An Unusually Rare but Interesting Co-Occurrence of Idiopathic Pulmonary Artery and Pulmonary Vein Thrombosis: A Case Report

Mohammadbagher Sharifkazemi, MD, MSc, Gholamreza Rezaian, MD, Mohammadhassan Nemati, MD, and Morteza Chehrevar, MD, *Shiraz, Iran*

INTRODUCTION

The endothelium of the pulmonary vascular bed is unique because of its strong fibrinolytic properties. The enhanced blood fluidity in the pulmonary circulation is mediated partly by the presence of a high ratio of tissue plasminogen activator to plasminogen activator-inhibitor 1 in this vascular system, which also benefits from the presence of endogenous heparin-like proteoglycans that contribute to the presence of a nonthrombogenic endothelial surface.[1-3] Any injury to these endothelial cells therefore could disrupt this fluidity and lead to the development of localized thrombosis. In situ pulmonary artery thrombosis is a rare event but may be seen in patients with chronic obstructive pulmonary disease,[4] primary pulmonary hypertension,[5] or any underlying cause of pulmonary inflammation.[6]

Although in situ pulmonary artery thrombosis is an unusual finding, its concomitant presence with pulmonary vein thrombosis (PVT) is even more uncommon. Here we present such an interesting case.

CASE PRESENTATION

A 44-year-old male welder with a history of heavy smoking was rushed to the emergency department of our university hospital with severe air hunger despite receiving nasal oxygen.

Six months earlier, he had developed pulmonary thrombosis and was treated elsewhere with heparin and finally discharged on oral rivaroxaban. A color Doppler study of the lower extremities showed no evidence of deep vein thrombosis, but computed tomographic angiography of the pulmonary arteries demonstrated complete thrombotic occlusion of the right pulmonary artery and its lobar and segmental branches.

The patient reported pleuritic chest pain and was found to be markedly dyspneic and orthopneic. His arterial oxygen saturation was found to be 57% while receiving 6 L of nasal oxygen. Emergency pulmonary computed tomographic angiography showed a large clot involving the main and right pulmonary arteries (Figure 1). Further investigations, including color Doppler study of the lower extremities, common and both iliac veins, renal veins, and inferior vena cava, failed to show any clot. Transesophageal echocardiography (TEE) revealed severe pulmonary hypertension associated with remarkable right atrial and ventricular dilatation accompanied by severe right ventricular systolic dysfunction. No clots could be detected in any cardiac chambers. However, there was a large fungating mass in the main pulmonary artery with extension to, mainly into, and nearly totally occluding the proximal part of right pulmonary artery (Figures 2 and 3, Video 1).

Interestingly, the proximal part of the right inferior pulmonary vein was also occupied by the same type of echogenic mass (Figure 4, Video 2). The remaining pulmonary veins, however, were patent.

Paraclinical investigations, including factor V Leiden, protein C, protein S, antithrombin III, anticardiolipin antibodies immunoglobulin G and immunoglobulin M, antiphospholipid antibodies, perinuclear antineutrophil cytoplasmic antibody, cytoplasmic antineutrophil cytoplasmic antibody, and antinuclear antibody HEp-Z, failed to guide us toward any etiologic cause. In addition, the results of all workups to detect any occult malignancy were negative.

Because of the patient's poor condition, he was taken to the operating room, where the pulmonary arterial mass was resected. In addition, the left atrium was opened, and the right inferior pulmonary vein mass was resected as well (Videos 3 and 4, Figures 5 and 6). Histopathologic examination of both masses showed organized thrombi.

The patient's postoperative course was uneventful and was characterized by impressive symptomatic alleviation. His oxygen dependency gradually disappeared, and he was discharged with oxygen saturation of 84.0% on room air.

DISCUSSION

The co-occurrence of pulmonary artery thrombosis and PVT in our patient is most probably the first of its kind reported in the literature. The diagnosis was made possible only after meticulous TEE. However, we were unable to detect the underlying etiology, and no primary site or cause of thrombosis was found.

Although pulmonary artery thromboembolic complications are quite frequent, PVT is very rare and has an unclear incidence.[7]

Isolated PVT has been reported in association with a variety of clinical conditions, such as lung transplantation,[8] radiofrequency ablation of atrial fibrillation,[9] primary or metastatic lung cancer,[7] and lobectomy.[10]

Lung transplantation is one of the main causes of PVT, most likely due to surgery-induced endothelial injury or blood stasis in the blind pulmonary vein stump.[8]

Catheter ablation is frequently used to establish sinus rhythm in patients with atrial fibrillation. However, ablation can be associated with both acute and chronic PVT and its associated complications.[9]

Primary lung cancers such as bronchogenic carcinoma and metastatic tumors have been reported to cause PVT.[7] Direct tumor extension

From the Department of Cardiology, Nemazee Hospital (M.S., G.R., M.C.), and the Department of Surgery, Shahid Faghihi Hospital (M.N.), Shiraz University of Medical Sciences, Shiraz, Iran.

