A 71-year-old nonsmoking woman presented to the surgical unit with a 16-h history of upper abdominal pain. The pain had developed suddenly in the periumbilical area and spread to the left upper abdomen. She had vomited twice after the onset of pain but had not apparently aspirated vomitus into her lungs, and the pain then radiated to the left side of the chest and shoulder. She had previously been well but had angina and hypertension treated with atenolol and bendroflumethiazide. Her pulse was 96 beats per minute, the blood pressure was 130/70 mm Hg, and the temperature was 36.5°C. There was tenderness on palpation of the left upper abdomen and there were crackles and diminished breath sounds at the left lung base. The WBC count was $10.8 \times 10^9/L$, the hemoglobin value was 12.7 g/dL, and the serum amylase level was mildly elevated at 415 U/L (normal, <220 U/L). An abdominal radiograph showed no abnormalities, and ultrasound scan showed a single gallstone in the gallbladder and no abnormalities in the biliary tree, liver, pancreas, and spleen. A chest radiograph (Fig 1) was interpreted as showing consolidation in the left lower lobe with associated air bronchograms and a pleural effusion. A diagnosis of pneumonia with referred abdominal pain was made, and she was transferred to the medical unit. Results of an ECG were normal. The PaO$_2$ was 53.2 mm Hg, the Pco$_2$ was 37.5 mm Hg, and the pH value was 7.37. She was treated with oxygen and intravenously administered cefuroxime and erythromycin, but her condition deteriorated. Another chest radiograph (Fig 2) 48 h after admission showed a large left hydropneumothorax. Aspiration of the pleural effusion yielded pus; a culture of this material showed coliform organisms. Her condition improved after intercostal drainage of the left pyopneumothorax, and a further diagnostic test was then performed.

What is this patient’s diagnosis?

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Diagnosis: Boerhaave’s syndrome.

A water-soluble contrast (Iopamidol) esophagogram demonstrated leakage from the esophagus into the mediastinum and left pleural space (Fig 3). Thoracotomy was performed with removal of food debris and pus. Primary suture of a 4.5-cm rupture of the left lateral wall of the distal esophagus was performed and jejunostomy feeding was established. The patient died 18 days later from cardiorespiratory failure due to sepsis.

Spontaneous rupture of the esophagus after vomiting was first described in 1724 by Boerhaave. It is a rare condition and diagnosis is notoriously difficult. During vomiting, the pyloric sphincter is closed and the intraesophagus pressure may rise steeply, particularly if the patient attempts to suppress vomiting by closing the pharyngeal sphincter. Rupture typically occurs in the lowest third of the thoracic esophagus where the muscle fibers are thinned as they fan out into the stomach and where the esophagus has no serosal covering and very little adjacent support. Rupture occurs at lower pressure if there is mucosal disease of the esophagus. Rupture is a more severe form of the Mallory-Weiss syndrome of mucosal tears produced by protracted retching and vomiting.

Following rupture of the esophagus, the mediastinum fills with air and gastric contents. A few hours later, the pleural membrane gives way and air and food debris pass into the pleural cavity. In a classic case of this syndrome, the patient experiences severe pain in the lower chest after vomiting, which is then followed some hours later by pleuritic pain and prostration. Crepitus may be palpable in the subcutaneous tissues of the neck. A chest radiograph typically shows pneumomediastinum (which was present on our patient’s initial radiograph [Fig 1]), followed by a hydropneumothorax. Mackler, therefore, emphasized a diagnostic triad of vomiting, chest pain, and subcutaneous emphysema. However, over-reliance on these classic features may lead to delays in diagnosis since they are often absent. The complete triad of vomiting, chest pain, and subcutaneous emphysema was present in only 1 of 14 cases reported by Walker et al. This series emphasized how confusing the clinical presentation may be. Pain was the commonest symptom but it was often abdominal and associated with rigidity. Two of the 14 patients had no history of vomiting and two vomited after onset of pain. Subcutaneous emphysema was present in only four of 14 patients. The chest radiograph was usually abnormal but the features were easily confused with those of pneumonia or spontaneous pneumothorax. The mean delay from presentation to diagnosis was 4 days, and the patients initially presented to a variety of specialists with misdiagnoses, such as pneumonia, pneumothorax, myocardial infarction, or perforated duodenal ulcer.

It is, therefore, important not to rely on classic features, one of which is subcutaneous emphysema, which were not present in our patient, but to have a high index of suspicion in patients presenting with vomiting and either chest or abdominal pain. The key investigation is an esophagogram which detects rupture of the esophagus in more than 90% of cases.

Primary surgical repair is the treatment of choice when the diagnosis is made within 24 h of rupture. Mortality increased markedly with delay in diagnosis, and patients referred late, with ongoing pleural and mediastinal contamination, pose a major management dilemma. In these cases, various options are available. For patients whose condition is stable, simple conservative management has been advocated by some and involves the following: nothing by mouth, nasogastric suction, pleural drainage, antibiotics, and feeding by enterostomy or parenteral nutrition. Where the pa-
tient's condition is unstable, thoracotomy allows thorough cleaning of the pleural cavity, debridement of devitalized tissue, and direct placement of chest drains. Esophageal tissue usually is edematous and friable if there has been delay in diagnosis so that there is a high leakage rate if primary repair is undertaken. A variety of alternative techniques, such as T-tube intraesophageal drainage, esophageal exclusion, and diversion procedures may be used.

REFERENCES