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Case Report

A rare cause of acute esophageal necrosis: A case report ☆,☆☆

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ABSTRACT

Acute esophageal necrosis (AEN) or black esophagus is a rare entity characterized by diffuse circumferential black pigmentation of the esophageal mucosa due to ischemic necrosis. It may be lethal, especially among elderly patients with multiple comorbidities and hemodynamic instability. Diagnosis is based on gastroscopy. Treatment consists of intravenous fluids, proton pump inhibitors, and additional therapies to treat the underlying illness. We report a rare case of a woman in her 50s with cervical cancer who presented with hematemesis and sepsis. Upper gastrointestinal endoscopy showed a black esophagus and an ulcerobudding duodenal process. Few days later, she developed abdominal distension with diffuse pain. Abdominal CT scan demonstrated perforation of gastroduodenal tumor. The treatment was based on resuscitation, proton pump inhibitors, antibiotics, and surgery of the perforated tumor. Unfortunately, the patient died 2 days later because of septic shock. The black esophagus is a fatal complication, thus diagnosis at an early stage and timely management may improve survival. This is the first case reported of AEN due to perforated duodenal tumor explained by septic shock leading to an ischemic esophageal injury.

Introduction

Acute esophageal necrosis (AEN) is an uncommon disease. The pathogenesis of AEN is multifactorial. The etiology includes a combined effect of ischemic injury, impaired esophageal mucosal defense systems, and backflow injury from gastric contents. Herein, we report a rare case of black esophagus due to a perforated gastroduodenal tumor and septic shock.

Abbreviations: AEN, Acute esophageal necrosis; CT scan, computed tomography scan; CRP, C Reactive Protein.

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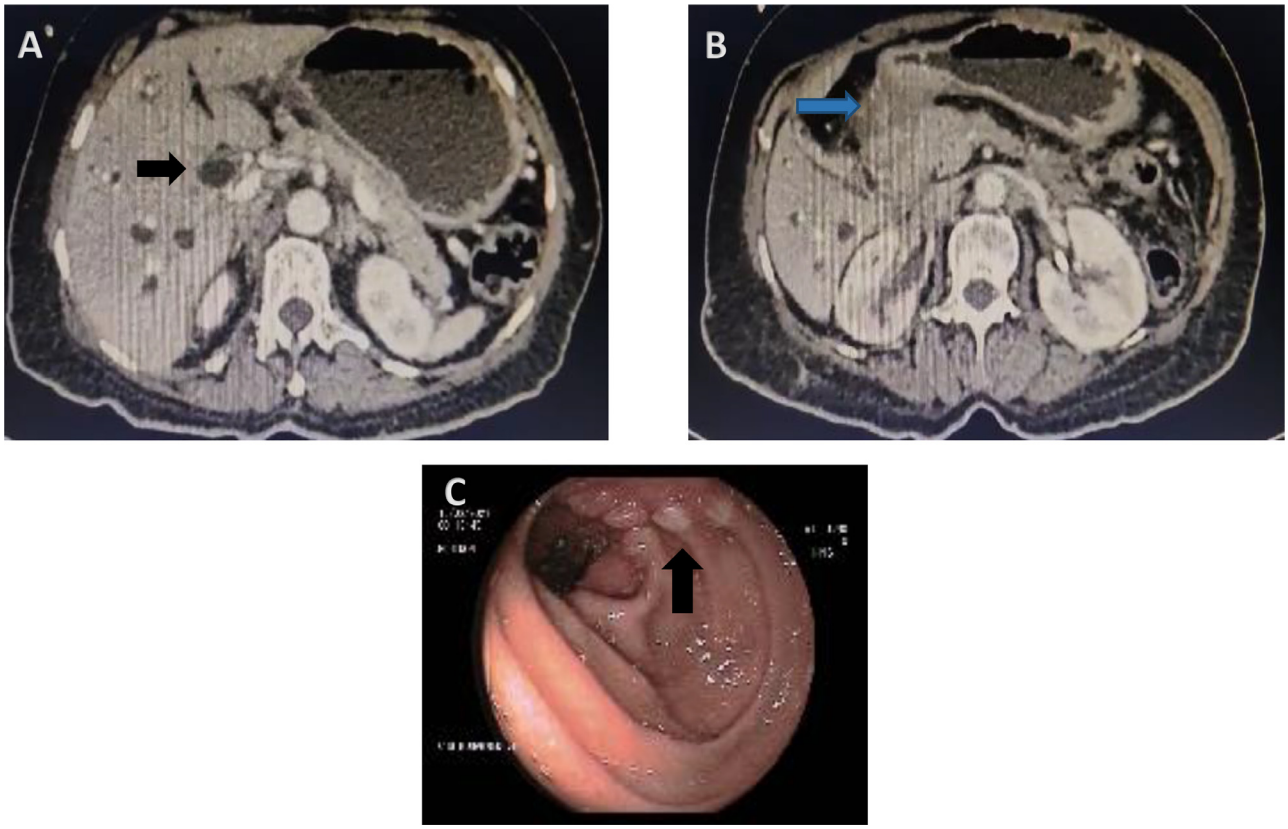

