





Managing Acute Esophageal Necrosis Secondary to Diabetic Ketoacidosis in a Rural Centre: A Case Report and Proposed Algorithm for Management

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ABSTRACT

Acute esophageal necrosis (AEN) is a rare phenomenon, usually presenting with hematemesis or melena in patients who are already unwell with concurrent pathology. We present a case of AEN in an acutely unwell 82-year-old man who presented in diabetic ketoacidosis (DKA). He was successfully managed in our rural centre with remote guidance from a tertiary centre. We propose an evidence-informed management algorithm to guide and assist clinicians caring for patients with AEN in resource-limited settings. Initial management of AEN should prioritize resuscitation and correction of any underlying pathology, such as DKA. Early endoscopic assessment is required for the diagnosis of AEN and quantification of the extent of necrosis. Early repeat endoscopy (48–72 h) is recommended to assess healing and is useful in guiding ongoing management. In resource-limited settings, early referral and transfer for patients with AEN is recommended in the absence of intensive care support, endoscopy, or limited blood products.

1 | Background

Acute esophageal necrosis (AEN) or "black esophagus [1]" is a rare phenomenon, with an estimated incidence up to 0.28%, occurring in critically unwell patients [2]. Presentations of AEN can be variable, from non-specific abdominal complaints, upper gastrointestinal bleeding to profound shock [3]. Currently, there is no unified consensus of terminology, disease process, severity grading or treatment. "Acute esophageal mucosal lesion (AEML)" [4] is an alternate term used to encompass spectrums of AEN, black esophagus and acute

necrotizing esophagitis. The hallmark for this syndrome is the development of diffuse circumferential black mucosal discoloration in the distal esophagus which may extend proximally. This black mucosa sharply demarcates at the gastroesophageal junction on endoscopy [1]. However, endoscopic appearances can be similar to reflux esophagitis Los Angeles classification grade D (RE-D)—a different entity [4, 5]. Therefore, clinical and endoscopic diagnosis of this condition can be difficult. Furthermore, despite the growing recognition of this condition, its pathophysiology remains poorly characterized. Its etiology is thought to be multifactorial, arising

 $\textbf{Abbreviations}: A EML, a cute \ esophage \ al \ mucos \ al \ lesion; A EN, a cute \ esophage \ al \ necrosis; CT, computer tomography; DKA, diabetic \ ketoacidosis; ICU, intensive \ care \ unit; NGT, nasogastric \ tube; PPI, proton pump inhibitor; RE, reflux \ esophagitis; UGI, upper gastrointestinal; UGIB, upper gastrointestinal \ bleeding.$

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Summary

- There is no consensus on terminology, pathogenesis, severity grading nor treatment relating to AEN.
- AEN should be suspected in elderly and co-morbid patients presenting with upper GI bleeding.
- Early endoscopic examination is recommended if available.
- Early referral to specialist upper GI service should be considered in resource limited settings to guide management for these patients.

from a combination of ischaemic insult, impaired mucosal barrier systems as well as backflow injury from gastric secretions [6, 7]. Diabetic ketoacidosis (DKA) is suggested as a common inciting factor [1, 8]. In the absence of DKA, states of profound hypoperfusion, have also been implicated in the development of AEN. Notably, AEN leads to a significant burden of morbidity, with an estimated mortality rate of 32% [6]. Complications may include stricture, perforation, mediastinitis, and sepsis [1].

Demands of managing patients with severe conditions that may lead to AEN, such as DKA or sepsis, may delay definitive diagnosis through endoscopy. In rural settings, these issues may be amplified due to resource limitations, unfamiliarity with the condition, and absence of evidence-based guidelines for the diagnosis and management. This case report and literature review describe a case of AEN in a rural setting and propose an evidence-informed algorithm for management.

2 | Case Presentation

2.1 | Case History and Examination

An 82-year-old man presented in DKA following an unwitnessed fall preceded by days of nausea with vague abdominal pain. His comorbidities included cerebrovascular disease, type 2 diabetes mellitus, and a previous diagnosis of ulcerative esophagitis. He was previously a smoker and consumed alcohol occasionally. Initial blood tests revealed a blood glucose of 29 mmol/L, pH7.18, and ketones of 7 mmol/L. Additionally, he had an acute kidney injury and electrolyte derangements manifesting in non-sustained ventricular tachycardia. He was commenced on insulin, electrolyte replacement, fluid therapy, and admitted to the intensive care unit (ICU). The patient developed hematemesis and melena on day 1 of admission.

