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CASE REPORT

The Dark side of the Esophagus: A Case report of Acute Esophageal Necrosis

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Abstract

Acute Esophageal necrosis (AEN) is a rare clinical entity, with fewer than 150 reported cases in literature and endoscopic prevalence ranging from 0.01 to 0.28%. First described by Goldenberg and later characterized by Gurvits in 2007, this condition predominantly affects the distal esophagus due to its relatively poor blood supply, resulting in its characteristic black appearance. AEN typically occurs in male patients with predisposing risk factors such as chronic liver disease, chronic kidney disease, alcoholism, malnutrition and cardiovascular comorbidities. It is also associated with sepsis and carries a high mortality rate. Upper gastrointestinal endoscopy remains the gold standard. Given the rarity of AEN, the standardized management guidelines are not well defined and the primary therapeutic focus remains on optimizing medical condition. We present the case of an 81-year old female who presented with septic shock, profound hypoglycemia with hypothermia and workup revealed Acute Esophageal Necrosis. The development of AEN in our patient was multifactorial, with esophageal ischemia secondary to septic shock induced hypotension playing a key role. The condition may have been exacerbated by reduced cardiac output and vasoconstriction of splanchnic blood vessels secondary to hypothermia. Furthermore, gastric reflux likely contributed to additional mucosal injury. The patient was managed successfully with conservative yet aggressive approach with inotropic support, antibiotics and total parenteral nutrition. We conclude that AEN should be considered in any patient with predisposing risk factors, particularly in presence of critical illness. This case highlights the urgent need for development of standardized guidelines to ensure timely diagnosis and optimal management for this rare but serious condition.

1. Introduction

Acute Esophageal Necrosis (AEN) was initially described by Goldenberg in 1990 as Necrotizing Esophagitis or Black Esophagus for its characteristic endoscopic appearance [1]. In 2007, Gurvitis et al. further characterized AEN as a distinct and rare clinical entity, later termed Acute Esophageal Necrosis Syndrome (Gurvits Syndrome) following review of case reports[2]. It predominantly affects male patients, typically in their mid-sixties, and arises from a combination of tissue hypo-perfusion, gastric acid exposure, and impaired mucosal defense mechanisms. The condition is more prevalent in patients with preexisting risk factors, including chronic liver disease (CLD), renal impairment, cardiovascular comorbidities (Hypertension and Diabetes Mellitus), and excessive alcohol consumption

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with distal esophagus being the most commonly involved site owing to its vasculature [3] Here we present the case of an 81 year old female who presented with septic shock and was subsequently diagnosed with AEN during endoscopic evaluation.

2. Case Presentation

An eighty-one (81) year old female was brought to the Accident and Emergency Department by paramedics with altered mental status, hypotension, recurrent non-bloody vomiting and profound hypoglycemia. Collateral history obtained from her daughter revealed excessive alcohol consumption and progressive deterioration over past few days. On arrival in resuscitation rooms, she was drowsy and confused. Point of care blood glucose testing revealed severe hypoglycemia (0.2 mmol/l). She was profoundly hypotensive (58/42mm Hg), tachycardiac (sinus at 120 beats per minute) and tachypneic (22 breaths per minute). Her oxygen saturation was low at 76% (room air) and body temperature was recorded as 36°C. She received intravenous (IV) thiamine followed by a 50% dextrose IV push for hypoglycemia correction, along with crystalloid fluids and active warming using a Bair Hugger to address profound hypotension and hypothermia. Routine investigations, including blood tests, blood cultures, urinalysis, and a chest X-ray, were requested to assess for potential underlying cause.

She had past medical history of CLD, Chronic kidney disease (CKD), Gastroesophageal Reflux Disease (GERD), dyslipidemia, recurrent falls history, hypothyroidism and diverticulosis.

