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

Sağ süperiyor vena kava olmaksızın izole persistan sol süperiyor vena kava: İki olgu sunumu

Levent Korkmaz, M.D., Ali Rıza Akyüz, M.D., M. Emre Erkuş, M.D., Cevat Topal, M.D.[#]

Cardiology Departments of Ahi Evren Chest, Heart and Vascular Surgery Training and Research Hospital, and [#]Numune Training and Research Hospital, both in Trabzon

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.

Özet – Sağ süperiyor vena kavanın eşlik etmediği persistan sol süperiyor vena kava çok nadir bir doğuştan anomalidir. İzole olarak bulunması daha da nadir bir durumdur. Persistan sol süperiyor vena kava genellikle semptomsuzdur ve tesadüfen ortaya çıkarılır. Bu yazıda, sağ süperiyor vena kava olmaksızın persistan sol süperiyor vena kava saptanan semptomsuz iki hasta (52 yaşında kadın ve 65 yaşında erkek) sunuldu. Her iki olguda da tanı kardiyak bilgisayarlı tomografi ile doğrulandı.

Persistent left superior vena cava is not uncommon. It is estimated to occur in 0.3-0.5% of the general population and 3-10% of the patients have congenital cardiac abnormalities.[1] It usually coexists with right superior vena cava, but rarely RSVC may be absent.[2] We present two cases of persistent LSVC with absent RSVC.

Abbreviations:

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

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

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Correspondence: Dr. Ali Rıza Akyüz. Akçaabat Haçkalı Baba Devlet Hastanesi, Kardiyoloji Kliniği, 61300 Akçaabat, Trabzon, Turkey.
Tel: +90 462 - 228 58 03 e-mail: dralirizaakyuz@gmail.com

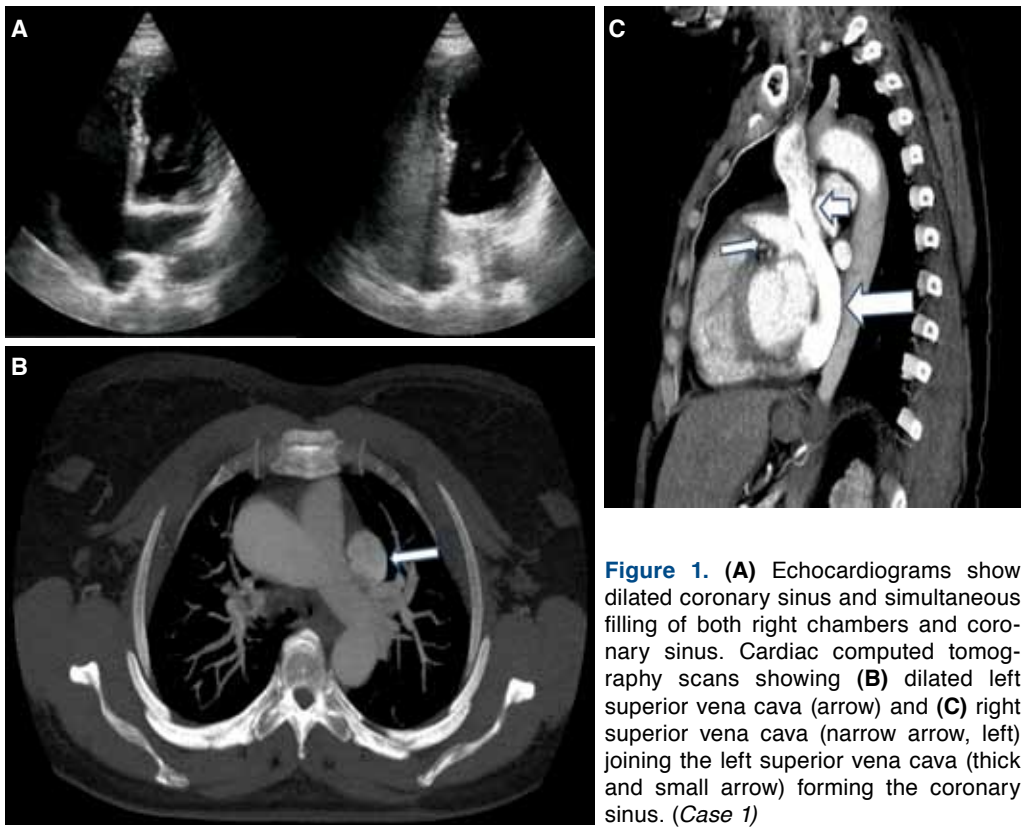

Figure 1. **(A)** Echocardiograms show dilated coronary sinus and simultaneous filling of both right chambers and coronary sinus. Cardiac computed tomography scans showing **(B)** dilated left superior vena cava (arrow) and **(C)** right superior vena cava (narrow arrow, left) joining the left superior vena cava (thick and small arrow) forming the coronary sinus. (*Case 1)*

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.

