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• CASE REPORT •

Acute esophageal necrosis and liver pathology, a rare combination

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Abstract

Acute esophageal necrosis (AEN) or 'black esophagus' is a clinical condition found at endoscopy. It is a rare entity the exact etiology of which remains unknown. We describe a case of 'black esophagus', first of its kind, in the setting of liver cirrhosis and hepatic encephalopathy.

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INTRODUCTION

Endoscopic discovery of a 'black esophagus' or 'acute esophageal necrosis' (AEN) that is unrelated to ingestion of caustic or corrosive agents is quite exceptional. It has been described only a few times in the past. Lacey *et al*. in 1991 reported 25 cases[13]. A similar report by Benoit *et al*.[3] in 1999 cited 27 cases after an extensive review of the literature. Since then there have been sporadic case reports. It is difficult to estimate its true incidence as we think this disease is largely underreported. The exact etiology of 'black esophagus' still remains unknown. We describe a case of 'black esophagus', first of its kind, in the setting of liver cirrhosis and hepatic encephalopathy.

CASE REPORT

A 59 year-old hispanic male with a known past medical history of hypertension, chronic alcohol abuse, liver cirrhosis, chronic pancreatitis and depression was admitted to the psychiatry service for aggressive behaviours, depression and suicidal ideation. On d 2, he was transferred to the medical floor after evaluation for a change in mental status with confusion, disorientation and impulsive behaviours.

A detailed history was elicited on further interrogation. Patient was known to have hypertension for 10 years for which he required monopril, metoprolol and clonidine. The history also revealed alcohol abuse for 45 years, still an active abuser, alcoholic liver cirrhosis diagnosed for 4 years and negative hepatitis B and C serologies. He had multiple previous admissions for hepatic encephalopathy and earlier in the same year underwent chemotherapy for a biopsy proven hepatocellular carcinoma. He denied smoking and illicit drug abuse.

On examination the patient was drowsy but arousable, responding to verbal commands, oriented to 'place and person', but not 'time'. He was afebrile and hemo-dynamically stable. General physical examination was normal. Cardiac, pulmonary, abdominal and neurological examinations did not show any abnormalities. Rectal examination was normal with a negative guaiac test.

Laboratory data on admission were: WBC 5.1 n/L with 48% granulocytes, hemoglobin (H)/ hematocrit (Hct) of 10.3/ 31, MCV of 80.3 fl, RDW of 15.4%, platelets of 174 /n.

Na 141 mE, K 4.5 mE, Cl 108 mE, CO_2 26.1 mE, BUN 32 mg/dL, Creatinine 2.1 mg, Gluc 134 mg, Total bili 0.6 (0.3 direct), albumin 2.8 gm, proteins 6.7 gm, alkaline phosp 217 U/L, SGOT 50 U/L, SGPT 39U/L, LD 225 U/L, PT 11.4, aPTT 27.6 s and INR 1.05 s, Ammonia 139.8 um.

Ethanol on admission was 56.2, serum Osm 312, normal TSH, syphilis(-), ferritin 593, TIBC 168, total iron 58 and iron saturation 35%, B12/folate 226/22.7. CT scan of the head was negative for bleed/metastasis or any mass. CT scan of the abdomen revealed a few scattered diverticuli and accessory spleen.

A diagnosis of hepatic encephalopathy, anemia and intra-vascular volume depletion was established based on the clinical and laboratory data. Hydration with normal saline at 125 cc/h and treatment with lactulose 445 cc qid (titrated to bowel movement) were given. The rest of the management included administration of resperidal, thiamine, folate and ativan (prn). His anti- hypertensive medications were continued and patient was placed under close observation for alcohol withdrawal.

During the following 2 d (d 4 and 5) the patient's mental status improved to full orientation without any signs of confusion, agitation and/or suicidal ideation. Ammonia level dropped to 85 um. A drop in H/Hct (9.7/29.7) was related to intravascular volume depletion.

