CASE REPORT

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PERSISTENT LEFT SUPERIOR VENA CAVA MIMICKING LEFT ATRIAL MASS ON ECHOCARDIOGRAPHY

I. N. Dimitrova[1], V. Groudeva[2]

[1]Department of Cardiology, University Hospital "Sv. Ekaterina", Medical University – Sofia, Bulgaria
[2]Department of Diagnostic Imaging, University Hospital "Sv. Ekaterina", Medical University – Sofia, Bulgaria

Abstract. *Persistent left superior vena cava is a rare congenital venous anomaly, occurring in approximately 0.2–3% of the general population and more frequently (1.3–11%) in patients with congenital heart disease. It results from failed regression of the left superior cardinal vein during embryonic development. We report the case of a 70-year-old asymptomatic male in whom routine transthoracic echocardiography revealed a large, echo-lucent, cystic-like structure projecting into the left atrium, initially mimicking a left atrial mass. Further evaluation with contrast-enhanced computed tomography confirmed the diagnosis of an isolated persistent left superior vena cava draining into a markedly dilated coronary sinus, associated with the absence of the right superior vena cava. Persistent left superior vena cava may coexist with other congenital cardiac anomalies, such as atrial septal defect, bicuspid aortic valve, or cor triatriatum. Recognition of this vascular variation is essential in order to avoid misdiagnosis and to prevent potential complications during invasive cardiac or thoracic procedures. This case highlights the importance of multimodality imaging for accurate diagnosis and the differential diagnostic challenges such anomalies may present.*

Key words: *persistent left superior vena cava, coronary sinus dilatation, echocardiography, vascular anomaly, cardiac computed tomography*

Corresponding author: *Iva N. Dimitrova, MD, PhD, Department of Cardiology, University Hospital Sv. Ekaterina, Medical University – Sofia, 52A, Pencho Slaveikov Blvd., 1431 Sofia, Bulgaria, tel.: +359882 838320, email: dimytrova@yahoo.com*

ORCID: *0000-0001-6198-7134*

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INTRODUCTION

Persistent left superior vena cava (PLSVC) is a rare congenital vascular anomaly that originates at the junction of the left subclavian and internal jugular veins and descends through the left side of the mediastinum, adjacent to the aortic arch. It most commonly drains into the right atrium via the coronary sinus (CS) [1]. During the eighth week of gestation, the right superior cardinal vein forms the normal superior vena cava (SVC), whereas the left counterpart, below the innominate vein, usually regresses to become the ligament of Marshall. Failure of this regression results in the formation of a PLSVC [2].

Although reports of the presence of a PLSVC probably date back to the 17th–18th centuries, the first detailed and systematic description of this anomaly was provided in 1850 by John Marshall (1818–1891), an English surgeon and anatomy lecturer at University College Hospital, London, who published the first comprehensive review of the great anterior veins of

the thoracic region, which included a detailed description of the PLSVC in humans and other mammals [3, 4].

The reported prevalence among the general population is approximately 0.2–3% and it is more common in patients with congenital heart disease, between 1.3 and 11% and no statistically significant sex-related difference has been observed. It is frequently detected incidentally during imaging or invasive procedures; therefore, the true prevalence of PLSVC may be underestimated and could, in fact, be higher than reported [1].

While PLSVC is asymptomatic in approximately 40% of patients, it may be associated with congenital cardiac anomalies, such as atrial septal defect, bicuspid aortic valve, cor triatriatum, etc. [2]. Depending on its drainage pattern and associated defects, PLSVC can also lead to arrhythmias, cyanosis, or paradoxical embolism [5]. Recognition of this anomaly is essential before procedures, such as cardiac resynchronization therapy, pacemaker implantation, or central venous catheterization, as it may complicate these interventions [6].

Herein, we report a rare case of isolated persistent left superior vena cava (IPLSVC) in a 70-year-old male with situs solitus and no other cardiac anomalies. The condition was discovered incidentally during routine echocardiographic evaluation, where it appeared as a cystic, mass-like structure adjacent to the left atrium, posing differential diagnostic challenges before being confirmed by computed tomography (CT).

CASE REPORT

A 70-year-old male with a medical history of hypertension, dyslipidemia, diabetes mellitus, and a prior inferior wall myocardial infarction treated with percutaneous coronary intervention and bare-metal stent implantation to the right coronary artery 15 years earlier presented for routine follow-up. The patient had not attended regular medical check-ups in recent years. He denied experiencing chest pain, dyspnea, palpitations, or syncope. Physical examination, including cardiac auscultation, was unremarkable, with stable vital signs and normal cardiovascular findings. All laboratory parameters were within normal reference ranges. The electrocardiogram demonstrated sinus rhythm at a rate of 75 beats per minute, a normal electrical axis, and Q waves in the inferior leads, consistent with a previous inferior wall myocardial infarction.

Transthoracic echocardiography (TTE) revealed situs solitus and preserved left ventricular systolic function, with mild inferoposterior wall hypokinesia. Mild mitral and tricuspid regurgitation were present, and the estimated right ventricular systolic pressure was 20 mmHg. Incidentally, an unusually large, ovoid, well-defined, echo-lucent structure measuring approximately 9 × 10 mm in diameter was visualized in apical 4-chamber view, projecting into the left atrium (Figure 1a). It had smooth contours and appeared to be connected to the free wall of the left atrium, protruding into the atrial cavity during left atrial systole without affecting mitral valve leaflet motion, and resembling a cystic formation. In the parasternal long-axis view, the structure was poorly delineated, raising uncertainty as to whether it was intracavitary or exerting external compression with indentation of the posterior wall of the left atrium. There was no evidence of pulmonary venous inflow obstruction or significant hemodynamic effect (Figure 1b).