Her GCS was 12 at the time of admission which improved after IV Dextrose. Physical Examination revealed dry mucous membranes and signs of malnourishment. Chest examination was unremarkable. Heart and bowel sounds were normal. Only mild epigastric tenderness was appreciated without any abdominal distention, guarding or organomegaly. Despite receiving fluids, her blood pressure (BP) remained critically low (85/50 mmHg). Inotropic support was commenced and patient was transferred to Intensive Care Unit (ICU) for hemodynamic stabilization. Her Chest X ray performed in Resuscitation room is shown in Figure 1

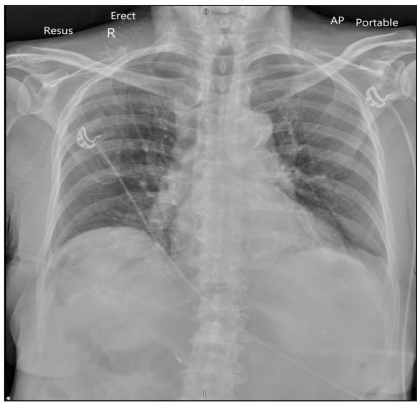

Figure 1. *AP view, Erect, showing no consolidation or pleural effusion. No pneumothorax or acute osseous abnormality identified. Thoracic Spine degeneration.*

Initial laboratory results are shown in Table 1

Table 1. *Blood results of the patients with normal reference ranges.*

Test and (unit)	Result	Reference
Hemoglobin (g/dl)	10.5	12.0- 15.0
Hematocrit (L/L)	0.38	0.36-0.46
White Cell Count (x10^9/L)	14	4.0- 10
Neutrophils (x10^9/L)	13.17	1.5- 8
Platelets (x10^9/L)	137	150 - 300
MCV (fl)	93.9	83-101
CRP (mg/l)	165.3	<5
PT (seconds)	14	11.5 – 16.0
INR	1.0	0.8 – 1.2
Urea (mmol/l)	14.7	3.20- 8.20
Creatinine (µmol/l)	314	49- 90
GFR (ml/min/1.73m2)	11.4	90
Sodium (mmol/l)	132	135- 146

Potassium (mmol/l)	4.4	3.50- 5.30
Chloride (mmol/l)	90	98- 110
Calcium (mmol/l)	2.36	2.10- 2.65
Lactate (mmol/l)	20	<1
ALT (U/L)	78	5-33
GGT (U/L)	96	6-42
ALP (U/L)	85	30-130
Total proteins (g/L)	67	60-80
Albumin (g/L)	33	35-50
Fecal occult blood	Negative	--
Urine Analysis	Commensals	--

She was empirically commenced on IV Piperacillin Tazobactam, 80mg IV bolus of Proton Pump Inhibitor (PPI) was given and High Flow Nasal Cannula (HFNC) was utilized to maintain her oxygen saturations. Her working diagnosis at this time was sepsis of unknown origin, acute kidney injury, alcohol excess and high anion gap metabolic acidosis. Owing to suspicion of bowel obstruction/mesenteric ischemia, an emergency contrasted Computed Tomography (CT) Thorax, Abdomen and Pelvis was booked which reported dilatation of distal esophagus with mural thickening at the gastroesophageal junction (GEJ). Small and large bowels were reported as of normal caliber. Endoscopy was recommended on the basis of the scan findings. The patient demonstrated hemodynamic improvement with rewarming and fluid resuscitation leading to BP stabilization, normalization of lactate, resolution of acute kidney injury with restored urine output, and inotropic support was successfully weaned off. Given the current clinical suspicion of sepsis on unknown origin and lactic acidosis, microbiology advised treatment with Piperacillin Tazobactam. Due to patients complains of dysphagia at this stage along with epigastric tenderness and CT scan findings, an esophagogastroduodenoscopy (EGD) was performed. Endoscopic examination revealed diffuse black mucosal thickening with coated mucosa involving the middle and lower third of esophagus and terminating at Gastroesophageal junction (EGJ). Also, hiatal hernia and atrophic gastritis was reported with a normal appearing duodenum. Biopsies were taken from non-necrotic regions to minimize the risk of perforation and bleeding as sampling could compromise the integrity of esophageal wall. This approach also facilitated the exclusion of other potential differential diagnoses.

