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Hitherto Unreported Pattern of Complex Obstructive Partial Anomalous Pulmonary Venous Drainage with Dual Drainage of Accessory Pulmonary Veins

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Abstract

Partial anomalous pulmonary venous drainage is a congenital cardiac disorder characterized by abnormal drainage of one or more pulmonary veins into the systemic circulation. It can be isolated or associated with other congenital cardiac anomalies, most commonly atrial septal defect and patent ductus arteriosus. The clinical presentation is variable and depends on the degree of shunting and associated cardiac anomalies. Many patients usually remain asymptomatic until late in life. In this article, we presented a complex case of obstructive partial anomalous pulmonary venous drainage with dual drainage of bilateral accessory pulmonary veins with intact interatrial septum in conjunction with a patent ductus arteriosus and a ventricular septal defect. This pattern is incredibly rare and to the best of our knowledge has not been previously reported. Computed tomography played a pivotal role in precisely elucidating the intricate anatomy in this case with a complex pattern of anomalous pulmonary venous drainage.

Keywords

- accessory pulmonary veins
- PAPVC
- dual drainage
- obstructive partial anomalous pulmonary venous drainage
- partial anomalous pulmonary venous drainage

Introduction

Partial anomalous pulmonary venous connection (PAPVC) is a congenital cardiac anomaly characterized by the abnormal drainage of one or more pulmonary veins into the systemic venous circulation, likely due to the incomplete involution of primitive pulmonary venous connections.[1] The most common anomalous patterns are isolated drainage of the right superior pulmonary vein (RSPV) into the superior vena cava (SVC) or right atrium (RA) and left superior pulmonary vein (LSVP) draining into the innominate vein or the coronary sinus.[2,3] Patients with PAPVC are usually asymptomatic and can be incidentally detected. The severity of symptoms depends on the degree of shunting and coexistent cardiac defects. It is associated with atrial septal defect (ASD) in 90% of cases.[4] Obstruction to the anomalous connection can result in elevated venous pressures, subsequently leading to pulmonary edema and respiratory distress. Obstruction is more commonly seen in infracardiac total anomalous pulmonary venous connection (TAPVC) cases and is rarely reported in supra cardiac PAPVC.[5] In this article, we presented an extremely rare case of complex obstructive supra

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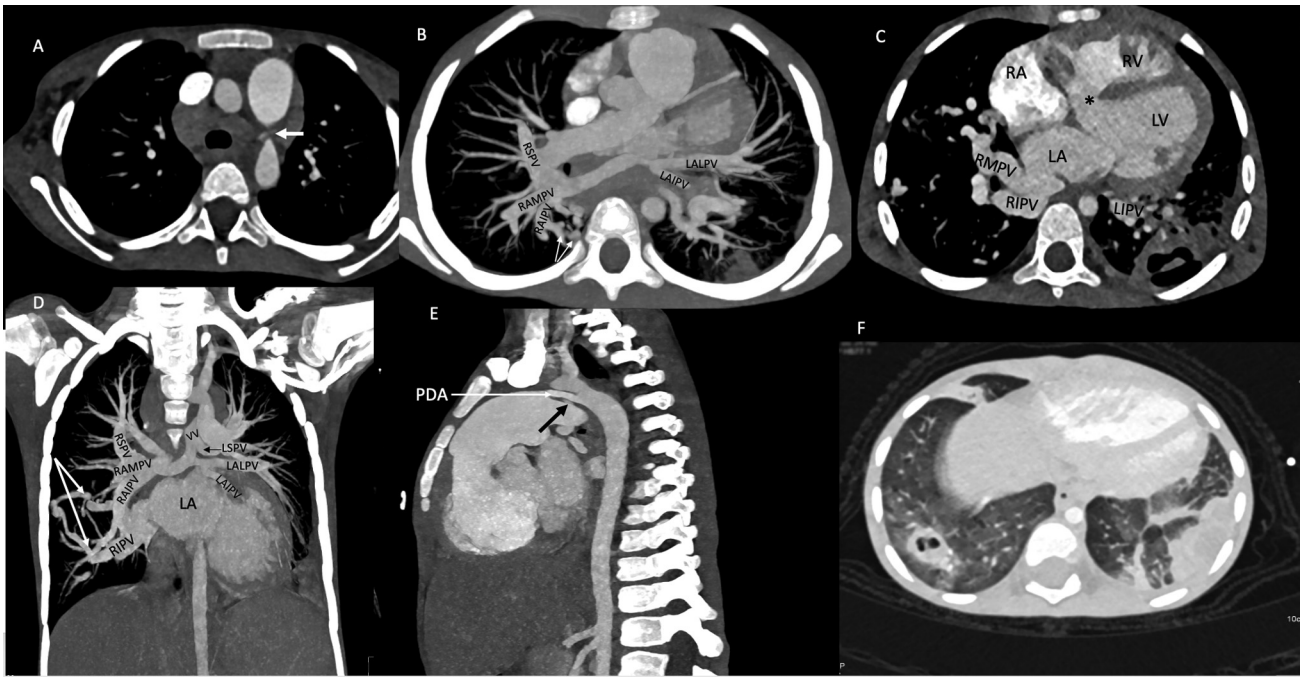

