Esophagoatrial Fistula with Previous Pericarditis Complicating Esophageal Ulceration*

Report of Two Cases and a Review of the Literature

Richard W. Snyder, M.D., F.C.C.P.; Peter R. Dumas, M.D.; and Byron E. Kolts, M.D.‡

Sixteen cases of nontraumatic left atrial-esophageal fistulas have been reported previously. These fistulas usually result from chronic peptic esophagitis or cancer. The diagnosis is suggested by the triad of chronic dysphagia, hematemesis, and acute neurologic signs. There may be cardiac manifestations such as pericarditis, atrial fibrillation, or shock. An unusual feature of these fistulas is systemic embolization of food, air, or septic necrotic debris which may result in sudden central nervous system symptoms. All reported cases resulted in death due to hemorrhage, although there was often a variable time interval between the onset of hematemesis and the patient’s death. The authors report two additional cases in which an episode of pericarditis preceded fistula development. Based on these 18 cases, the spectrum of esophagoatrial fistulas is reviewed, as well as the signs which may herald fistula development.

(CHEST 1990; 98:679-81)

Nontraumatic fistulas between the esophagus and the left atrium are rare events; only 16 cases have been reported. The authors had the unusual opportunity to review two cases.

CASE REPORTS

CASE 1

A 59-year-old woman presented with worsening pyrosis and history of gastric fundoplication four years earlier. Physical examination results were unremarkable. A barium swallow showed a hiatal hernia with a small ulcer at the esophagogastric junction, and an antireflux procedure was performed. Eight months later, the patient was readmitted for severe substernal chest pain, fever of 38°C, and a pericardial friction rub. An electrocardiogram was normal, and the echocardiogram showed a pericardial effusion. Symptoms resolved, and a diagnosis of idiopathic pericarditis was made. Upper endoscopy was performed which revealed free reflux of gastric contents into the esophagus with Barrett’s epithelium and two ulcers in the distal esophagus. Symptoms of stricture developed, and the patient was treated with periodic bougienage. Three years later, the patient presented with a vomiting episode followed by syncope. The next day after bougienage, the patient had a generalized seizure followed by profuse hematemesis and death.

The post-mortem examination revealed distorted esophageal anatomy with fibrosis at the esophagogastric junction due to prior surgery. In addition, a large, deep ulcer measuring 3.8 × 2.5 cm on the anterior wall of the distal esophagus was adherent to the wall of the left atrium. At the base of the ulcer was a perforation measuring 4 mm in diameter extending into the left atrium (Fig 1).

Histologic examination showed that sections of the esophagus were lined with gastric type glandular epithelium. At the deepest part of the ulcer, there was an area of necrosis and communication between the lumen of the left atrium and the ulcer. The brain had minimal atrophy without focal lesion.

CASE 2

This 74-year-old white man presented complaining of severe substernal burning aggravated by reclining or by eating large meals. He had a past history of peptic ulcer disease and mild chronic obstructive pulmonary disease. The physical examination results were unremarkable. A barium swallow and endoscopy showed a distal esophageal stricture and two esophageal ulcers. Two years later, a chest x-ray film revealed a solitary nodule, and a left upper lobectomy was performed which revealed adenocarcinoma of the lung with two positive hilar nodes. The patient subsequently received radiation therapy to the chest.

One year after thoracotomy, the patient returned to the hospital because of fever to 39.4°C, pleuritic chest pain, and a pericardial friction rub. An echocardiogram showed a small pericardial effusion, and a diagnosis of idiopathic pericarditis was made.

Four years after thoracotomy, the patient was brought to the emergency room in a disoriented state. He had been suffering from substernal chest pain for one month and had been receiving

*From the Department of Medicine, ‡Abington Memorial Hospital, Abington, PA, and the †University Hospital, Jacksonville, FL. Manuscript received December 7; revision accepted February 2.

Figure 1. Portmortem specimen from first case demonstrating a probe through the esophageal ulcer with tip in the left atrium.

