Pyopneumopericardium Attributed to an Esophagopericardial Fistula: Report of a Survivor and Review of the Literature

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Herein we describe a case of pyopneumopericardium that resulted from formation of an acquired esophagopericardial fistula in a patient with silent, benign esophageal ulcer disease. Atypical features on initial examination suggested congestive heart failure or a pneumonic process (or both). The delayed development of pneumopericardium disclosed on a chest roentgenogram led to the clinical recognition of the esophagopericardial fistula. Subsequent emergent pericardiocentesis relieved cardiac tamponade and enabled us to diagnose pyopneumopericardium. A radiographic contrast study with use of meglumine diatrizoate revealed the site of the fistula in the midesophagus. The esophagopericardial fistula was surgically closed, and our patient had a good final result. Formation of an esophagopericardial fistula is a relatively uncommon finding; of the 60 previously reported cases, only 10 patients have survived. As illustrated in the current case, early diagnosis and treatment, including pericardial drainage and intense antibiotic therapy followed by a well-planned operative closure of the fistula, are paramount for the successful management of esophagopericardial fistulas.

Esophagopericardial fistulas are uncommon findings and are often associated with unfavorable outcomes. Etiologic factors include both benign and malignant esophageal diseases; however, benign conditions are the most common causes of formation of esophagopericardial fistulas. Initial symptoms often include acute epigastric or retrosternal chest pain and dyspnea in a setting of previously diagnosed ulcer disease. Herein, however, we describe a case of pyopneumopericardium that resulted from spontaneous formation of an esophagopericardial fistula in a patient who had no history or complaints of gastroesophageal disease but who had symptoms compatible with congestive heart failure, pulmonary disease, or an infectious process.

REPORT OF CASE
A 79-year-old man, a retired cattle buyer, was referred to our institution for evaluation of recent onset of atrial fibrillation with a ventricular rate of 130 beats/min, dyspnea, and pleural effusions disclosed on a chest roentgenogram, as reviewed by his local physician. Previously, the patient had been healthy. Five weeks before referral, he underwent a routine general examination by his local physician; results were unremarkable, including normal findings on a chest roentgenogram and an electrocardiogram. Approximately 3 weeks before referral, the patient experienced the onset of malaise, myalgias, loss of appetite, cough productive of white phlegm, dyspnea on exertion, and mild peripheral edema. During this period, his abdominal girth increased, and he gained 3.6 kg. The patient denied having fever. No chest pain, orthopnea, palpitations, paroxysmal nocturnal dyspnea, nausea, vomiting, or diarrhea was noted. He had no past history of hypertension, diabetes, or rheumatic fever, and his family had no history of severe heart disease or

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malignant lesions. In addition, he had no history of tuberculosis, fungal infections, malignant tumors, collagen vascular disease, trauma, irradiation of the thorax, or pneumothorax. The patient was a nonsmoker and had no allergies. He consumed 1 ounce of alcohol per week, and his only medication was four to eight aspirins (325 mg) daily during the previous 14 to 21 days for persistent myalgias.

On admission to our institution, findings on physical examination revealed an alert elderly man in no immediate distress. The blood pressure was 140/70 mm Hg without pulsus paradoxus. The pulse rate was 130 beats/min with an irregularly irregular rhythm, respiratory rate was 20/min, and oral temperature was 37.8°C. Bibasilar rales were detected. The jugular venous pressure, measured at 5 cm above the sternal angle at 45 degrees, was only mildly increased at 10 cm of water. On palpation, the precordium was quiet, and the cardiac apex was palpable on the anterior axillary line at the fifth intercostal space. The first heart sound was normal, and a soft second heart sound was detected; no S4 or S3 was noted, but a grade 2 (on the basis of 1 to 6) apical systolic murmur was audible. The mildly distended abdomen was nontender, and mild ascites was present. The span of the nontender liver was approximately 10 cm by percussion at the right midclavicular line. Mild ankle edema (1+) was present bilaterally. The patient was oriented to person, place, and time, and no motor or sensory deficits were elicited.

