


Esophago-Pericardial Fistulae as a Sequela of Boerhaave Syndrome and Esophageal Stenting: A Case Report and Review of Literature

Journal of Investigative Medicine High
Impact Case Reports
Volume 9: 1–5
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Medical Research
DOI: 10.1177/23247096211036540
journals.sagepub.com/home/hic


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Abstract

Esophago-pericardial fistulae is a rare and dreaded entity. Most reported cases in the literature were described in association with advanced upper gastrointestinal malignancies, prior surgical procedures, and radiofrequency atrial fibrillation ablation. It has been rarely reported in association with benign esophageal conditions. Surgery had been the mainstay of treatment, but there are increasingly reported cases treated successfully with esophageal stenting and pericardial drainage. In this article, we report a novel case of an esophago-pericardial fistulae occurring as a sequela of esophageal stent placed for the management of Boerhaave syndrome.

Keywords

esophago-pericardial fistulae, esophageal stenting, Boerhaave syndrome

Case Presentation

A 36-year-old gentleman with a past medical history of major depressive disorder, non-insulin-dependent diabetes, and hypertension presented with recurrent vomiting and severe substernal chest pain for 2 days. On presentation to the emergency department, he was found to be hypoxic and hypotensive with biochemical evidence of diabetic ketoacidosis (glucose 600 mg/dL, bicarbonate 3 mmol/L, and anion gap 29 mmol/L). The chest radiograph on admission showed a left-sided tension pneumothorax (Figure 1A). A chest tube was emergently placed with drainage of 1.2 L of brownish-yellow drainage with elevated amylase levels. The patient was intubated for hypoxic respiratory failure and was started on broad-spectrum antibiotics. Computed tomography (CT) scan of the thorax showed mediastinal air and distal esophageal thickening suggestive of esophageal perforation (Figure 1B). He underwent emergent esophagogastroduodenoscopy (EGD), which showed a dusky distal esophagus 38 cm from the incisors and left-sided esophageal ulcer 2 cm proximal to the gastroesophageal junction. On insufflation, there was no air leak identified. Given the above findings, a decision was made to deploy a 23 × 150 mm EndoMAXX fully covered esophageal stent under fluoroscopic guidance.

He underwent another EGD 10 days later due to an increase in output and amylase levels from the chest tube coinciding with oral feeding initiation. The stent was found to have migrated partially to the stomach, the position was revised, and a percutaneous gastrostomy tube (PEG) was placed for nutrition. Repeat esophagogram showed no leak. He tolerated the initiation of tube feeds via PEG tube. Cultures from pleural fluid were growing polymicrobial *Candida* species. The patient was discharged to an acute rehabilitation facility after a 3-week hospitalization with a plan for follow-up as an outpatient for removal of the stent.

The patient presented 2 weeks later from the rehabilitation facility with worsening shortness of breath and low-grade fevers. He was noted to be tachycardic to a heart rate of 120 beats per minute and hypoxic, requiring supplemental

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Received April 28, 2021. Revised July 3, 2021. Accepted July 7, 2021.

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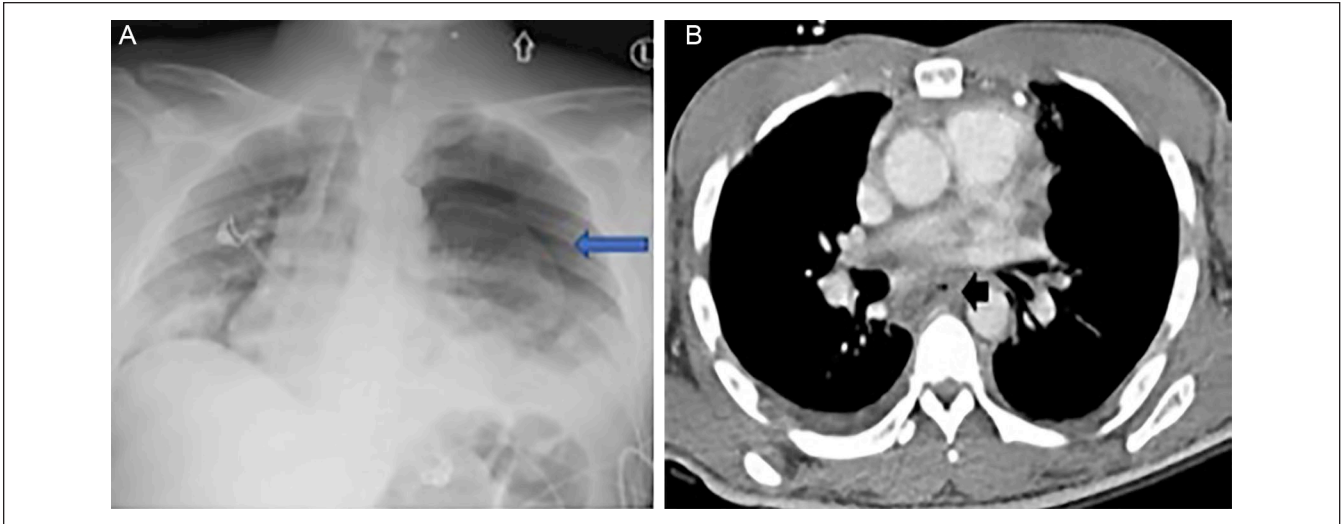