CHEST / 98 / 3 / SEPTEMBER, 1990 679
indomethacin plus antacids for ten days. The patient had an expressive aphasia and was clutching his head in pain. Within minutes, the patient began vomiting, became unresponsive, hypotensive, and died.

Post-mortem examination of the lungs showed no evidence of recurrent tumor. On the anterior wall of the esophagus, just below the bifurcation of the trachea, a large ulcer measuring 8 cm in its greatest dimension was found. The base of the ulcer was adherent to the wall of the left atrium. A defect in the left atrium, measuring 3 mm in diameter, communicated with the lumen of the ulcer (Fig 2).

Histologic evaluation of the esophageal ulcer showed granulation tissue and foreign body giant cells but no evidence of malignancy. The wall of the atrium adjacent to the atrial fistula was infarcted with only ghost-like remnants of muscle cells remaining. Examination of the brain was not allowed.

**Discussion**

The first case of a nontraumatic left atrial-esophageal fistula was reported in 1874 in a 24-year-old soldier with a mediastinal abscess which penetrated the esophagus and left atrium.¹ With the reports of 17 additional cases (including the authors' cases), the spectrum of this disorder can now be appreciated.

The average age of patients is 50 years, with a range from 13 to 80 years. The formation of an esophagoatrial fistula seems to require prolonged, chronic inflammation in order to obliterate the pericardial space with fibrosis, permitting the esophagus to adhere to the myocardium. The average duration of esophageal symptoms, in the 17 cases where this was recorded before fistula formation, was nine years.

The majority of cases were due to chronic reflux esophagitis (ten of 18) and were related to benign esophageal ulcers. Six of these ten cases had chronic strictures. Three cases of the 18 were due to esophageal cancer. Nearly half of the reported cases had either previous surgery (six patients) or irradiation (two patients).

Left atrial-esophageal fistula often presents with the clinical triad of hematemesis and CNS changes in a patient with chronic dysphagia. Of the 18 known cases, GI bleeding was present in 16 patients, and 12 had long standing dysphagia. Central nervous system symptoms have been recognized in 16 of 18 patients. Eight patients had focal CNS symptoms; and eight had diffuse, nonspecific changes. Systemic emboli were documented in seven subjects and suspected in four others. In five patients emboli were caused by food particles. One was due to tumor pieces, and another was caused by air. In four additional patients, the presence of either focal infarction or microabscesses suggested embolization, but the actual emboli were not found.

Systemic emboli may result from a temporary reversal in pressure differences between the left atrium and the esophageal lumen. The left atrium is a relatively low pressure chamber. During regurgitation, the intraesophageal pressure can transiently exceed the pressure in the left atrium permitting entrance of esophageal contents.³⁻⁸

The two cases reported by the authors are unique because both patients developed symptoms and signs consistent with pericarditis years before a fistula was identified. Prior reports have not emphasized cardiac involvement, but when reviewed with a focus on cardiac involvement, it was often present. Six patients in addition to the authors' cases had pericarditis at autopsy as evidence by more than 50 ml of pericardial effusion and/or inflammation of the pericardium. Six patients had atrial fibrillation. One patient developed an acute myocardial infarction at the time of fistula formation. It has been postulated that chronic inflammation is necessary to obliterate the pericardial space.⁸ This obliteration of the pericardial space allows an esophagoatrial fistula to form rather than a esophago-pericardial fistula. We postulate that this chronic inflammation of the pericardium and/or atrium may manifest itself clinically as pericarditis or atrial fibrillation.

Another feature of one of the authors' cases which should be emphasized is the finding of Barrett's epithelium. Studies estimate that 10 to 12 percent of patients with uncomplicated chronic gastroesophageal reflux develop the glandular metaplastic epithelium typical of this disorder. Of the 18 cases in this review, nine had chronic reflux symptoms, with five patients recognized as having Barrett's epithelium. This observation suggests that of this epithelial lesion may be associated with fistula formation.