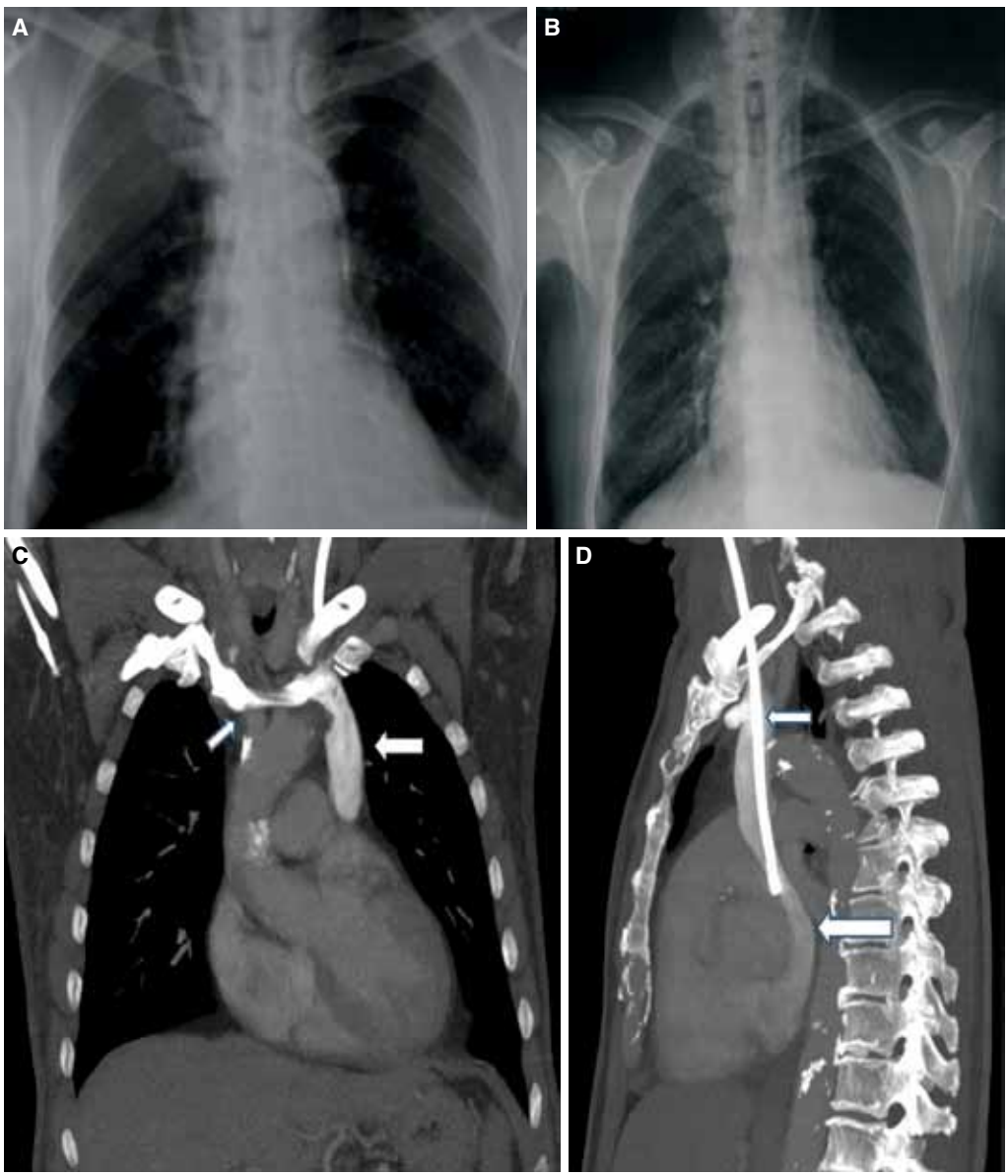

Figure 2. Chest radiographies showing **(A)** the dialysis catheter implanted via the jugular vein in inappropriate position after the first attempt and **(B)** the dialysis catheter in the left superior vena cava after the second attempt. Cardiac computed tomography scans showing **(C)** the right superior vena (narrow arrow) and dilated persistent left superior vein (thick arrow) and **(D)** the dialysis catheter implanted via the left jugular vein in the left superior vena cava, reaching the coronary sinus. (*Case 2)*

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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/anormallik.