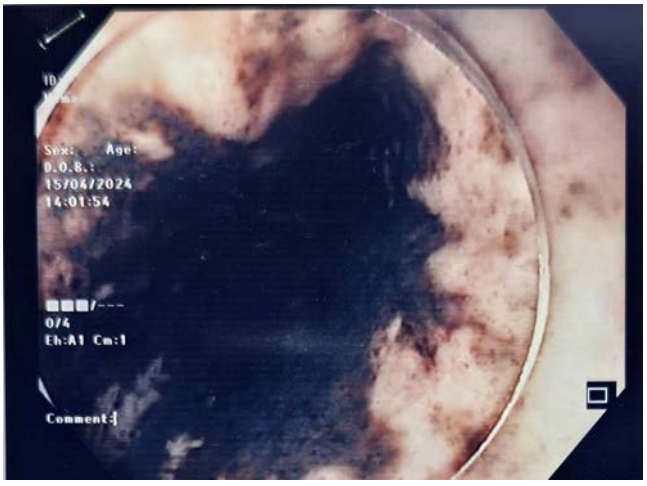
A dark and black esophagus

Flavio Scarilli, Davide Tizzani

Emergency Medicine 2, Ospedale San Giovanni Bosco, Torino, Italy



A 70-year-old Italian man presented to our emergency department for dyspnea and confusion. He was affected by diabetes mellitus type 2, but he stopped his medication 10 days before. At admission, his vital signs were blood pressure of 60/50 mmHg, heart rate of 130 bpm, peripheral oxygen saturation of 94% in the Venturi Mask FiO_2 40%, respiratory rate of 40/min, and body temperature of 36°C. Physical examination revealed hypoperfusion with a high Mottling Score and dark stools with a positive hydrogen peroxide reaction. He denied abdominal pain, nausea, and vomiting. Laboratory findings showed leucocytosis (White Blood Cells, WBC, 31,580/mm^3; N 91.6%), Hemoglobin (Hb) 14.5 g/dL, Hematocrit Test (Hct) 49%, hyperglycaemia (>700 mg/L), and normal coagulation time. Arterial blood gas documented a metabolic acidosis with pH 6.95, Partial Pressure of Carbon Dioxide (pCO2) 27.5 mmHg, HCO3 7.1 mmol/L, lactate 8.14 mmol/L (normal value <2), and elevated anion gap metabolic. He was first treated with IV therapy as follows omeprazole 80 mg, tranexamic acid 1 g, Ringer acetate 1000 cc, sodium bicarbonate 8.4% 100 mL, and magnesium sulfate 2 g. Then, a continuous IV infusion of Ringer 150 mL/h and omeprazole 8 mg/h was started. An Esophagogastroduodenoscopy (EGD) was arranged and showed circumferential blackening of the distal half of the esophagus.

Question

What disorder arises with this dark and black esophagus?

1. Caustic ingestion
2. Melanoma
3. Gurvits syndrome
4. Acanthosis nigricans

Correspondence: Flavio Scarilli, Emergency Medicine 2, Ospedale San Giovanni Bosco, Torino, Italy.
E-mail: flavio19sx@gmail.com

Key words: black esophagus, Gurvits, ketoacidosis, diabetes, AEN.

Contributions: all the authors approved the final version of the manuscript, agreed to be held accountable for all aspects, and equally contributed to the work.

Conflicts of interest: the authors declare no potential conflict of interest.

Funding: none.

Availability of data and materials: all data underlying the findings are fully available upon reasonable request to the Corresponding Author.

Ethics approval and consent to participate: not necessary. The patient provided consent for access to medical records at the time of admission.

Received: 21 June 2024.
Accepted: 3 July 2024.
Early view: 29 July 2024.

Emergency Care Journal 2024; 20:12738
doi:10.4081/ecj.2023.12738

Answer

The correct diagnosis is Gurvits Syndrome also known as Acute Esophageal Necrosis (AEN) or "black esophagus" due to the associated colour change.[1] Firstly described in 1990,[2] AEN is a rare syndrome with a male preponderance, with an incidence four times greater than that for women and a peak incidence during the sixth decade of life.[3] The prevalence is only 0.001~0.2%, while its mortality rate is up to 32%.[4] Most of the cases are fatal due to perforation, stricture, bleeding, and infections. The etiology is not completely clear. According to a "2 hit hypothesis", AEN can arise when an acute event occurs and causes local hypoperfusion and subsequently necrosis.[5] Combined effects of vascular compromise, impaired defence mechanism of the esophagus, and exposure to gastric acids all contribute to AEN.[6] The major risk factors include male sex, chronic diseases (diabetes mellitus with, hyper-

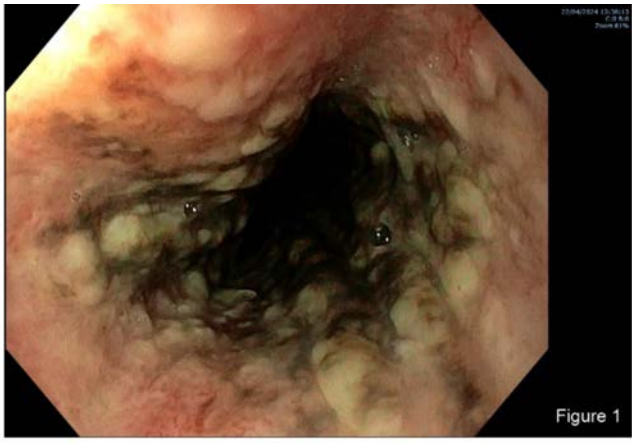


Figure 1. EGD showing a lighter esophageal mucosa with some white exudate spots.

