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CASE REPORT

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A rare sequelae of esophageal perforation: Fibrosing mediastinitis

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Abstract

Fibrosing mediastinitis (FM) is a rare disease caused by different causes. If left untreated, the prognosis is poor. The common causes of FM are *Tuberculosis* and *Histoplasma capsulatum* infection. Esophageal perforation is also a rare condition that is often easily under- and mis-diagnosed due to the lack of specificity of symptoms. Here we report a case of FM caused by esophageal perforation.

KEYWORDS
esophageal perforation, fibrosing mediastinitis, foreign bodies

INTRODUCTION

Fibrosing mediastinitis (FM) is a rare disease characterized by the proliferation of fibrous tissue in the mediastinum and compression of surrounding duct structures, including pulmonary arteries, pulmonary veins, and bronchus.[1] Compression of the pulmonary arteries and/or pulmonary veins can eventually lead to pulmonary hypertension, right heart failure, and death. The common causes of FM are tuberculosis and histoplasmic infection.[1]

Esophageal perforation is also a rare and severe complication of esophageal injury, with a mortality rate of 20%–30%.[2,3] Acute esophageal perforation is common and presents with the classic signs of vomiting, chest pain, and subcutaneous emphysema, which can lead to peri-esophagitis and mediastinitis.[2] Those who survive an acute episode are at risk of developing a chronic fistula that manifests itself with symptoms such as dysphagia.[2] However, gastric content, infection et al could triggered massive proliferation of fibrous tissues in mediastinum, which eventually lead to FM with compressing of bronchus and pulmonary vessels.[2,4,5] Here, we reported a rare case of esophageal perforation due to foreign body eventually developing FM.

CASE DESCRIPTION

A 30-year-old man was admitted for exertional dyspnea with a history of esophageal injury by foreign body 9 years ago. Esophagostomy and gastroesophagostomy were performed 6 years ago due to esophageal stenosis. He has no histories of cardiopulmonary disease, *Tuberculosis* infection, immune system disease, and radiotherapy.

On physical examination, his blood pressure was 91/49 mmHg, heart rate 80 beats/min, respiratory rate 20 breaths/min, oxygen saturation 89%, and temperature 36.5°C. His lips were cyanotic, jugular veins were flat, hepatojugular reflux were negative and lungs were clear. The heart was enlarged leftward and systolic murmur (grade 3/6) was found at tricuspid valve auscultatory area.

