

Two Cases with Persistent Left Superior Vena Cava with Absence of Right Superior Vena Cava

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Sağ Süperior Vena Kava Olmaksızın Persistan Sol Süperior Vena Kava Saptanan İki Olgu

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ABSTRACT

Persistent left superior vena cava (PLSVC) is the most common congenital venous anomaly of the thorax. PLSVC with absent right superior vena cava (RSVC) which is also termed as "isolated PLSVC" is a rare entity and estimated to be present 0.09-0.13% of patients. PLSVC with absent RSVC is usually asymptomatic but its presence can lead to serious complications. In this report, we describe two cases of PLSVC with absent RSVC and its clinical implications.

Keywords: contrast echocardiography, coronary sinus, echocardiography, persistent left superior vena cava

ÖZ

Persistan sol süperior vena kava (PSSVK) toraksın en sık doğumsal venöz anomalisidir. "İzole PSSVK" olarak da adlandırılan sağ süperior vena kava (SSVK) yokluğuyla birlikte olan PSSVK nadir görülen bir durumdur ve hastaların %0.09-0.13'nde bulunduğu tahmin edilmektedir. SSVK yokluğuyla birlikte olan PSSVK genellikle asemptomatiktir ancak varlığı ciddi komplikasyonlara yol açabilir. Bu yazıda, iki olguda saptadığımız SSVK yokluğuyla birlikte olan PSSVK'yi ve klinik önemini tanımladık.

Anahtar kelimeler: kontrast ekokardiyografi, koroner sinüs, ekokardiyografi, persistan sol süperior vena kava

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INTRODUCTION

Persistent left superior vena cava (PLSVC) is present roughly in 0.5-2% of population and 10% of these patients have other congenital heart abnormalities [1]. PLSVC with absent right superior vena cava (RSVC) also referred to as "isolated PLSVC" is a rare venous congenital anomaly and exists in 0.09%-0.13% of the patients with congenital heart disease [2]. Isolated PLSVC is usually asymptomatic and diagnosed incidentally but sometimes this condition can lead to serious complications during central venous access, device implantation, and cardiothoracic surgery. Isolated PLSVC is also associated with presence of other congenital anomalies and arrhythmias [3].

CASE PRESENTATION 1

A 63-year-old woman presented to our clinic with recurrent syncope episodes. Her previous medical history revealed hypertension and her medical therapy included daily doses of verapamil 240 mg plus trandolapril 4 mg and metoprolol 50 mg. Physical examination was unremarkable except heart rate of 40 bpm and blood pressure of 80/50 mmHg. The 12-lead electrocardiogram showed sinus bradycardia with a rate of 42 bpm. Transthoracic echocardiography (TTE) revealed normal left ventricular systolic function with a left ventricular ejection fraction of 60%, mild mitral and tricuspid regurgitation with normal systolic pulmonary artery pressure. The interatrial and interventricular septum were both intact.

