



PALLIATIVE CARE CASE OF THE MONTH

“Meaning and Management of Black Esophagus”

by
Reed Nerness, MD

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Case: DO is a 79-year-old man with end-stage renal disease on hemodialysis for over 4 years; renal cell carcinoma s/p nephrectomy; coronary artery disease s/p drug-eluting stent placement x3; heart failure with reduced ejection fraction; prior stroke without residual deficits; atrial fibrillation on anticoagulation; prostate cancer s/p androgen deprivation therapy; and recurrent small bowel obstructions previously on total parenteral nutrition now on oral diet; who has had recurrent admissions over the last four months with recurrent bacteremia and arteriovenous fistula dysfunction and bleeding resulting in placement of a femoral tunneled dialysis catheter, and who was transferred to a tertiary care hospital for higher level of care for acute upper gastrointestinal bleeding. Anticoagulation had been stopped, and he was transfused with 4 units pRBCs.

On admission day two, esophagogastroduodenoscopy (EGD) was performed and found “black esophagus”, or acute esophageal necrosis (AEN). DO’s diet was restricted to no intake, intravenous proton pump inhibitor (PPI) twice daily was started, and he was administered intravenous prophylactic antibiotics due to high risk for infection. Nasogastric (NG) tube was not placed due to risk of perforation. Gastroenterology recommended goals of care discussions given high risk of mortality related to AEN and possible indication for artificial hydration and nutrition. Palliative care consultation was obtained. Further investigation found DO had severe intermittent lower chest pain and was requiring about one-to-two units of platelets and red blood cells per day since admission.

Background: AEN is a rare gastrointestinal condition diagnosed via visual inspection with EGD. Characteristic findings are diffuse, circumferential blackening of esophageal mucosa with abrupt transition to normal mucosa at the gastroesophageal junction. It is thought that AEN is an underdiagnosed phenomenon. Available studies have an incidence of about 0.1-0.2 percent of individuals undergoing EGD for any reason.^{1,2} AEN is relevant for the palliative care practitioner as an indicator of an acute downward change in serious illness trajectory. A diagnosis of AEN should prompt early discussions of goals of care given high rates of morbidity and mortality.

Etiology

Mucosal ischemia, injury from gastric reflux, and impaired mucosal healing are thought to combine to cause AEN.³ The conditions that can lead to this pathophysiology are common in those with serious illness. Many conditions have been implicated, usually in combination, as contributors to the development of AEN.

These include malnutrition, uses of non-steroidal anti-inflammatory drugs (NSAIDs) and antihypertensive medications, alcohol use, local infections, diabetic ketoacidosis, malignancy, gastric outlet obstructions, arthroplasty surgeries, and solid organ transplantation.^{3,4}

Presentation

Older individuals with a combination of serious illnesses, typically admitted with evidence of upper gastrointestinal bleeding.

Disease Course

Various stages have been outlined to characterize the course of AEN, consisting of initial insult, healing stage with necrotic debris sloughing and mucosal exudates, followed by return to normal mucosa without relapse of the disease.³ Return to normal esophageal mucosa has been reported as early as one to two weeks from diagnosis,⁴ however a definitive time course has not been outlined. Nutritional status before diagnosis and during healing is thought to play pivotal role in recovery from AEN. Therefore, careful discussion about artificial hydration and nutrition is necessary.

Esophageal perforation due to fragile esophageal wall is the most serious complication. It occurs in < 7 percent of cases and requires surgical intervention.⁵ Sepsis is another more common complication in the acute period. Longer-term complications mainly are related to development of esophageal strictures, estimated to occur in 25 to 40 percent of patients who survive the initial phase.⁶

Prognosis

The most recent information available has found that mortality rates for those diagnosed with AEN are as high as 30 to 50 percent.⁷ One chart review found 31.8 percent of patients who were found to have AEN in the hospital died during that admission.⁵ Death most commonly occurs due to the combination of AEN and the underlying condition(s). Mortality rates attributed to AEN alone are lower at about 6 percent.⁵

Symptom Management

Severe chest pain is common in those with AEN. As no specific therapies exist, an understanding of the relevant pathophysiology can guide treatment of acute pain. Reduction of ongoing injury from gastric reflux should be started with intravenous proton pump inhibitor. Diet should be restricted to nothing by mouth to promote initial healing. Optimal timing of resumption of oral nutrition is unclear. After an initial period, some suggest starting sucralfate to aid in mucosal healing.⁴ NSAIDs should be avoided in the treatment of pain. Acetaminophen and short-acting analgesics are most commonly used.

Personal details in the case published have been altered to protect patient privacy.

For palliative care consultations please contact the Supportive and Palliative Care programs at PUH/MUH, 412-647-7243, pager # 8511, Shadyside, 412-647-7243, pager # 8513, Perioperative/ Trauma Pain, 412-647-7243, pager # 7246, UPCI Cancer Pain Service, pager 412-644-1724, Magee Women’s Hospital, pager 412-647-7243 pager # 8510, VA Palliative Care Program, 412-688-6178, pager # 296. Hillman Outpatient: 412-692-4724. For ethics consultations at UPMC Presbyterian-Montefiore and Children’s pager 412-456-1518

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Goals of Care

AEN is an indicator of severe underlying serious illness with a high mortality rate. In acute setting, discussions around enteral and parenteral nutrition will likely arise, as part of the initial treatment requires avoidance of oral nutritional intake, NG tube placement as well as nutritional support for mucosal healing. A new diagnosis of AEN may also provide an avenue to discuss the patient's overall health status and concerns about the patient entering a "new phase of illness" characterized by a more rapid decline. Some patients and/or their loved ones may have values that align with placing limits on surgical interventions, long-term artificial hydration and nutrition, and code status.

Case Conclusion: After goals of care conversations, it was clear DO and his family valued prioritizing prolonging DO's life. DO and his family were willing to undergo procedures, artificial hydration and nutrition, ongoing admission, and transfer to critical care unit to achieve these goals. DO and his family agreed with the recommendation from the palliative care service that undergoing cardiopulmonary resuscitation would be very unlikely to add meaningful time to DO's life. DO's code status was changed to DNR/DNI. After about a week an NG tube was placed and medications and blood product transfusions were continued. Overnight about 3 weeks into admission, DO had an acute worsening of encephalopathy and became hypotensive and tachycardic. Evaluation and treatment were initiated including resuscitation with blood products, however despite aggressive management DO died later that morning likely from an acute worsening of gastrointestinal bleeding.

Summary: AEN is a complication of severe illness and can be used by palliative care clinicians as a harbinger of more rapid decline. Acute symptom management should be addressed with attention to the underlying pathophysiology while initiating discussion of values to guide short- and long-term interventions.

References:

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