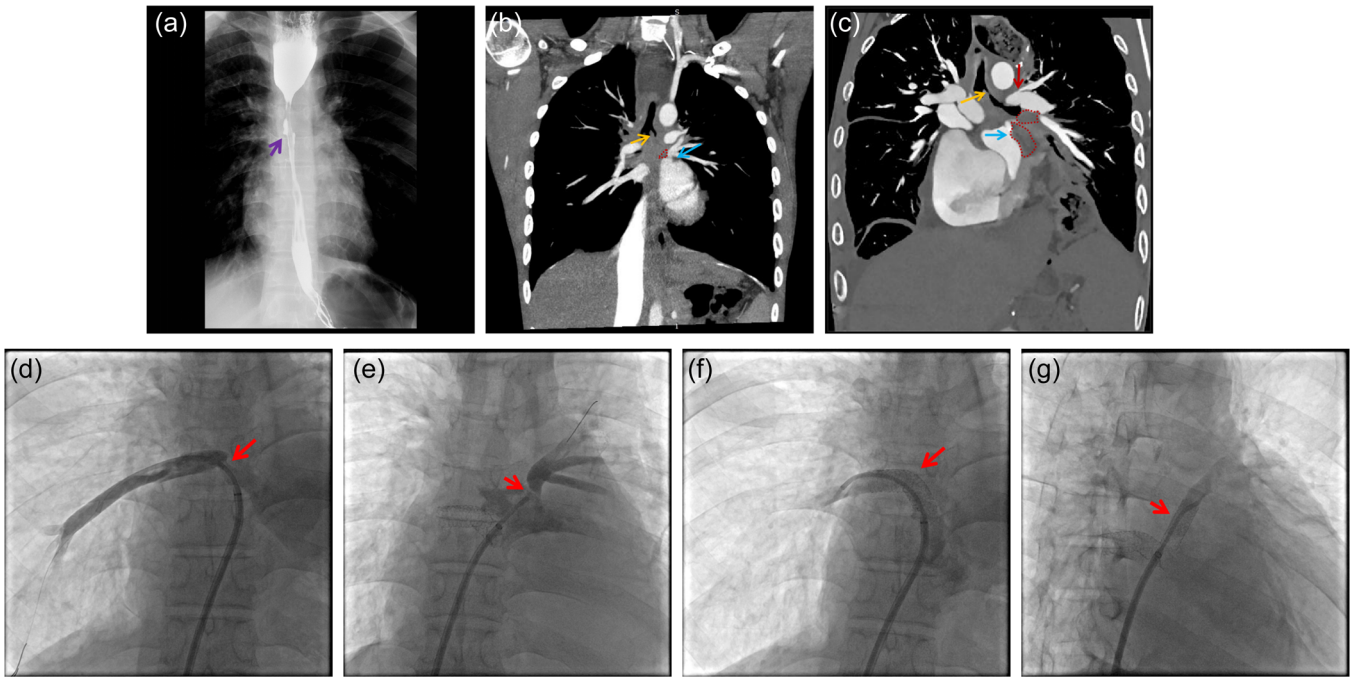

FIGURE 1 (a) Barium meal radiography shows an esophageal fistula 6 years ago (purple arrow). (b) Enhanced computed tomography 6 years ago showed stenosis of pulmonary vein (blue arrow) and bronchus (yellow arrow) compressed by proliferative fibrotic tissues (red dots) in mediastinum. (c) Computed tomography pulmonary angiography demonstrated stenosis of left superior pulmonary vein (blue arrow), left superior pulmonary artery (red arrow) and bronchus (yellow arrow) compressed by proliferative fibrotic tissues (red dots) in mediastinum. Thoracic stomach also can be found. (d) Selective pulmonary vein angiography showed right inferior pulmonary vein stenosis (red arrow). (e) Selective pulmonary vein angiography showed left superior pulmonary vein stenosis (red arrow). (f) Selective pulmonary vein angiography showed stenting in the right inferior pulmonary vein stenosis (red arrow). (g) Selective pulmonary vein angiography showed stenting in the left superior pulmonary vein stenosis (red arrow).

The second heart sound (S2) in pulmonary area was louder than in aortic area. Abdominal examination revealed no tenderness or organomegaly, and pulses in the extremities were symmetrical with no edema. Echocardiography revealed stenosis of left and right pulmonary veins, enlargement of the right heart, widening of the main pulmonary artery, massive regurgitation in tricuspid valve, 106 mmHg of estimated pulmonary artery systolic pressure. A barium meal examination and chest enhanced Computed tomography 6 years ago showed esophageal fistula (Figure 1a) and pulmonary vessel and bronchial stenosis (Figure 1b) caused by FM, respectively. The current CT pulmonary angiography demonstrated the enlarged right atrium and ventricle and stenosis of right inferior pulmonary vein and left superior pulmonary vein compressed by proliferative and fibrotic soft tissues in mediastinum (Figure 1c), which was eventually diagnosed as FM. Further laboratory testing showed no evidence of syphilis, hepatitis, TB, immune system disease, and IgG4 relative condition. Right heart catheterization showed that mean pulmonary artery pressure was 63 mmHg, pulmonary artery wedge pressure (PAWP) 12 mmHg, cardiac index was 1.97 L/(min.m^2), and pulmonary vascular resistance was 22.4 wood units, suggesting precapillary pulmonary hypertension. The pressure gradient across stenotic lesion was 38 mmHg in right inferior pulmonary vein and 15 mmHg in left superior pulmonary vein measured by guiding catheter. Subsequently, two stents were implanted in RIPV and LSPV, respectively (Figure 1d–f) The patient's symptoms were significantly relieved. Aspirin for 6 months and rivaroxaban for 1 year were administrated after stenting. The long-term efficacy of stent implantation has been monitored by regular follow-up. The entire course of the disease and the timeline of diagnosis and treatment for this patient are shown in the supplementary materials (Table 1).

