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Persistent Levoatriocardinal Vein in Association with Complete Duplication of SVC and IVC

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Abstract

Persistent levoatriocardinal vein (LACV) is a rare pulmonary venous anomaly characterized by persistence of embryonic pulmonary to systemic communication. It is commonly associated with obstructive lesions of the left heart, such as hypoplastic left heart syndrome and mitral atresia. Complete duplication of the superior vena cava (SVC) and inferior vena cava (IVC) is rare, and occurs due to aberrations in the multistep development of the cardinal venous system. Coexistence of double SVC and IVC has been reported in the literature; however, their occurrence with persistent levoatriocardinal vein is hitherto unreported. The current case describes a rare coexistence of these complex pulmonary and systemic venous anomalies: persistent levoatriocardinal vein in a child with double SVC and IVC. The embryological development and possible aberrations leading to these anomalies are also detailed.

Keywords

- congenital cardiac anomaly
- CT angiography
- IVC duplication
- levoatriocardinal vein
- SVC duplication

Introduction

Multiple communications are present in the embryo, between the pulmonary and cardinal venous systems. These normally regress, resulting in independent development of both these systems.[1] Persistent levoatriocardinal vein (LACV) is a rare pulmonary venous anomaly characterized by the persistence of one such communication. It is usually associated with congenital cardiac anomalies such as hypoplastic left heart syndrome. Complete duplication of the suprarenal segment of the inferior vena cava (IVC) is extremely rare, with a few reports in the literature. Duplication of the superior vena cava (SVC) is characterized by persistence of the left SVC. It is a common variation, and is usually associated with congenital heart diseases. The coexistence of the LACV with anomalies of the IVC has not been reported, to the best of our knowledge. We present the case of a 5-year-old child with mitral atresia, with incidentally detected persistent LACV, complete duplication of the SVC and the IVC.

Case Presentation

A 5-year-old child with cyanosis was diagnosed with mitral atresia and single ventricle physiology on transthoracic echocardiography. Anomalous pulmonary venous return was suspected on echocardiography, and the child underwent computed tomography angiography (CTA) for an evaluation of the cardiac and vascular anatomy. The scan was acquired on a dual source 2 × 192 slice CT scanner (Somatom Force, Siemens Medical Solutions, Forchheim, Germany). Intravenous nonionic iodinated contrast (iodine concentration 350 mgI/mL) was administered through the right

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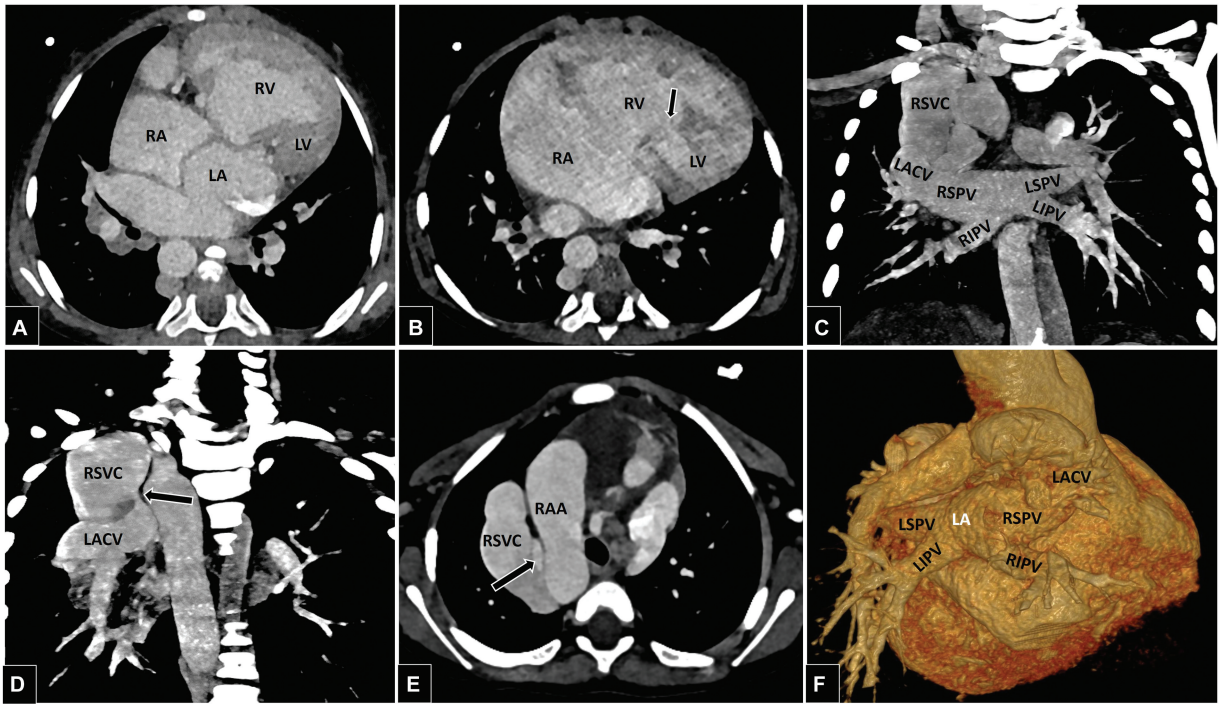