Fig. 1 – Radiology and endoscopic imaging of duodenal mass, (A) Abdominal CT scan dilatation of intrahepatic and extrahepatic bile ducts (arrow); (B) Contrast-enhanced abdominal CT scan showing irregular gastric wall thickening extending to the second part of duodenum (arrow); (C) Gastroscopy showing ulcero-budding duodenal process (arrow).

Case presentation

A woman in her 50s with a history of stage IVa cervical cancer treated by radiochemotherapy and nephrostomy for bilateral ureterohydronephrosis with a good outcome. The patient presented to the emergency room with jaundice, epigastric pain, nausea/vomiting, and deterioration of her general condition for the last 20 days. On clinical examination, she was icteric, and denutrished, and her performance status was 3 with epigastric tenderness. Laboratory tests revealed a total Bilirubin level of 11.40 mg/dL (reference range: 0.1-1.2 mg/dL), an elevated C-reactive protein (CRP) at 36 mg/dL (normal range: $<$ 0.3 mg/dL), and a high level of 19-9 CA at 233 UI/mL (normal range: $<$ 37 U/mL). A CT scan showed dilation of the intrahepatic bile duct and common bile in its hilar portion (Fig. 1A) upstream of an infiltrating process in the antropyloric region and second part of duodenum, measuring 41 $\times$ 26 mm and extending to 37 mm associated with peritoneal carcinomatosis (Fig. 1B). Then, the first line treatment with antibiotics was started to treat acute cholangitis grade II. Gastroscopy revealed an ulcerobudding duodenal process difficult to cross, extending to the pylorus (Fig. 1C). Biopsy was in favor of nonspecific subacute interstitial duodenitis. A novel gastroscopy with biopsy should be repeated. Meanwhile, the

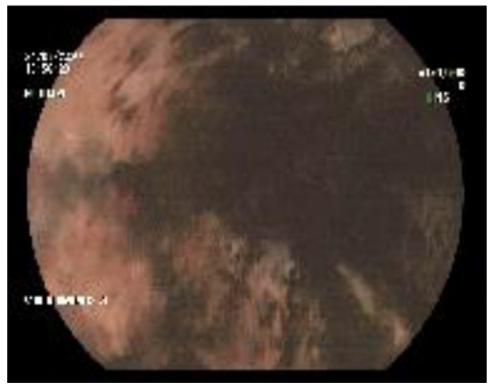

Fig. 2 – Upper gastrointestinal endoscopy shows a black esophageal.

patient presented a few days later with abdominal pain and hematemesis. Laboratory analysis revealed hemoglobin of 10 g/dL decreased to 5 g/dL and an increase in inflammatory markers, accompanied by a positive blood culture for *Escherichia coli* sensitive to third-generation ceftriaxone. After blood transfusion, gastroscopy was done showing a circum-

