Isolated agenesis of the right pulmonary veins with pulmonary sequestration

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ABSTRACT

We report two cases of agenesis of the right pulmonary veins (PVs) associated with sequestration of the right lung with systemic to pulmonary collateral. Both the children were referred for evaluation of recurrent lower respiratory tract infections. Transthoracic echocardiographic evaluation showed reversal of flow in the right pulmonary artery (PA) and absent right PVs. Cardiac catheterization confirmed the diagnosis of agenesis of the right PVs. Transcatheter closure of collaterals was performed in view of significant shunt from systemic to PA. Both the patients are asymptomatic on follow-up.

Keywords: Dextrocardia, pulmonary sequestration, pulmonary veins

INTRODUCTION

Embryologically, pulmonary veins (PVs) are formed when the splanchnic plexus around the lung buds fuses with the common PV from the posterior wall of the left atrium (LA). Defects during early embryonic stage may result in PV agenesis.[1,2] Recurrent lower respiratory infection (LRTI) is the most common presentation due to association with pulmonary sequestration and systemic to pulmonary collaterals.[2-4] Symptomatic patients may benefit from transcatheter closure of collaterals.[5] Here, we report our experience of two cases of isolated right PV agenesis.

CASE REPORTS

Case 1

A 15-month-old boy presented with symptoms of failure to thrive (weight < 5th percentile) and two episodes of LRTIs requiring hospitalization. Systemic examination was unremarkable. Chest X-ray revealed a shift of mediastinum with reduced pulmonary vascularity on the right side. Transthoracic echocardiogram (TTE) showed small right pulmonary artery (RPA: 'z' score -1.8) with complete reversal of flow on color flow mapping. Spectral Doppler showed a venous flow pattern in the RPA [Figure 1]. The right PVs were not visualized. Diagnostic cardiac catheterization showed normal PA pressures. The pulmonary angiogram revealed negative washout of contrast from RPA toward the left PA (LPA) [Figure 2]. Levophase showed a normal drainage of the left-sided PVs to LA and absence of right PVs. Aortogram showed multiple collaterals from the descending thoracic aorta supplying the right lung and subsequently filling the RPA. Levophase showed opacification of the RPA followed by contrast washout into LPA and subsequently left PVs into LA, leading to an arterioarterial circular shunt (physiological left-to-right shunt). This suggested complete absent of flow from the main PA to RPA. As the right lung was not contributing to oxygenation, the multiple aortopulmonary collaterals were closed using multiple Gianturco coils (Cook Medical, Inc., Bloomington, IN, USA).

Case 2

A 4-year-old boy was evaluated in view of recurrent LRTIs. Chest X-ray showed mediastinal shift to the right

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with differential vascularity [Figure 3]. TTE revealed hypoplastic RPA ('z' score -3.2). The right PVs were not visualized whereas the left PVs had an increased venous return. A collateral was seen originating from the celiac axis and supplying right lung. Computed tomography (CT) angiogram confirmed the findings of TTE. There was associated aberrant left subclavian artery and hypoplastic right lung with thickened interlobar fissure [Figure 4].

Cardiac catheterization showed O_2 step-up of 15% from mixed venous to LPA, with Qp: Qs of 1.8:1. The PA pressures were normal. Angiographic findings were similar to Case 1, except a single collateral arising from the celiac axis. It was closed with an 8-mm Amplatzer Vascular Plug 4 (St. Jude Medical, Inc., MN, USA) [Figure 5].

DISCUSSION

Atresia of unilateral PVs is a rare anomaly and nearly 50 cases were reported in the literature.[6] About 30%–50% are associated with other abnormalities, namely septal defects and complex anomalies such as single ventricle, veno-occlusive disease, and Scimitar syndrome.[7-9]

Embryologic basis

Pulmonary vasculature develops from splanchnic plexus surrounding the primitive lung buds. The splanchnic plexus during its development drains to systemic venous channels and establishes its communication with common PV by the end of 4th week. The connection between splanchnic plexus and systemic veins involutes subsequently. Faulty incorporation of PVs after the involution of their connections with systemic veins leads to atresia of PVs.[1,2]

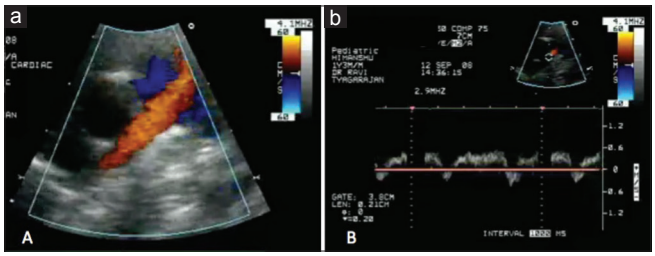

Figure 1: (a) Flow reversal with centripetal flow pattern on color flow mapping in the right pulmonary artery. (b) Spectral Doppler revealed a venous flow pattern in the right pulmonary artery

