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Case Report

Neonatology Section

Pulmonary Venous Atresia with Abnormal Drainage in a Neonate: A Case of Missing Vein

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ABSTRACT

Pulmonary Venous Atresia (PVA) is a rare congenital cardiovascular anomaly in which one or more pulmonary veins fail to establish a normal connection with the left atrium, resulting in obstruction of pulmonary venous return and progressive pulmonary venous hypertension. When associated with Partial Anomalous Pulmonary Venous Connection (PAPVC), pulmonary venous drainage is redirected into the systemic venous circulation, leading to abnormal haemodynamics and right-sided cardiac volume overload. Early recognition is essential because delayed diagnosis may result in severe cardiopulmonary complications. This case report describes a 30-day-old male neonate who presented with persistent respiratory distress, feeding difficulty, tachypnoea, and mild central cyanosis. Chest radiography demonstrated mild cardiomegaly with prominent pulmonary vascular markings. Transthoracic echocardiography revealed dilatation of the right atrium and right ventricle with a relatively small left atrium; however, pulmonary venous connections were not clearly visualised. Multidetector Computed Tomography (MDCT) angiography was therefore performed for detailed anatomical evaluation. Imaging demonstrated absence of the right pulmonary veins and the left lower pulmonary vein, consistent with PVA. The remaining left upper pulmonary vein drained anomalously into the left innominate vein through a vertical vein, representing PAPVC. The infant underwent surgical rerouting of the anomalous vertical vein to the left atrium with closure of the Atrial Septal Defect (ASD), successfully restoring physiological pulmonary venous drainage. Postoperatively, oxygen saturation improved and respiratory symptoms resolved. This case highlights the importance of MDCT angiography in accurately delineating complex pulmonary venous anomalies and guiding timely surgical management.

Keywords: Angiography, Cardiomegaly, Central cyanosis, Congenital vascular anomaly, Pulmonary hypertension

CASE REPORT

A 30-day-old male neonate presented to the Department of Neonatology with persistent respiratory distress and feeding difficulty. The infant was born at 38 weeks of gestation by normal vaginal delivery to a 26-year-old mother with an uneventful antenatal history. There was no history of maternal diabetes, hypertension, infections, or teratogenic drug exposure during pregnancy. Antenatal ultrasonography had not revealed any structural abnormalities. The infant had no siblings and there was no significant family history of congenital heart disease or genetic disorders. The neonate had a birth weight of 2.7 kg and a crown-heel length of 54 cm, with a calculated body surface area of approximately 0.2 m^2.

On examination, the infant demonstrated tachypnoea and increased work of breathing with mild central cyanosis. Oxygen saturation measured by pulse oximetry was 84% on room air and improved to 92% with supplemental oxygen. The respiratory rate was 64 breaths per minute and the heart rate was 152 beats per minute. Cardiovascular examination revealed a prominent right ventricular impulse and an accentuated pulmonary component of the second heart sound (loud P2). A soft systolic murmur was audible along the left lower sternal border. Mild hepatomegaly (approximately 2 cm below the right costal margin) was noted. No peripheral oedema or dysmorphic features were observed. Other systemic examinations were within normal limits. The infant was on exclusive breastfeeding, although feeding was reduced due to respiratory distress.

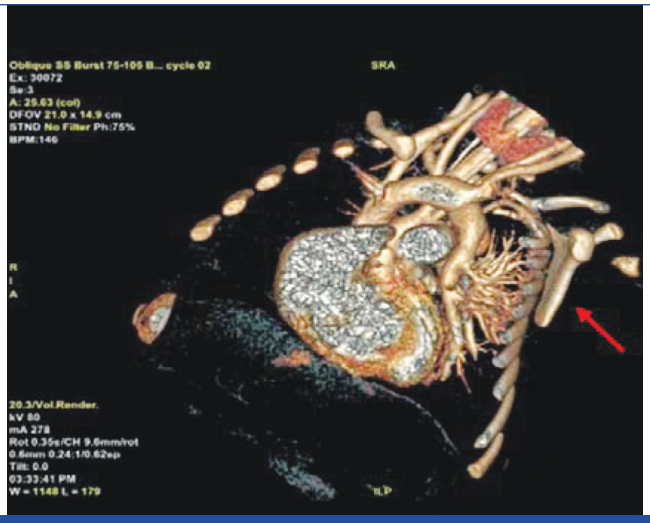
A preliminary chest radiograph demonstrated mild cardiomegaly with prominent pulmonary vascular markings, raising suspicion of an underlying congenital cardiac anomaly. Transthoracic echocardiography showed dilatation of the right atrium and right ventricle with a relatively small left atrium and left ventricle. A small ASD was also identified, although the pulmonary venous connections could not be clearly delineated.

