Case Report

Persistent Left Superior Vena Cava with Absent Right Superior Vena Cava: A Case Report and Literature Review

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A 71-year-old man presented with drowsiness three hours before admission. Stroke or hepatic encephalopathy was suspected but computed tomography (CT) scan of the brain revealed no abnormality. Cardiac auscultation discovered pansystolic murmurs at apex radiated to axilla. Plain chest roentgenogram showed markedly cardiac enlargement and electrocardiogram (ECG) revealed atrial fibrillation. Transthoracic echocardiography was performed to detect the etiology of cardiac enlargement, but it discovered a dilated coronary sinus instead. Finally, a very rare case of persistent left superior vena cava (PLSVC) with absence of right superior vena cava (RSVC) anomaly that is also known as isolated PVSVC was incidentally demonstrated by transesophageal echocardiography. The patient also had severe mitral and tricuspid regurgitations. Diabetes mellitus, dyslipidemia, and suspected cirrhosis of liver were also found. He was treated for hepatic encephalopathy, which is not related to PLSVC and he recovered uneventfully. PLSVC should be expected if any intracardiac venous procedures are going to be performed such as central venous catheterization, device implantation, or coronary artery bypass surgery.

Keywords: Persistent left superior vena cava, Congenital venous anomaly, Dilated coronary sinus, Echocardiography

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Persistent left superior vena cava (PLSVC) is a congenital venous anomaly and represents a remnant of the left anterior cardinal vein that typically obliterates during fetal development. The PLSVC drains cephalic venous return into the well-developed coronary sinus, which is subsequently enlarged. Consequently, there is no left-sided connection from the left axillary or subclavian vein to the right atrium (RA). This anomaly occurs in approximately 0.5% of the population with or without the presence of normal right superior cava and up to 10% those with established congenital heart disease[1]. A retrospective study at the Department of Pathology, Chulalongkorn University among the 362 perinatal and pediatric autopsies revealed 19 (5.2%) cases of PLSVC[2]. Seventeen cases of the PLSVC (89%) were associated with congenital heart diseases and eight cases were associated with specific syndromes, including heterotaxy syndrome, trisomy 18, trisomy 13, and Jacobsen syndrome. In addition, eight PLSVC cases were associated with hypoplastic left heart syndrome (HLHS). However, an isolated PLSVC is of no clinical significance but may pose some technical difficulty for placement of a pacemaker, implantable cardioverter defibrillator (ICD) lead, or a coronary artery bypass grafting, which may be needed in the future. It also can lead to intracardiac conduction abnormalities.

The PLSVC can be fortuitously diagnosed on chest radiography or echocardiography though this fetal anomaly that can be detected before birth. This condition is still rarely reported in Thai adult as an incidental finding. The authors wish to add another case report of isolated PLSVC in adult and perform the literature review.

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Case Report

A 71-year-old man previously diagnosed of cirrhosis of liver, was admitted to evaluate the etiology of drowsiness and stupor three hours

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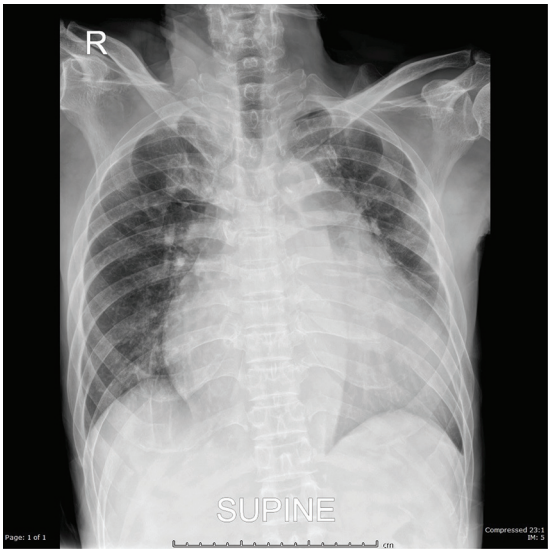

Figure 1. Plain chest X-ray revealed marked cardiomegaly; suspected mild pulmonary congestion. No blunting of costophrenic angle. Bony thorax is intact.

