Journal of Clinical and Investigative Surgery

https://doi.org/10.25083/2559-5555
https://proscholar.org/jcis/
ISSN: 2559-5555

Esophageal perforation due to acute esophageal necrosis: A case report and a comprehensive literature review

Carlos Petrola Chacón[1], Sandra Castro Boix[2*], Nivardo Rodríguez Conde[2], Amaia Gantxegi Madina[2], Mariona Jofra[2], Daniel Gil Sala[3], Manel Armengol Carrasco[1]

[1]*General and Digestive Surgery Department. Vall D´Hebron Hospital Universitari. Universitat Autonòma de Barcelona. Barcelona, Spain*
[2]*Upper GI Surgery Division. General and Digestive Surgery Department. Vall D 'Hebron Hospital Universitari. Universitat Autònoma De Barcelona. Barcelona, Spain*
[3]*Angiology and Vascular Surgery Department. Vall D'hebron Hospital Universitari. Universitat Autònoma De Barcelona. Barcelona, Spain*

ABSTRACT

Background. Acute esophageal necrosis is a rare and potentially lethal entity. The pathogenesis is multifactorial, generally presenting with symptoms of upper gastrointestinal bleeding. We present a case that presents atypically with initial esophageal perforation. *Case presentation*. A 46-year-old man with a history of alcoholism and cocaine use, an active smoker, and a ruptured celiac trunk aneurysm treated by embolization, who, after acute chest and epigastric pain, is diagnosed with a Stanford B thoracoabdominal aortic dissection, being repaired endovascularly by placing an aortic endoprosthesis. Due to clinical suspicion of mesenteric ischemia complicated with esophageal/gastric perforation, a postoperative tomography was performed, revealing perforation of the esophagus distal to the left pleura and ischemic cholecystitis. Transhiatal esophagectomy, cervical esophagostomy, Witzel-type decompressive gastrostomy, Witzel-type feeding jejunostomy, classic cholecystectomy, and mediastinum drainage were performed. During the postoperative period, the patient remained in critical condition, dying as a result of hypoxic encephalopathy. The histopathological study reported acute transmural esophageal ischemia. *Discussion*. Tissue hypoperfusion plays a dominant role in the pathogenesis of acute esophageal necrosis. Esophageal perforation is a serious complication and can occur in the early stages, with esophagectomy and deferred digestive reconstruction being the appropriate treatment. *Conclusion*. Ischemia is a fundamental mechanism of acute esophageal necrosis; its diagnosis must always be established in the various complications that may occur in patients with hemodynamic compromise, in order to obtain a timely treatment.

Category: Case Presentation

Received: April 01, 2022
Accepted: April 26, 2022
Published: May 15, 2022

Keywords:

acute esophageal necrosis, esophageal perforation, esophagectomy

***Corresponding author:**

Sandra Castro Boix,

Esophageal and Gastric Surgery Unit. Vall d´Hebron University Hospital. Universitat Autònoma de Barcelona. Passeig de la Vall d'Hebron, 119, 08035. Barcelona, Spain

E-mail: scastro@vhebron.net

Introduction

Acute esophageal necrosis (AEN) is a rare and potentially lethal entity, characterized by superficial circumferential necrosis of variable length, and predominantly affecting the distal esophagus, sparing the gastroesophageal junction [1-6]. Since its first description by Goldberg in 1990, less than 200 cases have been reported in the literature [7,8], with a prevalence that ranges from 0.2% to 10.3% in autopsy series reports [9-11]. It occurs in the absence of caustics or other injurious agents and its pathogenesis is multifactorial, but tissue hypoperfusion secondary to hemodynamic instability or large vessel arterial occlusion play a dominant role [7,8,12-14].

Typically, patients present at the emergency room with signs of upper gastrointestinal hemorrhage such as coffee-ground emesis, melena, or hematemesis, and conservative treatment is usually an acceptable modality [4,12,15]. Among reported complications, esophageal perforation is an acute and severe complication that occurs in less than 7% of patients and can lead to mediastinitis, abscess formation, or pneumomediastinum and requires emergent surgical repair or endoscopic placement of an esophageal stent [1,16,17].