For further anatomical evaluation, contrast-enhanced MDCT angiography was performed using a 128-slice CT scanner with a paediatric low-dose protocol. Non-ionic iodinated contrast (1.5 mL/kg) was administered intravenously at a rate of 1 mL/s, and image acquisition was performed using a bolus-tracking technique with the region of interest placed in the main pulmonary artery. Scanning was initiated once the attenuation reached 100 HU. ECG gating was not used. The estimated radiation dose for the study was approximately 1-2 mSv. Image acquisition was performed with thin-slice reconstruction followed by multiplanar reformations, maximum intensity projection, and three-dimensional volume-rendered reconstructions.

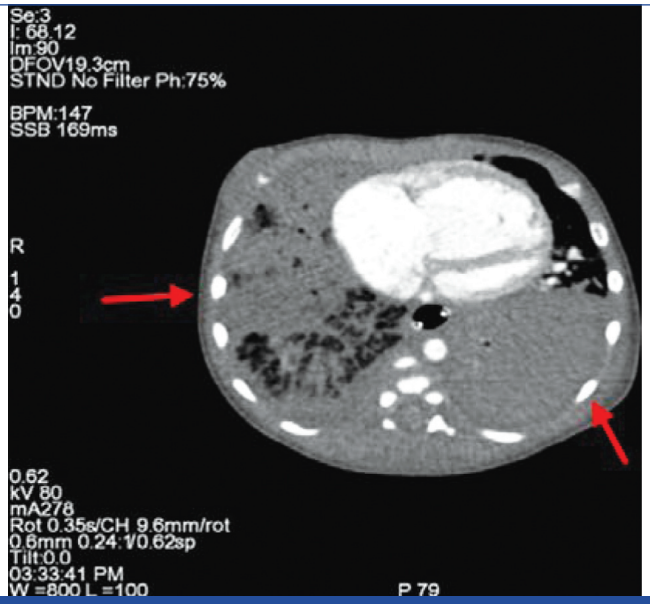
MDCT imaging demonstrated levocardia with situs solitus and preserved atrioventricular and ventriculoarterial concordance. The right atrium and right ventricle were markedly dilated, while the left atrium and left ventricle appeared relatively small, indicating right-sided volume overload. The left upper pulmonary vein drained via a vertical vein into the left innominate vein, consistent with PAPVC. The right pulmonary veins and left lower pulmonary vein were not visualised, suggesting PVA [Table/Fig-1,2].

Evaluation of the pulmonary venous system revealed absence of the right pulmonary veins and the left lower pulmonary vein, consistent with PVA. The left upper pulmonary vein formed a vertically oriented venous channel draining into the left innominate vein through a vertical vein measuring approximately 7.3 mm in diameter, representing Partial Anomalous Pulmonary Venous Connection (PAPVC). Absence of the right pulmonary veins and the left lower pulmonary vein supports the diagnosis of PVA with PAPVC [Table/Fig-3].

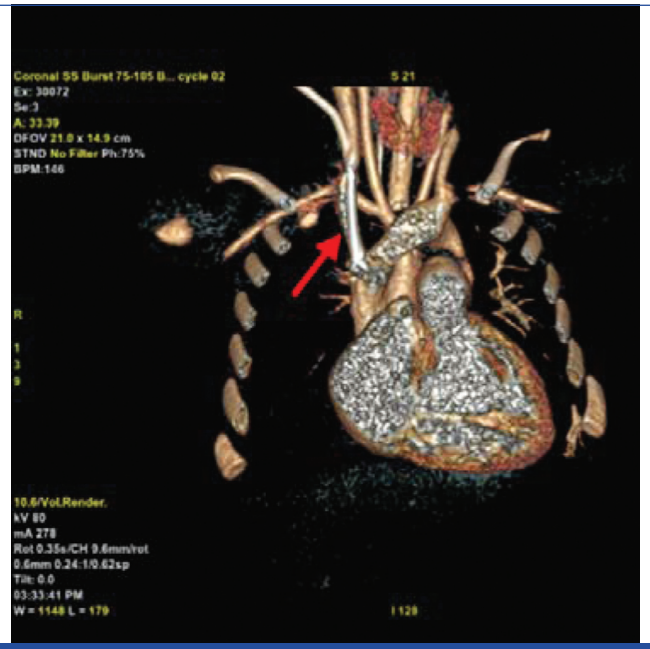
Three-dimensional CT angiographic reconstructions further illustrated the anomalous pulmonary venous drainage bypassing the left atrium and entering the systemic venous circulation through the innominate vein [Table/Fig-4].