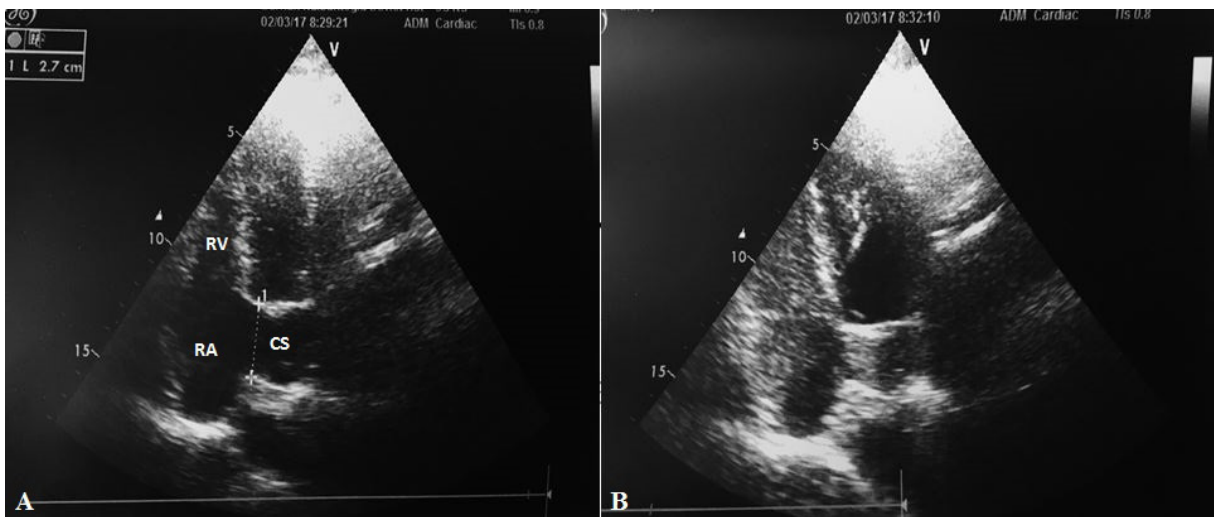


Figure 1. Transthoracic echocardiography showing dilated coronary sinus (Panel A) and coronary sinus opacification before right atrium (Panel B) (RA: right atrium, RV: right ventricle, CS: coronary sinus).

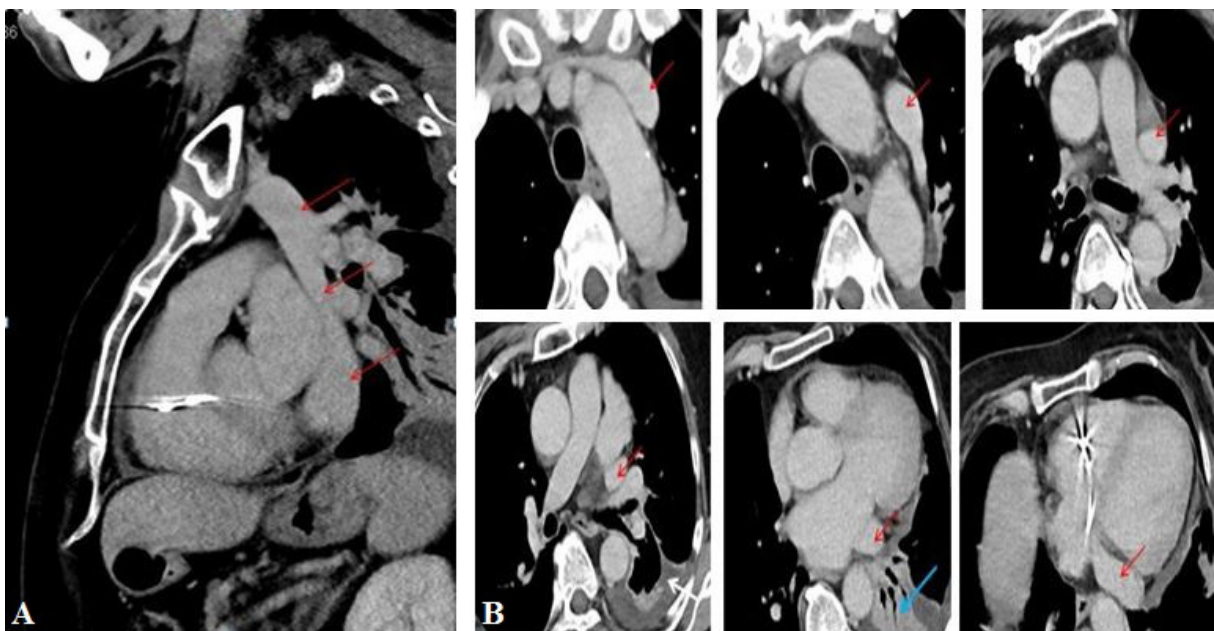


Figure 2. The sagittal oblique reformatted CT image demonstrates the drainage of persistent left superior vena cava to coronary sinus. (Panel A, arrows). Axial CT images show the absence of right SVC and the drainage of persistent left SVC to coronary sinus (red arrows). There is also left pleural effusion (white arrow) and left lower lobe atelectasis (blue arrow) (Panel B).