Figure 1. (A) Supine, portable, anteroposterior radiograph of the chest shows left tension pneumothorax (blue arrow). (B) Axial contrast-enhanced computed tomography at the level of the heart shows thickening of the esophagus with air foci anteriorly (black arrowhead).

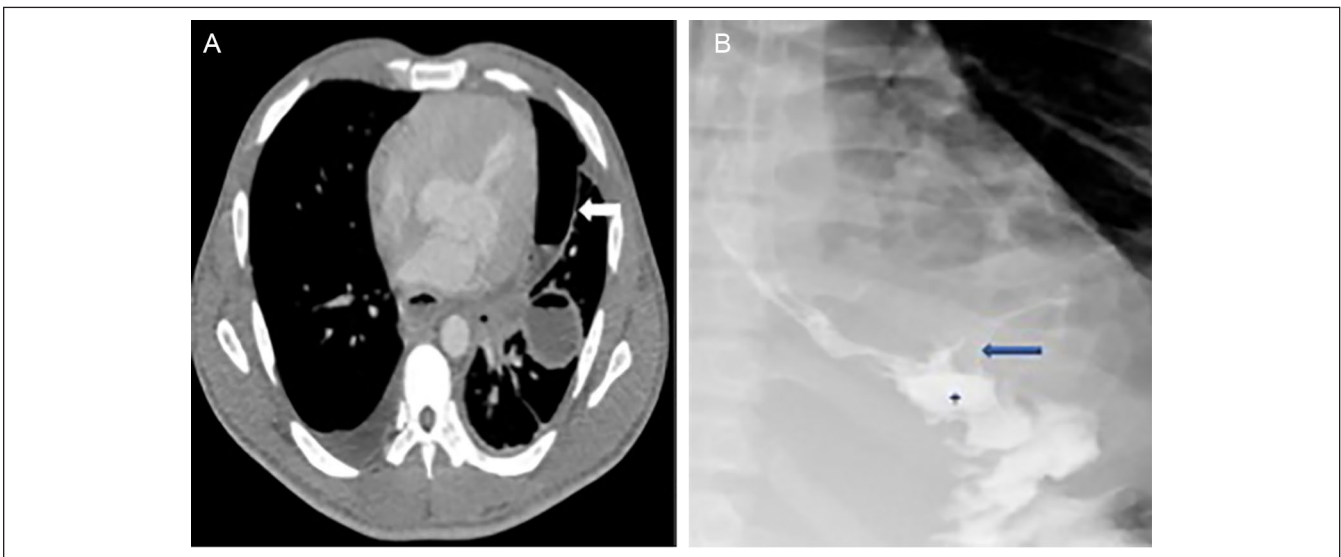


Figure 2. (A) Axial contrast computed tomography through the level of the heart demonstrates loculated pneumothorax (white arrow) and left para mediastinal fluid collection (*). (B) Upright, dynamic esophagogram showed contrast pooling above the diaphragm (+) and a contained contrast leak (blue arrowhead) in the lower esophagus posteriorly.

oxygen. The patient was started on broad-spectrum antibiotic and antifungal coverage. Repeat CT thorax showed a thick-walled left paramediastinal collection with air and fluid levels, new from the prior study (Figure 2A). He underwent CT-guided drainage of the collection; cultures resulted in no growth. Repeat esophagogram showed a contained esophageal perforation (Figure 2B). He underwent EGD with esophageal stent removal. The procedure was uncomplicated. His vitals normalized, and he remained afebrile for 72 hours after the stent removal.