Fig. 1 (**A**) Axial computed tomography (CT) image showing atretic left ventricle with mitral atresia. The interatrial septum was intact. (**B**) The right atrium and ventricle were dilated and there was a ventricular septal defect (*arrow*). (**C**) Coronal CT image showing the drainage of all four pulmonary veins into the left atrium. The right superior pulmonary vein was also draining into the levoatriocardinal vein (LACV). (**D, E**) The LACV coursed between the right SVC and right-sided aortic arch, and showed stenosis at its opening (*arrows*). (**F**) Virtual reconstructed three-dimensional image showing the drainage of all four pulmonary veins into the left atrium and the additional drainage of the right superior pulmonary vein into the LACV. LA, left atrium; LIPV, left inferior pulmonary vein; LSPV, left superior pulmonary vein; LV, left ventricle; RA, right atrium; RIPV, right inferior pulmonary vein; RSPV, right superior pulmonary vein; RSVC, right superior vena cava; RV, right ventricle.

antecubital vein (1.5 mL/kg at the rate of 1.5 mL/s, followed by 10 mL saline chase). Electrocardiogram-gated CTA of the thorax was done using the bolus-tracking technique (region of interest [ROI] in the ascending aorta), followed by a nongated scan of the thorax and upper abdomen. The reformatted images were viewed using vendor-provided software. On CTA, there was situs solitus with levocardia. The mitral valve was atretic, with a hypoplastic left ventricle. A large ventricular septal defect was present, with dilated right-sided cardiac chambers and double-outlet right ventricle, confirmatory of single ventricle physiology. The interatrial septum was intact. The right lower, left upper, and lower pulmonary veins normally drained into the left atrium. The right upper pulmonary vein showed anomalous dual drainage into the left atrium and into the right SVC through a dilated LACV. The LACV was compressed between the right SVC and right aortic arch and showed significant stenosis at its opening into the right SVC (►**Fig. 1** and ►**Supplementary Fig. S1** [available in the online version]). Two SVCs were seen with no intercommunicating vein. The right-sided SVC was draining into the right atrium, whereas the left-sided SVC was draining into the right atrium through the coronary sinus. Pulmonary arteries were confluent and good sized. The ductus arteriosus was patent, between the left brachiocephalic trunk and the left pulmonary artery. The aortic arch was right sided with a mirror-image branching pattern. Azygous and hemiazygos veins were dilated, and two IVCs were present on either side of the aorta. The right renal vein was draining into the smaller right IVC, which showed azygous continuation and drained into the right SVC. The left renal vein drained into the left-sided IVC, which showed hemiazygos continuation, and drained into the left SVC. Communications were seen between the azygous and hemiazygos veins (►**Fig. 2**). The hepatic segment of the IVC was absent, and the hepatic veins drained via the suprahepatic IVC into the right atrium. The patient was cyanosed with pulmonary venous hypertension because of the significant stenosis of the LACV–right SVC junction, leading to reactive pulmonary arterial hypertension. Considering the double-outlet right ventricle and future single ventricle palliation, it was imperative that the pulmonary pressures be brought down before embarking on such a route. A percutaneous balloon dilatation/stenting of the stenotic confluence of the LACV and the right SVC was performed to achieve the objective before a bidirectional Glenn procedure could be contemplated.