Coronary sinus (CS) was dilated in parasternal long axis and apical-4-chamber view. Contrast echocardiography was performed and agitated saline was infused through left antecubital vein and CS opacified just before right atrium (RA) which is consistent with persistent left superior vena cava (PLSVC). The

administration of agitated saline was repeated via the right antecubital vein. Contrast echocardiography performed via right antecubital vein demonstrated opacification of CS just before RA which is compatible with the absence of right superior vena cava (RSVC) (Figure 1, Panel A-B). Computed tomog-

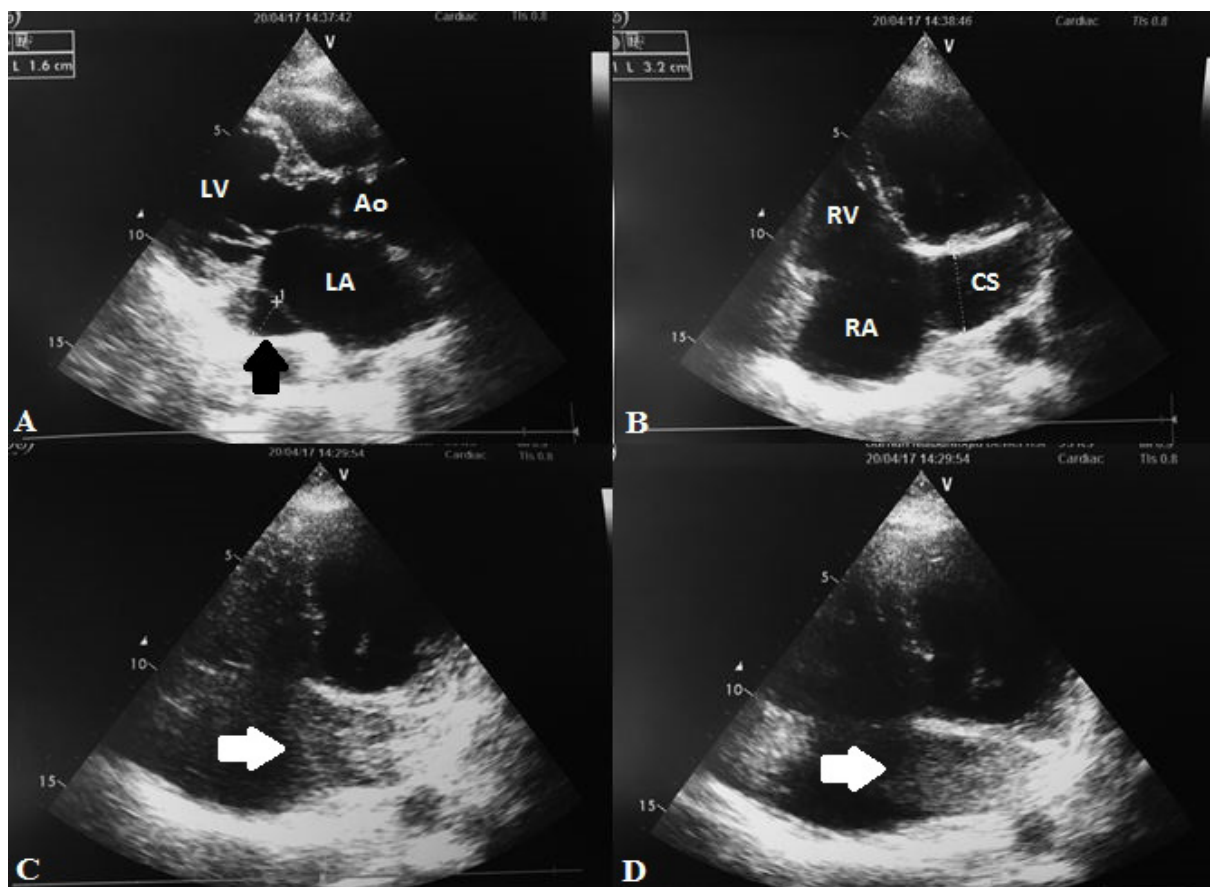


Figure 3. Transthoracic echocardiography parasternal long axis view (Panel A, black arrow) and apical view (Panel B) showing dilated coronary sinus and coronary sinus opacification before right atrium after agitated saline injection from both right and left arm (Panel C-D, white arrows) (LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle, CS: coronary sinus, Ao: aorta).

