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Pulmonary Vein Varix: A Diagnostic Enigma

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Abstract

We report a case of a 34-year-old woman who presented with chest pain, shortness of breath, and dry cough of 1 month duration. Contrast-enhanced computed tomography of the chest revealed an enlarged and tortuous right pulmonary vein that could not conclusively exclude a pulmonary arteriovenous malformation. Further evaluation with digital subtraction angiography excluded an arteriovenous malformation and confirmed the presence of a pulmonary vein varix. Accordingly, the patient was managed conservatively. This report highlights the need for catheter angiography in the diagnosis of pulmonary vein varix and provides a brief review of literature on the entity.

Keywords

- pulmonary vein varix
- rare pulmonary vascular anomaly
- pulmonary vein varix

Introduction

Pulmonary venous varix is a rare pulmonary vascular anomaly. It may be congenital or acquired and is characterized by focal aneurysmal dilatation of the pulmonary vein. Imaging appearances mimic pulmonary arteriovenous malformation (AVM) or pulmonary mass. As the management strategies differ, it is important to accurately diagnose the entity.

Case Report

A 34-year-old woman with symptoms of chest pain, shortness of breath, and dry cough of 1 month duration and with a diagnosis of pulmonary AVM on contrast-enhanced computed tomography (CECT) of the chest was referred to our institution for further evaluation and management. The chest pain was nonspecific. Her dyspnea progressed from grade 1 to grade 2 modified Medical Research Council dyspnea scale with no orthopnea and paroxysmal nocturnal dyspnea. Dry cough was associated with no diurnal or postural variation. She reported having similar symptoms intermittently during 3 years prior to presentation. On examination, the heart rate was 110 beats/min, respiratory rate was 22/min, blood pressure was 110/70 mm Hg, and oxygen saturation was 98% on room air. The laboratory workup showed anemia with low hemoglobin of 8.2 g/dL. Electrocardiography was normal. Echocardiography revealed mild pulmonary arterial hypertension. There were no other comorbidities. CECT of the chest (►**Fig. 1A**) revealed normal caliber (21 mm) main pulmonary artery with an enhancing dilated (16 mm) serpiginous vascular structure in the right lower lobe connecting the right interlobar pulmonary artery and the inferior pulmonary vein (►**Fig. 1B**). The inferior pulmonary vein was dilated. To rule out a pulmonary AVM, conventional pulmonary angiography was performed. The right pulmonary artery demonstrated a normal course, caliber, and branching pattern (►**Fig. 2A**). There was no early

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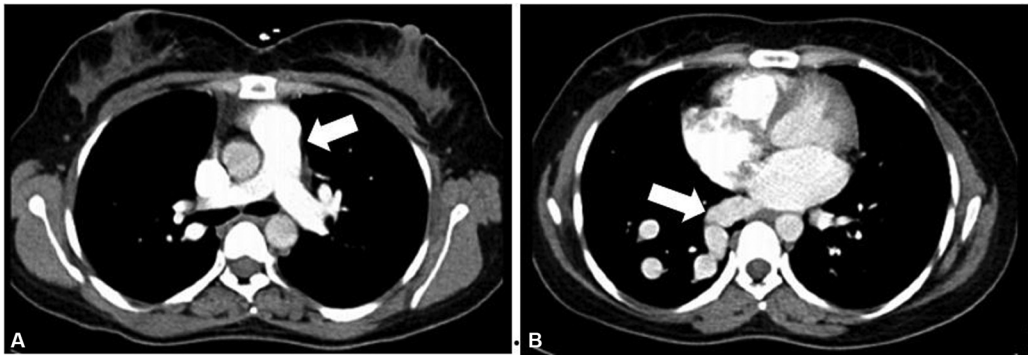

Fig. 1 Axial contrast-enhanced computed tomography of chest: (**A**) Normal caliber of main pulmonary artery (arrow). (**B**) Dilated and tortuous course of inferior pulmonary vein (arrow) noted in right lower lobe.

opacification of the pulmonary vein or any abnormal arteriovenous communication. The venous phase images of the right pulmonary angiography (►**Fig. 2B**) revealed a grossly dilated and tortuous anomalous right pulmonary vein representing a "pulmonary vein varix." The patient was treated conservatively for her symptoms and no intervention was performed for the venous varix. A follow-up echocardiography a year later revealed normal pulmonary artery pressures.