Discussion

Anomalous pulmonary venous return is generally classified as partial or complete, depending on the number of anomalous pulmonary veins.[2] The LACV is a distinct entity, characterized by persistent embryonic pulmonary to systemic venous communication. In the embryonic period, multiple connections are present between the pulmonary and cardinal venous systems; the LACV is one such connection.

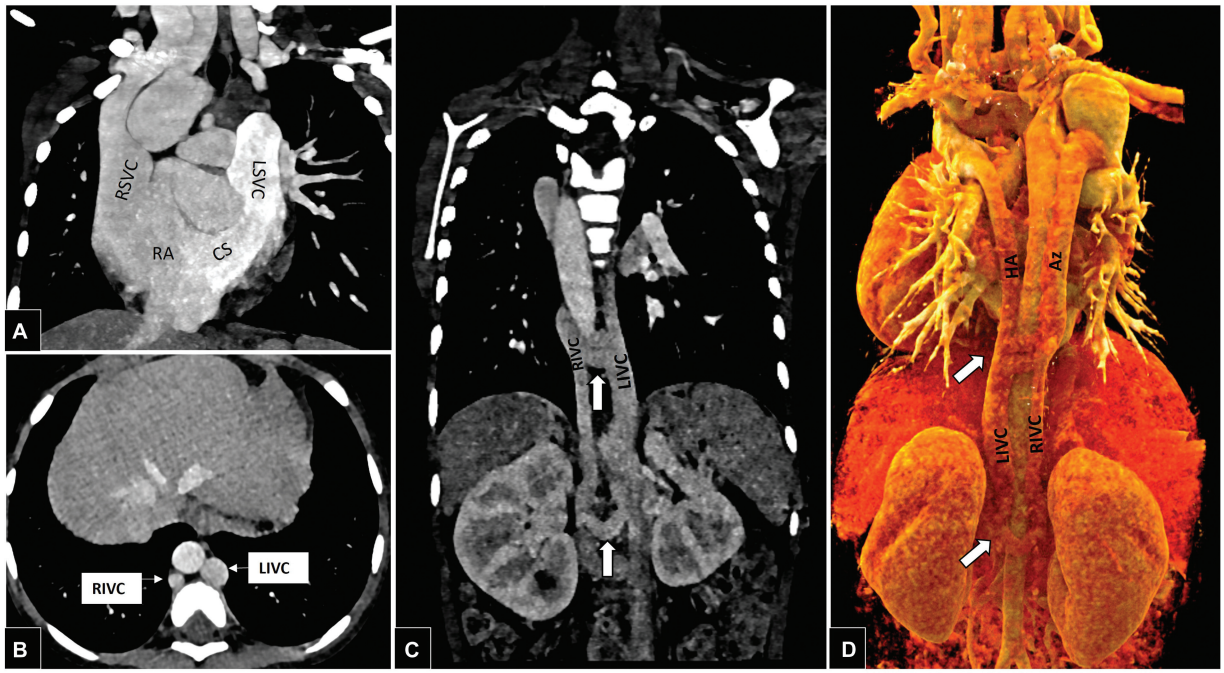

Fig. 2 (**A**) Coronal computed tomography (CT) image showing the drainage of the left superior vena cava (LSVC) through the coronary sinus (CS) into the right atrium (RA). The right superior vena cava (RSVC) was draining normally into the RA. (**B**) Axial image just above the diaphragm shows complete duplication of the IVC with the smaller right-sided IVC and larger left-sided IVC coursing along either side of the aorta. (**C**) Coronal maximum intensity projection and (**D**) virtual reconstructed image showing the supradiaphragmatic continuation of RSVC and LSVC as the azygous (Az) and hemiazygos (HA) veins, respectively, with *arrows* depicting the communications between them. LIVC, left inferior vena cava; RIVC, right inferior vena cava.