We present a case of esophageal perforation as the first clinical manifestation of acute esophageal necrosis, which occurred after an acute thoracic aortic dissection. This case report follows the SCARE guide related to surgical case reports [18].

To cite this article: Carlos Petrola Chacón, Sandra Castro Boix, Nivardo Rodríguez Conde, Amaia Gantxegi Madina, Mariona Jofra, Daniel Gil Sala, Manel Armengol Carrasco. Esophageal perforation due to acute esophageal necrosis: A case report and a comprehensive literature review. *J Clin Investig Surg*. 2022;7(1):104-108. doi: 10.25083/2559.5555/7.1.18

Case presentation

We present the case of a 46-year-old man with medical history of heavy alcohol and cocaine consumption, active smoker, pharmacologically treated hypertension, and a ruptured celiac trunk aneurysm treated with coil embolization on 2015. The patient presented in the emergency room of a regional hospital with sudden interscapular pain radiating to the central thoracic and epigastric region, being diagnosed of thoracoabdominal aortic dissection (Stanford Type B). He was transferred to our center to start with medical management of the Aortic syndrome in the intensive care unit. 24 hours after admission, the patient presented a clinical worsening with non-controlled thoracic and abdominal pain, dyspnea, and hemodynamic instability. An emergent computed tomography angiography (CTA) oriented to mesenteric malperfusion (congestion of proximal jejunum) due to compression of the superior mesenteric artery ostium because of the aortic dissection. An emergent thoracic endovascular aortic repair (TEVAR) was performed by the vascular surgery team, deploying a thoracic endograft RelayPro® in the proximal descending thoracic aorta covering the entry tear of the aortic dissection (Figure 1). The patient was transferred from the operating room to the ICU hemodynamically unstable, with high requirements of noradrenaline and dependent on invasive mechanical ventilation.

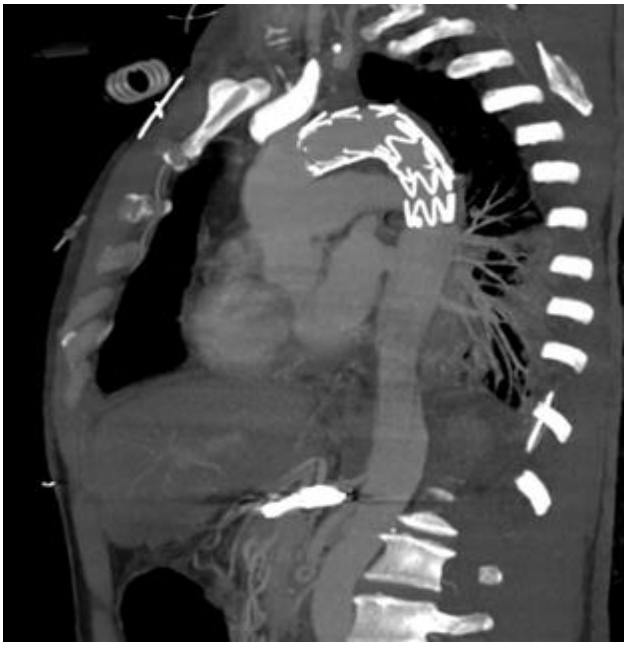

Figure 1. Thoracoabdominal multiphase tomography showing Relay® thoracic stent in situ and celiac trunk embolization coil occlusion.

On the first postoperative day, a CTA was performed, showing an improvement of the true aortic lumen compression and exclusion of the aortic dissection entry tear and pleural bilateral effusion, larger on the left pleural cavity. Due to suspected diagnosis of low-flow mesenteric ischemia, broad-spectrum antibiotic therapy was started and a nasogastric tube for gastrointestinal decompression was placed. Left pleural drainage was inserted to improve the patient's ventilatory mechanics. A biochemical test of the pleural fluid reported high levels of bilirubin. Due to the characteristics of the pleural fluid, methylene blue was administered through the nasogastric tube, objectifying it in pleural drainage. A second CTA was performed to rule out esophageal/gastric perforation, reporting disruption of the left posterolateral wall of the distal esophagus that directly communicated the esophageal lumen with the left pleural cavity, visualizing contrast output through it (Figure 2).