Current data suggest that there may be a variable interval of time between erosion into the atrium (associated with a sudden onset of neurologic symptoms and/or hematemesis) and the patient's death. This interval can range from less than an hour to ten
days, with an average of 55 hours, in the 15 cases where this can be estimated. Air in the atrium as seen by chest x-ray film16 may also be an important clue. An esophagram or fiberoptic upper endoscopy may be helpful in documenting the size, depth, and location of an esophagogastric ulcer with the potential of fistulization. Fistulas reported in this article all occurred on the anterior wall of the esophagus and were between the level of the carina and the esophagogastric junction. The ulcers were characterized as deep and at least 1 cm in diameter.

The report by West et al17 of four cases, as well as a review of 91 additional cases of benign ulcers penetrating the pericardium and heart, gives further perspective to the clinicopathologic features of this disorder. Similar predisposing factors are noted for other esophagocardiac fistula. These include previous surgery in the esophagogastric region and longstanding physical or chemical injury such as lye ingestion or irradiation. Pericarditis alone was noted in 44 cases, but a history of previous pericarditis was not noted in the 51 cases of atrial or ventricular perforation reviewed by this author.

Lehmann et al18 reported a 46-year-old man with a right atrial-esophageal fistula demonstrating the unique features of fistulous connection to the right rather than the left atrium, as well as successful surgical cure. Aortoesophageal fistula has also been reported19 with review20 revealing thoracic aortic aneurysm to be the most common cause of this disorder.

To the present time, no patient has survived the formation of left atrial-esophageal fistula. Early recognition and prompt surgery offers the only realistic chance for survival. The diagnosis should be suggested by the triad of chronic dysphagia, hematemesis, and CNS abnormalities. Previous history of pericarditis, as well as current findings of cardiac involvement, further support the diagnosis. The finding of an ulcer greater than 1 cm in diameter located on the anterior wall of the esophagus distal to the carina and possibly associated with Barrett's epithelium completes the diagnostic evaluation. Whether surgical intervention following this diagnostic approach will change the prognosis of this condition is unknown and awaits further study.

ACKNOWLEDGMENTS: The authors would like to acknowledge the secretarial efforts of Elaine Melair and the assistance of Ewa Grudziak, M.D., with reviewing the non-English language references.

REFERENCES
1 Gaz. Hebdomad. 1874; d459 Schmidt Jarib. BD. 167, d153
3 Prolla JC, Taebel DW, Kirser JB. Perforation of an esophagogastric ulcer into the left atrium. Gastroenterology 1967; 52:871-74
5 Ibashi HI, Granada I.O. Cerebral food embolism secondary to esophagocardiac perforation. JAMA 1972; 291:373-75
8 Mott JM, Austin GE. Cerebral embolization resulting from esophageal-atrial fistula. Arch Intern Med 1976; 136:718-20
10 Nadine M, Schaffer R, Wodacz R. Cerebral arterial air embolism due to an esophago-atrial fistula seen on CT. Neuroradiology 1979; 18:103-06
11 Murphy GF, Raymond AK, Scannell JG. Esophageal-atrial perforation due to recurrent esophagitis 18 years after esophageal bypass surgery. J Thorac Cardiovasc Surg 1979; 78:181-84
13 Brynjolfsson G, Kania R, Beckeris L. Gastroesophageal cardiac fistula due to perforation of an esophagogastric anastramotic ulcer into the left atrium. Hum Pathol 1980; 11:677-79
15 Fldmark P. Case of the summer season: spontaneous fistula between the esophagus and the left atrium. Sem Roentgenol 1983; 18:171-72
17 West AB, Nolan N, O'Brien, DS. Benign peptic ulcer penetrating pericardium and heart: clinicopathologic features and factors favoring survival. Gastroenterology 1988; 94:1478-87
18 Lehmann KG, Blair DH, Siskind BN, Wohlgelernter D. Right atrial-esophageal fistula and hypodensopneumopericardium after esophageal dilation. Chest 1987; 9:969-72