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Pulmonary Arteriovenous Fistula as a Cause of Chronic Hypoxemia in a Woman with Preserved Myocardial Function: Case Report

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ABSTRACT

Pulmonary arteriovenous fistulas are rare within the spectrum of vascular malformations. Dyspnea and fatigue are the most common symptoms. Their etiology is congenital in 90% of cases, and almost 85% are associated with Osler-Weber-Rendu syndrome (hereditary hemorrhagic telangiectasia). More than half of patients are asymptomatic, approximately 40% have neurological manifestations (cerebrovascular events, headaches, and seizures), and only 10% report dyspnea and cyanosis. This is a 50-year-old woman who presented with progressive dyspnea, dizziness, and fainting with oxygen saturation of 82% for a 6-month duration. She attended a private hospital where a pulmonary CT angiography was performed, which reported a dilatation of the right pulmonary trunk with an apparent fistula. She was referred to our cardiology center, where a sinus electrocardiogram reported a
normal heart rate with no signs of hypertrophy. Pulmonary computed tomography (CT) was performed, ruling out pulmonary thromboembolism. However, a fistula was found between the right basal pulmonary artery and the middle lobe to the ipsilateral pulmonary vein, causing a shunt of deoxygenated blood to the systemic circulation, causing desaturation in the patient (right to left). Echocardiographic scanning revealed no hemodynamic abnormalities in the cardiac chambers. Until the 1980s, treatment options in these cases were partial or total lobectomies. Percutaneous occlusion devices, primarily coils, began to be used with varying degrees of success. However, the choice of treatment option depends on the clinical presentation, the severity of symptoms, and the patient's wishes.

KEYWORDS: Pulmonary arteriovenous fistula, anomalous pulmonary venous return, cardiovascular abnormality

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INTRODUCTION

Pulmonary arteriovenous fistulas (PVAFs) are rare within the spectrum of vascular malformations, diagnosed in 1 in 5,000 to 10,000 patients. Dyspnea and fatigue are the most common symptoms **[1]**. Their etiology is congenital in 90% of cases, and almost 85% are associated with Osler-Weber-Rendu syndrome (hereditary hemorrhagic telangiectasia). The remaining approximately 10% are usually secondary to chest trauma, chest surgery, cirrhosis, cancer, mitral stenosis, infections, amyloidosis, and pregnancy, among others **[1,2]**. More than half of patients are asymptomatic, about 40% have neurological manifestations (cerebrovascular events, headaches, and seizures), and only 10% report dyspnea and cyanosis. Diagnosing this condition represents a diagnostic challenge for the physician **[2]**.

CLINICAL CASE

This is a 50-year-old woman who presented with progressive dyspnea, dizziness, and fainting with an oxygen saturation of 82%. She attended a private hospital where a pulmonary CT angiogram was performed, reporting a dilated right pulmonary trunk with an apparent fistula, without filling defects, and polycythemia in the laboratory. She was referred to our cardiology center where a sinus electrocardiogram was reported, with a normal heart rate and no signs of hypertrophy. A pulmonary CT angiogram was performed,

ruling out pulmonary thromboembolism with a fistula from the right basal pulmonary artery of the middle lobe to the ipsilateral pulmonary vein, resulting in a shunt of deoxygenated blood to the systemic circulation, causing desaturation in the patient. Echocardiographic scanning revealed no hemodynamic alterations in the cardiac chambers.