Fig. 1 (**A**) Axial image showing compression of vertical vein (white arrow). (**B**) Axial maximum intensity projection images depicting right superior pulmonary vein (RSPV), right accessory middle pulmonary vein (RAMPV), right accessory inferior pulmonary vein (RAIPV), left accessory lingula and accessory inferior pulmonary veins (LALPV, LAIPV) forming an anomalous common venous channel and presence of communicating collaterals(grouped white arrows). (**C**) Axial image showing ventricular septal defect (*) with right middle lobe vein (RMPV), right and left inferior pulmonary veins (RIPV, LIPV) draining into the left atrium(LA). (**D**) Coronal reformatted maximum intensity projection image showing accessory pulmonary veins and dual drainage via communicating channels (grouped white arrows). (**E**) Sagittal reformatted image showing obstruction of the vertical vein (VV) (black arrow) at the level of patent ductus arteriosus (PDA). (**F**) Axial image (lung window) showing consolidations in bilateral lower lobes with cavitation in right lower lobe. LSPV, left superior pulmonary vein; LV, left ventricle; RV, right ventricle.

cardiac PAPVC with intact interatrial septum, dual drainage of bilateral accessory pulmonary veins associated with the presence of a ventricular septal defect (VSD), and persistent ductus arteriosus (PDA).

Case Report

A 5-year-old child patient presented with chief complaints of fever, difficulty in breathing, and history of recurrent respiratory tract infections. On examination, pulse rate was 120/min, respiratory rate was 42/min, oxygen saturation was 94% at room air, and blood pressure was 96/58 mm Hg. Echocardiography revealed the presence of a perimembranous VSD. Computed tomography (CT) thoracic angiography showed perimembranous VSD and PDA with intact interatrial septum and distinct anomalous pulmonary venous drainage. Accessory pulmonary veins from the right middle lobe, lingula, and bilateral lower lobes were exhibiting a rare phenomenon of dual drainage into both systemic and pulmonary venous system. Bilateral superior pulmonary veins along with these accessory pulmonary veins formed the common vertical vein that was seen to ascend between descending thoracic aorta and left pulmonary artery, immediately inferior to PDA, eventually draining into the left innominate vein. Significant compression of the vertical vein was seen along its course between PDA and left pulmonary artery. The accessory veins via tortuous communicating channels also showed drainage into the left atrium, along with the right middle and bilateral inferior pulmonary veins (►**Figs. 1**–**3**). Coronary arteries were normal. No coarctation of the aorta was seen. Aortic arch was left-sided with normal branching pattern. Additionally, CT revealed the presence of cavitary consolidations in the bilateral lower lobes. Unfortunately, the child's condition deteriorated rapidly, and succumbed to the illness.

