Acute esophageal necrosis (“black esophagus”): Case series in forensic autopsies casuistry

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Abstract: Acute esophageal necrosis or “black esophagus” is a rare medical condition characterized by necrosis of the esophageal mucosa, most of the time limited to the distal esophagus. We present three such autopsied cases, all men with a history of chronic alcohol abuse, all from the same area in Mures County and in the same period of time (a few months apart). Histological examination confirmed necrosis in all cases, and a violent nature of the necrosis (acute intoxication with caustic substances) was excluded based on the strict limitation of the necrosis to the medium and inferior esophagus. Also, H-PAS stain was performed, to investigate possible fungal infections, often cause of esophageal necrosis in immunosuppressed patients. We found alcoholism and diabetes mellitus as risk factors for esophageal necrosis and believe that in establishing cause of death the forensic pathologist must consider other internal organs involvement more than the role of the injured esophagus.

Key Words: acute esophageal necrosis, black esophagus, forensic autopsy.

Acute esophageal necrosis (AEN), also known as “black esophagus” or “necrotizing esophagitis”, was considered, until recently, a rare entity. It has a prevalence of 0.01-0.28% in human cases [1], while in veterinary medicine is still an extremely rare finding [2]. It was described in 1967 by Brennan [3] and in 1977 by Lee [4] in post-mortem cases, related to hypothermia and to spontaneous thoracic aorta rupture respectively.

Acute esophageal necrosis is a condition marked by circumferential necrosis most frequently limited to the distal esophageal mucosal layer, with an abrupt transition to healthy mucosa at the gastro esophageal junction [5]. The microscopic examination usually reveals necrotic debris, mucosal and submucosal necrosis, and a local inflammatory response [6, 7]. Also, thrombosis of the blood vessels may be present.

The ethiopathogenesis of acute esophageal necrosis is still not completely understood. Possible etiologies include ischemia, severe viral infection, severe vomiting with corrosive injury and hypersensitivity to antibiotics [8]. Risk factors include diabetes mellitus, advanced age, male gender, hypertension, coronary artery disease, alcohol abuse, advanced malignancy [9]. Some authors reported cases associated with different pathogens, such as Klebsiella pneumoniae, Lactobacillus acidophilus, Cytomegalovirus, Candida, all encountered in immunosuppressed patients [10].

It is essential to exclude patients with caustic ingestion injury, and patients with small local necrotic areas, that may be secondary to infection [11]. Differential diagnosis for AEN includes melanosis, malignant melanoma, acanthosis nigricans and coal dust deposition [12].

In human patients, the most frequent clinical signs of AEN include abdominal pain, hematemesis and melena [1].
CASE REPORTS

We present three cases autopsied in the Institute of Legal Medicine within a short period of time (three months), geographically related (all from the north-eastern part of Mures County).

Case 1

The first case is of a 41-year-old male, with no history of disease, but a known chronic alcohol abuser. The patient presented, during the evening, epigastric pain, coffee ground vomiting and chills, for which he self administrated paracetamol and pantoprazole. He was found dead in his house the next day. The medico-legal autopsy revealed no external injuries. At the internal exam, 90% of the esophageal mucosa (excepting the proximal part) presented a blackened and friable aspect, with limited hemorrhagic areas (Fig. 1A). The blackened mucosa is demarcated clearly from the cardia at the gastroesophageal junction, with regular lining of the gastric mucosa (Fig. 1B). Also, 3500 ml ascites and hepatic fibrosis were found.

Histology (haematoxylin-eosin stain) confirmed the diagnosis of „Black esophagus”, with necrosis of the mucosal and submucosal layers (Fig. 2A). Also, the presence of neutrophils dissociating the muscle fibers was noted (Fig. 2B). Blood alcohol level was negative. The death was ruled as being nonviolent and was due to metabolic and hydroelectrolitic disorders associated to the liver cirrhosis and AEN.

Case 2

In our second case, a 61-year-old man, with a history of alcohol abuse and diabetes mellitus (last documented glycemic value dating 2 weeks back was 468 mg/dL), was found dead in his house. The autopsy revealed liver cirrhosis and blackened esophageal mucosa in the middle and lower third of the esophagus, clearly demarcated from the cardia (Fig. 3). The stomach content had a pH level of 5.5 and blood alcohol was negative.

Using hematoxylin - periodic acid Schiff stain (H-PAS), the result was weak positive and a fungal infection was excluded by the histological exam (Fig. 4A). The hematoxylin-eosin stain confirmed the necrosis of the mucosa and submucosa with marked inflammatory infiltrate and intravascular thrombus (Fig. 4B); also, the liver cirrhosis was histologically confirmed. The death was ruled nonviolent, due to metabolic disorders associated to the liver disease.

Figure 1. A – Black esophagus – Case 1; B – Gastric mucosa.