On d 6 of admission, the patient's clinical status deteriorated with a recurrence of confusion and lethargy. Patient was found to have tachycardia (heart rate of 120/min), a blood pressure of 105/70, a positive stool guaiac test, BUN/Creatinine of 68/3, ammonia of 105 um and a drop in his H/Hct (7.8/22). His management at this point included continuous intravenous hydration, blood transfusion with 3 units of packed red blood cells and intravenous proton pump inhibitors. His anti-hypertensive medications were held. The patient was transferred to a monitored setting for further observation and an esophageal gastro-duodenoscopy (EGD) was scheduled.

EGD revealed a continuous segment (15-35 cm) of necrosis with exudates, ulcerations and friable mucosa in the middle and distal parts of the esophagus. The gastro-esophageal (GE) junction showed no evidence of varices, stomach and duodenum linings were normal. A biopsy taken from the necrotic area confirmed the findings of fibrinoid necrotic debris with hemosiderin deposits and acute inflammatory cells on pathological examination of the specimen. An ultrasound of the abdomen revealed a medical renal disease without obstruction, liver texture consistent with cirrhosis with reversal of portal venous flow and mild ascites.

The patient made an uneventful recovery over the next 3 d. He was transferred to the medical floor from where he was discharged with a follow up in the outpatient clinic. No recurrences of any GI symptoms were reported in his 3-mo-follow-up.

DISCUSSION

'Black esophagus' has been described primarily in post-mortem studies[14,2]. Goldenberg *et al*.[8] gave us a detailed endoscopic description of 2 cases in the early 1990s. In a one-year prospective study conducted in Rouen University Hospital,

France, 8 (0.2%) cases of ' acute esophageal necrosis' were identified among the 3 900 patients who underwent EGD[6]. Since then there have been only a handful of case reports. The numbers reported in the literature may highly underestimate the real incidence of this condition as suggested by this prospective trial. Acute esophageal bleeding is the most common presentation. The condition is generally seen in the elderly and those having co-morbid conditions.

A variety of mechanisms have been proposed to account for the development of this condition, and low systemic perfusion seems to play the dominant role. Other mechanisms cited include direct toxic effect[21,25], as well as indirect mucosal breakdown and acid effect[1], however, none of these have been proved. The frequent involvement of distal 1/3 esophagus[24], absence of gastric lesions, presence of necrosis of mucosal/submucosal necrosis, presence of thrombus microscopically and rapid regression of disease after hemodynamic stability were similar to ischemic colitis[17,22] and strongly support the ischemic basis for the insult.

Other conditions associated with this disease entity were prolonged hypertension[10], ischemia[8,9], hyperglycemia[13], hypersensitivity to antibiotics[16], herpetic infection[5], gastric volvulus[11], posterior mediastinal haematoma and aortic dissection[14,18], anti-cardiolipin antibodies[4] and Steven Johnson syndrome[15]. In short, a multi-factorial etiology to absence of any disease (idiopathic) is possible. The pathogenesis of ischemic insult in our case can be accounted for by the low systemic perfusion with reversal of portal venous flow and worsening of hepatic encephalopathy.

Diagnosis is established with endoscopy with or without biopsy. The differential diagnosis included melanosis[23], pseudo-melanosis[12], and acanthosis nigrans[7].

The main reported complications of AEN were esophageal stenosis and stricture formation[13,3]. Lacy *et al*.[13] reported a 15% complication rate, whereas other case series have reported very low to none. Recurrence was less than 10%[6,20] and mortality ranged between 0-33%[6,13,19]. The prognosis depends on the patient's general status rather than the extent of local esophageal necrotic lesions.

Generally there is no standardized treatment but the overall consensus favours conservative treatment with intra-venous proton pump inhibitors and short-term parenteral nutrition. The main aim is to avoid extension of insult with time for a spontaneous and aided recovery.

In short, AEN is still uncommon with exact etiology and pathogenesis largely unclear. The majority do not have any long-term sequel, only a few develop strictures and stenosis. Prognosis varies, and a majority recover with conservative treatment with death being an uncommon outcome.

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