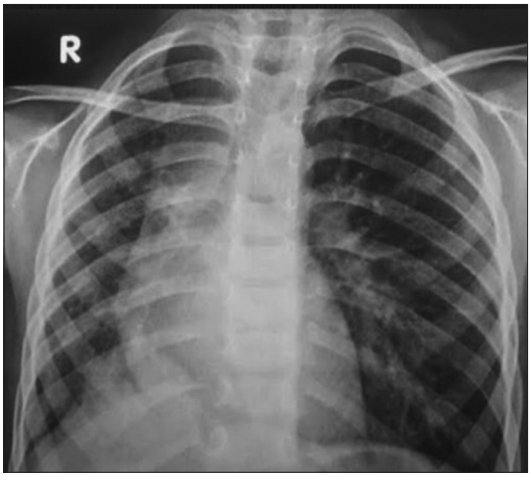

Figure 3: Dextroposition of heart. Reduced right lung vascularity than the left

Pathophysiology and clinical attributes

Failure of PVs to drain to LA leads to various histopathologic abnormalities, including intimal and medial thickening, fibrosis, and dilation of other PVs [Figure 5]. The ipsilateral PA and lung are underdeveloped and receive systemic collaterals, which can lead to left-to-right shunt. This may cause pulmonary hypertension, hemoptysis, or proneness to recurrent LRTIs. The clinical presentation may vary from asymptomatic state to severe pulmonary hypertension. This is essentially due to the balance between systemic collateral supply to affected lung and efficiency of its drainage and secondary changes in lung parenchyma due to associated congenital heart defects.[2,4]

Diagnosis

Chest X-ray (mediastinal shift) and TTE (reversal flow in the ipsilateral PA) provide subtle clues to the diagnosis. Definitive diagnosis can be made either on CT angiogram or on pulmonary arterial wedge angiography. Nonvisualization of confluence of PVs to LA, presence of systemic collateral, and dilated bronchial veins on CT angiogram are in favor of PV atresia.[3,5,10] Cardiac catheterization conclusively demonstrates atresia of PVs,

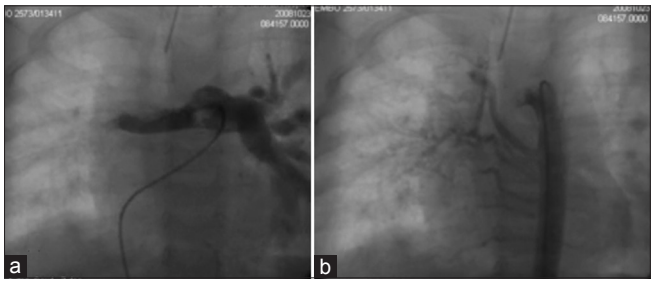

Figure 2: (a) Pulmonary angiogram revealed negative washout of contrast from right pulmonary artery toward the left pulmonary artery. (b) Aortogram shows multiple collaterals to the right lung

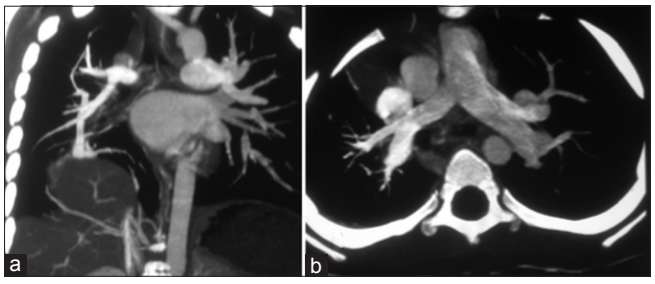

Figure 4: (a) Right pulmonary veins not seen. (b) Small right pulmonary artery than the left pulmonary artery

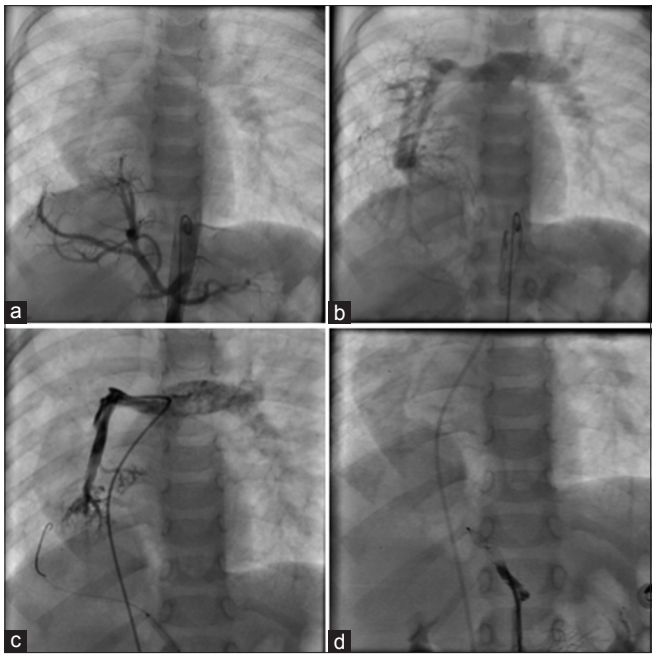

Figure 5: (a) Aortogram showing collateral from the celiac axis to the right lower lobe. (b) Subsequent opacification of the right pulmonary artery without pulmonary venous phase. (c) Pulmonary angiogram done with balloon occlusion of collateral showing agenesis of right-sided pulmonary veins. The right pulmonary artery is small. (d) Vascular plug occlusion to collateral

presence of systemic collaterals, degree of left-to-right shunt, and severity of pulmonary hypertension.[1,3,7]

Management

Various treatment modalities employed include conservative management, transcatheter closure of systemic collaterals, and pneumonectomy. Transcatheter closure of systemic collaterals is indicated in children with recurrent LRTI, presence of pulmonary hypertension, and recurrent episodes of hemoptysis.[5] Both our patients responded very well to transcatheter closure of collaterals. The rarity of the condition poses difficulty to fathom its natural history; however, high mortality to the tune of 50% has been reported in various case reports.[1] Timely intervention can improve symptomatic status and alleviate progression of pathologic changes.

Our report highlights the importance of appropriate diagnostic evaluation and management of isolated right PV agenesis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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