However, as the pulmonary venous connections with the left atrium develop, the LACV normally regresses. The development of the brachiocephalic vein and disappearance of the left SVC occur around the same period as the regression of the LACV. Hence, persistent LACV is classically associated with the development complex of small/absent brachiocephalic vein and persistent left SVC, as seen in the current case.[3–5] Persistent LACV is seen in association with obstructive pathologies of the left heart, such as hypoplastic left heart syndrome, coarctation of aorta, and cor triatriatum.[6,7] In such conditions, persistent LACV acts as an alternate pathway for drainage of some of the pulmonary venous blood and decompresses the left atrium, especially when the interatrial septum is intact. Usually, the LACV drains the left atrium or one of the upper lobe pulmonary veins into a systemic vessel, such as the innominate vein, jugular vein, or SVC. A study on the echocardiographic appearances of the LACV showed comparable involvement of the right and left upper lobe pulmonary veins.[8] Stenosis of the LACV is extremely rare and has been reported previously in a patient with mitral stenosis.[9]

Development of the IVC and azygous venous system is a multistep process comprising anastomosis formation and sequential regression of various components of the cardinal venous system. The hepatic veins and hepatic segment of the IVC are formed independently by the vitelline veins. The prerenal segment of the IVC is derived from the right supracardinal vein, while the renal segment is derived from the supra-subcardinal anastomosis. The azygous venous system is derived from the supracardinal veins.[10] Congenital anomalies of the IVC are rare, and include the left IVC, azygous continuation, complete duplication, circumaortic left renal vein, etc.[11] IVC interruption with azygos continuation occurs due to failure of formation of the right subcardinal–hepatic anastomosis and atrophy of the subcardinal vein. In such a situation, the suprarenal segment of the IVC is absent; as a result, as seen in this case, the hepatic veins drain into the right atrium, whereas the infrarenal IVC continues as the azygous vein. However, complete duplication of the IVC with azygous and hemiazygos continuation is extremely rare and the literature review revealed three such case reports. Among them, one case showed a supradiaphragmatic union of the azygous and hemiazygos veins and drainage into the SVC.[12] Another case showed normal course of the right IVC with drainage into the right atrium, while the left IVC showed hemiazygos continuation with SVC drainage.[13] None of these two cases had associated SVC anomalies. The third case report showed complete SVC and IVC duplication; however, the hemiazygos vein drained into the azygous vein at the D8 level.[14]

While reporting CTA of congenital cardiac anomalies, it is important to follow a step-wise approach to ensure that all findings are identified. One such checklist for CTA, which is widely followed, is essentially an extrapolation of the segmental approach proposed by Van Praagh.[15,16] It comprises the identification of the visceroatrial situs, bronchial situs, cardiac looping, cardiac position, systemic venous drainage (SVC, IVC normally draining into the right atrium), pulmonary venous drainage (normally draining into the left

atrium), atrioventricular and ventriculoarterial concordance (right atrium – right ventricle – pulmonary trunk and left atrium–left ventricle–aorta) or discordance, cardiac chamber size, septal defects, sizes of the aorta and pulmonary trunk, and any associated stenosis or atresia. Cardiac magnetic resonance imaging (MRI) can also help in the diagnosis of complex anomalies such as the LACV, although it has no superiority over CTA in its identification per se.[17] However, using phase-contrast sequences helps in establishing the direction of flow across the shunt and the shunt fraction (Qp:Qs). In addition, cardiac function can be calculated using MRI.

Conclusion

Coexistence of complex pulmonary and systemic venous anomalies is a unique occurrence, hitherto unreported. While the SVC and IVC anomalies may be asymptomatic and incidentally detected, they may have surgical implications. Persistent LACV may result in bidirectional pulmonary systemic flow, and can cause paradoxical embolism.[18] The LACV is usually surgically managed by simple ligation with/without extracardiac rerouting, depending on the venous pressures.[3,19] On cross-sectional imaging, it is essential to differentiate it from other common mimics including persistent left SVC, vertical vein, dilated superior intercostal vein, etc.[6] CTA helps in accurate delineation of the systemic and pulmonary venous anatomy, which is critical for surgical planning.

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Conflict of Interest
None declared.

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