Black oesophagus, upside-down stomach and cameron lesions: cascade effects of a large hiatal hernia

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SUMMARY

Acute oesophageal necrosis, black oesophagus (BE) or Gurvits syndrome (GS) is a rare form of severe oesophagitis appearing as a striking circumferential discolouration of distal mucosa with various proximal extensions abruptly terminating at the gastro-oesophageal junction. It is most commonly associated with acute exacerbations of medical comorbidities, while associations with altered gut anatomy are rare. We present a unique constellation of BE, Cameron ulcers (CU), and gastric volvulus from a large paraesophageal hiatal hernia. Our patient recently recovered from COVID-19 and was malnourished and frail, while the expanding paraesophageal hiatal hernia turned into an acute organoaxial gastric volvulus with accompanying outlet obstruction. In low-flow post-COVID coagulopathic states, compensatory mechanisms may lack against gastric stunning and sudden massive reflux on the oesophagus. We additionally performed a systematic review and discovered additional cases with coexistent volvulus and paraesophageal hernia, although there are no previous reports of BE with CU, which makes this study the first.

BACKGROUND

Acute oesophageal necrosis (AEN), black oesophagus (BE) or Gurvits syndrome (GS) is a rare but emerging cause of gastrointestinal (GI) bleeding first described in autopsy studies in the pre-endoscopic era.[1] However, its incidence has increased over the past two decades as case recognition, reporting and understanding have grown. It presents on endoscopy as striking black circumferential distal oesophageal mucosal discolouration with various proximal extensions and abrupt cessation at the gastro-oesophageal junction (GEJ), often seen in acutely ill patients.[2] The aetiology of BE is multifactorial—a combination of tissue hypoperfusion, massive reflux of gastric contents, and compromised local mucosal barriers. Diagnosis is endoscopic and does not require tissue sampling. Associated medical conditions may include sepsis, cardiovascular disease, diabetes mellitus with ketoacidosis, thromboembolic phenomena, alcohol abuse, oncologic disorders and malnourishment. Alterations in foregut anatomy seen in the setting of significant hiatal hernia may play a role in the pathogenesis of the syndrome. Mucosal recovery is common when underlying derangement is identified and corrected, while post-endoscopy care and surveillance schedules are individualised when strictures or stenosis complicate oesophageal recovery. All-cause mortality nears 32% due to comorbid medical conditions, but fatality specific to AEN is much lower at around 6%.[3] Our case represents an unusual case of AEN in post-COVID coagulopathy, altered local circulation, and outlet obstruction in the setting of a large volvulating paraesophageal hernia in a malnourished patient. We additionally performed a systematic search to parse out unique features of our case and define the common pathogenesis of this syndrome in such a setting.

CASE PRESENTATION

A 77-year-old woman was presented to our emergency department with abdominal pain, multiple emesis episodes and transient loss of consciousness. Her medical history included chronic obstructive pulmonary disease, gastro-oesophageal reflux disease, peptic ulcer disease, hiatal hernia, malnutrition and recent COVID-19 infection. She was haemodynamically stable but ill-appearing with diffuse tenderness on abdominal palpation. Laboratory analysis showed a remarkable count for white blood cell count at 18.5 k/μL, haemoglobin at 9.8 g/dL, serum creatinine at 1.4 mg/dL and serum albumin at 3.3 g/dL. A CT scan of the abdomen revealed large paraesophageal hiatal with organoaxial volvulation and distal oesophageal distention (figures 1 and 2). She was made nil-per-os, resuscitated with intravenous ringers lactate solution and started on continuous intravenous pantoprazole infusion. An oesophagogastroduodenoscopy revealed a large hiatal hernia, CU and necrotic discolouration of the mid-to-lower third of the oesophagus with an abrupt transition at the gastro-oesophageal junction (figures 3 and 4). The patient declined surgical intervention to correct the hernia and volvulus. She was eventually discharged home in stable condition tolerating oral intake with hospice services.