Keywords: In situ pulmonary artery thrombosis, Pulmonary vein thrombosis, Surgical clot removal

Conflicts of interest: The authors reported no actual or potential conflicts of interest relative to this document.

Copyright 2019 by the American Society of Echocardiography. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

2468-6441

https://doi.org/10.1016/j.case.2019.10.012

VIDEO HIGHLIGHTS

Video 1: TEE demonstrating a mass in the main pulmonary artery extending into the osteoproximal part of right pulmonary artery.

Video 2: TEE demonstrating an oscillating mass in the right inferior pulmonary vein.

Video 3: Main pulmonary artery and right pulmonary artery exploration, in addition to mass removal.

Video 4: Right inferior pulmonary vein exploration, in addition to mass removal.

View the video content online at www.cvcasejournal.com.

into the vein or epithelial damage secondary to tumor invasion may be the underlying pathophysiologic mechanisms in such cases.

Lobectomy, especially that of the left upper lobe, can be associated with PVT. In a retrospective study, Ohtaka *et al.*[10] detected PVT in 3.3% of all their patients who underwent lobectomy. The frequency, however, was 17.9% in those who underwent left upper lobectomy.

At times, however, no clear-cut etiology could be found, as in our patient, and the case may be labeled as idiopathic.[11]

Diagnosis and treatment of PVT are obviously difficult. However, an early diagnosis is of paramount importance to rescue the patient and prevent catastrophic complications.[10] Definite diagnosis requires multimodality studies, including computed tomographic pulmonary arteriography with venous phase and TEE. Little is known about the preferred treatment of patients with PVT, although in critically ill patients such as reported here, thrombectomy might be lifesaving.

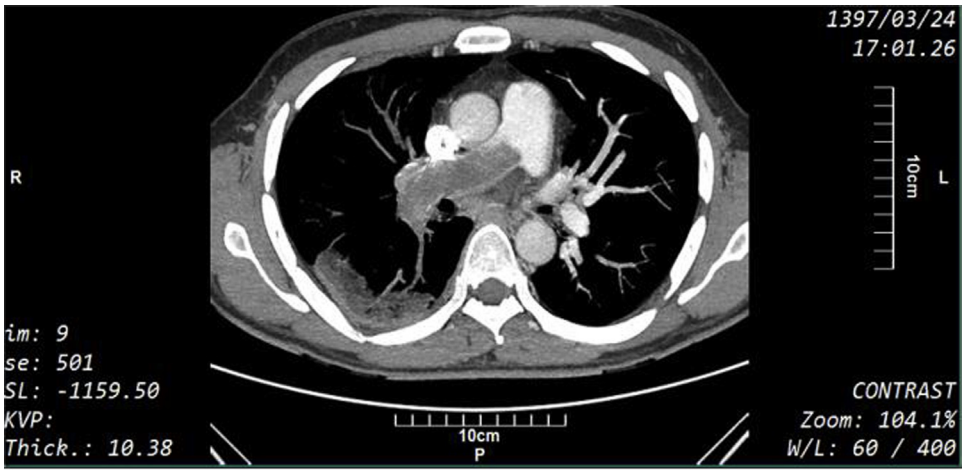

Figure 1 Spiral computed tomographic angiography of pulmonary vessels contrast (pulmonary thromboendarterectomy protocol) demonstrating a large filling defect starting from the main pulmonary artery (MPA) and extending to the proximal part of the right pulmonary artery (RPA).

