Absent right superior vena cava with persistent left superior vena cava: two case reports

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Persistent left and absent right superior vena cava is a rare congenital anomaly, which is usually asymptomatic. Persistent left superior vena cava (PLSVC) is generally coexistent with right superior vena cava (RSVC), but rarely associated with absent RSVC. Herein, we report two children referred to our department because of ventricular septal defect. We determined PLSVC with absent RSVC during the angiography.

Key words: persistent left superior vena cava, absent right superior vena cava.

Persistent left superior vena cava (PLSVC) is generally coexistent with right superior vena cava (RSVC), but rarely associated with absent RSVC. PLSVC is usually asymptomatic and discovered incidentally. Precise anatomical knowledge of the great vessels of the neck is essential for safe anesthesia, pacemaker implantation and cardiac surgery. Therefore, PLSVC must be recognized to avoid potential complications such as those associated with placement of central catheters and cardiac surgery.

Case Reports

Case 1

A four-year-old female patient was referred to our hospital for ventricular septal defect (VSD). Clinical examination revealed a grade 2/6 systolic murmur in the 3rd intercostal space. Transthoracic two-dimensional echocardiography showed visceroatrial situs solitus, muscular outlet VSD with two-sided shunt and mild pulmonary hypertension. Cardiac catheterization (Fig. 1a, 1b) revealed the absence of the RSVC, and PLSVC was documented with a right upper limb venogram during the cardiac angiography. We observed a large PLSVC draining into the coronary sinus. The LSVC joined by the right brachiocephalic vein was large and prominent. During the surgery, the preoperative diagnosis was confirmed, and thus the operation was completed without any complication.

Case 2

A five-year-old male child was admitted for VSD. Cardiovascular examination revealed a grade 3/6 systolic murmur best heard in the left third intercostal space. Transthoracic two-dimensional echocardiography showed visceroatrial situs solitus, muscular outlet VSD, and slight subvalvular pulmonary stenosis. Angiography revealed an absence of RSVC with PLSVC (Fig. 2a, 2b).

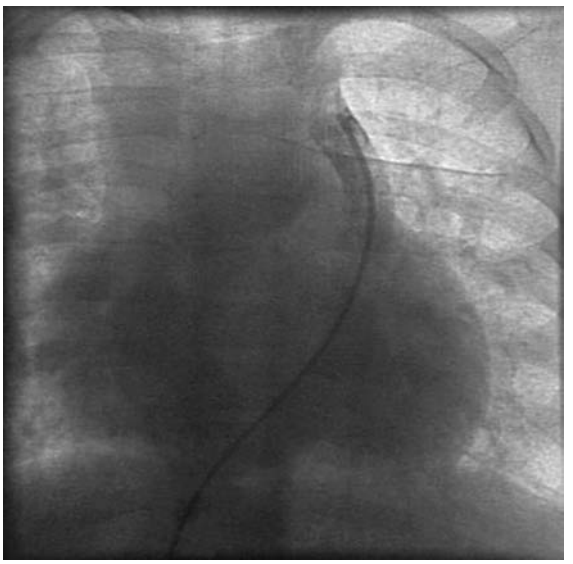

Figure 1a. Left superior vena cava (LSVC) draining into the coronary sinus.

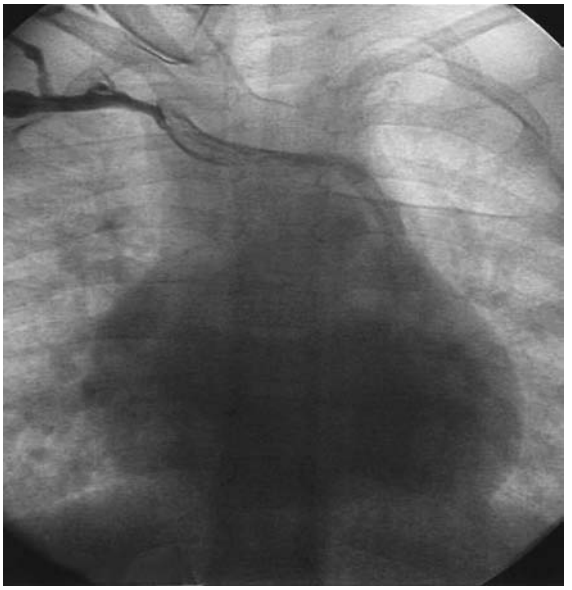

Figure 1b. Right upper limb venogram indicates absence of right superior vena cava and persistent left superior vena cava (PLSVC).