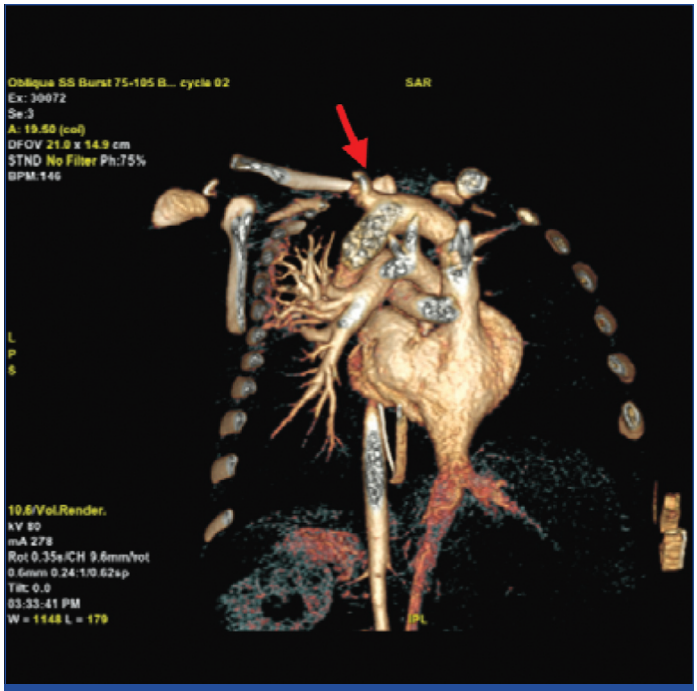
[Table/Fig-1]: Three-dimensional volume-rendered CT angiogram (left anterior oblique view) showing levocardia, markedly dilated right atrium and right ventricle, with absence of identifiable pulmonary veins consistent with PVA.



[Table/Fig-2]: Axial contrast-enhanced CT image at the level of the cardiac chambers showing dilated right atrium and right ventricle with a relatively small left atrium, consistent with right-sided volume overload due to anomalous pulmonary venous drainage.

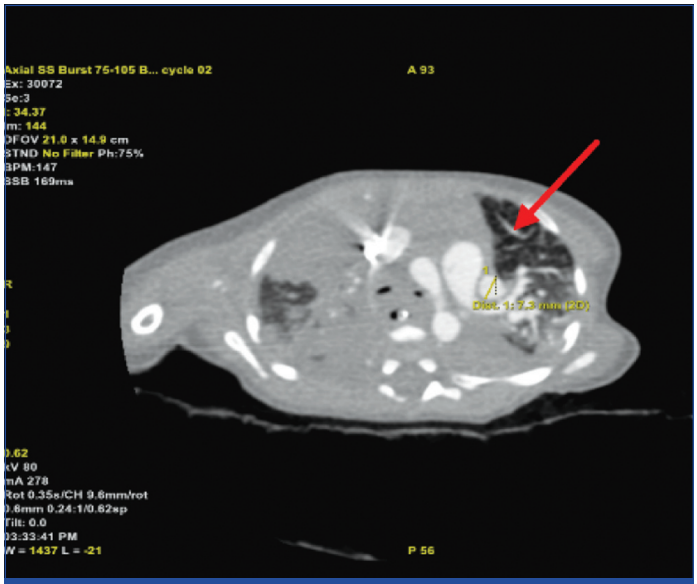


[Table/Fig-3]: Coronal three-dimensional CT reconstruction demonstrating the vertically oriented anomalous pulmonary vein draining into the left innominate vein, absence of the right pulmonary veins and left lower pulmonary vein.



[Table/Fig-4]: Oblique volume-rendered CT angiogram (right anterior oblique projection) illustrating anomalous pulmonary venous drainage bypassing the left atrium and entering the left innominate vein through the vertical vein.