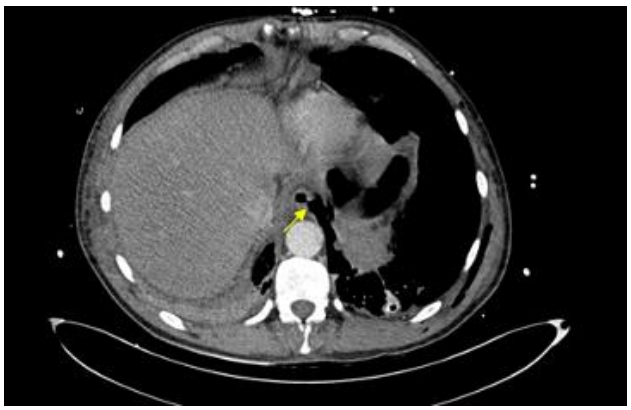

Figure 2. Abdominal multiphase tomography showing esophageal perforation (yellow arrow).

An emergent exploratory laparotomy was carried out by the members of the Esophagogastric Surgery Unit of our center with findings of distal esophageal ischemia (Figure 3) with esophageal perforation to the left pleura and ischemic cholecystitis. A transhiatal esophagectomy (incidental rupture of the gastroesophageal junction occurred), cervical esophagostomy, Witzel-type decompressive gastrostomy, Witzel-type feeding jejunostomy, classical cholecystectomy and drainage of the mediastinum were performed.

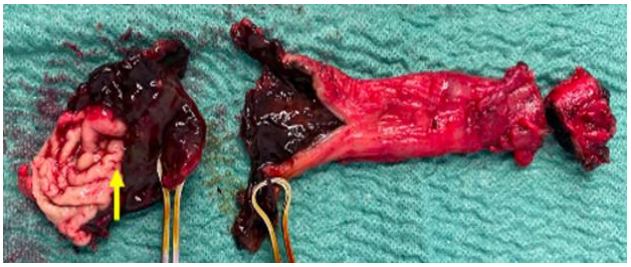

Figure 3. Esophagectomy specimen showing distal esophageal necrosis changes that spare gastroesophageal junction (yellow arrow)

During the postoperative period in the ICU, the patient remained in critical condition, with respiratory failure refractory to treatment and clinical signs of hypoxic encephalopathy. A brain computed tomography was

performed, reporting tonsillar and transtentorial herniation with radiological signs of generalized ischemia of the brain, resulting in the patient´s death.

The histopathological study reported acute transmural esophageal ischemia with wall perforation and acute cholecystitis with extensive necrosis of the gallbladder wall.

Discussion

We report the case of an esophageal perforation as an unusual presentation form of AEN in a patient with history of alcohol and cocaine consumption, hypertension, and previous endovascular embolization of the celiac trunk due to a ruptured aneurysm with recent diagnosis of thoracoabdominal type B aortic dissection treated with a thoracic aortic endograft.

AEN is a condition in which the esophagus, usually the distal portion, develops necrosis of the mucosa [1]. It is a life-threatening condition with a high mortality rate and it is considered to be underreported, in part due to low awareness of this condition as a differential diagnosis of upper gastrointestinal bleeding and in part due to it often being self-resolving [19].

