Case Reports

Cardiac Tamponade Secondary to Esophagopericardial Fistula

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Abstract

Esophagopericardial fistulas are an extremely rare structural defect that may arise from malignant or iatrogenic etiologies. This article reports the case of a patient with cardiac tamponade secondary to hydropneumopericardium from esophagopericardial fistula. Given the high morbidity and mortality of this condition, this article describes challenges in diagnosis and clinical decision-making to improve early identification and interdisciplinary management.

Keywords: Esophagus; fistula; cardiac tamponade; stents; carcinoma; pericardial effusion; pneumopericardium

Case Report

Presentation and Physical Examination

51-year-old man presented with new-onset crushing substernal chest pain. He was tachycardic to 117/min and hypotensive to 89/57 mm Hg but afebrile, with a normal respiratory rate and oxygen saturation on room air. He was not experiencing shortness of breath or chills. His initial electrocardiogram was notable only for sinus tachycardia (Fig. 1), but a computed tomography (CT) scan of his chest revealed hydropneumopericardium. A transthoracic echocardiogram showed moderate pericardial effusion with right atrial

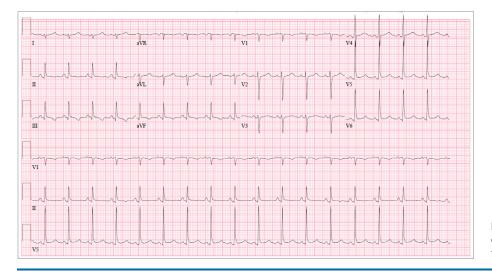


Fig. 1 Initial electrocardiogram shows sinus tachycardia.

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systolic collapse and right ventricular diastolic collapse consistent with tamponade physiology (Fig. 2). Urgent pericardiocentesis drained 320 mL of serosanguinous fluid with intermittent air bubbles, and a pericardial drain was placed.

Medical History

The patient's medical history was significant for Siewert III gastroesophageal adenocarcinoma diagnosed 4 years earlier. The patient underwent concurrent chemoradiation with leucovorin calcium (folinic acid), fluorouracil, and oxaliplatin (FOLFOX). Seven months later he underwent exploratory laparotomy and aborted esophagectomy when intraoperative findings revealed liver metastases. He underwent subsequent chemotherapy with leucovorin calcium (folinic acid), fluorouracil, and irinotecan hydrochloride (FOLFIRI), followed by palliative immunotherapy with pembrolizumab. He had been undergoing definitive chemoradiation for the past 3 years. He most recently received radiation therapy 12 months before and chemotherapy 3 weeks before. In addition, an episode of dysphagia 8 months before this episode prompted esophagogastroduodenoscopy (EGD), with dilatation of stricture, brush biopsy, and stent placement. His disease was restaged 2 weeks before this presentation for chest pain, with no evidence of disease activity.

Differential Diagnosis

The differential diagnosis for this patient's presentation included malignant pleural-pericardial effusion, chylopericardium secondary to thoracic duct abnormality, bronchopericardial or tracheopericardial fistula, and esophagopericardial fistula.



Fig. 2 Initial transthoracic echocardiogram shows a moderate circumferential pericardial effusion with suspected pericardial tamponade physiology.

Key Points

- Maintain a high index of suspicion for patients with a history of malignancy, prior esophageal stent, cardiac ablation, or esophageal injury (eg, caustic ingestion or radiation) because prompt diagnosis and treatment of esophagopericardial fistulas are needed to prevent sequelae such as tamponade or infection.
- Recognize that lack of expected contrast extravasation during esophagram does not necessarily exclude the presence of a fistula.
- When first-line diagnostic measures do not reveal a fistula but clinical suspicion remains high, biochemical analysis of the accumulating fluid may reveal the presence of an esophageal fistula.