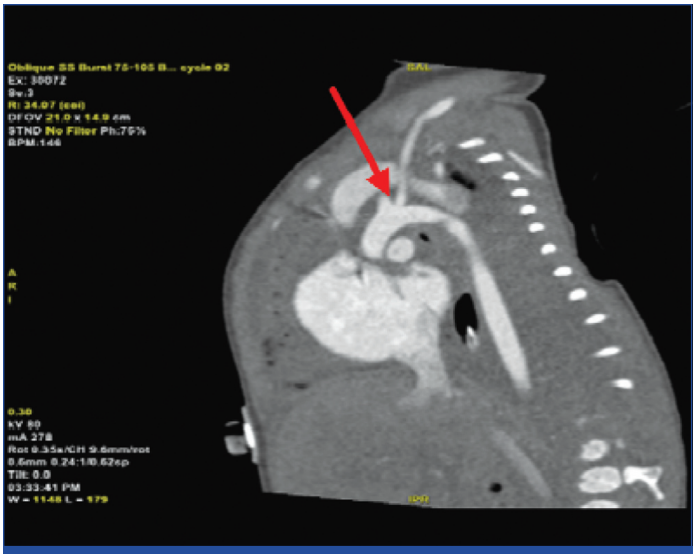
Axial CT images demonstrated the vertical venous channel coursing superiorly toward the innominate vein and associated pulmonary parenchymal changes, including consolidation and ground-glass opacities consistent with pulmonary venous congestion [Table/Fig-5].



[Table/Fig-5]: Axial contrast-enhanced CT image demonstrating the vertical vein measuring approximately 7.3 mm draining into the left innominate vein.

Sagittal CT reconstruction illustrated the spatial relationship between the aortic arch, vertical vein, and absence of direct pulmonary venous drainage into the left atrium [Table/Fig-6].

Quantitative imaging measurements demonstrated enlargement of the pulmonary arterial system, with the main pulmonary artery, right pulmonary artery, and left pulmonary artery showing elevated Z-scores, consistent with increased pulmonary arterial pressure. In contrast, the Sinotubular Junction (STJ) measured 7.7 mm with a Z-score of -0.98, which lies within normal limits. This relative preservation of the STJ dimension, despite significant right-sided cardiac dilatation, suggests that the volume overload is predominantly confined to the right heart and pulmonary circulation, without associated dilation of the systemic arterial outflow tract [Table/Fig-7] [1].



[Table/Fig-6]: Sagittal contrast-enhanced CT reconstruction showing the relationship between the aortic arch and the anomalous vertical vein, with absence of direct pulmonary venous drainage into the left atrium.

Parameters	Measured Value (mm)	Normal Range (mm)	Z-score
Main Pulmonary Artery (MPA)	13.0	5.8-10.8	+3.18
Right Pulmonary Artery (RPA)	6.0	3.3-6.4	+1.65
Left Pulmonary Artery (LPA)	7.5	3.1-6.0	+3.36
Sinotubular Junction (STJ)	7.7	7.0-10.4	-0.98
Distal aortic arch	6.1	4.2-7.9	+0.36
Vertical vein diameter	7.3	-	-

[Table/Fig-7]: Summary of imaging-based measurements [1].

Based on the clinical presentation and initial echocardiographic findings, several differential diagnoses were considered, including Total Anomalous Pulmonary Venous Connection (TAPVC), pulmonary vein stenosis, and congenital heart diseases associated with pulmonary hypertension. TAPVC was considered because of the presence of right-sided cardiac enlargement and suspected abnormal pulmonary venous drainage; however, this diagnosis was ruled out on MDCT angiography, which demonstrated absence of the right pulmonary veins and the left lower pulmonary vein rather than anomalous drainage of all pulmonary veins. Pulmonary vein stenosis was also considered but excluded because imaging showed complete non-visualisation of the affected pulmonary veins rather than narrowed venous channels. Other congenital cardiac anomalies causing right heart enlargement, such as large ASD or ventricular septal defect, were excluded based on echocardiographic findings. The combination of absent pulmonary veins and anomalous drainage of the remaining left upper pulmonary vein into the left innominate vein on MDCT angiography confirmed the diagnosis of PVA with PAPVC.

The initial management focused on stabilising the infant's respiratory status. Continuous Positive Airway Pressure (CPAP) and supplemental oxygen were administered to correct hypoxaemia. Supportive care included close monitoring of vital parameters, nutritional support, and infection prophylaxis with intravenous cefotaxime at a dose of 135 mg (50 mg/kg) every eight hours for five days. Following multidisciplinary evaluation by neonatology, paediatric cardiology, and cardiothoracic surgery teams, the infant underwent pre-anaesthetic evaluation, including assessment of airway patency, respiratory status, and cardiovascular stability. Baseline laboratory investigations, including haemoglobin, renal function, and coagulation profile, were reviewed. The infant was optimised with oxygen support and was deemed fit for surgery. The infant subsequently underwent surgical correction under cardiopulmonary bypass. The anomalous vertical vein carrying the left upper pulmonary venous return was surgically mobilised and anastomosed to the left atrium to establish physiological pulmonary venous drainage. The proximal connection of the vertical vein to the innominate vein was ligated to prevent residual shunting. The ASD was closed primarily. Intraoperative inspection confirmed that the right pulmonary veins and the left lower pulmonary vein were atretic and not amenable to reconstruction.

