Isolated persistent left superior vena cava with absent right superior vena cava in two cases

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Summary – Persistent left superior vena cava (LSVC) with absent right superior vena cava (RSVC) is a very rare congenital anomaly. Its isolated existence is even rarer. Persistent LSVC is usually asymptomatic and discovered incidentally. We present persistent LSVC with absent RSVC in two asymptomatic patients, namely, a 52-year-old woman and 65-year-old man. The diagnosis was confirmed by cardiac computed tomography in both cases.

CASE REPORT

Case 1– A 52-year-old woman underwent transthoracic echocardiography as part of evaluation of atypical chest pain of long history. She did not have any significant medical or surgical history. Her electrocardiogram and treadmill exercise test were normal. Echocardiography did not show any significant cardiac abnormality except for a large dilated coronary sinus of 30 mm in diameter. After saline injection into the left antecubital vein, the coronary sinus filled first then emptied into the right atrium. Then, after agitated saline injection into the right antecubital vein, there was again filling of the coronary sinus then emptying into the right atrium (Fig. 1a). Diagnosis of persistent LSVC with absent RSVC was thought. In order to demonstrate it more clearly, cardiac computed tomography was performed, which demonstrated persistent left LSVC with absent RSVC (Fig. 1b, c).

Case 2– A 65-year-old man required dialysis for chronic renal failure. As he did not have a dialysis fistula, a temporary dialysis catheter was implanted to the right internal jugular vein. A posteroanterior chest radiogram did not confirm its placement in the right position (Fig. 2a), so the catheter was pulled back and another catheter was placed into the left internal jugular vein. However, a subsequent chest radiogram again showed that it was not in the right position and it seemed to be in the LSVC rather than the left subclavian vein (Fig. 2b). After consultation with a nephrologist whether CT imaging with contrast would be necessary to confirm the diagnosis, CT imaging with contrast was performed, which demonstrated persistent left LSVC with absent RSVC (Fig. 2c).

Abbreviations:
CT Computed tomography
LSVC Left superior vena cava
RSVC Right superior vena cava

Received: December 7, 2010 Accepted: June 13, 2011
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be detrimental to the patient, CT examination was performed, which showed persistent LSVC with absent RSVC (Fig. 2c, d).

**DISCUSSION**

Persistent LSVC with absent RSVC is a very rare congenital anomaly. Persistent LSVC is usually asymptomatic and discovered incidentally.\(^3\) Electrocardiography is not specific and chest radiography may show a paramediastinal bulge below the aortic arch, presenting as a widened aortic shadow along the left upper cardiac border of the aortic arch toward the middle third of the left clavicle. Thanks to the modern imaging modalities including echocardiography, CT, and magnetic resonance imaging, diagnosis of this anomaly is clearly and easily confirmed.

Even though persistent LSVC is usually not associated with any negative hemodynamic effect, awareness of its existence is crucial because of potential problems in central venous catheterization,\(^4\) pacemaker implantation,\(^5\) or cardiopulmonary bypass, in which case a persistent LSVC prevents retrograde cardioplegia in case of absent RSVC.

Persistent LSVC may also cause rhythm disturbances such as sinus node dysfunction and atrioventricular block.\(^6\) These rhythm disorders may be related with fragmentation and stretching of the conduction tissue caused by enlargement of the coronary sinus.

Persistent LSVC may accompany other congenital cardiovascular defects such as tetralogy of Fallot and Eisenmenger’s syndrome.\(^7\) Other anomalies include coarctation of the aorta, atrial septal defect, ventricular septal defect, and endocardial cushion defect. Some of these anomalies (e.g., atrial septal defect, unroofed coronary sinus, or direct communication of the vein to the left atrium) impose great risk for paradoxical embolism, which is estimated to be as high as 50% to 70%.\(^8\) None of our cases had these abnormalities.

In conclusion, clinicians must be aware of and alert to the possibility of central venous variations such as persistent LSVC with absent RSVC and their clinical consequences in order to avoid possible complications in routine clinical practice. Diagnosis of persistent LSVC with or without RSVC can be easily made by noninvasive imaging modalities.
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Conflict-of-interest issues regarding the authorship or article: None declared

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Key words: Heart defects, congenital; tomography, X-ray computed; vena cava, superior/abnormalities.

Anahtar sözcükler: Kalp defekti, doğuştan; bilgisayarlı tomografi; vena kava süperiyor/anormallık.