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CASE REPORT

CLINICAL CASE

Hydropneumopericardium Due to a Traumatic Esophageal-Pericardial Fistula

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ABSTRACT

Esophago-pericardial fistula is a rare, life-threatening condition, usually arising as a complication of benign esophageal disorders or iatrogenic causes. Prompt diagnosis via multimodality imaging is crucial, with computed tomography being the most sensitive. Management varies based on severity, with a growing trend toward early endoscopic interventions, which result in improved outcomes. (J Am Coll Cardiol Case Rep 2024;29:102357) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 75-year-old man presented to the emergency department with a 2-day history of fever, shortness of breath, productive cough, and sharp left-sided chest pain. On arrival, his temperature was 101°F, heart rate of 104 beats/min, blood pressure of 90/60 mm Hg, and oxygen saturation of 90% on room air. On physical exam, he appeared diaphoretic and tachypneic,

LEARNING OBJECTIVES

- To identify the clinical features and presentation of an EPF and choose the right imaging tests so that a correct diagnosis may be made without delay.
- To appreciate the multimodality imaging findings in EPF and differentiate them from other causes of pericardial pathology to ensure appropriate treatment.

and coarse crackles were auscultated in both lung bases. He had normal heart sounds without a gallop, murmur, or friction rub. His extremities were warm, without signs of peripheral edema or jugular venous distention.

PAST MEDICAL HISTORY

He had a history of ulcerative esophagitis due to severe gastroesophageal reflux disease complicated by complex esophageal strictures requiring regular dilations with Savary and balloon dilators, with the latest one occurring 2 weeks before this presentation and was apparently without incident. Previous esophageal biopsies ruled out malignant or infectious causes. He had no known cardiac history and transthoracic echocardiography (TTE) 6 months prior showed left ventricular ejection fraction of 55% to 60% with no significant valvular or wall motion abnormalities.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

ABBREVIATIONS AND ACRONYMS

AEF = atrio-esophageal fistula

CT = computed tomography

- EP = esophageal perforation
- EPF = esophago-pericardial fistula

TTE = transthoracic echocardiogram

DIFFERENTIAL DIAGNOSIS

The initial differential diagnosis included respiratory infections, early systemic inflammatory response syndrome, heart failure, and ischemia. Given his history of esophageal dilations, other possibilities were aspiration pneumonia or an esophageal perforation (EP).

INVESTIGATIONS

Laboratory testing was notable for a white blood cell count of $15.0 \times 10^3/\mu$ L, an elevated lactic acid level of 4.8 mmol/L (normal 0.5-2.0 mmol/L), an elevated C-reactive protein of 355.5 mg/L (normal <10.0 mg/L), and troponin-I 0.014 ng/mL (normal <0.04 ng/mL). Electrocardiogram showed low voltage and sinus rhythm with ST-segment elevation in leads II, III, aVF, and V₃-V₆ (Figure 1). TTE identified a large circumferential pericardial effusion with fibrinous material but no signs of tamponade (Figure 2, Video 1). A computed tomography (CT) scan of the chest demonstrated hydropneumopericardium and pneumomediastinum with a paraesophageal foci of air as well as a distended esophagus (Figure 3, Video 2).

MANAGEMENT

He was initiated on vasopressors, piperacillintazobactam, and fluconazole, and underwent urgent pericardiocentesis with pericardial drain placement and 375 mL of straw-colored fluid evacuated. His hemodynamics improved, and vasopressors were weaned over the next 12 hours. Given his history of esophageal ulcerations/strictures requiring frequent dilation and identification of hydropneumopericardium, there was a concern for esophago-pericardial fistula (EPF), prompting a contrast esophagram on day 2. This identified mucosal irregularities and contrast extravasation outside the esophageal border, suggestive of an EP, and given the TTE findings, a fistula to the pericardial space was diagnosed (Figure 4B). He was taken later that day for esophagogastroduodenoscopy; this identified ulcerations and severe esophagitis 23 cm from the incisors as well as a stricture that could be traversed only with a neonatal scope (Figure 5). A fully covered 23 mm imes 120 mm esophageal stent (Endomaxx, Merit Medical) was placed across the stricture and areas of ulceration. A repeat esophagram was obtained the following day, which did not identify any contrast extravasation (Figure 4C).