Postoperatively, the infant was monitored in the neonatal intensive care unit. Oxygen saturation improved to 97% by the third postoperative day and respiratory distress gradually resolved. The postoperative period was complicated by transient renal dysfunction, which resolved with supportive management, including carefully titrated intravenous isotonic (normal saline) fluid therapy. Although fluid restriction is typically recommended in post-cardiac surgery neonates, a relatively higher maintenance fluid rate of 110 mL/kg/day for 48 hours was administered in this case to ensure adequate renal perfusion and support urine output during the period of transient renal impairment. Fluid balance and cardiorespiratory status were closely monitored to avoid volume overload. The infant showed progressive clinical improvement and was discharged after 12 days of hospitalisation with stable cardiorespiratory status. Low-dose aspirin therapy was initiated as antiplatelet therapy for thromboprophylaxis at a dose of 8.1 mg (3 mg/kg) once daily.

At one-month follow-up, repeat imaging demonstrated unobstructed flow through the reconstructed pulmonary venous pathway with improvement in cardiac chamber dimensions. The infant remains clinically stable and continues regular follow-up with the paediatric cardiology team. The patient was last followed-up at three months postoperatively and remained clinically stable.

DISCUSSION

The PVA is a rare congenital cardiovascular anomaly characterised by the absence or failure of one or more pulmonary veins to establish a normal connection with the left atrium. This results in obstruction of pulmonary venous return and leads to pulmonary venous hypertension, pulmonary congestion, and progressive pulmonary vascular disease [2]. Depending on the severity and number of pulmonary veins affected, patients may present with respiratory distress, cyanosis, pulmonary hypertension, and right-sided cardiac enlargement during the neonatal period. PAPVC is another rare congenital anomaly in which one or more pulmonary veins drain into the systemic venous circulation instead of the left atrium, producing abnormal pulmonary venous return and right heart volume overload [3]. When PVA occurs in association with PAPVC, the haemodynamic consequences can be more complex because pulmonary venous drainage is partially obstructed and partially redirected into systemic circulation [4].

In the present case, three pulmonary veins (right upper, right lower, and left lower pulmonary veins) were not visualised, consistent with PVA, while the remaining left upper pulmonary vein drained into the left innominate vein through a vertical vein, representing PAPVC. This unusual anatomical configuration resulted in abnormal pulmonary venous return and significant right-sided cardiac dilation.

Previous reports have described cases of common pulmonary vein atresia in neonates and emphasised that early diagnosis is essential because severe cyanosis and circulatory compromise can occur soon after birth [5]. Rare neonatal variants of PVA may initially mimic TAPVC, highlighting the diagnostic difficulty of this condition [6]. Isolated unilateral PVA has also been reported presenting with respiratory symptoms and pulmonary hypertension, underscoring the importance of early identification of pulmonary venous anomalies [7].

Unilateral pulmonary vein atresia may present with life-threatening complications such as haemoptysis, and surgical intervention can improve clinical outcomes in selected patients [8]. It may also manifest as recurrent pulmonary infections and respiratory symptoms due to chronic pulmonary venous obstruction [9].

The PAPVC is a rare congenital anomaly in which one or more pulmonary veins drain into systemic veins, producing a left-to-right shunt and right heart enlargement [10]. It has been reported to have a prevalence of approximately 0.4-0.7% in autopsy series and may remain asymptomatic or present with symptoms related to right-sided volume overload [11].

The PAPVC can be associated with pulmonary hypertension, and early recognition is important to prevent long-term complications [12]. PVA may also present with diverse clinical manifestations, including recurrent pleural effusion, reflecting the variability in presentation [13].

The MDCT angiography plays a crucial role in accurately delineating pulmonary venous anatomy and identifying anomalous venous drainage patterns [14]. Cross-sectional imaging provides detailed visualisation of anomalous pulmonary venous pathways and facilitates surgical planning [15].