OUTCOME AND FOLLOW-UP

Despite eventual weaning off mechanical ventilation and advance on an oral diet, the patient declined surgical intervention for correction of hiatal hernia and was eventually discharged home in stable condition tolerating oral intake. She was found to be well after her 6 month follow-up but refused repeat endoscopy and surgical intervention.

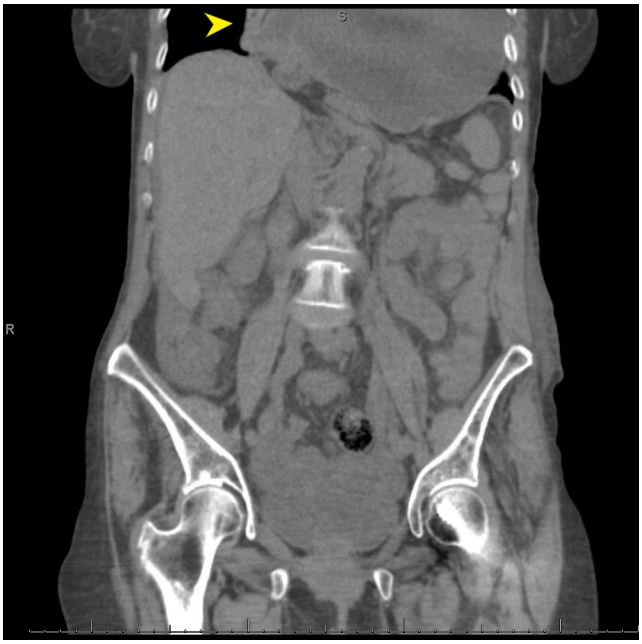

Figure 1 CT abdomen reveals complete herniation of the stomach into the posterior mediastinum (arrows).

DISCUSSION

The incidence of AEN or GS ranges between 0.01% and 0.30%, although these estimates are under-reported as autopsy studies were the primary method of diagnosis in the pre-endoscopy era. At the same time, sicker patients at the highest risk for AEN would often forgo or delay endoscopy due to their tenuous states.[2 3] Gurvits staging system was introduced in 2007 to describe the natural history of AEN from the endoscopic and histopathologic analyses[2], subdividing it into black appearing mucosa with tissue necrosis in Stage 1, transition changes in Stage 2 and normal mucosal appearance with microscopically present granulation tissue in Stage 3. Rapid recovery from Stage 1 may be seen as early as the first week of diagnosis[3] and effectively underestimate AEN incidence. The diagnosis of GS requires the exclusion of pseudomelanosis, malignant melanoma and caustic injuries.[4] On performing a systematic search of other similar presentations, only a handful of AEN cases with a concurrent hernia and/or gastric volvulus have been reported.[5–10] Our study is the first to report the case of AEN associated with severe mechanical features of a large hiatal hernia, including CU. These constellations of findings are unique and may imply a common underlying pathophysiological mechanism that stems from large hiatal hernias and their effect on the surrounding structures.[11 12]

The occurence of necrosis seen in AEN is thought to be a consequence of multiple derangements in the setting of an acute illness and long-standing comorbidities. [2 3]An important factor is a sudden state of hypoperfusion or vascular compromise that preferentially affects watershed areas of the distal oesophagus with various proximal extension with colloquial hindering mucosal regeneration and the inability to protect