raphy confirmed the absence of RSVC and showed the drainage of PLSVC into CS (Figure 2, red arrows). During follow-up period, treatment with verapamil 240 mg plus trandolapril 4 mg was stopped and patient's heart rate and blood pressure values normalized, so she was discharged daily treatment with ramipril 2.5 mg without any complication.

CASE PRESENTATION 2

A 61-year-old woman presented to our clinic with symptoms of shortness of breath and palpitation. Her previous medical history revealed hypertension, hyperlipidemia and her daily medical treatment included perindopril 5 mg, amlodipin 5 mg, bisoprolol 5 mg and rosuvastatin 20 mg. Her blood pressure was 155/90

mmHg and heart rate was 110 bpm. The 12-lead electrocardiogram showed sinus tachycardia with a rate of 114 bpm without any ischemic changes. TTE revealed concentric left ventricular hypertrophy and normal left ventricular systolic function with a left ventricular ejection fraction of 65% and dilated CS (Figure 3, Panel A-B). Contrast echocardiography was performed and agitated saline injected from both left and right antecubital veins demonstrated opacification of CS before right atrium (Figure 3, Panel C-D). Computed tomography and three-dimensional reconstruction images showed absence of right SVC and the venous drainage via left sided SVC and azygous venous system (Figure 4, arrows). Patient was discharged with daily treatment of perindopril 10 mg, amlodipin 10 mg, rosuvastatin 20 mg and furosemide 40 mg.

