Absence of Right Pulmonary Artery With Pulmonary Valve Stenosis and Partial Connection of Descending Aorta—A Case Report

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Case report

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Abstract

Background: Unilateral absence of pulmonary artery is a rare congenital disorder that can remain asymptomatic until adulthood. Absence of right pulmonary artery (ARPA) has been reported in two-thirds of this patient population. It may be isolated, but it is more often associated with other congenital cardiac defects such as aortic coarctation, tetralogy of Fallot, atrial septal defect and patent ductus arteriosus.

Case presentation: We are the first to introduce an adult case of ARPA with pulmonary valve stenosis (PVS) and associated with partial connection of descending aorta. We investigated the condition by right heart catheterization, right ventricle angiography, pulmonary arteriography and aorta angiography. According to the above results, we suggest that surgical treatment should be positively considered.

Conclusions: The case reminds that echocardiography combined with detailed imaging examination is very important for the diagnosis of complex congenital heart disease and provides the basis for the selection of the best treatment.

Introduction

Unilateral absence of the pulmonary artery (UAPA) that normally arises from the sixth aortic arch is a very rare congenital malformation with a prevalence of 0.005%. UAPA is more common on the right side¹. UAPA may present in infancy with respiratory distress, pulmonary hypertension and congestive heart failure¹. When severe pulmonary hypertension does not develop in infancy, the condition may remain asymptomatic until adulthood. This report presents a rare case of UAPA with PVS and partial connection of descending aorta and underscores the role of imaging modalities in identifying this condition and providing evidence for further treatment.

Case Presentation

This case involves a 52-year-old female with presenting symptom of worsening shortness of breath after activity 1 year ago. No history of hypertension, diabetes or smoking. Clinical workup revealed right heart failure. Echocardiography indicates congenital heart disease, moderate and severe pulmonary valve stenosis and regurgitation (Fig. 1). In order to explore the disease and choose the appropriate treatment, our team performed right heart catheterization, right ventricle angiography, pulmonary arteriography and ascending aorta angiography. Right heart catheterization examination indicated right ventricular pressure (SBP 142 mmHg, diastolic pressure 8 mmHg, mean pressure 42 mmHg), right atrium pressure (SBP 20 mmHg, diastolic pressure 6 mmHg, mean pressure 10 mmHg), pulmonary artery pressure (SBP 104 mmHg, diastolic pressure 27 mmHg, Mean pressure 52 mmHg), PCWP 6 mmHg, total pulmonary resistance 10.64 wood and minor pulmonary resistance 9.41 wood, and pulmonary hypertension was considered. Right ventricular angiography showed muscular trabecular hypertrophy, no stenosis of right ventricular outflow tract, pulmonary valve hypertrophy, limited opening, "vault sign" and "spray column sign" (Fig. 2, video 1). Pulmonary angiography indicated that the right pulmonary artery was not developed,

and the left pulmonary artery and its branches were dilated. ARPA was considered (Fig. 2, video1). Aortography suggested that multiple lateral branches of the descending aorta supplied the right lung field (Fig. 2, video1). Based on the above results, the clinical diagnosis is as follows: congenital heart disease, PVS, ARPA, pulmonary hypertension, collateral circulation of descending aorta supplying right lung. Combined cardiac surgery and thoracic surgery are recommended.

Discussion

UAPA is a rare anomaly and usually occurs in association with other cardiovascular malformations. It is associated with respiratory symptoms, such as dyspnea or hemoptysis. Pulmonary valve stenosis occurs in isolation in 8%-10% of congenital heart disease but is often associated with other congenital lesions². PVS are most often congenital and can be associated with genetic syndromes including Noonan, Alagille and Williams syndromes as well as congenital rubella. UAPA and PVS were present in this case, which is very rare. Cardiac ultrasound is more sensitive to congenital heart valve disease, and there is a possibility of missed diagnosis for ARPA. Therefore, CT examination or invasive imaging examination is necessary in patients with pulmonary hypertension. Especially combined with a variety of complicated congenital heart disease, echocardiography combined with cardiac catheterization, ventricle and large angiography is very helpful to explain the condition. In this case, there was PVS and blood return. Percutaneous pulmonary valve balloon dilation may aggravate blood reflux, that is not appropriate. The patient presented with postactive dyspnea without hemoptysis, and there was no evidence of the need for vascular embolization despite the presence of collateral flow from the descending aorta to the right lung. According to the relevant data of UAPA and PVS, surgical treatment may be a better choice for PVS and UAPA³. We suggest that surgical treatment should be positively considered in this case.

Conclusions

A 52-year-old woman with only a one-year history of post-activity dyspnea was diagnosed with UAPA and PVS with pulmonary hypertension and descending aortic collateral flow to the right lung by echocardiography, cardiac catheterization, and invasive imaging techniques. It reminds that echocardiography combined with detailed imaging examination is very important for the diagnosis of complex congenital heart disease and provide the basis for the selection of the best treatment.

Abbreviations

ARPA: Absence of right pulmonary artery; PVS: Pulmonary valve stenosis

UAPA: Unilateral absence of pulmonary artery; CT: Computed tomographic

Declarations

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Not applicable.

Authors'contributions

JWL drafted the manuscript. CFZ organized the study. JWL and CJ participated in the treatment. JWL contributed to the development of methodology. All authors read and approved the final manuscript.

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Ethics approval and consent to participate

Not applicable.

Consent for publication

Written informed consent of clinical detail and image publication was obtained from the patient.

Competing interest

None declare

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Figures

Figure 1

Echocardiography indicates moderate and severe PVS and regurgitation.

Figure 2

Right ventricular angiography showed "vault sign" and "spray column sign". Pulmonary angiography indicated that ARPA, and the left pulmonary artery and its branches were dilated.

Supplementary Files

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