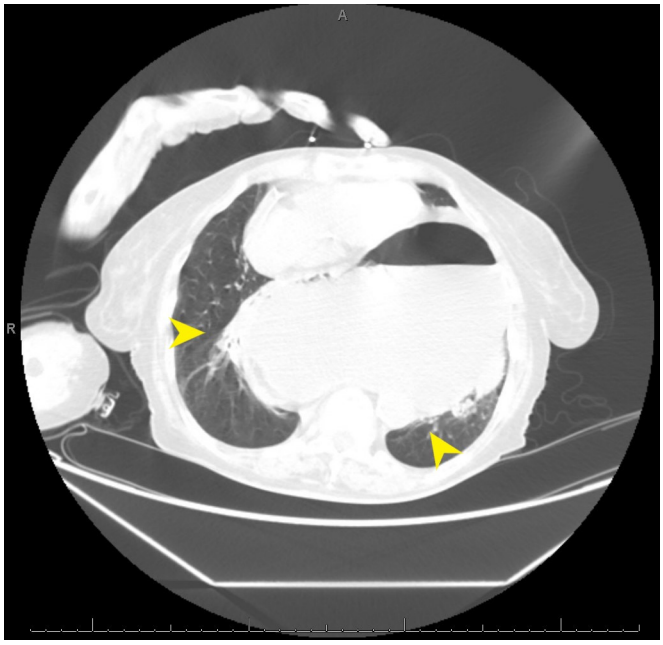

Figure 2 CT abdomen reveals herniation of the stomach into the posterior mediastinum (arrows), oesophageal dilatation, and gastric distension.

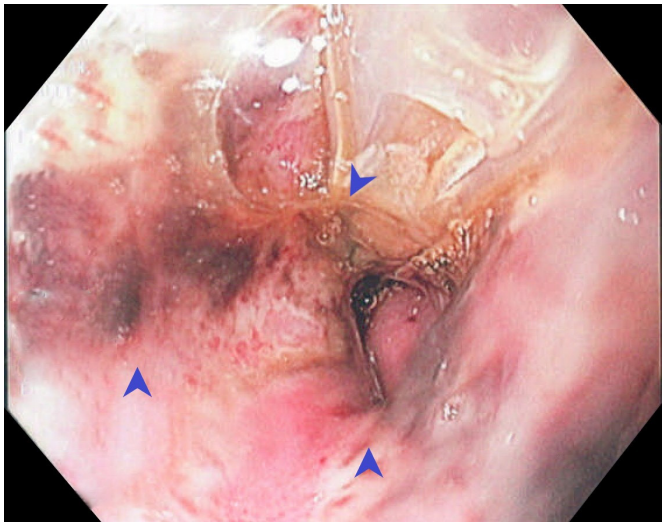

Figure 3 Esophagogastroduodenoscopy (EGD) demonstrating necrotic lesions along the oesophageal lumen (arrowheads).

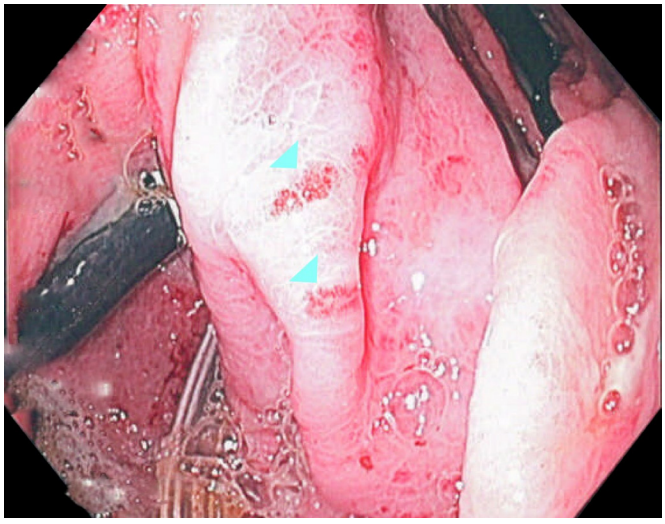

Figure 4 Esophagogastroduodenoscopy (EGD) demonstrating cameron ulcers (arrowheads).

