Fever and Pneumopericardium in a Patient with Systemic Sclerosis*
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A 66-year-old woman was admitted with three days of fever, chills, and pleuritic chest pain. She had a 14-year history of systemic sclerosis with esophageal involvement. Five years prior to admission, she had undergone a hiatal hernia repair for reflux esophagitis, following which she required periodic bougienage for dilation of a distal esophageal stricture. On admission the patient was acutely ill. Her temperature was 37.1°C and blood pressure 78/50 mg Hg. Examination revealed thickened skin, telangiectasias, dullness to percussion and rales at the left lung base, and a loud splashing sound heard diffusely over the precordium corresponding with systole. The ECG showed ST segment elevation in the anterior and lateral leads. Her anteroposterior chest x-ray film (Fig 1) showed a pleural density at the left base, a left lower lobe infiltrate, and an air-fluid level in the esophagus (arrow). A decubitus view (Fig 2) demonstrated pneumopericardium (arrow), an air-fluid interface in the esophagus (open arrow), and small bilateral free-flowing pleural effusions (arrowheads).

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Diagnosis: Esophageal perforation with esophagopericardial fistula and hydropneumopericardium

The patient was treated with IV fluids and antibiotics. A meglumine diatrizoate swallow (Fig 3) showed direct passage of contrast material from the distal esophagus into the pericardium. Flexible esophagoscopy demonstrated a small anterolateral perforation of the distal esophagus just proximal to the squamocolumnar junction. Purulent pericardial fluid was drained with a tube placed into the pericardium via a subxiphoid pericardial window. The patient improved with continued therapy, although she had intermittent fevers. Twenty days later, she underwent a transhiatal esophagectomy with cervical esophagogastric anastomosis and feeding jejunostomy. The patient improved with continued therapy, although she had intermittent fevers. Twenty days later, she underwent a transhiatal esophagectomy with cervical esophagogastric anastomosis and feeding jejunostomy. Unfortunately, following surgery, the patient developed septic shock and died. Pathology of the resected esophagus demonstrated a fistulous tract with ulceration and chronic inflammation of the esophageal wall. Autopsy findings included organizing fibrinous purulent pericarditis.

In patients with systemic sclerosis, peptic reflux esophagitis is exceedingly common. There is, however, only one previously reported case of esophagopericardial fistula formation due to peptic esophagitis in a systemic sclerosis patient. Esophagopericardial fistula due to all causes is rare; a recent review found only 49 cases reported in the world literature. The most common causes were benign: esophagitis with ulceration, foreign bodies (including an occupational accident in a sword swallow), iatrogenic (instrumental perforations and breakdown of anastomotic sites), and caustic ingestion. Esophageal carcinomas were responsible in about one fourth of the cases.

Characteristic features of the clinical presentation include pleuritic chest pain, fever, and a splashing sound over the precordium during systole, known as the "water wheel murmur" (bruit de moulin). Electrocardiographic findings of pericarditis may be present.

The most common radiologic features on plain roentgenogram are pneumopericardium and hydropneumopericardium. Other findings include enlargement of the cardiac silhouette, pleural effusions (especially left-sided), bilateral or left-sided pulmonary infiltrates, and pneumomediastinum. In the series reported by Cyrlak et al, either a fistulous tract or gross filling of the pericardium was seen in 80 percent of patients who had a contrast study of the esophagus.

The initial therapeutic approach involves supportive measures, antibiotics, and early pericardial drainage. After stabilization, a variety of more definitive surgical procedures has been attempted. In the past, the outcome among the reported cases was always fatal. However, several recently reported cases have survived, indicating that the prognosis for the rare patient with esophagopericardial fistula is no longer so uniformly dismal.

REFERENCES