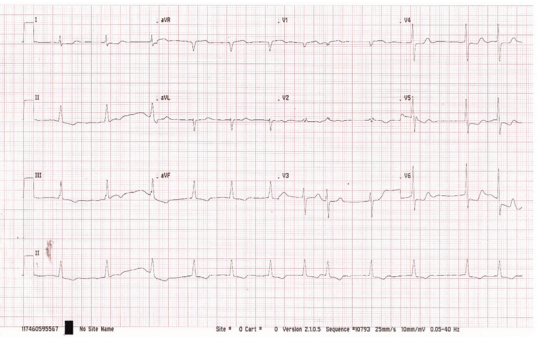

Figure 2. Electrocardiogram showed atrial fibrillation with ST-depression and T wave abnormality. Inferior wall ischemia should be considered.

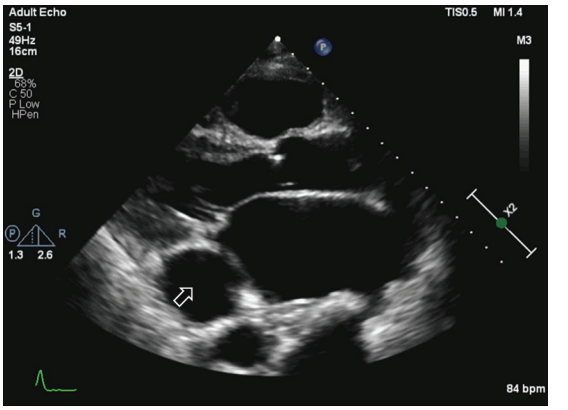

Figure 3. Transthoracic echocardiogram (TTE) revealed a markedly dilated coronary sinus (arrow).

before admission. One day earlier, his speech and movement were slow, but he was still conscious. He lived alone in his house and was transferred to the hospital because of drowsiness and stupor. Upon admission, his temperature was 36.0°C, blood pressure was 144/89 mmHg, pulse rate was 75 beats/minute, and respiratory rate was 19 breaths per minute. The Glasgow Coma Scale was $E_4V_1M_3$. General appearance revealed he was anemic and icteric. He was stuporous and did not respond to verbal command. Though he had no hemiplegia, ischemic stroke or hepatic encephalopathy was initially suspected but computed tomography (CT) scan of the brain revealed no intracerebral abnormality. Cardiac auscultation discovered pansystolic murmurs at apex radiated to axilla, plain chest roentgenogram showed markedly cardiac enlargement (Figure 1) and electrocardiogram revealed atrial fibrillation (Figure 2). Laboratory data showed hematocrit of 22%, thrombopenia, HbA_1C 9.2%, transaminase 51 U/L, alkaline phosphatase 159 U/L, total bilirubin of 1.69 mg/dL, albumin 2.6 g/dL, triglyceride 227 mg/dL, total cholesterol 227 mg/dL, and HDL-cholesterol 35 mg/dL. Ultrasonogram of upper abdomen revealed suspected cirrhosis of liver and dilatation of inferior vena cava and hepatic veins suspicious of heart failure.

Transthoracic echocardiography was performed to find the etiology of cardiac enlargement, but it revealed a dilated coronary sinus (CS) instead (Figure 3). The left ventricular ejection fraction was 68% and the estimated peak systolic pulmonary artery pressure was high due to left-side heart disease. Severe mitral regurgitation and severe tricuspid regurgitation were also detected. The inter-atrial and inter-ventricular septum were both intact. All four pulmonary veins drained into the left atrium (LA). The thickness of the pericardium was normal with no effusion. An agitated saline injection was made from the left antecubital vein. The bubble appeared first in the CS and then in the RA (Figure 4). The injection was repeated from the right antecubital vein, which also demonstrated first appearance of the contrast in the CS and afterwards in the RA (Figure 5). Computerized tomography of thoracic venous drainage showed a bridging vein draining the right jugular and right subclavian veins; it joined the left brachiocephalic vein and formed the PLSVC, which descended at the left side of the mediastinum leftward of the pulmonary artery and LA before draining into the RA via a dilated CS (Figure 6-8). The right superior vena cava (RSVC)