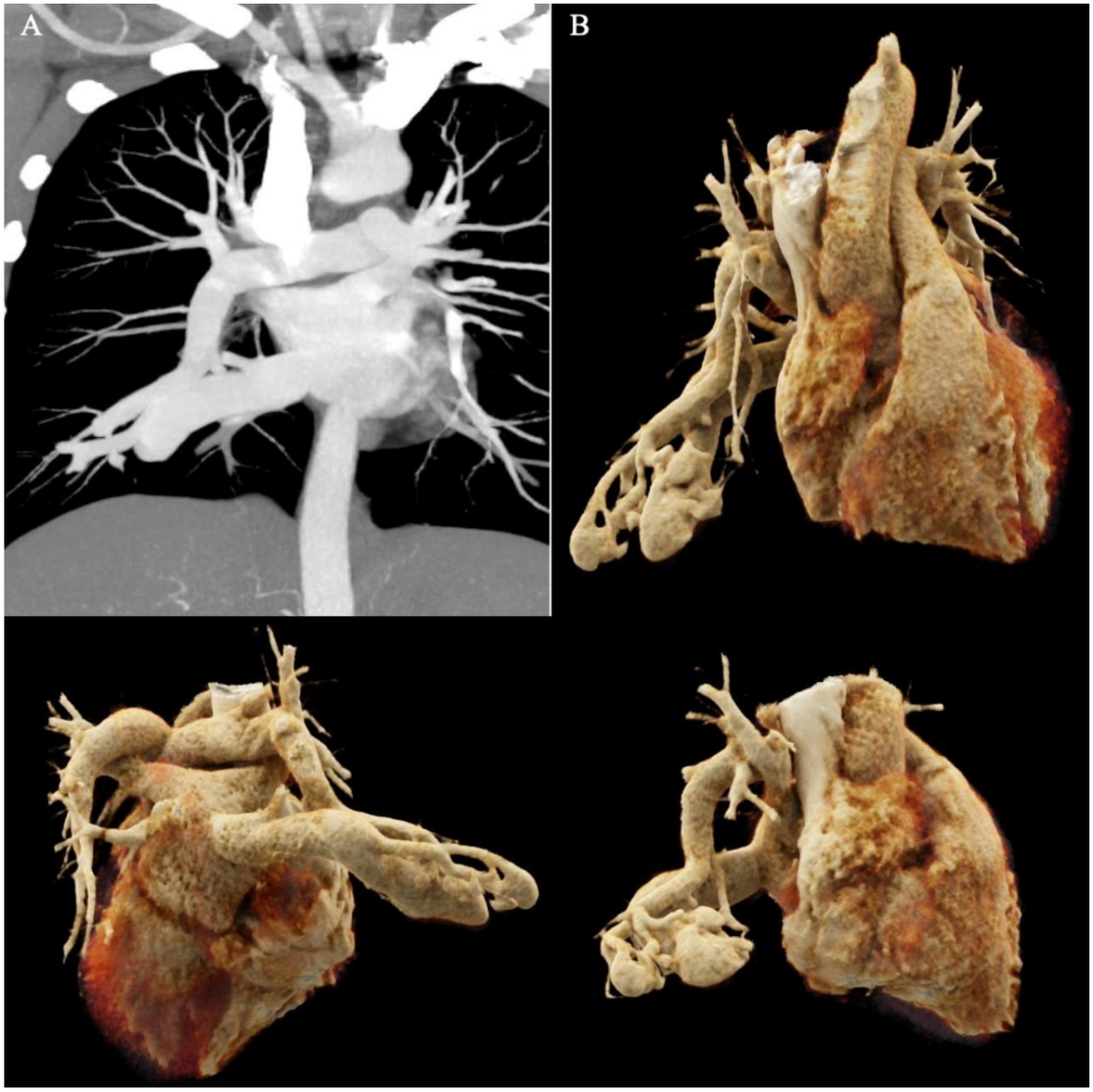

Figure 1A, B). Contrast-enhanced pulmonary angiography with 3D reconstruction showing the fistula from the right basal pulmonary artery of the middle lobe to the ipsilateral pulmonary vein, causing a right-to-left shunt.

DISCUSSION

VAPF comorbidities occur in 60% of patients, and when left untreated, mortality can rise from 26% to 50%. Depending on the magnitude of the right-to-left shunt, determined primarily by the size of the fistula, they can cause hypoxemia and present symptoms such as dyspnea, cyanosis, polycythemia, and clubbing **[3]**. Therefore, the diagnosis of this entity represents a diagnostic challenge for the physician.

Diagnosis of this entity represents a diagnostic challenge for the physician. Histologically, the simple type presents as a gle aneurysmal sac and contains a single segmental artery that feeds the malformation. Complex PAVFs involve multiple abnormal vessels and consist of one or more lobulated venous sacs of varying size supplied by more than one feeding artery, often arising from adjacent segmental branches of the pulmonary artery. Complex PAVFs may involve entire lung segments or an entire lobe **[4]**.

Until the 1980s, treatment options in these cases were partial or total lobectomies. Percutaneous occlusion devices, primarily coils, began to be used with varying degrees of success. In recent years, Amplatzer devices have been the preferred device for this type of occlusion and, in general, for vascular malformations **[5]**. However, the choice of treatment

option depends on the clinical presentation, the severity of symptoms, and the patient's wishes.

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