Hamman’s Crunch: Boerhaave Syndrome

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Objectives: To understand a rare but deadly beast. Check out our podcast on this topic at iTunes, Soundcloud, or Spotify.

What is it?
Boerhaave Syndrome, or effort rupture of the esophagus, is a rare but deadly condition of esophageal perforation. It is associated with high morbidity and mortality. Untreated it is nearly universally fatal at 95%, even if treated mortality can still reach 40%.

Where does it occur? Effort rupture of the esophagus most frequently occurs in patients with a normal esophagus, in the left posterior aspect of the distal thoracic esophagus, as a result of a sudden rise in intra-esophageal pressure. It may also occur in the presence of esophageal pathology, such as Barrett’s esophagus, eosinophilic esophagitis, or esophageal ulcers. It may also occur in the intra-abdominal portion of the esophagus, but this is rare and will not be encountered on board exams.

What are the results of perforation? Rupture into the mediastinal cavity, with the expulsion of gastric contents into the mediastinum, leading to a cascade of mediastinitis, infection, necrosis, and the rapid onset of overwhelming sepsis.

What is the presentation? Boerhaave syndrome classically presents in a male, middle aged alcoholic, with a history of recent alcohol consumption, presenting with the acute onset of excruciating chest pain, radiating to the back, with emesis. But up to 1/3 of patients have no history of vomiting. In the modern era, esophageal perforation most commonly occurs following upper G.I. endoscopy.

Physical Exam:
Chest pain, neck pain, severe odynophagia, fever, tachypnea, tachycardia and hypotension. SIRS criteria develop rapidly, within the first hour. Sepsis follows quickly.

Chest wall crepitus on palpation → “Hamman’s Crunch”. Pathognomonic of the syndrome, though not often present and cannot be relied upon. Pleural effusions have also been described, classically on the left.

Investigation. Although the definitive test is a water-soluble contrast (Gastrografin) esophagram, practically speaking, a rapidly obtained CT will be the most available test in the emergency department. The esophagram does a better job of localizing the perforation, but is more cumbersome to obtain.

Plain films may show air in the soft tissues, pleural effusion, mediastinal widening, or sub-diaphragmatic air if the perforation is in the gastric portion of the esophagus. They can however, in the early stages, be normal and therefore have no reliable specificity or sensitivity for assisting in diagnosis.

Labs: worthless. Leukocytosis may be present, but labs are generally not helpful. Blood cultures and blood gases should be drawn as sepsis is a major complication.

Differential Diagnosis
The usual suspects of chest pain presentations, myocardial infarction, pulmonary embolus, pneumothorax, and aortic dissection.
Perhaps the closest in presentation is Mallory-Weiss syndrome, which are longitudinal tears in the distal esophagus. These are incomplete tears, and are most commonly due to repetitive vomiting or retching, classically in alcoholics. These patients do not have esophageal perforation, and do not show signs of acute mediastinitis. They are managed symptomatically and require no radiographic studies or further testing. They can often safely be discharged with symptomatic management.
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Management
NPO (#obvi)
Fluid resuscitation, preferably lactated Ringers
Intravenous proton pump inhibitors
Early intravenous broad-spectrum antibiotics. (piperacillin/tazobactam is an excellent choice)
Rapid surgical consultation

Not all patients require surgery. Small leaks may be managed medically. Obviously, this decision is above our paygrade and surgery should always be called in cases of suspected esophageal perforation.

Prognosis: Morbidity and mortality are high. Up to 45% mortality in some studies. Early recognition and intervention is essential.

References