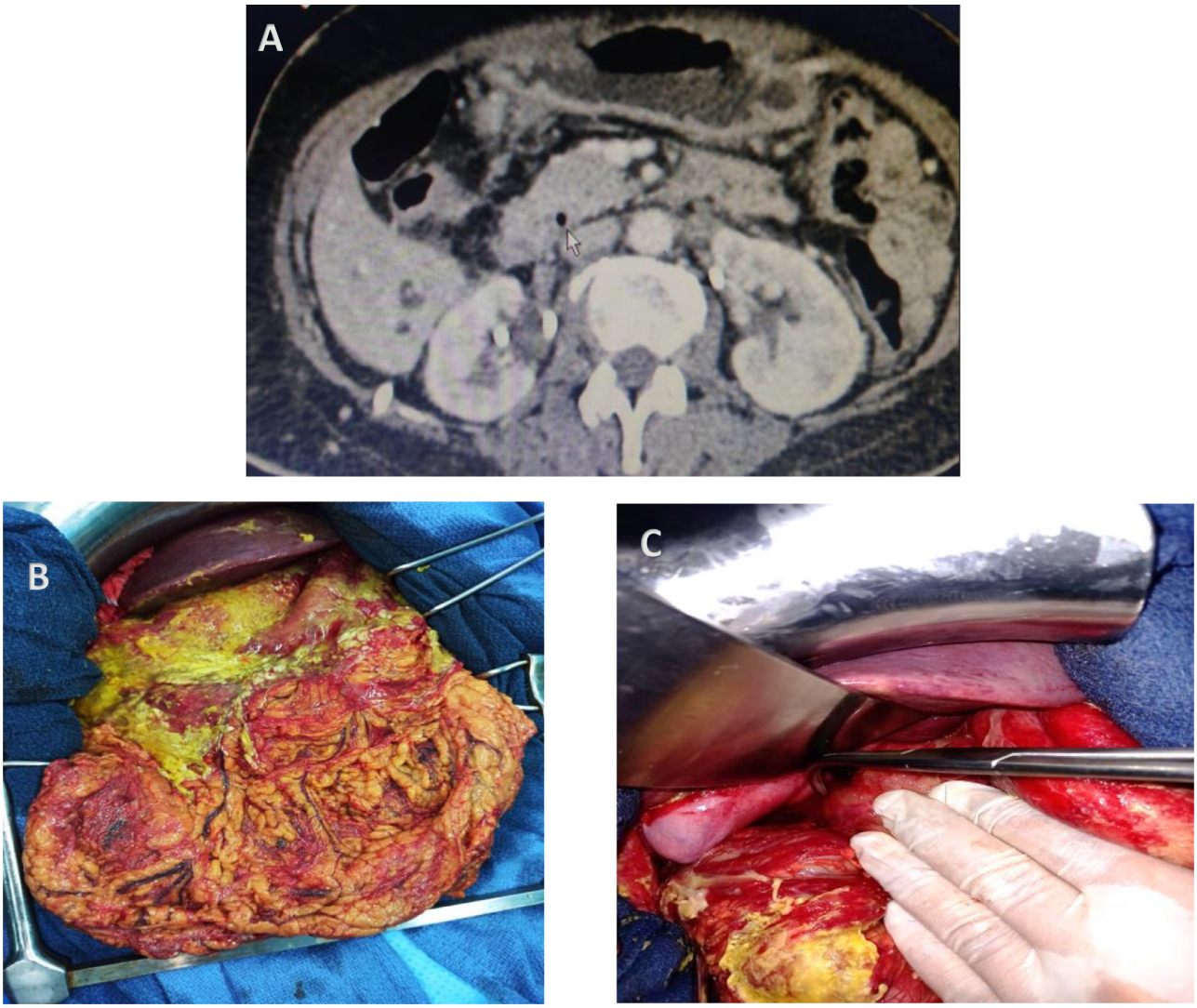

Fig. 3 – Perforation of gastroduodenal tumor; (A) Abdominal CT scan revealing a perforated gastroduodenal. tumor (arrow). (B) Purulent general peritonitis after an exploratory laparotomy. (C) Exploratory laparotomy showing a perforation in the second part of duodenum.

ferential black-colored mucosa from the Killian mouth to the lower esophagus that was fragile and bleeding on contact reason why the exam was interrupted (Fig. 2). After this, our patient developed an abdominal distension with diffuse abdominal pain and conscious disorder. Physical examination found a confused patient with hypotension tachycardia and abdominal contracture. The blood tests revealed an elevated white blood cell count of 15000/m3 (reference range between 4,000 and 10,000/mm^3) and an increase of CRP level at 300 mg/dL. An emergent abdomen computed tomography was performed demonstrating acute generalized peritonitis on perforated gastroduodenal tumor (Fig. 3A). Therefore, the patient underwent surgery, and a perforated tumor in the second part of the duodenum was diagnosed (Figs. 3B and C) and repaired with drainage, cleaning, and feeding jejunostomy. The patient was resuscitated with intravenous fluids, vasoactive drugs, 3 blood transfusions, antibiotics, and proton pump inhibitors, but she died 2 days later because of septic shock.