Abbreviations

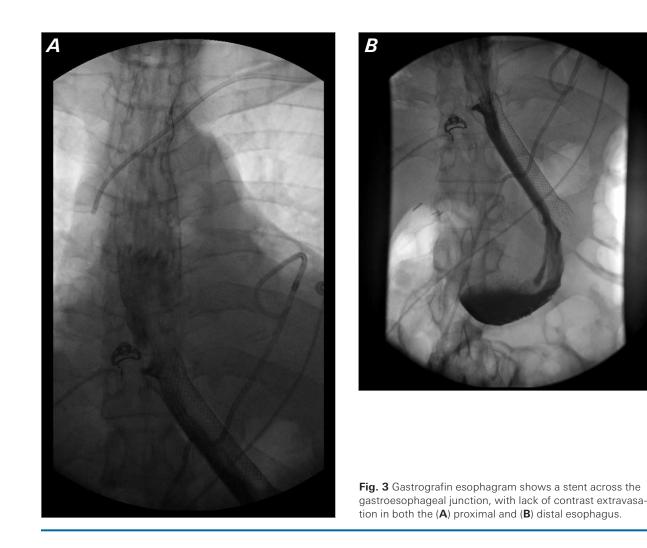
CT, computed tomography EGD, esophagogastroduodenoscopy

Technique

Because of concern for pericardial effusion secondary to esophageal fistula based on CT findings, the patient underwent EGD and esophagram for further evaluation. No obvious leak was observed on intraprocedural fluoroscopy imaging (Fig. 3), but there was tissue invasion into the esophageal stent. It was noted that the leak may have originated adjacent to this original stent and therefore may have been inaccessible by endoscopic means. Given that the original stent was embedded, it could not safely be entirely removed endoscopically. Therefore, a new stent was placed within the original stent to ensure tissue necrosis and safe removal 6 weeks later. The pericardial drain continued to have greater-than-expected output, which consisted mostly of brown, murky fluid with occasional food particles.

The patient subsequently underwent contrast CT studies and repeat EGD, but again, no extravasation of contrast was seen (Fig. 4). With continued suspicion that an unseen fistula may be present, the patient was given drinking water with FD&C Blue #1 Dye. Only when the pericardial drain output revealed blue fluid was the presence of a fistula confirmed. Laboratory analysis of the fluid further confirmed that the accumulations were of gastrointestinal origin, revealing an amylase of 13,000 U/L and fluid pH of 4.2. This fluid analysis and blue dye finally verified the suspected esophageal to pericardial flow. Notably, fluid cytology was negative for malignancy and infection.

After clinical and laboratory confirmation that this patient had a pericardial effusion secondary to esophageal fistula leading to cardiac tamponade, the tentative plan



was to proceed with esophagectomy with diversion. An additional CT scan revealed a small amount of free contrast and air within the pericardial space. Another EGD was performed to reposition or replace the 2 existing stents to try to gain control of the fistula. During the procedure, both esophageal stents were removed and revealed a 30-mm fistula in the lower one-third of the esophagus. A new 23-mm × 15.5-cm WallFlex covered stent (Boston Scientific) was placed under fluoroscopic guidance.

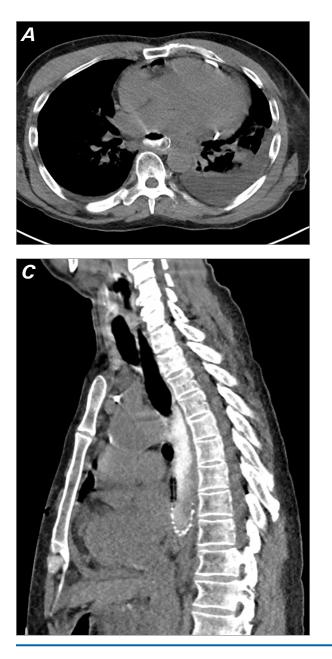
In the days after this EGD, the fistula output remained high. The difficult decision was made to proceed with esophagectomy. Given the patient's poor nutritional status, it was decided to perform a 3-hole esophagectomy with diversion rather than concomitant reconstruction.

Outcome and Follow-Up

When the esophageal stent was removed, a 3.5-cm to 4-cm fistula was seen just above the gastroesophageal junction, and the pericardial drain and the heart itself were visible. During the abdominal portion of the operation, densely adherent tissues were revealed in the upper abdomen, and the spleen could not be dissected free from the stomach. Ultimately, the patient underwent exploratory laparotomy, esophagectomy, partial gastrectomy, partial splenectomy, gastrostomy, and jejunostomy 1 month after his initial presentation. After an extended postoperative course, the patient was discharged home with interdisciplinary follow-up.