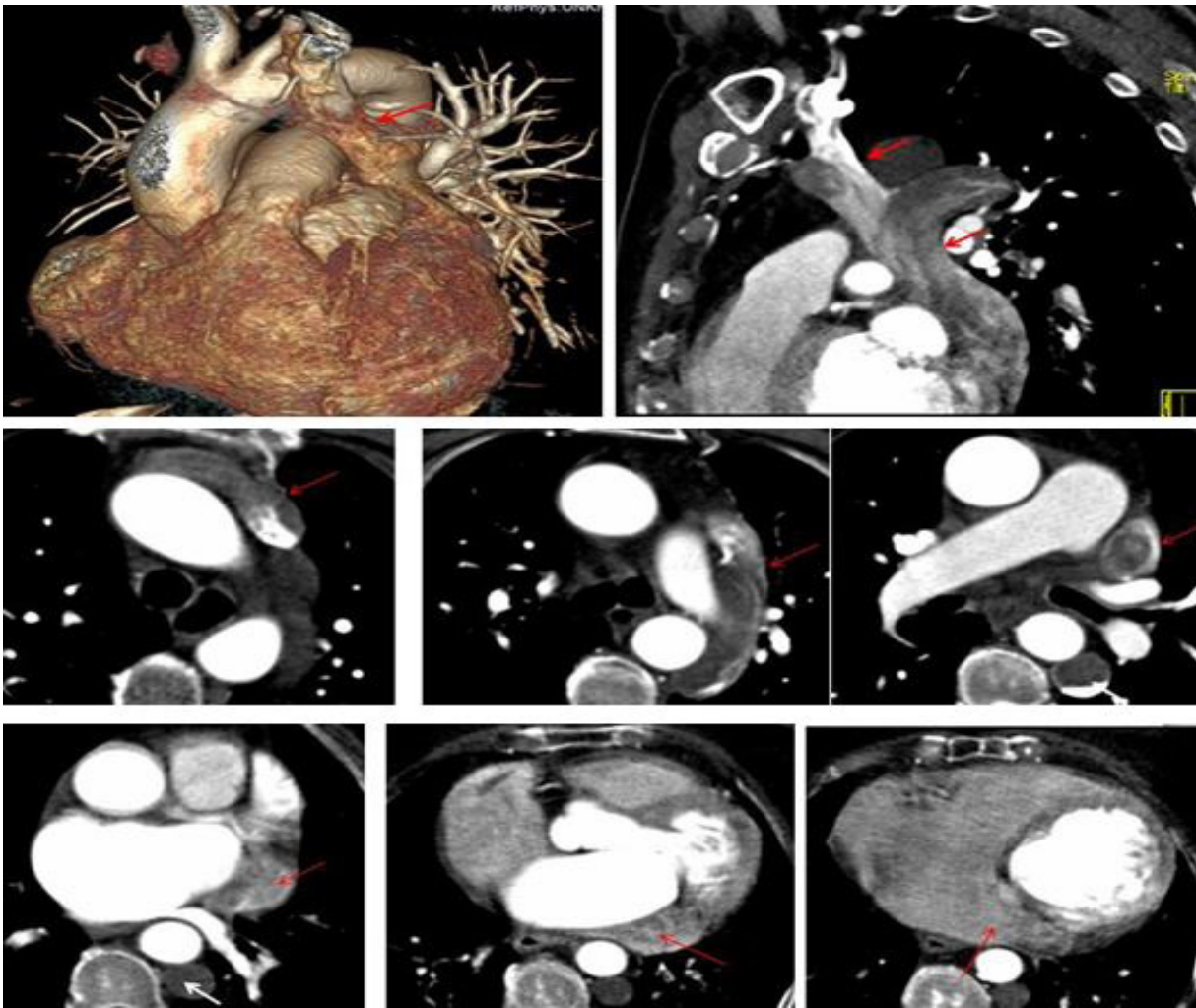


Figure 4. The three-dimensional volume rendered CT image and sagittal oblique reformatted CT image shows the persistent left superior vena cava and axial contrast enhanced CT image shows absence of right superior vena cava (SVC) and the venous drainage via left sided SVC (red arrow) and azygous venous system.

DISCUSSION

Isolated PLSVC is a rare congenital venous anomaly and its diagnosis is generally as an incidental finding during central venous catheterization, device implantation, cardiovascular imaging or surgery [4]. PLSVC with absent RSVC is usually asymptomatic but some technical and procedural problems may be seen in accessing the right heart during catheterization or pacemaker (PM) / implantable cardiac defibrillator (ICD) implantation via left subclavian approach in the setting of isolated PLSVC and rarely it can lead to serious complications such as shock, cardiac arrest or angina [5-7]. In a large proportion of cases PLSVC

drains into the right atrium via a dilated CS. Rarely, PLSVC drains into the left atrium [8]. PLSVC should be suspected in patients with a dilated CS on TTE. The possible causes of dilated CS includes PLSVC, total anomalous pulmonary venous return to CS, coronary arteriovenous fistula with drainage into the CS, connections between hepatic veins and CS, severe right heart failure with pulmonary hypertension [9]. The diagnosis of PLSVC can be made by contrast echocardiography with administration of agitated saline via both left and right antecubital veins. In normal circumstances, agitated saline injected via either antecubital vein should first opacify the right atrium and then right ventricle. In the presence of PLSVC with

RSVC, agitated saline injection via left antecubital vein directly drains into the CS and bubbles can be seen first in the CS before opacification of the right heart and agitated saline injection via right antecubital vein normally opacifies the right atrium and then right ventricle. In the setting of isolated PLSVC, bilateral agitated saline administration through both left and the right antecubital veins directly drains into CS and opacifies CS before right atrium as in our cases^[5].

To conclude, isolated PLSVC in adult patients is a rare congenital vascular anomaly and it may cause complications during right heart catheterization or PM / ICD implantation and lead to cardiac arrhythmias such as atrial and ventricular fibrillation^[7,10]. Clinicians and echocardiographers must be aware of this rare entity and should be suspected in patients with a dilated CS on TTE.

Conflict of interest: None.

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