Discussion

AEN is a rare medical disorder characterized by diffuse, circumferential, black mucosal pigmentation of the esophagus found on gastroscopy with an abrupt transition to normal mucosa [1]. The prevalence of this rare disease varies between 0.06 and 0.28% in the literature [2]. It is more frequent in elderly men with comorbidities such as diabetes mellitus, hypertension, kidney disease, alcoholism, malnutrition, and cardiovascular diseases [2].

The etiology of AEN is multifactorial; esophageal hypoperfusion with ischemia, an impaired mucosal defense barrier, and injury related to reflux of gastric contents are the principal hypotheses that explain the pathogenesis of this disease [1]. In our case, we suggest that perforated gastroduodenal tumors complicated with sepsis and hypovolemia are the main risk factors for the emergence of acute esophageal necrosis.

In most case studies, the clinical presentation of AEN ranges from an asymptomatic state to a wide spectrum of clinical manifestations. The main symptom is hematemesis (85%), according to a recent systematic review published in 2020 [3]. Other symptoms, such as syncope/hypotension, acute abdominal pain, fever/infection, dysphagia, anemia, acute pancreatitis, or multiorgan failure due to sepsis, have been reported in some cases [4]. Our patient presented with a variety of symptoms, especially hematemesis, anemia, and signs of sepsis.

The diagnosis of AEN is made by gastroscopy identifying diffuse circumferential black discoloration of the esophagus mucosa with abrupt demarcation at the Z-line; rarely, upper endoscopy shows stricture, tracheoesophageal fistula or perforation of the esophagus as complications of AEN [5]. Histological examination is not required for the diagnosis; meanwhile, it allows differentiation between AEN and esophageal pigmentation due to mucosal melanosis, pseudomembrane esophagitis, acanthosis nigricans, ingestion of coal dust or dye, and ruling out other etiologies such as malignancy, Helicobacter pylori, cytomegalovirus, herpes simplex virus or fungal infection [6].

The mainstay of management of AEN is supportive measures by resuscitation with intravenous fluids, blood transfusion, and treatment of underlying illness [6]. Parenteral nutrition may be needed in malnourished patients to improve their nutritional status and expedite healing, while nasogastric tubes should not be used to avoid perforation [5]. Medical management of AEN includes proton pump inhibitor infusion to control gastric acidity and protect against further upper gastrointestinal bleeding, which should be continued to prevent stricture formation [5,7]. Empiric antibiotics should be initiated when esophageal perforation and unexplained fevers are suspected in immunocompromised patients [4]. Following the results of esophageal cultures and biopsy specimens; specific antimicrobial therapy should be adapted [2]. Surgical intervention in patients with AEN is reserved for perforated esophagus with resultant mediastinitis and abscess formation [8].

In a more recent study, the mortality rate was 12.5% and was related especially to patient characteristics such as serious medical history, older age, and higher incidence of malignancy. The death of our patient was mainly caused by the underlying disease and septic shock.

Another particularity of our case was the perforation of a duodenal tumor, which is a rare malignant tumor, accounting for less than 1% of all gastrointestinal neoplasms [3]. Despite the negative histological result, the initial endoscopic and imaging findings, along with the results of the exploratory laparotomy, indicated the existence of a perforated duodenal tumor. This tumor was identified as the primary cause of septic shock, AEN, and the subsequent fatality of our patient, despite the implementation of both surgical and various medical treatments.

Conclusion

AEN is an uncommon disorder characterized by a poor prognosis with a high mortality rate dependent on the severity and extent of esophageal involvement, as well as the underlying disease. This lethal case of AEN, attributed to a perforated gastroduodenal tumor, underscores the association with septic shock and ischemic esophageal injury. The primary objective of this clinical case is to encourage physicians to consider AEN as a potential diagnosis when confronted with a confluence of sepsis and hematemesis. Early recognition of this correlation offers the potential for accelerated diagnoses and, ultimately, enhanced rates of survival.

Patient consent

The daughter of the patient provided written informed consent for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Ethics approval and consent to participate

Not applicable.

Authors contributions

KR cared for the patient and write the manuscript. AD, RJ contributed in the surgical management of this case. OE, HK, AZ, ZI contributed to the patient management. GK read and approved the final version of the case. All the authors gave final approval of the manuscript.

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