DISCUSSION

FM is a benign and fibroproliferative disease in the mediastinum that compresses the bronchi, pulmonary arteries, and pulmonary veins, leading to pulmonary hypertension and right heart failure.[1] Common triggers

TABLE 1 Timeline for the patient.

Date	Events
February 2011	Esophagus being injured by foreign body during eating.
	Gastroscopy: scarring esophageal stenosis.
June 2014	Presenting with dysphagia for one year.
	Gastroscopy: scarring esophageal stenosis.
	No treatment was given.
January 2015	Progressive worsening of dysphagia and dyspnea
	Gastroscope: failed to passe through.
	Bronchoscopy: bronchial stenosis.
	Barium meal: esophageal fistula and esophageal stenosis
	Enhanced chest CT: proliferative soft tissues in mediastinum, stenosis of bronchus and pulmonary vessels.
	Undergoing esophagectomy for esophageal stenosis.
	Upper gastrointestinal contrast study: no leak and no relevant stenosis postoperative.
January 2021	Admitting for accelerated dyspnea.
	Echocardiography: stenosis of left and right pulmonary veins, enlargement of the right heart, pulmonary hypertension.
	Computed tomography pulmonary angiography: enlarged right atrium and ventricle, stenosis of right inferior pulmonary vein and left superior pulmonary vein compressed by proliferative fibrotic tissues in mediastinum.
	Undergoing percutaneous transluminal pulmonary vein angioplasty with stents.

of FM include *Tuberculosis* and *Histoplasma capsulatum* infection, radiotherapy, surgery, and so forth.[1] These factors trigger abnormal immune response in the mediastinum and abnormal proliferation of fibrous tissue to replace adipose tissue, eventually leading to mediastinal fibrosis.[1] Due to the lack of awareness and specificity of symptoms, FM is often under- and mis-diagnosed.[1]

Esophageal perforation is also a very rare clinical condition. Acute perforation of the esophagus can lead to acute infection, peri-esophagitis, and mediastinitis, with a high mortality rate.[2] A small number of patients with chronic esophageal perforation may present with esophageal stenosis.[2] For this case we reported, imaging supported the diagnosis of FM with pulmonary vein stenosis, bronchial stenosis, and pulmonary artery stenosis, and there was no history or laboratory evidence of *Tuberculosis* infection, *Histoplasma capsulatum* infection, sarcoidosis, auto-immune system condition, or IgG4-related disease. Although surgical procedures can also lead to FM, this patient's pulmonary vein stenosis occurred before surgery. Accordingly, the reason for the FM in this patient may be that the gastric contents stimulation and periesophageal infection after esophageal perforation caused the proliferation of fibrous tissue in the mediastinum, and finally compressed the pulmonary blood vessels and bronchus.[2,4] Acute mediastinal inflammation caused by esophageal perforation is the direct result of gastric content irritation and infection. However, chronic mediastinitis may be caused by an abnormal immune response caused by the above factors. Of course, the specific mechanism still needs further study. Moreover, we cannot completely rule out the possibility of FM caused by other factors. Nevertheless, this case at least suggests that for patients with esophageal injury, in the later follow-up process, we should pay close attention to whether there is pulmonary hypertension and right heart enlargement. If shortness of breath, pulmonary hypertension, and right heart enlargement occur, the possibility of FM should be vigilant. One thing worth noting is that the patient's PAWP was normal, but at the upper limit. This may be related to the original diameter of the narrowed pulmonary veins or to the fact that adequate diuretic therapy has been given before surgery.[6,7]

Finally, we have to acknowledge the limitations of this case. First of all, we did not conduct very comprehensive screening tests for pathogens, such as aspergillosis, and there are some reports that these rare pathogens can also cause FM. Second, we cannot

completely rule out that surgical stimulation accelerates the process of fibrous tissue proliferation.

AUTHOR CONTRIBUTIONS

Yunshan Cao conceived and designed this case report. Jingwen Zhang searched the literature and wrote the first draft of the manuscript. Mingwang Ding made the figures. Aiping Tang searched and sorted the literature. All authors read and approved the final manuscript.

CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

ETHICS STATEMENT

The patient gave written consent to publish the case study.

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