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Case Report
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Pulmonary Arteriovenous Fistula with Pulmonary Hypertension – To Close or Not to Close?

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A 37-year-old female presented to our facility suffering from exertional dyspnea for 2-3 months. Her chest x-ray showed a nodular shadow in the right lower lung, and chest CT revealed a pulmonary arteriovenous fistula (PAVF). Subsequent echocardiographic examination detected no intra-cardiac shunt but did indicate pulmonary hypertension as evidenced by a tricuspid regurgitation flow velocity of 4.17 M/sec. Contrast echocardiography with antecubital vein injection of agitated normal saline demonstrated visualization of the left heart chambers compatible with PAVF. At cardiac catheterization, pulmonary arterial pressure was 59/26 mmHg, mean 34 mmHg. Because there was no intra-cardiac communication detected, primary pulmonary hypertension was tentatively diagnosed. Pulmonary angiography demonstrated a PAVF arising from the lower right pulmonary artery, forming a secular structure on its course in draining into the left atrium through a long pulmonary vein. In this particular anomaly, a concurrence of PAVF with pulmonary hypertension, we judged that the PAVF might serve as a safety valve for pulmonary hypertension and should not be closed. We therefore left the PAVF untreated and thereafter provided medical management for this patient.

The concomitant presence of PAVF and pulmonary hypertension is a rare clinical condition. The ultimate treatment strategy for this uncommon condition should be carefully considered.

Key Words: Computed tomography • Contrast echocardiography • Pulmonary angiography • Pulmonary arteriovenous fistula • Pulmonary hypertension

INTRODUCTION

Pulmonary arteriovenous fistula (PAVF) is a rare vascular anomaly with a direct communication between pulmonary artery and pulmonary vein without an intervening capillary bed. The prevalence of PAVF has been estimated to be approximately 2 to 3 cases per 100,000 population.[1] Most cases of PAVF are congenital, which are closely associated with Osler-Weber-Rendu disease or hereditary hemorrhagic telangiectasia.[1,2] Nonetheless, there are still sporadic cases of isolated PAVF reported.[3,4]

Patients with PAVF may be asymptomatic, but more frequently they present with mild symptoms such as dyspnea on exertion and manifestations of systemic hypoxemia – cyanosis, polycythemia, and clubbing of the fingers and toes.[3]

With the direct connection of pulmonary artery to a low pressure pulmonary vein, pulmonary arterial pressure is normal in most cases except for the presence of concomitant underlying pathological conditions that are associated with elevation of pulmonary arterial pressure.[5]

Herein we report a case of PAVF associated with pulmonary hypertension. In this patient, the causes of pul-

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monary hypertension were not sufficiently determined after extensive investigation. The patient was therefore tentatively diagnosed with primary pulmonary hypertension. The management strategy for this special clinical condition appears to be somewhat of a medical dilemma. In this case, we chose not to treat this PAVF with invasive interventions.

CASE REPORT

A 37-year-old female was referred for evaluation and management of a PAVF diagnosed at another hospital. She reported feeling well until 2-3 months before admission, when she began suffering from exertional shortness of breath. The patient visited a local hospital where chest x-ray showed a nodular shadow in the right lower lung field. Chest computed tomography (CT) revealed a PAVF involving the right lower lung, and she was then referred to our facility for further treatment.

At presentation, she appeared with no lip cyanosis, and no clubbing of the fingers and toes. Her blood pressure was 98/70 mmHg, with a regular pulse of 84/min. There was no heart murmur detected at auscultation and no abnormal sound audible upon examination of the patient's chest. Additionally, no telangiectasia could be detected on the skin or mucosa. However, her chest x-ray revealed an abnormal nodular shadow in the lower right lung. The electrocardiogram showed right axis deviation and clockwise rotation suggesting right ventricular hypertrophy. Hemograms showed mild elevation of red blood cell count ($5.25 \times 10^6/mm^3$) with mild elevation of hemoglobin (15.7 g/dL) and hematocrit (45.1%).

Echocardiographic imaging demonstrated dilated right side chambers with moderate tricuspid regurgitation (TR). The TR flow velocity was 4.17 M/sec, representing an estimated pressure gradient of 70 mmHg across the tricuspid valve. There was no atrial or ventricular septal defect nor patent ductus arteriosus detected. Contrast echocardiography with injection of agitated saline from the antecubital vein showed visualization of echo contrast in the left heart, compatible with the presence of an intra-cardiac right-to-left shunt.

A chest CT with contrast enhancement showed a PAVF arising from the right lower pulmonary artery, proceeding into a saccular structure, then connecting to a pulmonary vein before draining into the left atrium (Figure 1). The PAVF was about 1.7 cm at the largest diameter of the sinusoidal structure.

The patient then underwent cardiac catheterization. The procedure revealed that the mean pulmonary capillary wedge pressure was 9 mmHg, and the main pulmonary artery pressure 59/26 mmHg, with a mean of 34 mmHg; the aortic pressure was 91/65 mmHg and the left ventricle pressure 98/13 mmHg. There was no oxygen step-up in the right heart and the oxygen saturation in the ascending aorta was 92%. Pulmonary angiography showed a PAVF arising from the right lower pulmonary artery and drained into the left atrium through a long pulmonary vein (Figure 2A, B). No intervention was performed for this PAVF and medical treatment was suggested. The patient was later put on sildenafil 20 mg 3 times a day to control her pulmonary hypertension.