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FIGURE 2 Transthoracic Echocardiography: Subcostal 4-Chamber View



A moderate pericardial effusion (echolucency) is present (arrows).

Pericardial fluid cultures grew *Streptococcus vestibularis* and *Candida albicans*. His pericardial drain was removed after 6 days when the output fell to <50 mL/24 h and after a repeat TTE showed no significant residual effusion.

DISCUSSION

EPF is a rare and diagnostically challenging disorder with a significant mortality risk, especially if the diagnosis is delayed and complicated by tamponade.¹ Most EPFs are attributable to non-neoplastic esophageal disorders, including foreign body ingestion, Boerhaave syndrome, and chronic esophagitis (**Table 1**).² With the frequent use of endoscopic procedures, iatrogenic causes have now become the leading cause of EP.² A recent multinational study (POTTER-AF [PrOgnosis following oesophageal fis-Tula formaTion in patients undergoing cathetER ablation for AF]) that analyzed 553,729 catheter ablation procedures for atrial fibrillation revealed that



⁽A) Axial, (B) coronal, and (C) sagittal views, showing pneumopericardium (white arrow), a complex pericardial effusion (asterisk), and pneumomediastinum (yellow arrow).



(A) Three weeks before hospitalization: proximal esophageal dilatation and distal narrowing (arrow) due to esophageal stricture, with no evidence of extraluminal contrast to suggest perforation. (B) Day 2 of hospitalization: esophageal mucosal irregularity with extravasation of contrast (arrow) suggestive of perforation. (C) Day 3 of hospitalization: esophageal stent in place (arrow) without evidence of contrast extravasation.

the post-procedural incidence of esophageal fistulas was 0.025%.³ Of these, atrio-esophageal fistulas (AEFs) accounted for 95.8% of cases, whereas EPFs were seen in just 3.4% of patients.³ The study noted a high overall mortality rate of 65.8% (for AEF cases; there was 0% mortality in the 4 EPF cases in this registry), with 18.8% of patients suffering long-term complications.³ Most of the published data on EP have combined cases of EPF, AEF, and perforation into the mediastinum without fistula formation;



(A) Three weeks before hospitalization showing a distal esophageal stricture (arrow). (B) Day 2 of hospitalization revealing severe esophagitis (arrowhead) and ulceration (asterisk) in the distal esophagus, likely the site of perforation.

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TABLE 1 Various Causes of Esophagopericardial Fistula	
Category	Common Causes
latrogenic	Complication of ablation procedures for atrial fibrillation or ventricular tachycardia (most common cause), esophagogastroduodenal endoscopic procedures, esophageal surgery (esophagectomy, Nissen fundoplication), or radiotherapy
Foreign body ingestion	Fish bone, caustic substances, very hot liquids
Trauma	Penetrating or blunt chest injury
Neoplastic	Esophageal tumors (primary or metastases), compression from tumors originating from adjacent structures
Barotrauma	Severe vomiting (Boerhaave syndrome), severe straining (childbirth, weightlifting)
Inflammatory	Esophagitis, esophageal ulcers, systemic disorders affecting the esophagus (eg, scleroderma)
Infectious	Esophageal infections (eg, human immunodeficiency virus, tuberculosis, candida) and complications of esophageal infections (eg, abscess formation)
Miscellaneous	Barrett's esophagus, achalasia, and esophageal diverticulum

however, EPF is significantly rarer.³ Consequently, our understanding of EPF's presentation, prognosis, and management is somewhat limited. AEF typically presents with hematemesis and neurological symptoms due to embolism, while EPF may present with fever, cough, dyspnea, or nonspecific symptoms; as a result, diagnosis may sometimes be delayed by 1 to 2 weeks. As mentioned, EPF has a lower mortality rate compared with AEF, and nonsurgical management might be effective in some cases of EPF.^{3,4}