Discussion

PAPVC is a rare congenital cardiac disorder, constituting approximately 0.4 to 0.7% of cardiovascular conditions, and likely arises from persistent primitive pulmonary venous connections.[6] During the embryological course of development, the primitive lung buds in the foregut are surrounded by the splanchnic venous plexus that drains into the bilateral cardinal veins and umbilical -vitelline vein. A primitive pulmonary vein develops as an outpouching in relation to the posterior wall of the left atrium and establishes communication with the splanchnic plexus. Eventually, the pulmonary venous communication with the cardinal veins and umbilical-vitelline vein involutes, and the primitive pulmonary vein incorporates into the atrial wall, resulting in the formation of four pulmonary veins that drain into the left atrium.[7]

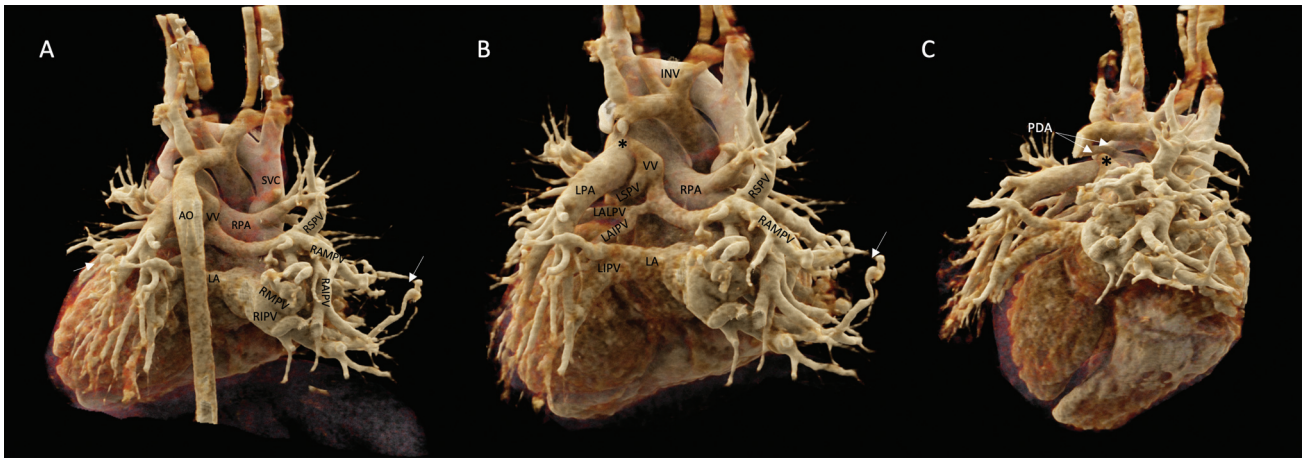

Fig. 2 (A–C) Volume-rendered computed tomographic images showing anomalous drainage of pulmonary veins with obstruction (*) of the vertical vein (VV) at the level of patent ductus arteriosus (PDA) (grouped white arrows) and dual drainage of accessory veins into vertical vein and via communicating collaterals (single arrows) into LA. AO, aorta; INV, innominate vein; LA, left atrium; LAIPV, left accessory inferior pulmonary vein; LALPV, left accessory lingular pulmonary vein; LIPV, left inferior pulmonary vein; LPA, left pulmonary artery; LSPV, left superior pulmonary vein; RAIPV, right accessory inferior pulmonary vein; RAMPV, right accessory middle pulmonary vein; RIPV, right inferior pulmonary vein; RMPV: right middle pulmonary vein; RPA, right pulmonary artery; RSPV, right superior pulmonary vein; SVC, superior vena cava.

