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

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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,1 primary pulmonary hypertension,2 or any underlying cause of pulmonary inflammation.3

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 demonstrated complete thrombotic occlusion of the right pulmonary artery (Figures 2 and 3, Video 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 an echogenic mass in the left atrial appendage and a newly developed left atrial thrombus (Figure 4, Videos 2 and 3). A lung perfusion scan revealed a segmental right lower lung perfusion defect. The remaining pulmonary veins, however, were patent.

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 another echogenic mass in the left atrial appendage and a newly developed left atrial thrombus.

The diagnosis was made possible only after meticulous TEE. However, 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, radiofrequency ablation of atrial fibrillation, primary or metastatic lung cancer, 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...
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. 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. Diagnosis and treatment of PVT are obviously difficult. However, an early diagnosis is of paramount importance to rescue the patient and prevent catastrophic complications. 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).
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.

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SUPPLEMENTARY DATA

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