Given his clinical improvement, bolus feeds were restarted via the PEG tube. Shortly after the bolus tube feed initiation, the patient developed severe chest pain, 10/10 in severity, radiating to his back. Pain improved with leaning forward. He was noted to be tachycardic with heart rate of 130 beats per minute with a new oxygen requirement to maintain $SpO_2 > 92\%$. He remained afebrile and normotensive. Electrocardiogram showed sinus tachycardia and ST-segment elevation in anterior leads, V1, V2, and V3 (Figure 3). Troponin levels were within the normal range. Laboratory

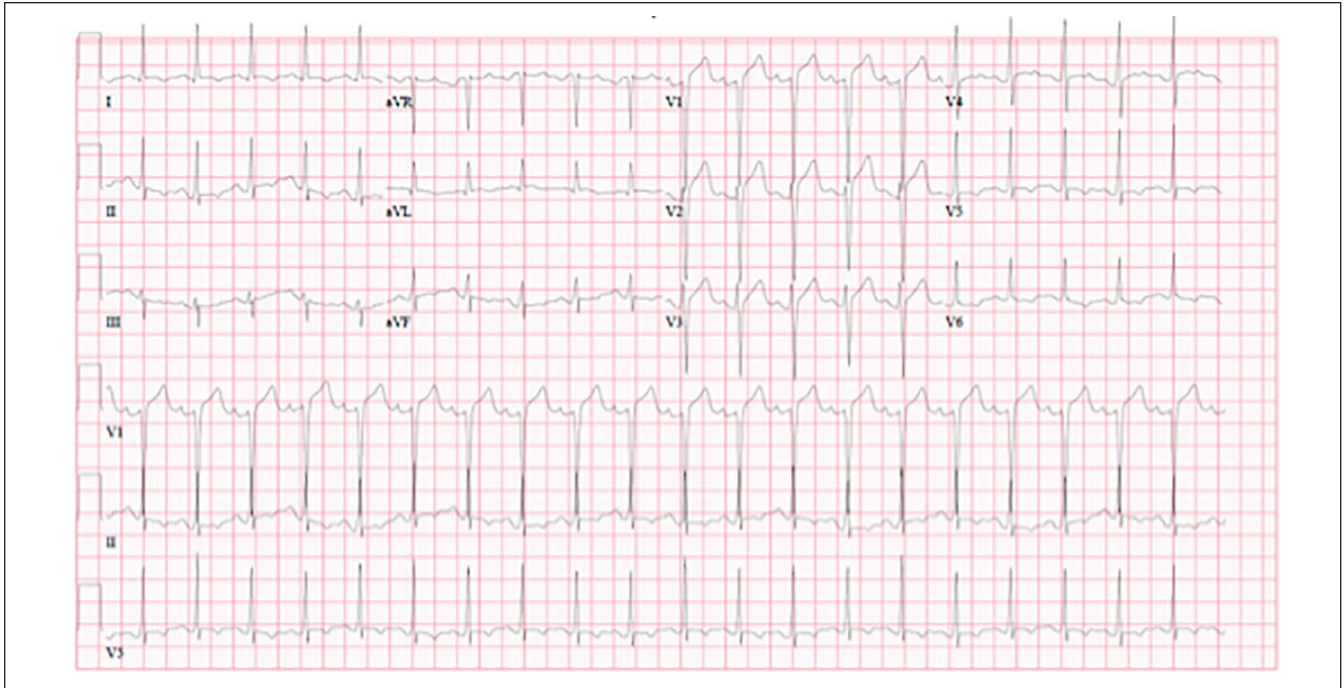


Figure 3. Twelve-lead electrocardiogram showing sinus tachycardia, ST elevation in leads VI, V2, and V3, fulfilling left ventricular hypertrophy criteria.