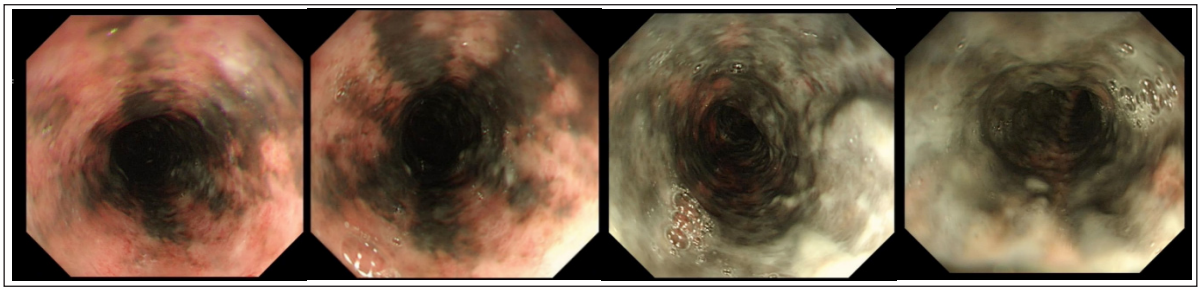

Figure 2. *Esophagogastroduodenoscopy showing Acute Esophageal Necrosis (AEN)*

The diagnosis of Acute Esophageal Necrosis (AEN) was made endoscopically on observation of necrosed esophageal mucosa. Given the patients concurrent septic state, a conservative yet aggressive treatment approach was adopted. This included IV antibiotics, proton pump inhibitors, bowel rest, total parenteral nutrition (TPN), and close monitoring. Histopathology of EGJ and mid-esophageal biopsy specimens is illustrated in Figures 3 and 4

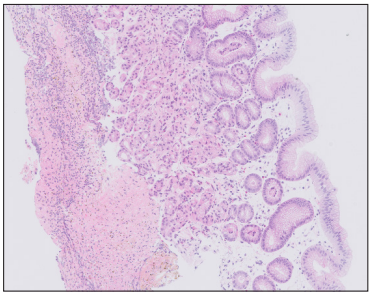

Figure 3. *Hematoxylin and Eosin stain (10X magnification) showing gastric cardiac mucosa with ulceration*

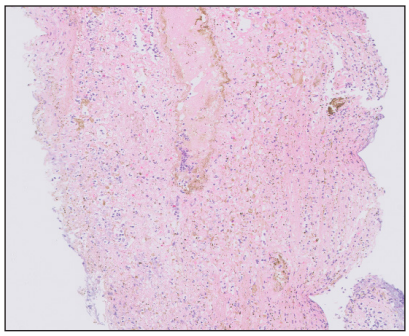

Figure 4. *Hematoxylin and Eosin stain (10X magnification) showing ulcer slough, acute inflammation and fibrin only. No esophageal mucosa seen*

The patient received multidisciplinary inpatient care which included inputs from dietitians, speech and language therapy team and was discharged to rehabilitation facility. The patient and her family didn't agree with interval endoscopy for reassessment. The development of Acute Esophageal Necrosis (AEN) in this patient was likely multifactorial. Esophageal ischemia due to hypotension in the context of septic shock played a key role. Additionally, AEN may have been exacerbated by low cardiac output and profound vasoconstriction secondary to hypothermia. Reflux of gastric contents likely contributed to further mucosal injury. The diagnosis was further supported by the presence of multiple risk factors, including CKD, CLD, alcoholism, advanced age, and malnutrition.

3. Discussions

Acute esophageal Necrosis (AEN) is a rare clinical condition with an estimated prevalence of approximately 0.2% in autopsy series, with even lower rates reported in endoscopic evaluations (ranging from 0.01–0.28% of cases). AEN predominantly affects males, with studies indicating male to female ratio of up to 4:1 Our case involved an elderly female patient in her eighties, which is relatively uncommon given the male predominance in previous studies [4]. The esophageal blood supply is segmental, with the upper esophagus receiving circulation from superior and inferior thyroid arteries, the mid-esophagus being supplied by branches of bronchial, right intercostal and descending aorta and lower esophagus deriving its blood supply from left gastric, left inferior phrenic and splenic arteries [5]. The lower esophagus has poor blood supply, making it prone to ischemia during low blood flow states like shock. This area, known as a watershed zone, is highly exposed to acidic reflux, affecting up to 97% of AEN cases. AEN occurs due to a 'two hit hypothesis' process: first, ischemic injury from low blood flow, followed by mucosal damage from acid reflux, worsening tissue death, especially in high risk individuals [6].