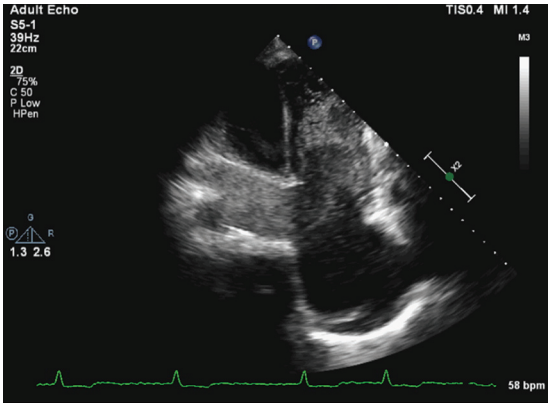

Figure 4. TTE: contrast given from the left arm opacified the coronary sinus (CS) before the right atrium (RA).

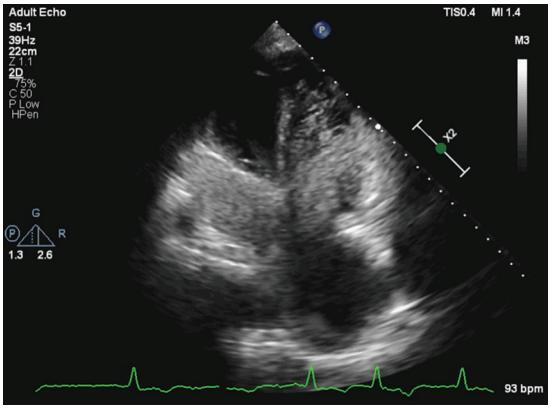

Figure 5. TTE: repeated contrast given from the left arm opacified the coronary sinus (CS) before right atrium (RA).

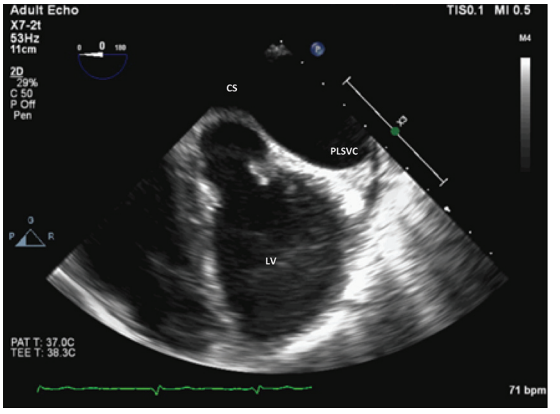

Figure 6. TEE at mid-esophagus view demonstrated the presence of dilated coronary sinus (CS) which was connected to persistent left superior vena cava (PLSVC).

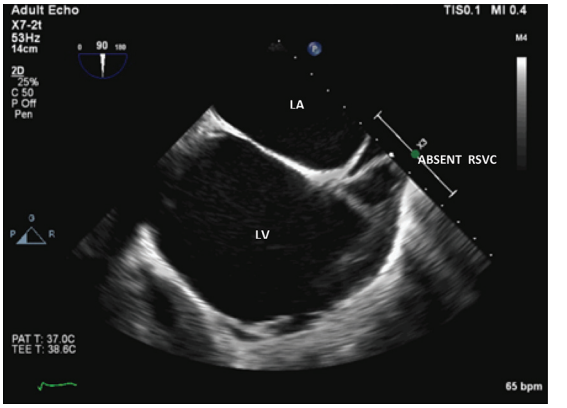

Figure 7. TEE in bicaval view demonstrated the absence of right superior vena cava (RSVC).