2.2 | Differential Diagnosis, Investigation, and Management

Repeat blood testing showed that the hemoglobin decreased from 121 to 82 g/L. In the setting of suspected new upper gastro-intestinal bleeding (UGIB), the patient was referred to General Surgery for consideration of urgent gastroscopy, resuscitated with packed red cells, and commenced on high-dose intravenous proton pump inhibitor (PPI).

Gastroscopy identified 12 cm of circumferential necrotic mucosa at the distal esophagus (Figure 1). There was no evidence of active bleeding. The patient was kept nil by mouth and referred to a tertiary center for advice. He subsequently became febrile with concerns for possible mediastinitis and was commenced on broad-spectrum antimicrobials-Piperacillin/Tazobactam and Caspofungin. Computer tomography (CT) imaging of the chest and abdomen demonstrated bilateral pleural effusions and bi-basal atelectasis without evidence of mediastinitis. A repeat gastroscopy performed at 48 h demonstrated slough at the distal esophagus and healing of the esophageal mucosa without extension of the necrosis (Figure 2). No biopsies were taken due to the improved appearances of the esophagus. A nasogastric tube (NGT) was inserted to allow for enteral feeding. The patient fully recovered from his DKA; however, he had a prolonged admission with delirium and urosepsis. His follow-up gastroscopy at the 6-week mark demonstrated complete resolution of the AEN (Figure 3).

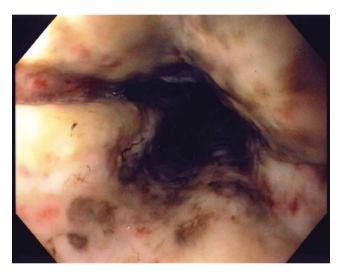




FIGURE 2 | Endoscopic appearances of healing distal esophageal mucosa at 48 h.

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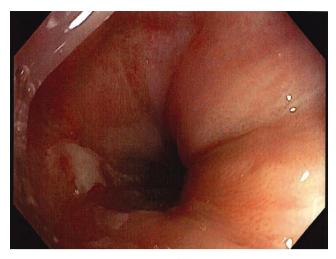


FIGURE 3 | Resolution at 6-week follow-up gastroscopy.

3 | Discussion

We present an interesting and rare case of AEN in the setting of DKA. The mechanisms underlying AEN in DKA remain undefined in the literature. Of note, several studies describe increased age, male sex, poor clinical condition with multiple medical comorbidities such as cardiovascular disease, diabetes mellitus, chronic kidney disease, and alcohol abuse, as well as malignancy as risk factors for developing AEN [1, 4]. Currently, there are no best-practice guidelines for the work-up and management of AEN. Our experience highlights that the work-up should include screening of comorbidities, biochemical screening, as well as early endoscopy. Management of AEN is largely conservative. Surgical intervention is usually reserved for severe cases with evidence of complications including perforation or mediastinitis. In contrast to other mucosal pathologies, such as those of the colon, there is a significant gap in the literature as to how best to grade the severity of AEN, with no existing scoring systems for triage and prognostication. Herein, we summarize the salient evidence-informed components of management for AEN, with nuanced consideration into coordination and delivery of safe care in resource-poor settings (Figure 4).

3.1 | Non-Operative Approaches

Management is primarily conservative following the formalization of diagnosis by endoscopy [9]. Management priorities should consider resuscitation, correcting the underlying pathology, and optimization of perfusion and nutrition.

3.2 | Resuscitation

Goal-directed fluid resuscitation should occur in reference to evidence-based guidelines that consider the patient's hemodynamic status, comorbidities, and biochemical derangements. Regular monitoring with real-time approaches is recommended, particularly in the setting of instability. Administration of blood products is recommended in the setting of bleeding or decrementing hemoglobin levels [10]. Early hematology consultation

or transfer to a tertiary center may be warranted if blood product resources are limited.

High-dose PPI should be considered for all patients with UGIB for the protection of esophageal mucosa from acid reflux [10]. In the absence of sepsis, prophylactic antibiotics are not recommended. Despite this, judicious monitoring of the patient should occur, and in the event of rapid deterioration associated with fever, rigors, or shock, broad-spectrum antibiotics and antifungals should be considered [8, 11].

There are mixed opinions about the role of enteric feeding tubes, that is, NGT insertion for decompression and enteral nutrition, as well as further prevention of gastric reflux, due to the risk of esophageal perforation [1, 8, 12]. Our case highlights that NGT insertion is safe in the setting of mild mucosal injury. However, for cases of more extensive esophageal necrosis, nasogastric intubation should be avoided or placed with endoscopic guidance. Parenteral nutrition should be considered as an alternative avenue for nutrition if nasogastric intubation is avoided.