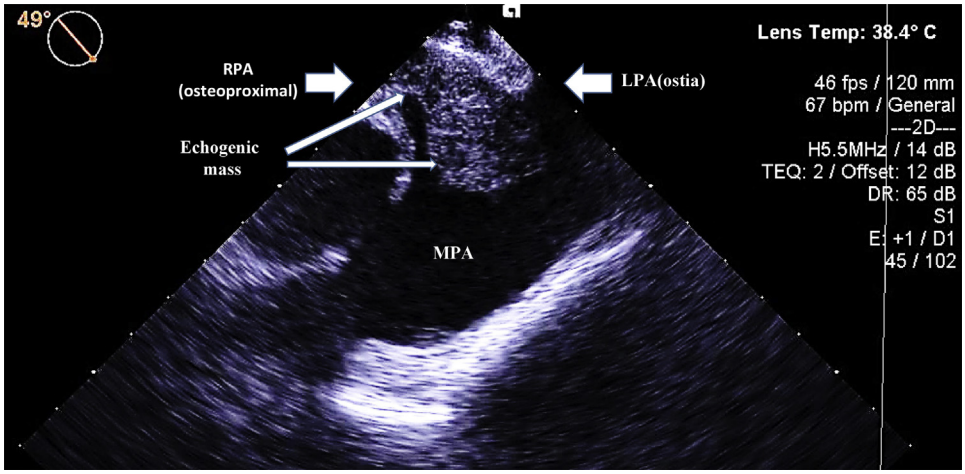

Figure 2 TEE (midesophageal level, transducer angle 49°) demonstrating the presence of an echogenic mass involving the main pulmonary artery (MPA), partially obliterating the ostium of the left pulmonary artery (LPA) and filling the proximal part of the right pulmonary artery (RPA).

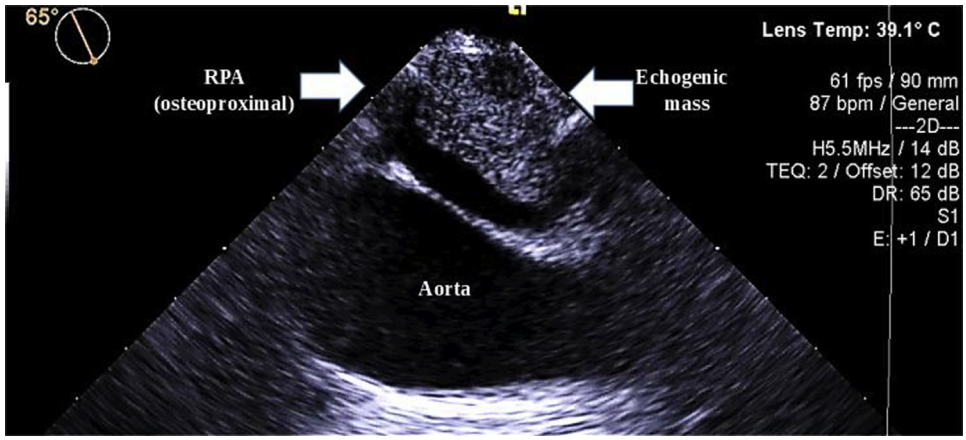

Figure 3 TEE (midesophageal level, transducer angle 65°) demonstrating the presence of an echogenic mass that filled the osteoproximal right pulmonary artery (RPA).

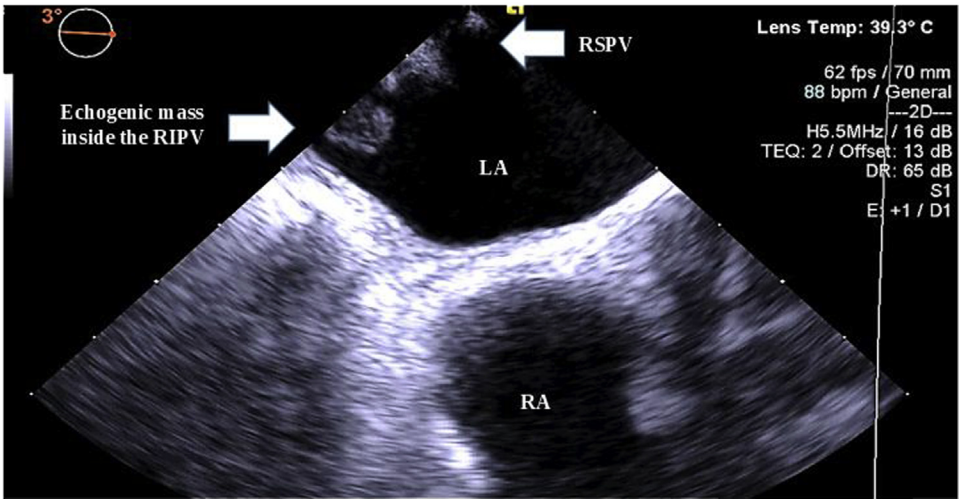

Figure 4 TEE (upper esophageal level, transducer angle 3°), demonstrating the presence of an echogenic mass inside the right inferior pulmonary vein (RIPV). *LA*, Left atrium; *RA*, right atrium; *RSPV*, right superior pulmonary vein.

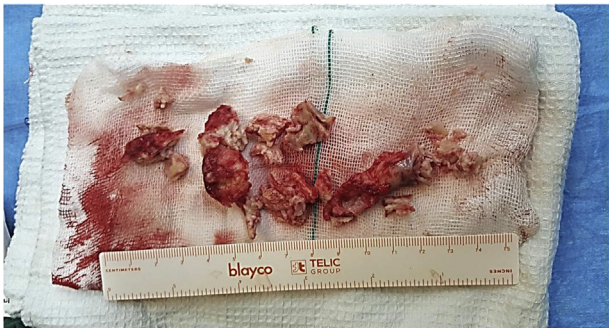

Figure 5 Photograph of the particles of mass surgically resected from the main and right pulmonary arteries.