An electrocardiogram indicated atrial fibrillation with a ventricular rate of 140 beats/min, nonspecific T-wave changes in inferior and lateral leads, and frequent unifocal premature ventricular complexes. A chest roentgenogram (Fig. 1) disclosed cardiac enlargement, small bilateral pleural effusions, and prominent pulmonary vasculature. On admission, laboratory studies yielded the following results: leukocyte count, 16,600/mm³ (normal, 4,100 to 10,900) with a differential count of 66% neutrophilic segments (normal, 42 to 75%), 13% neutrophilic bands (normal, 0 to 3%), 9% lymphocytes (normal, 16 to 52%), and 11% monocytes (normal, 1 to 11%); hemoglobin, 11.4 g/dl (normal, 12.9 to 16.6); platelet count, 313,000/mm³ (normal, 184,000 to 370,000); serum sodium, 119 meq/liter (normal, 135 to 145); potassium, 4.6 meq/liter (normal, 3.6 to 4.8); creatine kinase, 52 U/liter (normal, 52 to 336); and erythrocyte sedimentation rate, 101 mm in 1 hour (normal, 0 to 22). Renal, liver, and thyroid function tests and amylase levels were normal.

Thoracentesis yielded serous-appearing fluid with a protein content of 2.6 g/dl (plasma protein, 5.6 g/dl), leukocyte count of 300/mm³ (86% neutrophils), pH of 7.48, lactate dehydrogenase of 80 U/liter, and negative findings for cytology, gram staining, and cultures. Blood cultures were also negative. Two-dimensional echocardiography performed on the second hospital day demonstrated a mildly dilated left ventricle, mild reduction in systolic function with an ejection fraction of 42%, and generalized hypokinesis. Prolapse of the mitral valve and moderate mitral regurgitation were noted. A small anterior pericardial effusion was also present.

On the fourth hospital day, left ventricular angiography, selective coronary arteriography, and right ventricular biopsy were performed. The left ventriculogram demonstrated generalized hypokinesis, mild left ventricular dilatation, and reduced left ventricular function. The coronary arteriogram showed mild coronary artery disease. The right ventricular biopsy specimen had focal dense endocardial fibrosis, no evidence of myocarditis, and a mean of 2 lymphocytes per high-power field (normal, less than 5), and it was negative for amyloid and iron staining.

On the seventh hospital day, the patient's condition remained stable, but the leukocyte count had increased to 39,500/mm³, and progression of the left pleural effusion was evident on a chest roentgenogram. On the next day, a chest roentgenogram (Fig. 2) revealed the presence of pericardial air. Computed tomography of the chest (Fig. 3) indicated the presence of air anterior, lateral, and posterior to the heart. Repeated echocardiography showed a moderate anterior pericardial effusion and multiple minute densities with the
motion and appearance of microbubbles. Elective pericardiocentesis was attempted but was unsuccessful in removing a substantial amount of pericardial fluid.

On the ninth hospital day, the patient’s clinical status deteriorated abruptly; hemodynamic findings suggested cardiac tamponade. Bedside echocardiography revealed a large pericardial effusion with microdensities and evidence of cardiac tamponade in conjunction with systolic collapse of the right atrium (Fig. 4). After emergent repeated pericardiocentesis yielded 650 ml of purulent material, the hemodynamics immediately improved. Broad-spectrum antibiotic coverage was initiated, and subsequent culture results indicated polymicrobial (anaerobic and aerobic) pericarditis. A gastrointestinal contrast study with use of meglumine diatrizoate (Fig. 5) demonstrated a midanterior esophageal fistula, and endoscopy showed an exudative midanterior esophageal ulcer with an esophagopericardial fistula at 32 cm from the incisors.

The patient underwent an operation on the 12th hospital day. Through a left thoracotomy, purulent material was encountered in the pericardial space, and the fistula was detected superior to the left inferior pulmonary vein in the adjacent anterior wall of the esophagus. The pericardium was resected, and the defect in the esophageal mucosa was closed. The left anterior serratus muscle was then mobilized, transposed through the third intercostal space, and wrapped around the esophagus to complete the repair of the esophagopericardial fistula.

The patient’s postoperative course was complicated by the following conditions: persistent pleural effusions, respiratory failure that necessitated a tracheostomy, a bleeding duodenal ulcer for which surgical intervention was necessary, and acute urinary retention attributed to benign prostatic hypertrophy that necessitated a transurethral resection of the prostate. After receiving antibiotic therapy for 45 days and completing a 1-month period in the rehabilitation unit, the patient was dismissed on the 135th hospital day.