The pathogenesis and etiology of AEN are multifactorial, such as backflow of gastric contents causing esophageal injury, disruption of the vascular supply leading to hypoperfusion and ischemia, and impaired protective barrier systems due to a weakened immune system and/or a hemodynamic instability [20-22]. Another possible hypothesized mechanism of hypotension-induced distal esophageal necrosis is the concept of the 'two-hit' hypothesis; that is, the initial low vascular state event, predisposing the lower esophageal mucosa to severe injury of the esophageal linings by reflux of pepsin and acid, which can then lead to a rapid onset of necrosis unless the circulation is restored timely [23]. To date, ischemia due to low-flow rates or shock is the most widely accepted pathophysiological mechanism, being a fact that argues in favor of an ischemic etiology, the predominance of necrosis in the distal third of the esophagus which is more prone to ischemic injury since this part is less vascularized in anatomical studies and angiographic examinations compared with other parts of the esophagus [4,12,22,24-26]. Minatoya et al. published a case of transmural necrosis of the esophagus secondary to total aortic arch replacement with a Dacron graft due to acute aortic dissection, showing in a postoperative CTA, a complete thrombosis of the false lumen in the descending aorta, and concluded that the feeding artery of the esophagus originated from the false lumen [27].

A wide variety of conditions and risk factors have been associated with AEN. In a systematic review of the literature performed by Abdullah et al. [19] that included 130 cases, the most common associated comorbidities were diabetes in 38%, hypertension in 37%, and alcohol abuse in 25%. Despite the high prevalence of alcohol abuse in these patients, AEN associated with active alcohol drinking is a rare entity, and only ten cases have been reported in the current literature [21]. Alcohol ingestion has been reported to reduce lower esophageal sphincter pressure and esophageal peristalsis and can cause irritation of the gastric mucosa and the accumulation of a large volume of gastric secretions; thus, increased acid reflux, gastric stasis, and decreased mucosal protection are likely to be responsible for the development of AEN associated with alcohol abuse [28-30]. Cocaine use has previously been associated with AEN. Five cases have previously been described in association with cocaine use [31-35]. Cocaine is known to cause serious vasoconstriction, and it is thought that can be a precipitating factor in the development of AEN by compromising the blood supply to the esophagus, especially in predisposed patients [33].

In 70-90% of AEN cases patients present with signs of upper gastrointestinal bleeding associated with symptomatology related to their underlying disorder and signs of sepsis [4,12,15,36,37]. Generally, uncomplicated AEN follows an indolent clinical course with spontaneous resolution and the treatment is mainly supportive and consists of maintaining hemodynamic stability through adequate volemic resuscitation and minimizing acid exposure with intravenous proton pump inhibitors [15,35,38]. Early and late complications can reach up to 11% of the cases and surgical intervention is required in cases where there is massive necrosis with esophageal perforation, and herniated gastric volvulus [15,35].

Perforation of the esophagus, with a reported rate of 7% is one of the most feared complications of AEN [19]. Perforation can be seen in the initial stages of the disease, and it may lead to rapid clinical deterioration, mediastinitis, mediastinal abscess formation, empyema, and generalized sepsis; prompt recognition, intravenous antibiotics, and surgical intervention are life-saving [12,15]. Esophagectomy, decortication, lavage, and delayed reconstruction may be performed; primary closure of the perforated esophageal tissue should not be attempted [12,15].

Overall AEN mortality rates range from 13% to 36% and are largely due to the older age and underlying disease [12,15,24,37]. However, mortality specific to the AEN is 6%, and known risk factors include esophageal perforation, diabetic ketoacidosis, and compromised immune system [1,12,15].

The strength of our work lies in the fact that we present an unusual form of presentation of AEN, such as esophageal perforation (7% of cases), providing clinical evidence that allows this complication to be considered in the future as a form of presentation in patients with risk factors and diagnostic suspicion of AEN.

Conclusions

Acute esophageal necrosis is a serious clinical entity that can initially present complicated with esophageal perforation, which is a life-threatening condition. With multifactorial pathogenesis and ischemia as the fundamental mechanism, its diagnosis must always be present in the variety of complications that can occur in patients with hemodynamic compromise to achieve timely treatment. Being an infrequent entity, there is still no scientific evidence necessary to generate guidelines for its diagnostic algorithm and therapeutic protocol and most recommendations are based on single-center experiences.

Conflict of interest disclosure

There are no known conflicts of interest in the publication of this article. The manuscript was read and approved by all authors.

Compliance with ethical standards

Any aspect of the work covered in this manuscript has been conducted with the ethical approval of all relevant bodies and that such approvals are acknowledged within the manuscript.

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