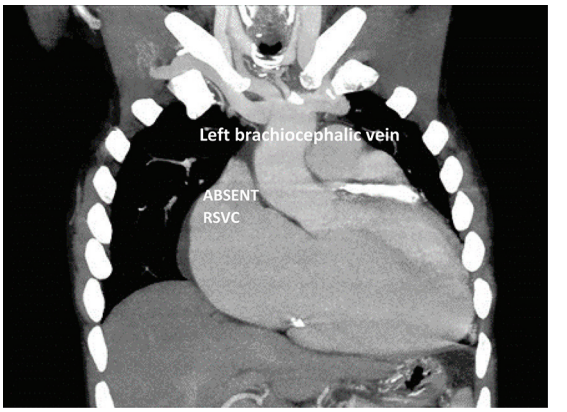

Figure 8. CT multiplanar reformatted image demonstrated the absence of right superior vena cava and a bridging vein (BV) drained between the right jugular and subclavian veins, which then joined the left brachiocephalic vein to form persistent left superior vena cava (PLSVC).

was absent and the PLSVC drained all venous blood from the head, neck, and upper extremities into the CS and then RA (Figure 9, 10). He was treated for hepatic encephalopathy and his consciousness fully recovered after seven days of hospitalization.

Discussion

The authors report a rare case of PLSVC with absent RSVC in an elderly previously diagnosed of cirrhosis of liver. The PLSVC is a very rare venous malformation either isolated or associated with RSVC. This venous malformation itself causes no hemodynamic disturbance and is usually diagnosed incidentally as happened in the present case who has been previously diagnosed with cirrhosis of liver, was

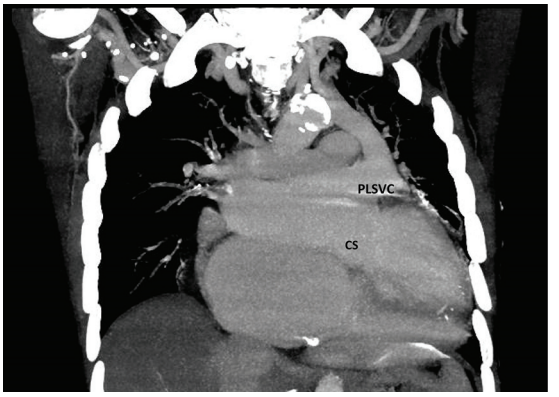

Figure 9. CT multiplanar reformatted image revealed the persistent left superior vena cava (PLSVC) draining into a dilated CS.

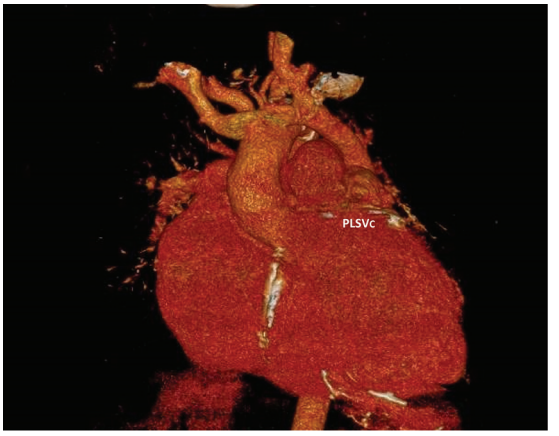

Figure 10. Axial 3D reconstruction (frontal plane) from multislice computed tomography displayed the isolated persistent left superior vena cava (PLSVC).

anemic at the time of admission, and had severe mitral and tricuspid regurgitations. All these conditions were not associated with PLSVC and the patient had no history of any hemodynamic disturbance or cardiac failure before. On admission, he had certain degree of congestive heart failure as shown by pulmonary congestion on chest roentgenogram and dilated inferior vena cava by ultrasonogram of upper abdomen. The cause of heart failure was most likely due to severe mitral and tricuspid regurgitations in addition to anemia.

As usual, transthoracic echocardiography serves as a simple and excellent modality to diagnose PLSVC. The characteristic finding is a dilated CS on parasternal long-axis view. The normal diameter of the CS is smaller than 1 cm. Isolated PLSVC severely increases venous flow and causes a truly giant CS[3,4]. The presence of isolated PLSVC can be confirmed by performing a bilateral "bubble study" with injection of agitated saline from both the left and the right antecubital veins[5].