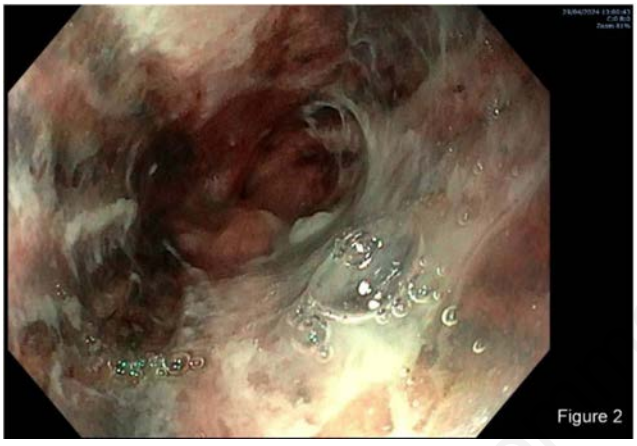


Figure 2. EGD documenting fibrin deposition associated with reepithelialisation.

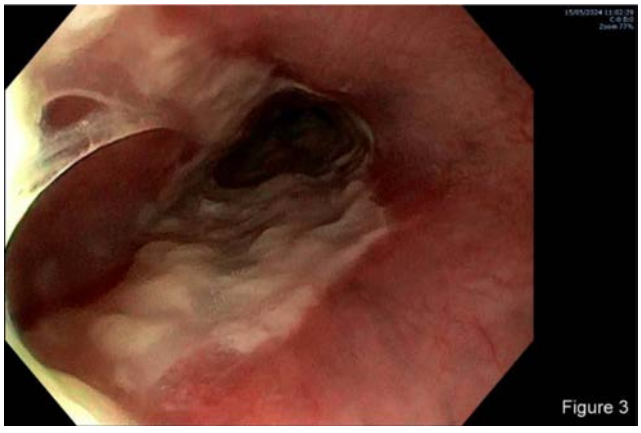


Figure 3. EGD after two weeks showing total resolution of AEN with no indication of stricture development or perforation.

tension, renal failure, vascular diseases), alcohol use disorder and/or recreational drug use (*e.g.*, cocaine), malnutrition, and cancer.[1] In a recently published systemic review, alcohol and hypertension were common comorbidities, and the overall mortality rate amounted to 32%.[7] The most common presenting symptoms are hematemesis, shock, melena, abdominal, or substernal pain. Diagnosis is made by EGD that shows circumferential dark pigmentation of the distal third of the esophagus, which is relatively less vascular, and necrosis that stops abruptly at the gastroesophageal junction. Colour change depends on the location of the lesion, degree of necrosis, and pattern of necrosis. Biopsy is recommended but not required for a definite diagnosis. AEN has no specific treatment. Antibiotics, antifungals and nutritional support should be considered on an individual basis. Candidiasis may occur in conjunction with AEN, whilst it is not thought to be causative, treatment is considered prudent given the poor prognosis associated with this condition.[8]The patient was admitted to the High Dependency Unit and was treated with IV fluid and amine support, piperacillin-tazobactam, oxacillin, fluconazole, proton pump inhibitors, and total parental nutrition. One week later, an EGD showed a lighter esophageal mucosa with some white exudate spots (Figure 1). After two weeks, EGD documented fibrin deposition associated with reepithelialisation (Figure 2). The patient was released after two more weeks, when an EGD showed total resolution of AEN with no indication of stricture development or perforation (Figure 3).

References

1. Gurvits GE, Shapsis A, Lau N, et al. Acute esophageal necrosis: a rare syndrome. J Gastroenterol 2007;42:29-38.
2. Goldenberg SP, Wain SL, Marignani P. Acute necrotizing esophagitis. Gastroenterology 1990;98:493-6.
3. Gurvits GE. Black esophagus: acute esophageal necrosis syndrome. World J Gastroenterol 2010;16:3219-25.
4. Akaishi R, Taniyama Y, Sakurai T, et al. Acute esophageal necrosis with esophagus perforation treated by thoracoscopic subtotal esophagectomy and reconstructive surgery on a secondary esophageal stricture: a case report. Surg Case Rep 2019;5:73.
5. Ziegler PE, Rosario Lora D, DeMeo M. An unusual case of black esophagus. ACG Case Rep J 2023;10:e01202.
6. Averbukh LD, Mavilia MG, Gurvits GE. Acute Esophageal Necrosis: a case series. Cureus 2018;10:e2391.
7. Abdullah HM, Ullah W, Abdallah M, et al. Clinical presentations, management, and outcomes of acute esophageal necrosis: a systemic review. Expert Rev Gastroenterol Hepatol 2019;13:507-14.
8. Beg S, Rowlands D. Acute Oesophageal Necrosis: a case report and review of the literature. BJMP 2015;8:a829