The common patterns of anomalous drainage involve isolated drainage of the RSPV into the SVC or RA and LSVP draining into the innominate vein or the coronary sinus. PAPVC can be associated with other cardiac defects such as ASD, PDA, and heterotaxy syndrome. ASD is associated with almost 90% cases.[2–4,6] In addition to deviations in drainage, inconsistencies in embryological venous development can also lead to other anatomical variations such as accessory or supernumerary pulmonary veins. Patients with PAPVC are often asymptomatic and may be incidentally detected or may develop symptoms later in life. PAPVC acts as a left-to-right shunt, causing gradual remodeling of the pulmonary vasculature due to increased pulmonary flow, ultimately resulting in pulmonary hypertension. Obstruction to the anomalous connection can result in increased pulmonary venous pressure, right-to-left shunt across associated ASD or VSD, resulting in cyanosis, pulmonary edema, and the need for urgent intervention.[7,8] Obstruction is most commonly described in infradiaphragmatic type of TAPVC and is extremely rare in PAPVC. Present case shows extremely rare coexistent anomalies, including obstruction in case of PAPVC at the level of PDA, an intact interatrial septum, presence of VSD, and dual accessory pulmonary venous drainage.

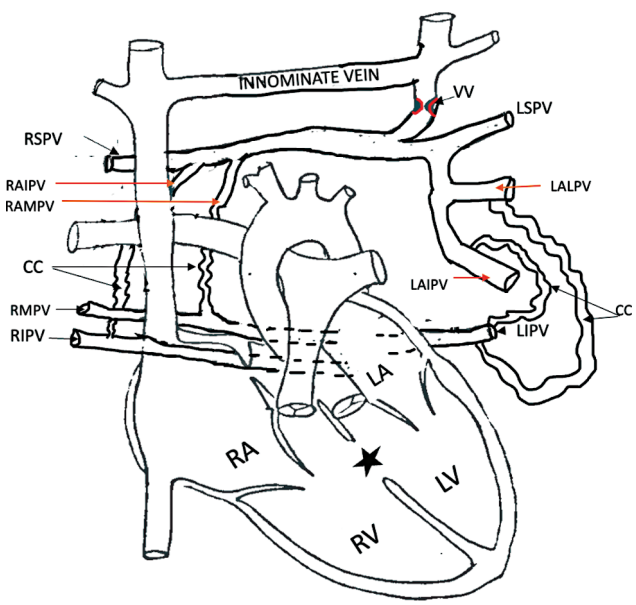

Fig. 3 Schematic diagram depicting the complex anomalous pulmonary venous drainage with obstruction at the level of patent ductus arteriosus (PDA) (narrowing of the vertical vein is marked in red), accessory pulmonary veins (orange arrow), dual drainage via communicating channels (CC), and ventricular septal defect (star). LA, left atrium; LAIPV, left accessory inferior pulmonary vein; LALPV, left accessory lingular pulmonary vein; LIPV, left inferior pulmonary vein; LSPV, left superior pulmonary vein; LV, left ventricle; RAIPV, right accessory inferior pulmonary vein; RAMPV, right accessory middle pulmonary vein; RA, right atrium; RIPV, right inferior pulmonary vein; RMPV, right middle pulmonary vein; RSPV, right superior pulmonary vein; RV, right ventricle; VV, vertical vein.

Modern multidetector CT scans excellently depict the anatomy of vascular structures in the thorax, with the advantages of rapid scans, elimination of motion artifacts, and multiplanar reformat and three-dimensional reconstruction images. It can be extremely helpful in identifying and delineating complex cardiac anomalies, particularly in cases where echocardiography provides an incomplete evaluation due to limited window. This enables the surgical team to formulate preoperative surgical strategies, anticipate potential challenges or complexities during the procedure, and develop contingency plans.[9,10] Although PAPVC is generally asymptomatic, the presence of pulmonary venous obstruction can lead to an early onset presentation with tachypnoea, respiratory infections, pulmonary edema, or cyanosis with poor prognosis.

Conflict of Interest
None declared.

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