DISCUSSION

Anatomically, the esophagus is adjacent to the cardiovascular system from the level of T-3 to T-11. Therefore, perforation of the esophagus can involve the pericardium and can result in death from sepsis, cardiac tamponade, or massive hemorrhage subsequent to atrial or ventricular perforation. The development of an esophagopericardial fistula is relatively rare and is associated with high mortality, particularly with the occurrence of pyopneumopericardium. A search of the literature revealed 60 cases of acquired esophagopericardial fistula and an in-hospital mortality of 83%. Ten survivors have been described. Benign esophageal conditions are the most common cause (Table 1) and account for 77% of the cases. Esophageal ulcer disease and reflux esophagitis contribute most substantially to this group (46% of benign etiologic factors). Perforation by an ingested
foreign object, the second most common benign cause, oc­curs predominantly in the pediatric age-group. Iatrogenic conditions are the third most common benign cause and include perforation after bougienage, breakdown of an anas­tomotic site after an operation, and perforation of the esopha­gus after radiation therapy. Esophageal malignant tumors constitute approximately a fourth of all reported cases of esophagopericardial fistulas.

Formation of an acquired esophagopericardial fistula occurs in patients with a known history of gastroesophageal disease, and the most commonly associated symptoms or signs are precordial pain, dyspnea, fever, and the presence of an audible systolic murmur over the precordium, similar to the splashing of a waterwheel (bruit de moulin). Esophago­pericardial fistulas have also manifested as pericarditis because of the development of purulent pericarditis.4,12,13

The patient described herein is atypical in that the history, review of systems, and findings on physical examination and initial invasive and noninvasive studies suggested congestive heart failure. Leukocytosis, however, indicated the possibility of an infectious process, but blood cultures were negative. Subsequent chest roentgenography revealed the fundamental finding of pneumopericardium, and computed tomography of the chest and echocardiography substantiated the presence of pericardial effusion. The abrupt development of cardiac tamponade necessitated echocardiographic-guided pericardiocentesis, which provided hemodynamic stabilization and identified purulent pericardial fluid. After pyopneumopericardium was diagnosed, both esophago­graphic and esophagoscopic studies were performed to determine the site of the fistula. No contrast dye was apparent in the pericardium of our patient, but the fistula was identi­fied with oral use of meglumine diatrizoate in a radiographic contrast study (Fig. 5). The esophagopericardial fistula was then surgically repaired by transposition of the left anterior serratus muscle through the third intercostal space to envelop the esophagus.

In our patient, the cause of the esophagopericardial fistula was silent, benign midesophageal ulcer disease and erosion and perforation of the esophagus. Although he had no prior history of ulcer disease, he had a history of substantial consumption of aspirin before hospitalization. On initial examination, his symptoms and signs suggested decompensated heart failure with recent onset of atrial fibrillation, pleural effusions, and cardiomegaly as disclosed on a chest roent­

Table 1.—Etiologic Factors for Esophagopericardial Fistulas
(Compiled From Cases Described in the Literature)

<table>
<thead>
<tr>
<th>Cause</th>
<th>Cases</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign esophageal conditions</td>
<td>46</td>
<td>77</td>
</tr>
<tr>
<td>Esophageal ulcer and reflux esophagitis</td>
<td>21</td>
<td>35</td>
</tr>
<tr>
<td>Ingestion of a foreign object</td>
<td>8</td>
<td>13</td>
</tr>
<tr>
<td>Iatrogenic*</td>
<td>6</td>
<td>10</td>
</tr>
<tr>
<td>Tuberculosis</td>
<td>4</td>
<td>7</td>
</tr>
<tr>
<td>Esophageal diverticula</td>
<td>3</td>
<td>5</td>
</tr>
<tr>
<td>Ingestion of caustic substances</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Barrett's esophagus</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Achalasia</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Malignant esophageal carcinoma</td>
<td>14</td>
<td>23</td>
</tr>
<tr>
<td>Total</td>
<td>60</td>
<td>100</td>
</tr>
</tbody>
</table>

*Includes two cases of formation of atrial-esophageal fistula.
genogram, dyspnea on exertion, weight gain, ascites, peripheral edema, and hyponatremia. The increased leukocyte count and low-grade fever, however, also suggested a systemic infectious process, but blood cultures were negative. Survival of our patient was clearly related to prompt pericardial drainage and early surgical repair of the fistulous tract. Medical management alone is not commonly associated with a favorable result. Spontaneous closure of the fistula without surgical repair has been reported in only four cases.\

CONCLUSION

The current case summary describes one of the relatively few patients who survived a pericardial complication of esophageal ulcer disease that resulted in formation of an esophagopericardial fistula and consequent pyopneumopericardium. Early diagnosis and treatment including pericardiocentesis and intermittent drainage of the pericardium, intense, full-course antibiotic therapy, and well-planned, early surgical intervention substantially improved the outcome of our patient. On initial examination, few clues indicated the development of pyopneumopericardium. This case report reinforces the difficulty of this diagnosis and perhaps the need for clinical awareness and inclusion of this entity in a differential diagnosis.

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REFERENCES