Black esophagus, or acute necrotizing esophagitis, is a blackening of the esophagus that is usually distal with a sharp demarcation at the gastroesophageal border. Black esophagus is known to the gastroenterology community; however, to our knowledge it is virtually unknown in the pathology literature with only a single instance described in 1967. It is thought to occur as a poorly elucidated ischemic phenomenon. We report a case of black esophagus in a 45-year-old woman with a history of cocaine and alcohol abuse who was found unresponsive after a vague 2-day illness. On autopsy examination, the esophagus was black with ischemic necrosis of the mucosa, submucosa, and muscularis propria including a diffuse acute inflammatory infiltrate and brown pigmentation limited to the mucosa. Positive periodic acid–Schiff and negative iron stains suggest that the pigment is lipofuscin, likely secondary to ischemia.

(Arch Pathol Lab Med. 2011;135:797–798)

Black esophagus, or acute necrotizing esophagitis, is a blackening of the esophagus that is predominately distal with a sharp demarcation of abnormality at the gastroesophageal border. Histologically, the mucosa is necrotic with disrupted muscle fibers and no histologically identifiable causative organisms or agents. Black esophagus is known to the gastroenterology community and carries a mortality rate as high as 38% in the literature; however, to our knowledge it is virtually unknown in the pathology literature with only a single instance described in 1967. It is thought to occur as a manifestation of a poorly elucidated ischemic phenomenon.

REPORT OF A CASE

We present the case of a 45-year-old woman with a history of cocaine and alcohol abuse who was found unresponsive after a vague 2-day illness. Emergency medical services responded and she was pronounced dead at the scene. External examination, at autopsy, was significant for nasal septal perforation. On internal examination, the esophagus was black with ischemic necrosis of the mucosa, submucosa, and muscularis. Microscopically, there was a diffuse acute inflammatory infiltrate and a brown pigmentation limited to the mucosa. A positive periodic acid–Schiff with and without diastase and negative iron stain suggest that the pigment was lipofuscin, which has not been previously described, likely secondary to ischemia. The liver had steatosis and increased fibrosis, consistent with chronic alcohol use. Toxicology showed cocaine metabolites. In light of the circumstances surrounding the death and autopsy findings, the cause of death was considered to be acute esophageal necrosis.

PATHOLOGIC FINDINGS

Grossly, the mucosal surface of the esophagus was remarkable for diffuse, uniform, black discoloration extending the entire length of the esophagus (Figure 1). Submucosal edema was also present. The mucosa and submucosa measured 0.7 cm in thickness, with a readily apparent, grossly unremarkable underlining muscularis propria. Discoloration extended precisely to the gastroesophageal junction. There an apparent transition to unremarkable gastric mucosa was observed, which extended throughout the stomach, excepting focal punctate areas of gastritis.

Microscopic examination demonstrated acute esophageal necrosis characterized by widespread mucosal sloughing, necrosis of residual mucosa, extensive submucosal edema, and an acute inflammatory infiltrate. Brown, granular amorphous pigmentation was present and found exclusively in the mucosa (Figure 2). The muscularis propria was architecturally intact but ischemic. The inflammatory infiltrate extended through the muscular layer and into the adventitia. To further characterize the pigment, special stains including a periodic acid–Schiff with and without diastase and iron stain were performed. The pigment was periodic acid–Schiff with and without diastase positive and iron negative, which suggests the pigment is lipofuscin.

COMMENT

Black esophagus, or acute necrotizing esophagitis, is a rare entity with a reported incidence of 0.0125% to 0.2% in patients undergoing upper endoscopy. The mean age is 68.4 years with a slight male predominance. The most common presenting symptom is gastrointestinal bleeding. Although its true etiology is yet to be fully explained, the leading hypothesis attributes this unusual condition to hypoperfusion-associated ischemia. The most common established risk factor is duodenal ulcer, but also a clear association has been shown in those who abuse alcohol, particularly “binge” drinkers. Additional proposed pre-
disposing factors include gastric outlet obstruction, malignancy, and/or a combination of the previously mentioned factors.\textsuperscript{5,6,7} The mortality rate ranges from 0% to 38%.\textsuperscript{1-3,4} If found early by endoscopy, treatment is supportive.\textsuperscript{6}

Black esophagus is characterized by its hallmark discoloration, with a grossly necrotic, friable, and ulcerated esophagus in the mid and distal portions and an abrupt transition at the gastroesophageal junction to unremarkable stomach.\textsuperscript{4} Given these gross findings, the diagnosis can be made without biopsy.\textsuperscript{4} However, the differential diagnosis includes melanosis, pseudomelanosis, coal dust deposition, corrosive ingestion, and acanthosis nigricans.\textsuperscript{4,6,8} It can be distinguished from melanosis and pseudomelanosis using melanin and iron stains, if suspected. Melanosis will have a positive melanin stain and pseudomelanosis will have a positive iron and melanin stain.

**CONCLUSION**

Acute necrotizing esophagitis is a rare entity, nearly exclusively described in gastroenterology literature previous to this case as a phenomenon identified during endoscopy. A single example of this entity was described in the pathology literature in 1967 and has not been updated or revisited since.\textsuperscript{2} Patients with this diagnosis often present with bleeding and several associated risk factors, including duodenal ulcers and/or a history of substance abuse.\textsuperscript{1} Our patient, a 45-year-old woman, was found unresponsive following a vague 2-day illness. Her contributing risk factors included a history of cocaine and alcohol abuse, the former confirmed by cocaine metabolites in her toxicology screen. The external examination finding of a perforated septum suggested a long-standing history of insufflation of illicit substances, and the microscopic findings of steatosis and fibrosis in the liver were consistent with her history of chronic alcohol use. These ancillary findings support the hypothesis that acute necrotizing esophagitis occurs at a higher rate in those with significant substance abuse history.

We thank Santino Lamancusa, BS, and John Maksem, MD, for their digital imaging expertise.

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**References**