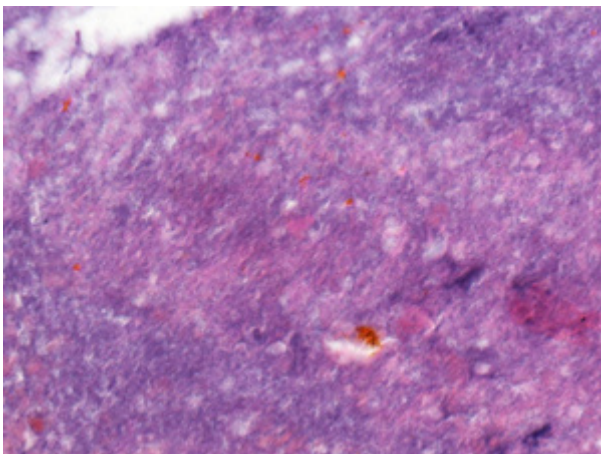
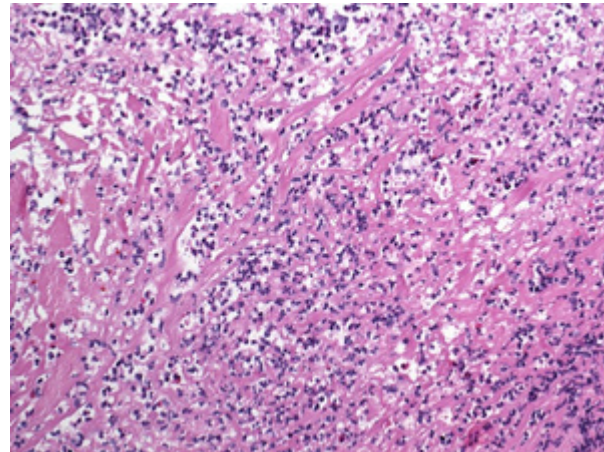
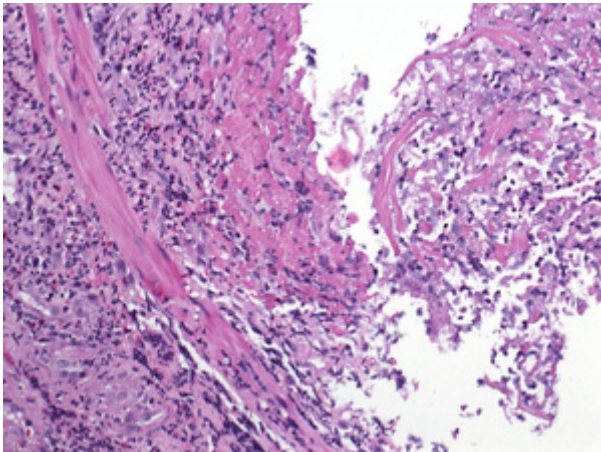
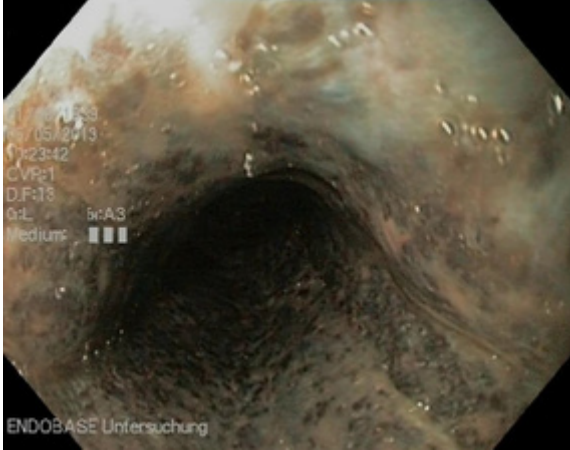


# September 2013

Esophageal biopsies from a 73-year-old male with known ankylosing spondylitis (under therapy with leflunomide) who presents with acute onset epigastric pain.

What is your diagnosis?



## Diagnosis

Acute esophageal necrosis ("black esophagus").

## Comment

Endoscopy (Panels A and B) shows circumferential, black discoloration of the esophageal mucosa, extending the entire length of the organ, yet with accentuation in the lower half. There is an abrupt demarcation to uninvolved cardiac mucosa at the Z-line. Upon histology, the mucosa is diffusely necrotic, with diffuse ulceration, widespread sloughing, and absence of viable squamous epithelium. There is massive acute inflammation (Panels C and D). Focally, brown, granular to amorphous pigmentation is observed within the necrotic debris which is negative for iron and melanin stains (Panel E).

Black esophagus is a rare disease with about 100 cases having been described in the literature. Upon gross inspection the lesion has to be differentiated from malignant melanoma, (pseudo)melanosis, acanthosis nigricans, caustic injury, coal dust deposition, and dye ingestion. Men are four times more commonly affected than women. Although the disease may be observed at any age, the peak incidence is in the sixth to seventh decade of life.

Patients mainly present with upper gastrointestinal bleeding (hematemesis and/or melena are present in >70% of cases), but may also report on epigastric pain, dysphagia, nausea or vomiting. Most patients have at least one severe comorbid condition of which diabetes mellitus, cardiovascular disease, renal insufficiency, malignancy, chronic alcohol abuse and/or malnutrition are the most common. Of note, mortality is remarkably high (up to 38%) and is commonly related to these underlying medical conditions, e.g. ultimately due to sepsis or multiorgan failure, overall reflecting impaired physiological reserve due to critical illness and/or general deconditioning. Mortality is not directly related to the esophageal disease, except in the rare cases of perforation, mediastinitis, and esophageal infection, particularly in immunocompromised individuals.

The etiology of acute esophageal necrosis is believed to be multifactorial and results from a combination of ischemic insult (tissue hypoperfusion in hemodynamic compromise and low-flow states), decreased function of mucosal defense barrier systems and reparative mechanisms, and chemical injury due to massive influx of gastric contents (gastric outlet obstruction or gastroparesis) on the already compromised esophageal mucosa. The reported positivity for periodic acid-Schiff (with and without diastase) suggests the iron-negative pigment to be lipofuscin.

Treatment of the disease should focus on supportive therapy which includes optimizing perfusion, minimizing gastroesophageal reflux, and treating esophageal infection if present.

## For further reading

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