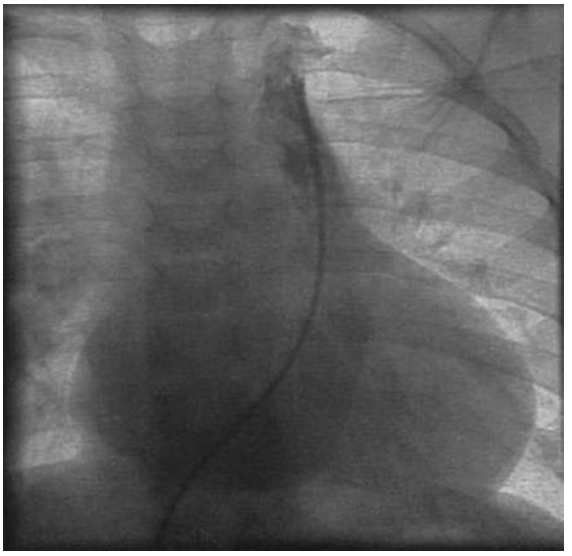

Figure 2a. Left superior vena cava (LSVC) draining into the coronary sinus.

Discussion

Persistence of the left superior vena cava (PLSVC) has an incidence of 0.3-0.5% in the general population. In the presence of another congenital heart disease, the incidence of PLSVC increases to 3-10%[1-3]. The absence of RSVC is a rare anomaly, with an incidence of 0.07-0.13%[4,5]. When LSVC is present, RSVC is usually functional[6]. The combination of PLSVC

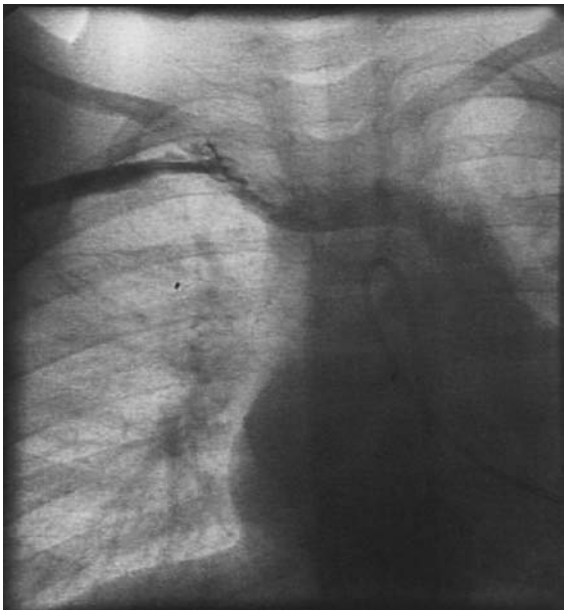

Figure 2b. Right upper limb venogram indicates absence of right superior vena cava and persistent left superior vena cava (PLSVC).

in the absence of RSVC is unusual and rare[4,6].

During normal cardiac development, the left-sided anterior cardinal vein atrophies, leaving only the coronary sinus and a remnant known as the ligament of Marshall. Failure of closure of the left anterior cardinal vein results in PLSVC. In the majority of cases, the PLSVC drains into the right atrium via an enlarged coronary sinus. On rare occasions, the PLSVC enters directly into the left atrium, resulting in partial anomalous systemic venous return [7].

The anomaly is generally asymptomatic and found incidentally and may be even higher in people without heart disease[4,5,8,9]. Bartram et al.[5] reviewed 121 patients with PLSVC and absent RSVC. The most common malformations were atrial septal defect (16%), endocardial cushion defect (11%) and Fallot's tetralogy (9%). However, the majority of these patients (n=65, 54%) did not have heart malformation, thus confirming that absent RSVC and PLSVC can often be seen unaccompanied by other malformations[5]. Absent RSVC with PLSVC is very rare in the setting of visceroatrial situs solitus, but is seen in visceral heterotaxy and associated atrial situs inversus[4,5]. Our two patients had visceroatrial situs solitus with VSD.

In absent RSVC with PLSVC, a large and stretched coronary sinus causes conduction disturbances[4]. It is also associated with disturbances of cardiac impulse formation and conduction, such as ectopic atrial focus, heart block, sinoatrial node dysfunction, and ventricular tachycardia[4,5,10].

The patients with absent RSVC with PLSVC were reported to have normal hemodynamics, because the PLSVC drains into the right atrium via an enlarged coronary sinus. Therefore, patients are mostly asymptomatic[4,9]. The anomaly is often discovered incidentally, while undergoing thorough tests for other heart conditions, intravenous pacemaker insertion, or during autopsy[5]. While catheterizing our two patients because of VSD, absent RSVC with PLSVC was detected coincidentally.

Modern imaging techniques including echocardiography, computed tomography and magnetic resonance imaging (MRI) provide precise diagnosis of this anomaly. Di Cesare and Anselmi[11] described an asymptomatic 22-year-old man with persistence of the LSVC in the absence of the RSVC using a dynamic MRI angiographic technique.

The diagnosis of absent RSVC is important in order to avoid management difficulties such as during transvenous pacemaker implantation, catheter insertion and vena cava cannulation. Moreover, it may cause difficulties and serious complications during total or partial cavopulmonary connection and cardiopulmonary bypass[5,12,13].

In conclusion, detailed and accurate procedures may identify this rare combined congenital defect, thereby preventing future complications during surgery.

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