**Title page**

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 that 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 a treatment algorithm to guide management of those presenting to a resource limited centre with AEN.

Initial management of AEN should prioritise resuscitation and correction of any underlying pathology, such as DKA. Early endoscopic assessment is required for diagnosis of AEN and quantification of the extent of necrosis. Early repeat endoscopy (48-72hrs) 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.

**Keywords**

Acute esophageal necrosis, Gurvits syndrome, diabetic ketoacidosis, necrotising esophagitis, endoscopic surgery, rural surgery

**Main text**

**Background**

Acute esophageal necrosis (AEN) is a rare phenomenon, with an estimated incidence up to 0.28%, occurring in critically unwell patients(1). Diagnosis is difficult, as presentations of AEN can be variable, from non-specific abdominal complaints to upper gastrointestinal bleeding to profound shock(2). Gold-standard diagnosis is the identification of circumferential discoloured black mucosa with sharp demarcation at the gastro-esophageal junction on endoscopy(3). Despite the growing recognition of this condition, its pathophysiology remains poorly characterised. Diabetic ketoacidosis (DKA) is suggested as a common inciting factor(3, 4). 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 associated mortality rate of 32%(5). Complications may include stricture, perforation, mediastinitis and sepsis(3).

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 in the context of resource limitations, unfamiliarity with the condition and absence of evidence-based guidelines for the diagnosis and management of this condition. This case report and literature review describes a case of AEN in a rural setting and proposes an algorithm for management with a focus on resource-poor settings.

**Case presentation**

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 29mmol/L, pH 7.18, and ketones of 7mmol/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 day 1 of admission.

Differential Diagnosis, investigation and management

Repeat blood testing showed that the hemoglobin decremented from 121g/L to 82g/L. In the setting of suspected new upper gastrointestinal 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 was performed which identified 12cm 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 centre 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 hours demonstrated slough at the distal esophagus and healing of the esophagus mucosa without extension of the necrosis (Figure 2). No biopsies were taken due to improved appearances of the esophagus. A nasogastric tube (NGT) was inserted to allow for enteral feeding. The patient fully recovered from his DKA, however 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).

**Discussion**

Currently there are no “best-practice” guidelines on the management of AEN. Management of AEN is largely conservative. Surgical intervention is usually reserved for severe cases with evidence of complication 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 summarise the salient components of management and propose a treatment algorithm for AEN (Figure 4), with nuanced consideration into coordination and delivery of safe care in rural and remote settings.

*Non-operative approaches*

Management is primarily conservative following the formalisation of diagnosis by endoscopy(6). The management priorities are resuscitation, correcting the underlying pathology and optimisation of perfusion and nutrition.

*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, particularly in the setting of high bleeding and decrementing of hemoglobin levels(7). Early hematology consultation or transfer 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(7). 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 and in these cases, broad-spectrum antibiotics and antifungals should be considered(4, 8).

There are mixed opinions about the role of enteric feeding tubes i.e. NGT insertion for decompression and enteral nutrition as well as further prevention of gastric reflux, due to the risk of esophageal perforation(3, 4, 9). 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.

*Restoring and maintaining homeostasis*

Nutrition is a fundamental consideration, with particular focus on quality of nutrition to improve mucosal healing, as well as 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 optimise healing(4, 10)

*Tertiary centre 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 centres who 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.

*Operative approaches*

*Endoscopic*

Endoscopy remains the gold-standard for diagnosis of AEN. Regarding endoscopic interventions, in settings of acute esophageal bleeding, endoscopic guided submucosal injections of adrenaline may be helpful(4). There is very limited evidence to support the placement of metal-covered stents or usage of balloon tamponade in the setting of bleeding associated with AEN, with one case describing perforation associated with balloon tamponade(11). Further research is required to establish safety of endoscopic interventions.

*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(11). 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.

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(4). Repeat endoscopy in 4 weeks should be considered for patients in all cases to assess 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(9).

**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 specialist upper GI service should be considered in resource limited settings to guide management for these patients.

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**Informed consent statement**

Informed consent for publication of the patient's clinical data and images was obtained from the patient.