Figure 2. A - Mucosal and submucosal necrosis, HE stain, 50X; B – Neutrophils among the muscle fibres, HE stain, 50X.
Case 3

A 64-year-old man, with a history of alcohol abuse and diabetes mellitus was found dead in his house, with no external traumatic injuries. The autopsy revealed blackened mucosa more pronounced in the middle third of the esophagus (Fig. 5), liver cirrhosis and hemorrhagic lines in the pancreas, suggestive of acute pancreatitis. Fourth degree of coronary atherosclerosis was noted on the left anterior descending branch of the left coronary artery, along with left ventricular hypertrophy and myocardiosclerosis. The histology confirmed the necrosis of the esophageal mucosa (hematoxylin-eosin stain). The death was ruled as nonviolent and due to the advanced cardiac pathology. The limited acute esophageal necrosis did not contribute to the cause of death.

DISCUSSION

The three reported cases were all males and lived in the same area of Mures County, a few villages apart from each other. We found no precise explanation to the geographical relationship, it might be coincidental. In all cases chronic alcoholism was noted in their history and in two cases diabetes mellitus was previously diagnosed. Both have been described before as risk factors for AEN. The type of alcohol used by all cases consisted mostly from a high-concentration distillate, home-made spirit.

From a medico-legal point of view, the first and most important thing is to exclude an acute intoxication with caustic substances (caustic esophagitis) that would classify the death as being violent. In the three presented cases, we based our decision to rule the death as being nonviolent and not due to caustic ingestion, on the clearly demarcated esophageal necrosis at the cardia and the absence of changes on the oro-pharyngeal and gastric mucosa. In case of caustic intoxication, although many patients survive the acute phase, the effects and evolution of the injury are serious with esophageal and gastric stenosis, leading to malignant lesions being the most probable outcome of caustic ingestion [13].
The H-PAS stain used in the histological exam was negative in case 1 and 3, but weakly positive in case 2; although a fungal etiology of the AEN could not be fully confirmed, it was still possible that fungi might have intervened as associated infection in the 61-year-old diabetic patient with poor glycemic control and liver cirrhosis; often, a fungal infection such as Candida may be the cause of AEN in immunosuppressed patients [10, 14].

In a recent literature review including 112 cases of AEN [15], the authors concluded that 78% of the cases involved male patients, and the mean age was 68 years. No obvious associated disease was observed but, in 15% of cases, a liver involvement was noted (from alcohol cirrhosis to acute fatty liver of pregnancy), followed by infectious causes in 7% of cases. Death rate was approximately 38%, while resolution occurred in 49% of the reviewed cases.

Upper gastro-intestinal (GI) bleeding is the most common symptom, varying from 65% to 100% in different studies [15, 16]. Another literature review comprising of 88 cases reports a death rate of 31.8% [6].

The endoscopic examination may reveal different types of lesions, from white plaques similar to those of pseudomembranous colitis [17] to blackened mucosa with adherent yellow exudates and ulcerations, which is considered the most common finding in AEN [18].

Regarding the course of treatment, it is recommended to treat the coexisting diseases and to correct the anemia if present [1]. Recommended medication includes proton pump inhibitor or histamine receptor blocker, preferably administered intravenously, with monitoring of normal motility via esophageal manometry and pH monitoring for acid reflux, according to some studies [19, 20].

In only two cases described in the literature, the authors considered the cause of death as being acute esophageal necrosis. The first case [21] of a 45-year-old woman with a history of cocaine and alcohol abuse, and the second case [22] of a 80-year-old male, chronic alcohol user and with a history of cardiac disease. Gurvits reacted to these statements [23, 24], bringing arguments that in both cases, it is wrong to rule the death as due to AEN. In the first case, the alcohol and cocaine abuse are more likely to have caused the death, while in the second case, the advanced age, heart problems and alcohol abuse are more likely to be the cause of death. We support that the prognosis of the patient depends mainly on the age and associated comorbidities, rather than on the extent of esophageal lesions. Concerning thanatogenesis, we also support the idea that the cause of death must be established considering other organs involvement (in our cases it was liver and heart) more than AEN itself. Among our cases, only in case 1 could we state a contribution of the esophageal necrosis, by eliciting prolonged vomiting.

CONCLUSIONS

Although black esophagus is a rare condition, it should be considered when facing an elderly patient with gastrointestinal bleeding, multiple associated diseases and alcohol abuse.

From the forensic pathologist’s point of view it is important to differentiate AEN from caustic esophagitis, the latter being the result of an acute intoxication and determining the nature of death to be violent. It is also important to establish the cause of death, which is mainly due to associated pathological conditions rather than to the esophageal necrosis itself.

Conflict of interest. The authors declare that they have no conflict of interest concerning this article.

References