DISCUSSION

Hemodynamically significant PAVFs are frequently

Figure 1. *Chest computed tomography with contrast enhancement. A dilated pulmonary artery (thin white arrows) connects to a dilated sinus-like chamber (x mark), which then connects to a long pulmonary vein (thick white arrows) before draining into left atrium.*

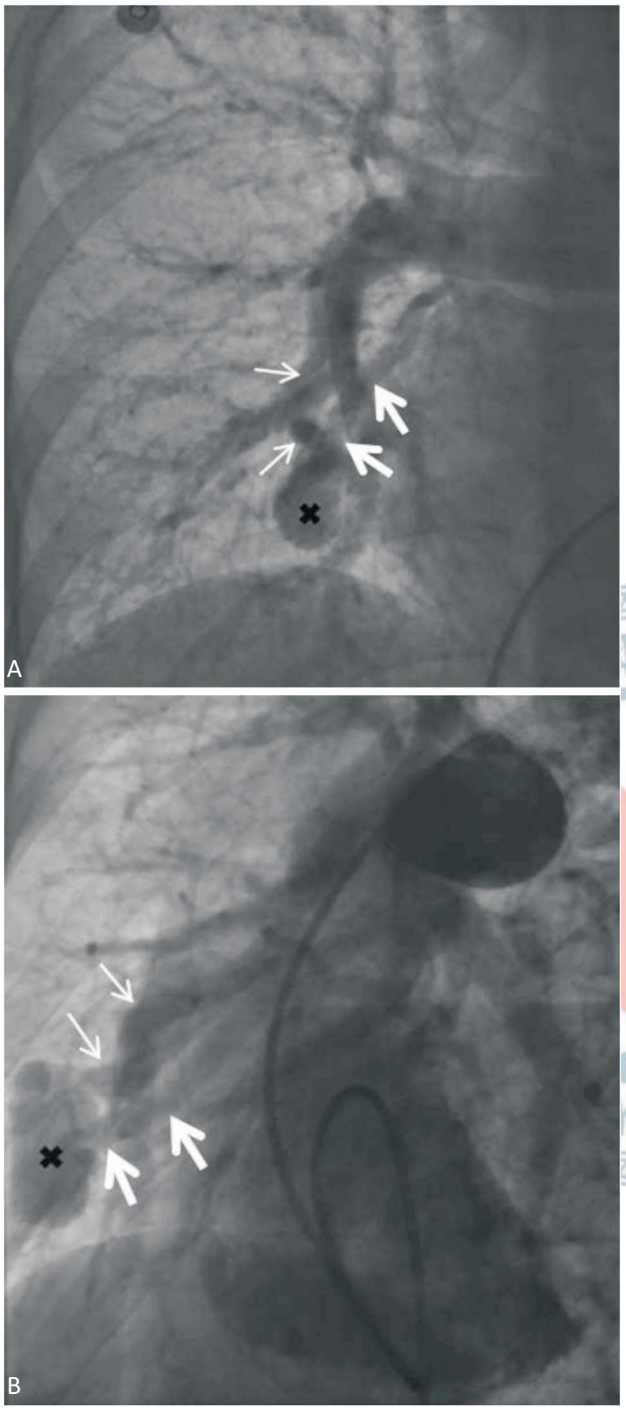

Figure 2. *Pulmonary angiograms. (A) AP view; (B) Lateral view. An anomalous pulmonary artery (thin white arrows), sinus-like structure (x mark) and a pulmonary vein (thick white arrows) are shown.*

symptomatic and are typically indicated for treatment. Before the percutaneous treatment modality was developed, surgery was the standard regimen.[6,7] Device occlusion for PAVF was adapted as an alternative regimen and gained popularity soon after its development because of its less invasive nature.[3,8,9] Both surgery and device occlusion were reported to have high success rates and low rates of complication.

In most PAVF cases, pulmonary arterial pressure is normal, or only mildly elevated. It is worth noting that le Roux et al.[5] reported an 11-year-old African female who had a large PAVF and moderate pulmonary hypertension (mean pressure 50 mm Hg) with elevated pulmonary resistance. This patient underwent surgery for resection of the fistula. Unfortunately, during this surgical procedure, acute heart failure developed. Resuscitation was not effective and the patient died 12 hours after operation. Pathological examination of the young patient's resected lung specimen showed the features of bilharzial granuloma and angiomatous changes with vascular sclerosis.

Our patient had a similar circumstance of moderate pulmonary hypertension overlayed with PAVF. Although the PAVF of our patient was relatively small in size, it was regarded as hemodynamically significant because of the presence of polycythemia and systemic hypoxemia. Through extensive investigation, including physical examination, contrast-enhanced lung CT, echocardiography and contrast echocardiography and catheterization with angiography, there was no demonstrated intra-cardiac shunt. The etiology of pulmonary hypertension in this patient was therefore obscured and was tentatively diagnosed as primary pulmonary hypertension. Due to our concern that the PAVF in this patient may be functioning as a safety valve for the concomitant pulmonary hypertension, we decided not to close this fistula.

With our patient, we did not attempt a transient occlusion of the fistula (such as balloon occlusion of the PAVF) to test the response because we believed that even we observed no immediate adverse response to this maneuver. However, the delayed response was still unpredictable and the risk of delayed serious sequele still could occur if we closed the PAVF merely according to the results of the transient occlusion.

A relevant therapeutic strategy has been proposed by Sandoval et al.[10] for patients with severe pulmonary hypertension. They demonstrated that, in patients with severe pulmonary hypertension, the creation of an artificial inter-atrial shunt might offer a significant palliative effect for these patients.

CONCLUSIONS

Although hemodynamically significant PAVF should be treated, either by surgery or device closure, in a particular condition in which pulmonary hypertension is present, the appropriateness of fistula closure should be carefully evaluated.

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