The clinical manifestations vary depending on the cause, location, size of the perforation, and the duration since the initial injury, with symptoms potentially emerging weeks later, ranging from mild and nonspecific symptoms to severe and life-threatening complications like pericardial infection and cardiac tamponade.^{1,2,4} Typical symptoms of EPF include chest or epigastric pain along with fever, dysphagia, subcutaneous emphysema, cough, and dyspnea.^{4,5} In more severe cases, systemic signs may involve fever, tachycardia, and hypotension.^{1,4,5} The air and blood mixing produces a "bruit de moulin" or water wheel murmur, heard on auscultation in the cases of hydropneumopericardium or intracardiac air.⁶

Multimodality imaging is vital in both the visualization and diagnosis of EPF. TTE is crucial for the initial assessment of air and/or fluid in the pericardium and imaging features of tamponade but lacks direct visualization of EP or fistulous tracts, necessitating additional imaging.² Transesophageal echocardiography should not be performed to assess for pericardial pathology (including EPF) or endocarditis if there is a possibility of EP. Pneumopericardium may demonstrate a radiolucent rim, the "continuous diaphragm sign," on plain radiographs, but it has limited sensitivity and specificity for EPF.² Contrast esophagography is usually the initial imaging modality for suspected EP, but has an overall false negative rate of 10%.² CT with oral and intravenous contrast is the most sensitive diagnostic modality for detecting EP, with a sensitivity of 59% to 100% and specificity of 80% to 100%.² CT is also crucial for evaluating the extent of involvement with adjacent structures and guiding treatment.²

In our case, although direct communication between the esophagus and pericardium was not visualized on the esophagram, EPF is very highly likely due to the presence of esophageal leak, air in the pericardium, and isolation of oral flora from pericardial fluid cultures. We postulate that repeated trauma from esophageal dilatations, along with underlying chronic inflammation from gastroesophageal reflux disease, led to EPF development. To the best of our knowledge, this case represents the first reported occurrence of an acute EPF following esophageal dilatation for an esophageal stricture.

Management varies depending on the severity of presentation, with surgical intervention recommended for cases of EPF associated with extensive mediastinal and pericardial food-material contamination, ongoing bleeding or hematoma accumulation, or abscess formation.⁷ However, there is a growing trend toward using a combination of endoscopic stenting and pericardial drainage, particularly when cases are diagnosed early and the patient is stable. When cardiac tamponade complicates the picture, urgent pericardial decompression, either surgical or percutaneous, is necessary.¹ Conservative measures may not be adequate except in cases of contained micro-perforations. The POTTER-AF study showed that patients who underwent either surgical or 6

endoscopic treatment had significantly lower mortality rates than those who received conservative management only (51.9% vs 56.5% vs 89.5%, respectively).³ Prompt initiation of broad-spectrum antibiotics to cover gram-positive, gram-negative, and anaerobic bacteria is crucial to prevent complications when EP is suspected.⁸ The choice and duration of antibiotics should be guided by culture results, with consideration for the addition of antifungal therapy if needed.⁸

FOLLOW-UP

Our patient was discharged to a skilled nursing facility with a treatment plan consisting of a total duration of 4 weeks of antibiotic (ampicillin/sulbactam) and antifungal (micafungin) therapy. At his 1-month follow-up, he had gained 2 pounds and was doing better without recurrence of esophageal symptoms.

CONCLUSIONS

EPF can be a complication of several esophageal pathologies and may be difficult to diagnose, requiring a high index of suspicion and tailored radiological imaging of the esophagus and cardiac structures with echocardiography, CT, and contrast esophagography. Because of the high mortality rate, prompt diagnosis of EPF is critical to initiate management and prevent complications.

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APPENDIX For supplemental videos, please see the online version of this paper.