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**Disclosures**

**List of abbreviations**

Acute esophageal necrosis; AEN, Diabetic ketoacidosis; DKA, Intensive care unit; ICU, Proton pump inhibitor; PPI, Computer tomography; CT, nasogastric tube; NGT, Upper gastrointestinal; GI, Upper gastrointestinal bleeding; UGIB

**Ethics approval and consent to participate**

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 for publication**

The patient provided informed consent that was written and signed for generation and publication of this manuscript using their de-identified medical information

**Data availability statement**

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

**Competing interests**

Nil

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Nil

**Authors’ contributions**

Annie Jiao Wang – Conceptualisation, methodology, validation, formal analysis, investigation, data curation, resources, writing – original draft, writing – review and editing, project administration

Khang Duy Ricky Le – Methodology, validation, formal analysis, investigation, data curation, resources, writing – original draft, writing – review and editing

Shasha Haycock – writing – review and editing

Brendan Desmond – Conceptualisation, writing – review and editing

Matthew Shears – supervision

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**Key clinical message**

In suspected cases of AEN in acutely unwell patients with UGIB or other abdominal complaints, 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.

**Figure legends**

Figure 1. Initial gastroscopy - acute necrosis of the distal esophageal mucosa

Figure 2. Endoscopic appearances of healing distal esophageal mucosa at 48hrs

Figure 3. Resolution at 6-week follow-up gastroscopy

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

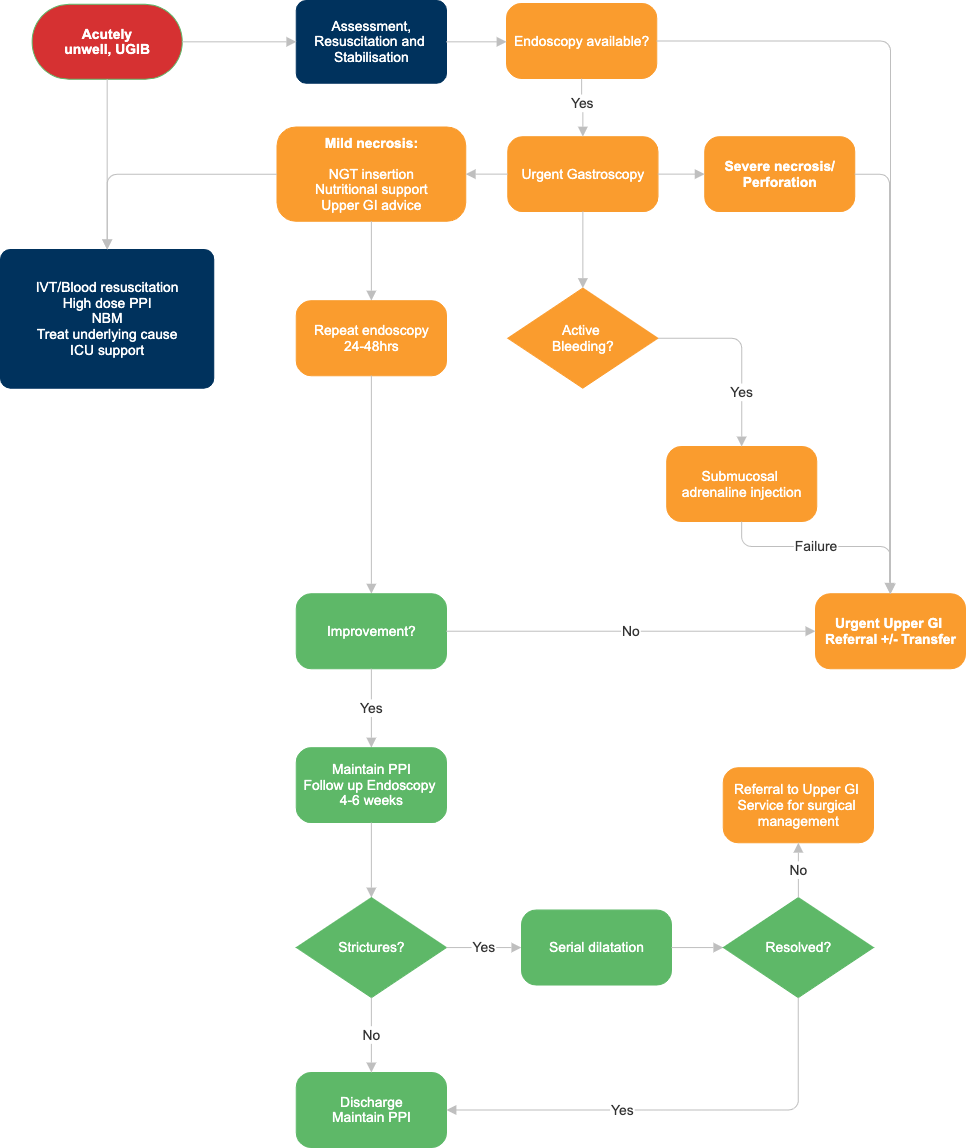
**Figures**

**Figure 1**

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| **Figure 3** | |
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**Figure 4**



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