Discussion

Pericardial effusions and subsequent tamponade are not an uncommon finding in clinical practice. Causes of pericardial effusions include history of recent myocardial infarction, uremia from end-stage kidney failure, autoimmune conditions, connective tissue disease, and malignancy.¹ Although many clinical reviews of the diagnosis, etiology, and management of pericardial effusions and tamponade exist, pericardial





effusions secondary to esophagopericardial fistulas are so rare that they are not even mentioned in the majority of the current review articles.¹⁻⁴

Little data exist on the incidence and prognosis of esophagopericardial fistulas. Most of the current literature consists of case reports discussing esophagopericardial fistulas stemming from various etiologies, such as cardiac ablation⁵; esophageal procedures, including surgery or stenting⁶⁻⁸; direct injuries, such as from caustic ingestion or radiation^{7,9,10}; and malignancy.^{11,12} Although esophageal fistulas are an established complication of esophageal carcinoma, these fistulas are

Fig. 4 Computed tomography scan of the chest with oral contrast shows no evidence of contrast extravasation or communication with the pericardium on (**A**) axial, (**B**) coronal, and (**C**) sagittal views.

most commonly esophageal-respiratory^{8,10,13} with only a few case reports discussing esophagopericardial fistulas.^{7,11,12,14} One literature review found that only 70 cases of esophagopericardial fistulas have been reported since 1931, with only 20% of those being secondary to malignancy.¹⁴ Given the low incidence of this extremely rare diagnosis, data are insufficient to compare the incidence of esophagopericardial fistulas with and without prior esophageal intervention or instrumentation. Therefore, although a history of esophageal instrumentation could raise suspicion for a possible esophagopericardial fistula, lack of prior esophageal intervention certainly does not exclude this possibility. Esophagopericardial fistulas most often present with new fever or chest pain and may show hydromediastinum or pneumomediastinum, hydropericardium or pneumopericardium, or leukocytosis. Although complications of infection and inflammation, such as recurrent pneumonia or pericarditis, are common sequelae for all esophageal fistulas,¹³ an esophagopericardial fistula presents the unique and life-threatening possibility of cardiac tamponade.

Diagnosis of esophageal fistulas is often confirmed on imaging modalities such as radiography, esophagography, endoscopy, CT, or magnetic resonance imaging. Diagnosis can also be made by direct visualization intraoperatively. A previous study showed that although pneumopericardium and hydropneumopericardium are the most common radiographic findings in the presence of an esophagopericardial fistula, they are seen on chest x-ray in up to 50% of cases.¹⁵ Therefore, this method may not be a reliable way to confirm the presence of a fistula.

When using endoscopy to diagnose an esophageal fistula, it is important to note any suspicion for connection to the heart or pericardium. Endoscopy is avoided when there is a suspected esophageal-intracardiac fistula to prevent insufflation air from entering the atria and causing an air embolus that may lead to stroke or even death.¹⁶ In contrast, when there is a suspected esophagopericardial fistula, insufflation could lead to pneumopericardium and hemodynamic instability. In these cases, the pericardium should be drained beforehand and a pericardial drain left in place.¹⁷

Furthermore, esophagography is the most common diagnostic tool for esophageal wall pathologies such as fistulas and is typically expected to reveal extravasation of contrast into the bronchial tree or mediastinum. As seen in this patient and in other case reports,¹⁸ however, lack of extravasation cannot always exclude the existence of a fistula. In these cases, laboratory analysis of drain output can be a vital tool for clinicians. In fact, these labs were critical in determining the etiology of the accumulating pericardial fluid in this patient, because 2 successive EGDs did not reveal extravasation of contrast. Fluid analysis showing low pH and high amylase levels confirmed that the fluid was gastrointestinal in origin and led to the diagnosis of an esophagopericardial fistula. If imaging and endoscopy have failed to reveal the presence of an esophageal fistula or extravasation yet clinical suspicion remains high, these laboratory studies

should be performed on the fluid to aid in diagnosing the underlying cause of fluid accumulation.

Because of the rarity of esophagopericardial fistulas, there are no firm management recommendations aside from anecdotal evidence in previous case reports that have been managed both surgically and nonsurgically. Emergent pericardiocentesis is imperative in cases of tamponade, as is source control with antibiotics and drainage in the case of purulent pericardial fistulas. After stabilization, management of the fistula varies greatly based on suspected etiology and patient history. Procedural management includes surgical closure, esophageal stenting, or esophagectomy. In contrast, there is evidence that these fistulas can be managed more conservatively with broad-spectrum antibiotics, continuous pericardial drainage, esophageal stenting, and prolonged fasting.^{19,20} Regardless, the paucity of existing guidance underscores the need for current interdisciplinary approaches in parallel with rigorous prospective and retrospective clinical investigations.

Article Information

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