CONCLUSION

PVT is a rare clinical condition with nonspecific symptoms. It can contribute to the development and persistence of symptoms of respiratory failure in patients with concomitant pulmonary artery thrombosis. Any delay in diagnosis can be life threatening and contribute to the development of chronic respiratory failure unresponsive to usual medical treatments, right heart failure, peripheral embolization, and stroke. TEE is an easily available, noninvasive diagnostic evaluation of choice that can lead to early diagnosis and treatment decision-making.

ACKNOWLEDGMENT

We thank Hossein Sharifkazemi for his invaluable assistance in condensing and collating visualizations for this article.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2019.10.012.

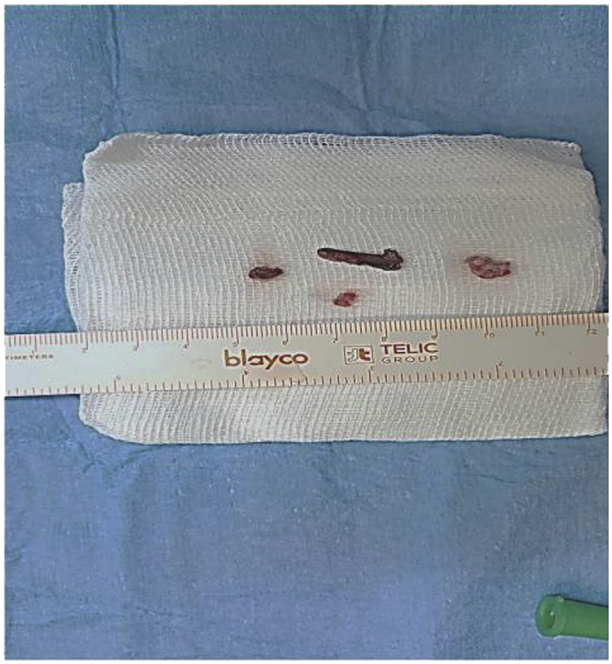

Figure 6 Photograph of the particles of mass surgically resected from the right inferior pulmonary vein.

REFERENCES

1. Kumar NG, Clark A, Roztocil E, Caliste X, Gillespie DL, Cullen JP. Fibrinolytic activity of endothelial cells from different venous beds. J Surg Res 2015;194:297-303.
2. Rosenhek R, Korschineck I, Gharehbaghi-Schnell E, Jakowitsch J, Bonderman D, Huber K, et al. Fibrinolytic balance of the arterial wall: pulmonary artery displays increased fibrinolytic potential compared with aorta. Lab Invest 2003;83:871-6.
3. Ryan US. Metabolic activity of pulmonary endothelium: modulations of structure and function. Annu Rev Physiol 1986;48:263-77.
4. Russo A, De Luca M, Vigna C, De Rito V, Pacilli M, Lombardo A, et al. Central pulmonary artery lesions in chronic obstructive pulmonary disease: a transesophageal echocardiography study. Circulation 1999;100:1808-15.
5. Agarwal PP, Wolfsohn AL, Matzinger FR, Seely JM, Peterson RA, Dennie C. In situ central pulmonary artery thrombosis in primary pulmonary hypertension. Acta Radiol 2005;46:696-700.
6. van Langevelde K, Flinterman LE, van Hylckama Vlieg A, Rosendaal FR, Cannegieter SC. Broadening the factor V Leiden paradox: pulmonary embolism and deep-vein thrombosis as 2 sides of the spectrum. Blood 2012;120:933-46.
7. Akiode O, Prakash G. Pulmonary vein thrombosis associated with metastatic carcinoma. Fed Pract 2014;31:26-8.
8. Uhlmann EJ, Dunitz JM, Fiol ME. Pulmonary vein thrombosis after lung transplantation presenting as stroke. J Heart Lung Transplant 2009;28:209-10.
9. Hanna M, Dahal Kh, Watti H, Swaminathan PD. A rare case of fatal acute pulmonary vein thrombosis after radiofrequency catheter ablation of atrial fibrillation. J Am Coll Cardiol 2018;71(11 suppl).
10. Ohtaka K, Hida Y, Kaga K, Takahashi Y, Kawase H, Hayama S, et al. Left upper lobectomy can be a risk factor for thrombosis in the pulmonary vein stump. J Cardiothorac Surg 2014;9:5.
11. Mumoli N, Cei M. Idiopathic pulmonary vein thrombosis. J Emerg Med 2012;42:182-3.