Transesophageal echocardiography is more sensitive than transthoracic studies for the diagnosis of PLSVC and associated cardiac anomalies, including atrial septal defect (ASD), anomalous connection of pulmonary veins, and PLSVC draining into the LA[6]. PLSVC and absence of RSVC can be well visualized by this method. In mid-esophageal views, the PLSVC is seen near to the left atrial appendage and left upper pulmonary vein. In the bicaval view, the absence of RSVC was demonstrated. Other techniques such as venous angiography[7], CT[7], and magnetic resonance imaging (MRI)[8] can directly visualize the venous anatomy and hence can also confirm the diagnosis.

In a literature review of published articles in the English literature between 1995 and 2018. The authors found 13 patients whose ages were older than 18 years, were diagnosed with persistent left superior vena cava and absent right superior vena cava. The characteristics of these patients are shown in the Table 1. There were eight males and five females. Seven cases were incidentally found when a pacemaker implantation was being performed. PLSVC is typically asymptomatic when it is not accompanied with other heart defects and its presence is usually discovered during central venous catheterization or pacemaker implantation.

However, a higher incidence of arrhythmias and conduction system abnormalities has been described in patients with PLSVC. There are two proposed mechanisms for this association, a dilated CS stretches the atrioventricular nodal tissue, which prepares a substrate for re-entrant tachycardia or the early conduction tissue has close proximity to the cardinal venous tissue, and this leads to sinus node dysfunction[9].

In the absence of a RSVC, central venous access should be made from the femoral vein in patient with PLSVC. When implanting a permanent pacemaker, the left subclavian vein is preferred. There is an acute angle between the CS ostium and the tricuspid valve, therefore, the lead should be looped in RA to enter the right ventricle[10].

In conclusion, the authors reported a case of PLSVC with absence of RSVC, which is an incidental finding in an elderly when a dilated CS was detected by transthoracic echocardiography.

Table 1. Characteristics of 13 adult patients reported with isolated PLSVC

First author, year of publication	Country	Sex	Age at diagnosis (year)	Heart defect	ECG	Symptomatic
Ozgul, 2010	Turkey	Female	30	No	-	-
Levent, 2011	Turkey	Female	52	-	-	-
Levent, 2011	Turkey	Male	65	-	-	-
Szymczyk, 2013	Poland	Male	72	No	-	-
Giuseppe, 2013	Italy	Female	71	No	LBBB	Asymptomatic
Jacques, 2014	Canada	Female	73	No	CHB	Fatigue
Azeem, 2014	U.K.	Male	66	ASD	NSR	Chest pain
Yusuke, 2015	Japan	Male	65	-	-	-
Jiri, 2015	Czech	Male	62	No	SSS	Syncope
Mahmadulla, 2016	India	Male	70	No	SSS	Syncope
Abraham, 2016	India	Male	43	No	SSS	Dizziness
Hatice, 2017	Cyprus	Female	63	No	SSS	Syncope
Zahra, 2019	U.K.	Male	28	No	-	-

ASD=atrial septal defect; ECG=electrocardiogram; LBBB=left bundle branch block; CHB=complete heart block; NSR=normal sinus rhythm; SSS=sick sinus syndrome

This condition is rarely known in adult in Thailand. The authors also described the diagnostic technique by injecting agitated saline from the left or right arm to reveal the opacification of the CS before the RA. The awareness of this thoracic venous anomaly is useful for clinicians, radiologists, sonographers, interventionist, anesthesiologists, and cardiothoracic surgeons to take precaution before performing any procedure involving the cardiac venous system to avoid any possible complication.

What is already known on this topic?

A case of PLSVC with absence of RSVC is rarely reported in adult in Thailand.

What this study adds?

This add another case of PLSVC with absence of RSVC in the elderly and was an incidental finding when a dilated CS was detected by transthoracic echocardiography. The awareness of this thoracic venous anomaly is useful for clinicians, radiologists, sonographers, interventionist, anesthesiologists, and cardiothoracic surgeons to take precaution before performing any procedure involving the cardiac venous system to avoid any possible complication.

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Authors' contributions

All authors had a role in writing and edited the report.

Conflicts of interest

The authors declare no conflict of interest.

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