The risk factor for AEN include cardiovascular disease (47.3%), Diabetes Mellitus (36.4%), Alcoholism (28.2%), Liver disease (17.3%), Chronic Kidney Disease (15.5%), advanced age, male sex, chronic pulmonary disease, GERD, post-operative states, septic shock, diabetic ketoacidosis, medications (Terlipressin, Anti-hypertensive, NSAIDs, Bisphosphonates), sudden hemodynamic compromise (in setting of trauma or infection), malnutrition, viral infections and blunt thoracic trauma [7]. A comparable pathophysiological mechanism was seen in our patient, who had sepsis of unknown origin complicated by hypotension. The resultant hypo perfusion was further aggravated by hypothermia induced vasoconstriction, leading to significant reduction in esophageal blood flow. Furthermore, the presence of multiple predisposing risk factors likely contributed to the severity of mucosal injury and progression to esophageal necrosis.

The clinical presentation of acute esophageal necrosis is usually upper GI bleeding in the form of hematemesis (66%) or melena (33%), shock, syncope, fever, epigastric or retrosternal pains (28%), dysphagia (12%) while 13% of patients had sepsis on presentation [8]. Unlike the typical presentation of AEN, which commonly includes hematemesis, our patient did not exhibit any upper GI bleeding. Instead, the clinical picture was dominated by sepsis and hypothermia. Throughout the inpatient stay, hemoglobin levels remained stable. Literature also suggests that AEN can often be overlooked in patients who do not present with hematemesis [9]

The important differentials of acute esophageal necrosis include pseudo melanosis, esophageal melanosis, malignant melanoma, acanthosis nigricans, ingestion of coal and dust and infections [10,11].

Esophagoduodenoscopy (EGD) remains the gold standard for diagnosis. While biopsy is generally recommended, it is not necessary to confirm the diagnosis [12]. The primary purpose of biopsy in

such cases is to exclude differential diagnosis and superimposed infections through bacterial, viral and fungal cultures. EGD typically shows diffuse circumferential progressive black discoloration of the esophagus with abrupt demarcation at the Z line. Radiology can partially aid in demonstrating pathology but the characteristic endoscopic appearance is sufficient to make the diagnosis. [13] In our patient, biopsy results effectively rule out alternative diagnoses, while CT findings played a crucial role in prompting the decision to proceed with EGD.

Given the rarity of AEN, standard management guidelines remain undefined. The goal of therapy should be focused on treating the coexisting medical illness. The primary therapeutic approach focus on stabilizing the patient's overall medical condition. Key treatment strategies includes maintaining hemodynamic stability, suppressing gastric acid secretion, restricting oral intake, initiating total parenteral nutrition (TPN), ensuring bowel rest and administering antibiotics (in case of infection). Red blood cell transfusion may be necessary in patients with significant hemoglobin decline. Surgical intervention, such as esophagectomy is generally reserved for cases involving full thickness esophageal perforation. However existing literature suggests that the increased rate of operative intervention haven't been associated with improved patient outcomes. [13,14]. In our patient, clinical improvement was achieved through conservative management including antibiotics, high dose proton pump inhibitors and TPN followed by reintroduction of oral feeding.

In most uncomplicated cases, AEN follows a predictable healing trajectory. The diffuse black esophageal mucosa typically begins to show signs of resolution, and depending on overall patient's condition, endoscopic appearance may return to normal within two to four weeks' time. Relapse is uncommon; however, there has been a documented case of duodenal necrosis occurring four months after the initial episode. [15].

In our case, the patient and her family didn't opt for a follow-up endoscopy but were willing to contribute her case for reporting. The most severe complication of AEN is perforation, which may lead to mediastinitis, mediastinal abscess, empyema and sepsis. Stricture formation and stenosis has also been reported. [16]

AEN carries a poor prognosis, with an estimated mortality rate of 32% [17]. Our patient remained stable and discharged without complications, and has no reported issues to date.

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