CASE REPORT

A Case of Uncommon Hematemesis from Acute Esophageal Necrosis

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ABSTRACT

Acute esophageal necrosis (AEN) is a rare and severe condition characterized by extensive tissue necrosis of the esophagus. This condition predominantly affects the distal part of the esophagus. Reports estimate the prevalence of AEN to be up to 0.2% in autopsy studies, while endoscopic series report a prevalence ranging from 0.01% to 0.28% of cases. It primarily affects patients with poor nutritional health and multiple comorbidities often as a result of underlying systemic condition. Patients present with upper gastrointestinal bleeding, such a hematemesis and melena. We report a case of AEN in a female patient who had presented with hematemesis.

INTRODUCTION

Acute esophageal necrosis (AEN) is a rare and severe condition characterized by extensive tissue death of the esophagus, which results in a striking, diffuse blackened appearance of the esophageal lining. This condition predominantly affects the distal part of the esophagus and is often associated with significant inflammation and potential for tissue destruction.[1] Reports estimate the prevalence of AEN to be up to 0.2% in autopsy studies, while endoscopic series report a prevalence ranging from 0.01% to 0.28% of cases.[2,3] Men are over four times more likely to develop this condition than women, with the average age of diagnosis being 68 years.[1,2,3] It primarily affects patients with poor nutritional health and multiple comorbidities often as a result of underlying systemic condition.[4,5] AEN is also linked to malignant tumors in about 10% of cases and has been reported in patients experiencing toxicity due to chemotherapy.[6] Around 70% of patients present with upper gastrointestinal bleeding, such a hematemesis and melena which can develop rapidly, often within 18 hours of the triggering event.[2,3,4] Other gastrointestinal symptoms may include dysphagia, epigastric pain, and chest pain. We report a case of acute esophageal necrosis in a female patient who had presented with hematemesis.

CASE REPORT

A 52-year-old woman diagnosed with a Myeloproliferative Syndrome was hospitalized in the Hematology Clinic referred to our Endoscopy unit for urgent Esophagogastroduodenoscopy (EGD) due to a primary complaint of hematemesis. Upon arrival, she was hypotensive with low SpO2 levels. Initial laboratory tests revealed a hemoglobin level of 8.0 g/dL, hematocrit of 24.2%, mean corpuscular volume (MCV) of 73.2, leukocytes at 116,000/μL, and thrombocytes at 64,000/μL. During the gastroscopic examination, circumferential necrosis of the esophageal mucosa was observed, presenting a darkened appearance extending to the distal esophagus. Three haemorrhagic lesions were identified in this region. The stomach mucosa appeared hyperaemic and edematous. with hemosiderin-stained blood, alongside two ulcerative lesions were noted without active bleeding (Figures 1 and 2). Haemostasis was achieved through infiltration using diluted adrenaline (1:10,000) and placement of hemoclips on all three haemorrhagic esophageal lesions. Biopsies were not taken due to active bleeding and unstable patient condition. The patient was transferred to the Gastroenterology Clinic for continued care, where she was administered parenteral proton pump

inhibitors (PPI) antibiotics (due to the risk of infection/ sepsis), mucoprotective agents, and received fresh blood and plasma as indicated by her hemogram and clinical status. Laboratory parameters, including acid-base status, were monitored regularly. Her medical history included treatment with Imatinib for her myeloproliferative syndrome, type 2 diabetes mellitus was managed with insulin therapy, palliative splenectomy six months prior, and hysterectomy with adnexectomy two years earlier. Over the following days, the patient showed no clinical or laboratory evidence of recurrent bleeding. Consequently, preparations were made for her transfer back to the Haematology Clinic for further management of her hematologic condition. A follow-up endoscopy was recommended to monitor for any potential late esophageal complications.

DISCUSSION

The management of AEN is guided primarily by clinical experience due to limited evidence-based recommendations.

Laboratory findings in AEN are typically nonspecific and reflect the underlying condition, such as lactic acidosis, hypoalbuminemia, anaemia, renal insufficiency, and hyperglycaemia.

Esophagogastroduodenoscopy (EGD) is the preferred diagnostic tool for AEN. It reveals characteristic findings such as circumferential black discoloration of the esophagus, predominantly in the distal segment, with clear demarcation from unaffected proximal tissue.[4] The endoscopic appearance can vary widely. In patients with an uncertain diagnosis, esophageal biopsies can be useful due to their characteristic histologic findings.[10] Findings may include active bleeding or blood clots in the esophagus, "coffee ground" material in the stomach, and esophageal or gastric ulcers may also be observed.[1] Submucosal adrenaline injections are commonly used to control bleeding.[7] In our case we decided to put also hemoclips, in absence of other modalities to archive haemostasis. Treatment focuses on addressing underlying clinical conditions, stabilizing hemodynamics, maintaining nil per os (NPO) status, and providing supportive care including blood transfusions and intravenous PPI to minimize further esophageal tissue damage. In most cases, patients respond well to conservative management, though a high mortality rate (approximately 32%) has been reported, primarily attributable to underlying comorbidities.[1,8] Prognostic factors for mortality include advanced age, elevated pulse rate, low hemoglobin, and hypoalbuminemia at presentation.[9] Acute esophageal necrosis is now increasingly recognized as a spectrum ranging from reversible superficial ischemia limited to the mucosa, as likely occurred in our case, to severe transmural necrosis associated with perforation, mediastinitis, and death. While no formal staging system currently exists, this case likely represents an earlier stage of AEN, with prompt endoscopic hemostasis and supportive care preventing progression to more fulminant forms. A classification system reflecting the graded nature of ischemic injury to the esophagus could help guide clinical decision-making and prognostication.

Further research is needed to refine diagnostic and therapeutic strategies and improve outcomes for patients with AEN.

CONCLUSION

AEN is a rare but significant condition that should be considered as a differential diagnosis in patients with multiple comorbidities who present with upper gastrointestinal bleeding. The condition spans a spectrum from superficial mucosal ischemia to full-thickness necrosis, and timely identification of its stage is essential. Early identification through endoscopy and timely intervention can improve patient outcomes.

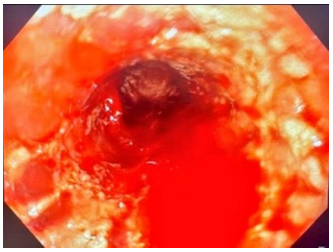

Figure 1 circumferential necrosis of the esophageal mucosa

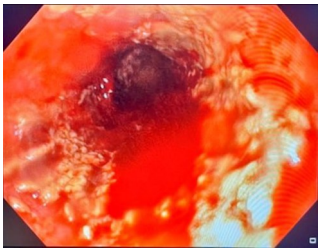

Figure 2 circumferential necrosis of the oesophageal mucosa

CONFLICT OF INTEREST

No conflict of interest from all authors!

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