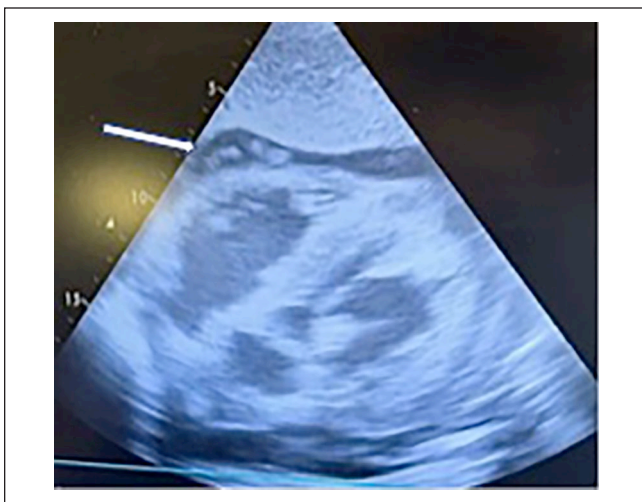


Figure 4. Bedside transthoracic echocardiogram parasternal short-axis view showing a moderate pericardial effusion with multiple echo dense debris (white arrow), new from prior study.

workup was significant for a new leukocytosis with white blood cell count of 14 000 cells/ μ L increased from 7000 cells/ μ L the day prior. Bedside echocardiogram showed a moderate pericardial effusion with echo dense floating debris (Figure 4), which was new compared with a recent study obtained 3 days earlier. There were no features of tamponade seen (Figure 5). CT thorax with intravenous contrast showed a new pneumopericardium concerning for an



Figure 5. Axial contrast computed tomography through the level of the heart demonstrates pneumopericardium (blue arrows), with loss of fat plane between the posterior pericardium and esophagus (white arrow). Additionally, a large pericardial effusion (*) is seen, with bilateral pleural effusions and a loculated left pneumothorax.

esophago-pericardial fistula (EPF). The patient subsequently underwent an emergent pericardial window with partial left lung decortication. Operative findings included an inflamed pericardium with the removal of 600 cc of brownish-yellow pericardial fluid. The pericardial fluid was sent for analysis and was found to be exudative. Cultures grew multiple *Candida* species, including *Candida albicans*. Ten days later, he underwent interval left thoracotomy, left lung decortication, and repair of esophageal perforation via

a diaphragmatic flap. The patient recovered well and was discharged home after 7 days of postoperative recovery on an oral diet.

The post-hospital course was complicated by readmission 1 month later with chest pain, shortness of breath, and difficulty swallowing solid food. He was found to have loculated left pneumothorax and high-grade distal esophageal stricture. He required chemical pleurodesis via pigtail chest tube and serial EGD dilatation with a resolution of both. As of his most recent clinic visit, 3 months following index EPF repair, he remains symptoms-free and tolerating solid food.

Discussion

Not only is EPF rare and challenging to diagnose, but the condition is also associated with high morbidity and mortality that increases exponentially when the diagnosis is delayed. Typically, EPF arises as a complication of esophageal surgery, esophageal injury, or ablation of atrial fibrillation. Rarely, the condition is reported after benign esophageal diseases, such as esophageal ulcers secondary to reflux, caustic ingestions, foreign bodies, and complicated diverticula. It can present anywhere from weeks to months after an initial esophageal insult.¹⁻³

Its symptoms and signs are nonspecific, and include epigastric pain, nausea, vomiting, odynophagia, and dysphagia. Chest pain, fever, and sepsis are usually present.⁴ Depending on how quickly air and fluid accumulate, cardiac tamponade can occur. Acute pericarditis has also been reported and is evident through changes in repeat electrocardiograms. Given these considerations, the best diagnostic modalities are CT with oral and intravenous contrast or magnetic resonance imaging of the esophagus. Typical CT findings include pneumomediastinum, which occurs with or without pericardial effusion. Even if an initial CT scan does not suggest a perforation of the esophageal wall, a repeat scan should be conducted within a few days if clinical suspicion remains high.^{4,5}

Transthoracic echocardiogram has a limited role in the diagnosis of EPF in the available literature.^{1,4} In our case, given the acute changes in our patient's clinical status, it was critical to obtain a bedside echocardiogram quickly as it revealed a newly formed moderate pericardial effusion with multiple free-floating echo-dense particles. We were then able to promptly confirm the presence of the EPF through a CT thorax with intravenous contrast. Most likely, those echo-dense particles represented early fibrinous exudative pericardial debris.