In the present case, MDCT angiography played a critical role in identifying the absence of multiple pulmonary veins and demonstrating the anomalous drainage of the left upper pulmonary vein into the left innominate vein. Surgical rerouting of the vertical vein to the left atrium successfully restored physiological pulmonary venous return and resulted in clinical improvement.

CONCLUSION(S)

The PVA associated with PAPVC is an extremely rare congenital vascular anomaly that may present in the neonatal period with respiratory distress and right-sided cardiac enlargement. This case highlights the diagnostic value of MDCT angiography in accurately delineating complex pulmonary venous anatomy when echocardiography is inconclusive. Early recognition of these anomalies is essential for appropriate surgical planning and timely intervention. Surgical rerouting of the anomalous pulmonary venous pathway can successfully restore physiological pulmonary venous return and improve clinical outcomes. Therefore, a high index of suspicion combined with advanced imaging and multidisciplinary management is crucial for the diagnosis and treatment of rare pulmonary venous anomalies in neonates.

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REFERENCES

[1] Cantinotti M, Giordano R, Scalese M, Murzi B, Assanta N, Spadoni I, et al. Nomograms for two-dimensional echocardiography derived valvular and arterial dimensions in Caucasian children. J Cardiol. 2017;69(1):208-15.
[2] Sim YW, Park J, Park B, Lim JK, Shin KM, Kim YS. Unilateral pulmonary vein atresia initially presenting as interstitial lung disease in an elderly female: Serial chest radiograph changes and its literature review. J Korean Soc Radiol. 2022;83(2):372.
[3] Choi MH, Greathouse F, Darki A. Medical management of isolated partial anomalous pulmonary venous connection with symptomatic right ventricular failure and pulmonary hypertension. Cureus. 2024;16(6):e62779.
[4] Seecheran RV, Dookie T, Seecheran VK, Persad SA, Marsang BL, Rampersad F, et al. Partial anomalous pulmonary venous connection: A great imitator? J Investig Med High Impact Case Rep. 2020;8:2324709620933425.
[5] Perez M, Kumar TKS, Briceno-Medina M, Alsheikh-Ali M, Sathanandam S, Knott-Craig CJ. Common pulmonary vein atresia: Report of three cases and review of the literature. Cardiol Young. 2016;26(4):629-35.
[6] Nakamura Y, Miyaji K, Miyata Y, Kitagawa A. An extremely rare variant of pulmonary venous atresia. Ann Thorac Surg. 2016;101(6):2382-84.
[7] Narayanan R, Shankar B, Paruthikunnan S. Isolated unilateral pulmonary vein atresia. Lung India. 2016;33(5):571.
[8] Dell'Amore A, Campisi A, Congiu S, Giunta D, Dolci G, Niro F. Unilateral pulmonary vein atresia without anomalous connection in adult patient with recurrent severe hemoptysis. J Vis Surg. 2018;4:111.
[9] Patil PP. Right pulmonary venous atresia: A rare cause for recurrent unilateral pneumonia. J Clin Diagn Res. 2017;11(9):SD01-SD02.
[10] Kawasaki H, Oshiro Y, Taira N, Furugen T, Ichi T, Yohena T, et al. Partial anomalous pulmonary venous connection coexisting with lung cancer: A case report and review of relevant cases from the literature. Ann Thorac Cardiovasc Surg. 2017;23(1):31-35.
[11] Türkvatan A, Tola HT, Kutlutürk N, Güzeltaş A, Ergül Y. Low-dose computed tomographic imaging of partial anomalous pulmonary venous connection in children. World J Pediatr Congenit Heart Surg. 2017;8(5):590-96.
[12] Pizzini A, Sonnweber T, Frank R, Theurl M, Weiss G, Tancevski I, et al. Clinical implications of partial anomalous pulmonary venous connection: A rare cause of severe pulmonary arterial hypertension. Pulm Circ. 2020;10(1):2045894019885352.
[13] Wang Y, Jiang S, Yang J, Sun R. Unilateral pulmonary vein atresia presenting with recurrent hydrothorax in an adult: A case report. Respir Med Case Rep. 2022;39:101711.
[14] Verma AK, Sethi S, Kohli N. Partial anomalous pulmonary venous connection: State-of-the-art review with assessment using a multidetector computed tomography angiography. Pol J Radiol. 2022;87:e549-e556.
[15] Fujii T, Machida K, Daisuke S, Yasuhiro N, Takano T. A rare case of partial anomalous pulmonary venous connection. Cureus. 2023;15(12):e50913.

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