3.3 | Restoring and Maintaining Homeostasis

Nutrition is a fundamental consideration, with particular focus on the quality of nutrition to improve mucosal healing, as well as the route of nutrition in the context of mucosal injury. In the immediate setting, enteral or parenteral nutrition should be considered with dietitian input to prevent further injury associated with oral intake and to optimize healing [8, 13].

3.4 | Tertiary Center Referral and Transfer

In situations where resources for ongoing care of the patient are not available, such as access to ICU, endoscopy, or specialist upper gastrointestinal (GI) surgical care, transfer to tertiary centers that are able to provide this support should be considered. This should occur in a timely manner to avoid further deterioration of the patient due to complications of AEN, namely perforation.

3.5 | Operative Approaches

3.5.1 | Endoscopic

Endoscopy remains the gold standard for the diagnosis of AEN. Regarding endoscopic interventions, in settings of acute esophageal bleeding, endoscopic-guided submucosal adrenaline infiltration may be helpful [8]. There is very limited evidence to support the placement of metal-covered stents or the usage of balloon tamponade in the setting of bleeding associated with AEN, with one case describing perforation associated with balloon tamponade [14]. Further research is required to establish the safety of endoscopic interventions. Importantly, in distinguishing AEN from severe reflux esophagitis (grade D), AEN is expected to improve relatively quickly

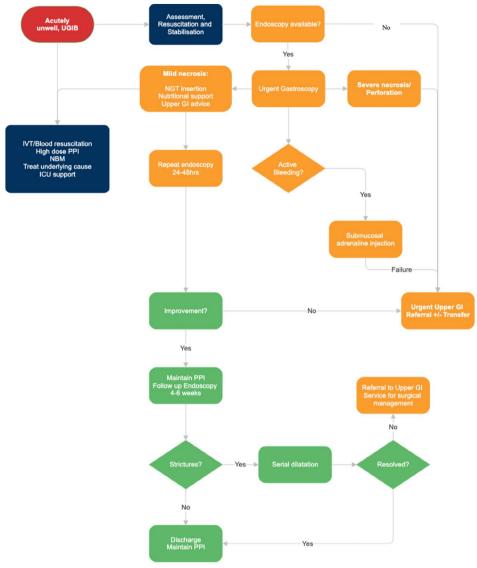


FIGURE 4 | Proposed management algorithm for acute esophageal necrosis in a rural centre.

upon the treatment of comorbidities and stabilization of the hemodynamic state [4].

3.5.2 | Surgical Management

Surgical intervention for AEN is reserved for severe complications such as perforation and downstream sequelae including mediastinitis. Emergent esophagectomy would be considered in the first instance followed by elective reconstruction [14]. Therefore, the role of timely triage and transfer of patients from rural or resource-poor settings to a tertiary centre for definitive management is strongly recommended.

3.6 | Follow-Up

With appropriate management of underlying conditions in the absence of operative intervention, the esophageal mucosa will

likely recover within the period of a month [8]. Repeat endoscopy in 4 weeks should be considered for patients in all cases to assess the extent of recovery as well as complications of AEN, such as strictures. In the setting of strictures, serial balloon dilatation may be required to prevent significant dysphagia and malnutrition [12].

4 | Conclusion

AEN is a rare phenomenon occurring in the acutely unwell and comorbid, which requires early recognition as it may have significant clinical implications. The mainstay of management is largely conservative. Early referral to a specialist upper GI service should be considered in resource-limited settings to guide management for these patients. Larger studies are required to further characterize best-practice care models for AEN, which in turn may help clinicians with developing scoring systems as well as refining clinical practice guidelines.

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Author Contributions

Annie Jiao Wang: conceptualization, data curation, formal analysis, investigation, methodology, project administration, writing – original draft, writing – review and editing. Khang Duy Ricky Le: data curation, formal analysis, investigation, methodology, resources, validation, writing – original draft, writing – review and editing. Shasha Haycock: writing – review and editing. Brendan Desmond: supervision, writing – review and editing. Matthew Shears: resources, supervision.

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The authors have nothing to report.

Ethics Statement

The case report generation process was discussed with our local ethics and governance team. No formal ethics approval was required following the discussions and therefore was waived. The patient provided consent for the de-identification and use of their medical information and data for the generation and publication of this case report.

Consent

The patient provided informed consent that was written and signed for the generation and publication of this manuscript using their deidentified medical information.

Conflicts of Interest

The authors declare no conflicts of interest.

Data Availability Statement

Data can be requested from the corresponding author when required. All relevant data have been provided in the generation of this manuscript, which is intended for open access publication.

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