While clinical practice guidelines used to recommend surgical treatment to manage EPF, increasingly, patients are being treated with a combination of endoscopic stenting and pericardial drainage, especially when diagnosed early.^{1,6,7} In a case series on managing EPF following atrial fibrillation ablation, Eitel and colleagues recommended draining of

the pericardial cavity before stenting to reduce the risk of pneumopericardium and hemodynamic instability that could arise as the result of air insufflation during the procedure.¹ If loculations are identified from radiographic or echocardiographic findings, a minimal anterolateral thoracotomy will allow for adequate debridement and drainage.² To enable the fistulae to heal, it is vital to maintain primary source control combined with antimicrobials. Similar to our case, a previous case report by Matta and colleagues isolated the pathogenic yeast, *Candida albicans*, in cultures from the pericardial cavity.⁸ This underscores, in our opinion, the importance of adding empiric antifungal coverage to an EPF patient's treatment plan.

Although there is no definitive evidence of a direct link between esophageal stenting and EPFs, there have been a few reported cases in association with esophageal stenting that have been left in situ for a prolonged period of time in the context of malignancy.⁹ Self-expandable esophageal stents are being increasingly used in a wide array of benign and malignant esophageal conditions. They used to have flared ends to assist with anchoring, but the majority now have finished edges.¹⁰ Even so, if left in place for a prolonged period of time, the radial forces that are required to maintain patency combined with cardiac contractility and esophageal peristalsis could, in theory, create a sheering force that weakens or causes damage to the underlying tissue. In our case, we believe that stent retrieval, which preceded the acute presentation, in addition to the recurrent stent migration and repositioning in already inflamed para-esophageal tissue, all contributed to the development of an EPF through direct esophageal injury.

Conclusion

Esophago-pericardial fistulae is rare and challenging to diagnose. Maintaining a high index of suspicion is key in early diagnosis and prompt treatment. In the appropriate clinical settings, bedside transthoracic echocardiogram could be a valuable tool in the early recognition of the condition. CT with oral and intravenous contrast or magnetic resonance imaging of the esophagus remains the gold standard in diagnosing EPF. With the increasing use of self-expandable esophageal stents in a wide array of benign esophageal conditions, it is important to recognize EPF as a potential complication, and yet there is a paucity of literature on this topic. Raising awareness of this dreaded complication is critical to ensure close outpatient follow-up and timely stent removal, which could potentially help prevent similar cases in the future.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

Ethics Approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed Consent

Verbal informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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References

1. Eitel C, Rolf S, Zachaus M, et al. Successful nonsurgical treatment of esophagopericardial fistulas after atrial fibrillation catheter ablation: a case series. *Circ Arrhythm Electrophysiol.* 2013;6:675-681.
2. Farkas ZC, Pal S, Jolly GP, Lim MMD, Malik A, Malekan R. Esophagopericardial fistula and pneumopericardium from caustic ingestion and esophageal stent. *Ann Thorac Surg.* 2019;107:e207-e208.
3. Luthi F, Groebli Y, Newton A, Kaeser P. Cardiac and pericardial fistulae associated with esophageal or gastric neoplasms: a literature review. *Int Surg.* 2003;88:188-193.
4. Dagues N, Kottkamp H, Piorkowski C, et al. Rapid detection and successful treatment of esophageal perforation after radiofrequency ablation of atrial fibrillation: lessons from five cases. *J Cardiovasc Electrophysiol.* 2006;17:1213-1215.
5. Ha FJ, Han HC, Sanders P, et al. Challenges and limitations in the diagnosis of atrioesophageal fistula. *J Cardiovasc Electrophysiol.* 2018;29:861-871.
6. Wang Y, Liou DZ, Tsai EB, et al. Treatment of a benign esophagopericardial fistula with an esophageal stent—a case report. *Ann Esophagus.* 2020;3. doi:10.21037/aoe.2020.03.05
7. Seo JM, Park JS, Jeong SS. Pericardial-esophageal fistula complicating atrial fibrillation ablation successfully resolved after pericardial drainage with conservative management. *Korean Circ J.* 2017;47:970-977.
8. Matta A, Elenizi K, AlHarthi R, et al. A rare case of *Candida* pericarditis associated with esophagopericardial fistula. *Am J Case Rep.* 2019;20:975-979.
9. Homann N, Noftz MR, Klingenberg-Noftz RD, Ludwig D. Delayed complications after placement of self-expanding stents in malignant esophageal obstruction: treatment strategies and survival rate. *Dig Dis Sci.* 2008;53:334-340.
10. Hindy P, Hong J, Lam-Tsai Y, Gress F. A comprehensive review of esophageal stents. *Gastroenterol Hepatol (N Y).* 2012;8:526-534.