Discussion

Pulmonary vein varix is a rare pulmonary vascular anomaly characterized by a focal abnormal dilatation of the pulmonary vein. The incidence is not known with less than 100 cases published in the literature. There is no gender predilection, and it affects all age groups. Pulmonary venous varix was first described by Puchet in 1843 during the autopsy of a child who died from digestive hemorrhage and presented with multiple multisystemic varices.[1] Mouquin et al reported the first angiographic description in a living patient in 1951.[2] Because of preferential direction of retrograde flow, the most common location of a pulmonary vein varix is the right lower lobe followed by the left upper lobe. Pulmonary varices are classified into three groups according to their appearance: saccular, tortuous, and confluent.[3] These could be congenital or acquired (secondary to mitral valve disease, etc.). The acquired forms are more common. The etiology of congenital form is unknown. They are considered to develop as a collateral pathway in the presence of a pulmonary venous stenosis or atresia.[4] They are usually asymptomatic. Rarely, complications such as systemic emboli caused by the release of thrombi from the varix or, even more infrequently, hemoptysis or hemothorax caused by rupture of the varix may occur. Onteddu et al described a case of a pulmonary vein varix near the pulmonary hilum with an associated superior pulmonary vein stenosis.[4]

On imaging, pulmonary varices may mimic AVMs, and case reports of pulmonary varices misdiagnosed as AVMs on CT scan have been documented.[5] The appearance of pulmonary vein varix on CT angiography is in the form of a tortuous vascular structure of increased caliber of a pulmonary vein that empties into the left atrium. CT angiography is often limited by flow artifacts and filling of pulmonary veins can be simultaneous with the arteries that may lead to an incorrect

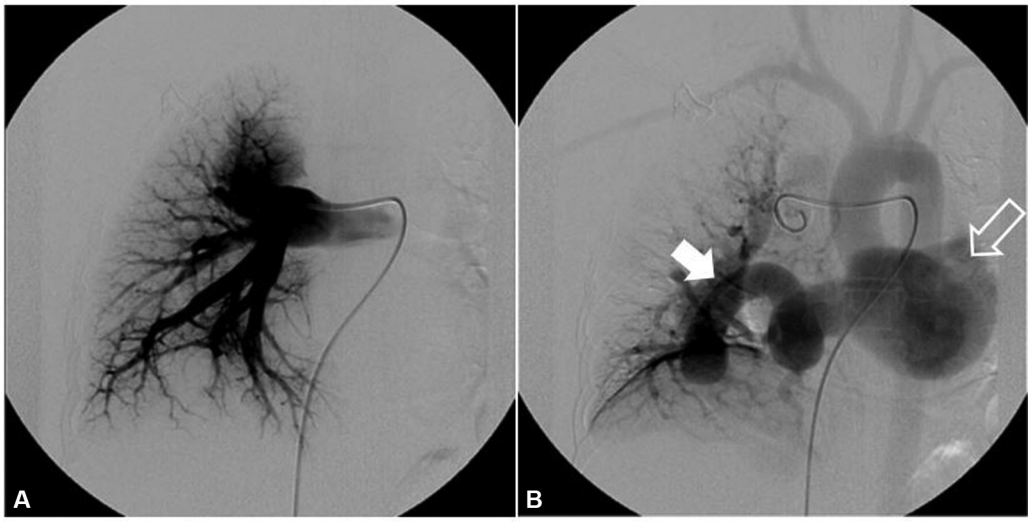

Fig. 2 Selective right pulmonary angiography. (**A**) Late arterial phase shows normal course, caliber, and branching pattern of right pulmonary artery. (**B**) On delayed phase, grossly dilated and tortuous right pulmonary vein (white arrow) is seen draining into left atrium (open arrow), confirming that this is a pulmonary vein varix.

diagnosis of a pulmonary AVM. A time-resolved magnetic resonance angiography can be used to assess the pulmonary venous anatomy and exclude a pulmonary AVM. The gold standard for diagnosis of pulmonary varix is pulmonary angiography. Batram and Strickland reported specific criteria to diagnose pulmonary venous varix on angiography[2] normal pulmonary arteries, no evidence of pulmonary arteriovenous fistula, simultaneous appearance of varicose veins and normal pulmonary veins, varicose veins draining into the left atrium, prolonged emptying of varicose veins compared with normal veins, and dilated and tortuous varicose veins are central, near the hilum, with normal peripheral veins.

The differential diagnosis of pulmonary varices includes other conditions with dilatation of the pulmonary veins, such as the hepatopulmonary and scimitar syndromes. Management of pulmonary vein varix includes observation in asymptomatic individuals and treating associated comorbidities. Resection is considered if the size of varix increases or if the patient is symptomatic.

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Conflict of Interest
None declared.

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