrium and manes	Syncope, pain in the epigastrium
Endoscopic Findings	Commitment of the middle and distal third of the esophagus	Commitment of the middle and distal third of the esophagus	Commitment of the entire esophagus
Treatment	Management of basic pathology + IBP	IBP	IBP
Results	Esophageal Stenosis	Asymptomatic	Asymptomatic

Table 1

In our case review, two patients presented with signs of digestive bleeding and one with syncope, all of them underwent endoscopic diagnosis that showed the typical characteristics of NEA, showing a black esophageal mucosa in all its circumference and in all cases it was affected. the distal third of the esophagus with an abrupt transition at the level of the gastric esophageal junction. In addition, in two patients a tomography was performed where a concentric thickening of the esophageal walls was observed. In two patients, biopsies were taken showing classic histological changes suggestive of NEA, and one of them also observed fungal hyphae and spores compatible with candida. All were managed with intravenous fluids, proton pump inhibitor plus the management of the underlying pathology, two of them needed intensive care with satisfactory evolution. In the subsequent controls, only one had esophageal stenosis as a complication that was managed with endoscopic dilation.

Conclusion

In conclusion, we can say that it is a rare clinical syndrome classically characterized by a black esophageal mucosa that stops abruptly in the gastroesophageal junction and suspicion is a key factor in the diagnosis of NEA, especially in elderly patients with comorbidities and the evidence of upper digestive bleeding. In addition, esophageal biopsy is probably not necessary if the clinical picture and the endoscopic findings are consistent with NEA. The goal of treatment should be directed to the treatment of coexisting medical diseases. The prognosis in general is poor, with mortality of almost a third of the patients due to the underlying critical illness.

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