Table 1 Summary of previous cases of acute oesophageal necrosis (AEN) associated with gastric volvulus and hiatal hernias medical literature

Author/Year	Article type	Presenting symptoms	Gastric volvulus	Hiatal hernia	Segment affected	Management	Outcomes
Kram *et al*[5] 2000	Full manuscript	Coffee-ground emesis	✓	✓	Distal	Laparotomy and oesophagectomy	Full recovery and alive
Hwang[16] 2007	Full manuscript	Coffee-ground emesis	X	✓	Pan-oesophageal	Laparoscopy, hernia repair and percutaneous endoscopic gastrostomy (PEG)	Alive and removal of PEG and resolution of acute oesophageal necrosis (AEN) on repeat endoscopy
Matsumoto *et al*[6] 2009	Full manuscript	Vomiting and epigastric pain	✓	✓	Distal	Laparotomy, oesophagectomy and total gastrectomy	Alive and oesophageal reconstruction
Garas *et al*[17] 2011	Full manuscript	epigastric pain	✓	✓	Distal	Esophagectomy with gastric pull-up	Multiorgan failure and death
Mclaughlin[18] 2011	Full manuscript	Coffee-ground emesis, nausea, and chest pain	X	✓	Middle-distal	Nissen fundoplication	Alive with AEN resolution on repeat endoscopy
Nunes *et al*[19] 2017	Case Image	Coffee-ground emesis and epigastric pain	✓	✓	Middle-distal	Laparotomy and Nissen fundoplication	Alive and discharged without any complications
Moore *et al*[9] 2018	Full manuscript	Nausea and abdominal pain	✓	X*	Middle-distal	Laparotomy with volvulating laparoscopic band removal	Discharge and AEN resolution on repeat endoscopy
Chowdhury[7] 2019	Abstract	Dysphagia	✓	✓	Middle-distal	Laparoscopy and PEG	Alive and discharge

*volvulus secondary to slipped laparoscopic gastric band

against massive reflux of gastric contents. In our patient, various mechanisms can account for AEN, including post-COVID hypercoagulable state, compromise to the blood flow to the lower oesophagus traversing through the lesser gastric curvature during gastric rotation and increase in esophageal intraluminal pressure on the mucosal and submucosal vessels with effective compression.[13] Gastric outlet obstruction, partial or complete, as seen in the gastric volvulation, leads to massive reflux of corrosive secretions to the distal esophagus overwhelming local mucosal barriers and local buffering systems that were already relatively compromised in this chronically malnourished elderly patient. Interestingly, under normal circumstances, the oesophagus may compensate for this sudden reflux by increasing blood flow, but this compensatory mechanism is lost in a coexistent low-flow state.[4]

A laparoscopic transabdominal repair was proposed to our patient; however, this was declined by her. Traditionally, surgery can be approached from the thorax or abdomen, and although no head-tohead studies exist—laparoscopic transabdominal is increasingly used in practice as thoracic approaches can be challenging. Inpatient sample studies have demonstrated lower readmission, reoperation and complication rates in laparoscopic approaches.[14 15]

Increased recognition of AEN in the 20th century has led to its ascent as the fourth leading cause of acute haemorrhage from the upper intestinal tract and a valid differential for GI bleeding.[1] Among known cases of GS, a direct effect from alteration of upper GI anatomy is rare; its management and prognosis are less defined, placing importance on case reports and nonrandomised studies to increase awareness (table 1). Future explorations may provide additional insight on pathogenesis and management with a clear goal of improving morbidity and mortality of such patients with AEN.

Learning points

- Acute esophageal necrosis (AEN) is gaining recognition as an emergent cause of upper gastrointestinal hemorrhage in patients presenting for hospital care.
- Alteration in gastrointestinal anatomy, including paraesophageal hiatal hernias, are rare but essential causes of AEN.
- AEN in the setting of a large paraesophageal hiatal hernia may represent regional compression of distal esophageal vasculature.
- Gastric volvulation may lead to massive reflux of corrosive secretions to the distal esophagus, contributing to AEN development.
- Future studies can explore AEN incidence within cases of altered gut anatomy.

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