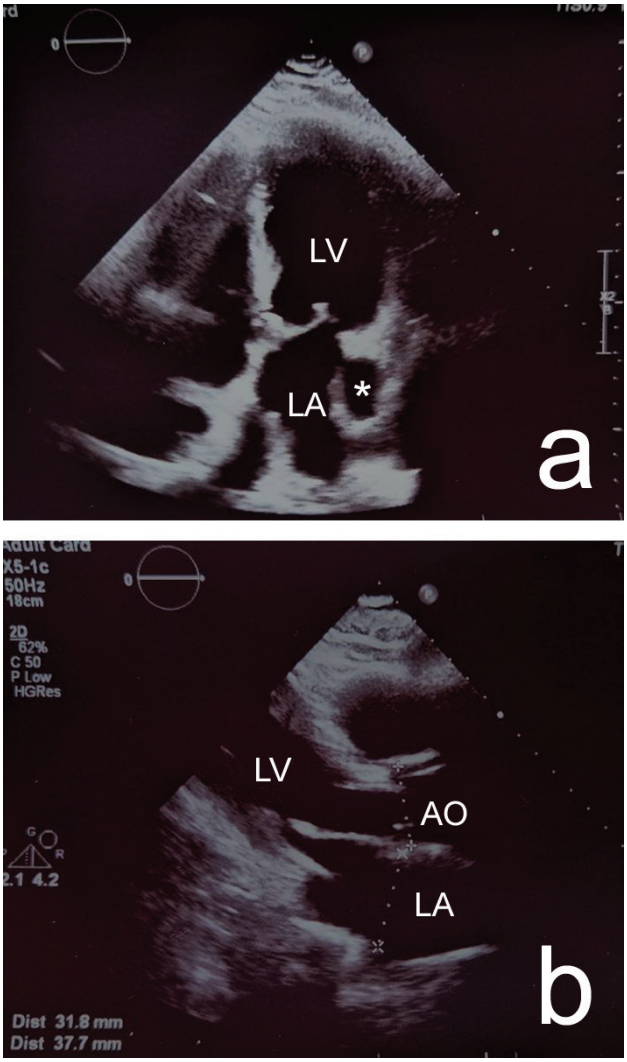

Fig. 1. a) TTE in apical 4-chamber view showing a huge echo-lucent structure (9 × 10 mm in diameter) projecting into the left atrium (*); **b)** TTE in left parasternal long-axis view- the structure is not delineated, neither intracavitary nor paracardiac. Other chambers visible are the left ventricle (LV), aorta (AO), and left atrium (LA)

This finding prompted consideration of the following differential diagnoses: dilated or aneurysmatic CS, vascular anomaly, pericardial or bronchogenic cyst, producing focal indentation and invagination of the atrial wall, encapsulated pericardial effusion in the atrio-ventricular groove, or an intracardiac cystic lesion, such as a fresh thrombus or myxoma with cystic degeneration, intracardiac blood cyst or even cor-triatrium or supramitral membrane, etc.

To further characterize the lesion, a contrast-enhanced cardiac CT was performed. It demonstrated normal pulmonary venous drainage with intact interatrial and interventricular septum. The right cardiac chambers, main pulmonary artery, and its major branches appeared normal in size. CT imaging noted a markedly dilated CS (diameter approximately 26 mm) (Figures 2a, 2b), receiving drainage from a PLSVC (Figure 2c) with invagination and compression of the posterior wall of the left atrium (Figure 2d). The right SVC was absent (Figure 2e, f). The CS drained normally into the right atrium. No other cardiac anomalies were identified.

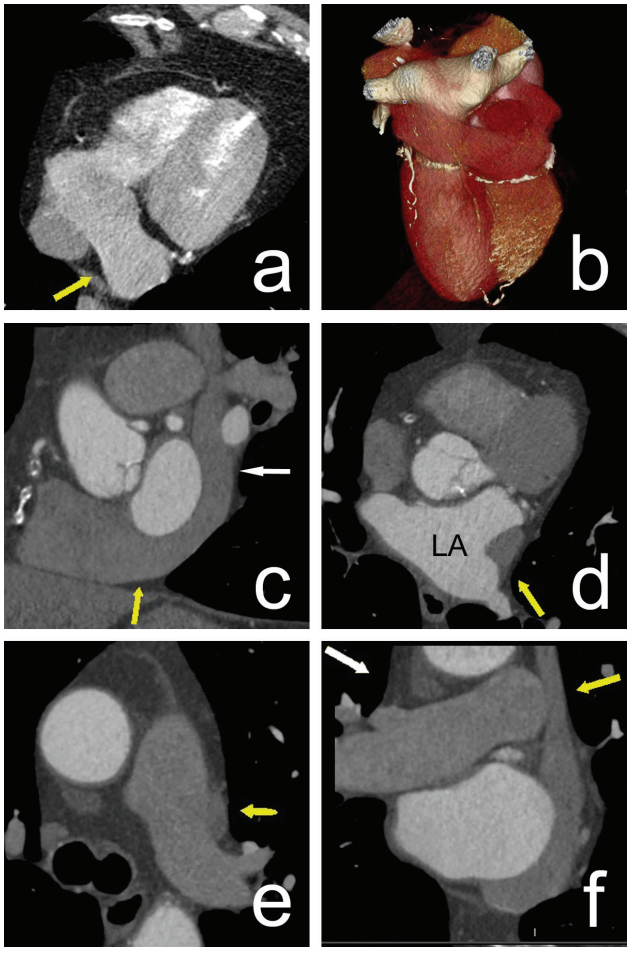

Fig. 2. Contrast-enhanced cardiac computed tomography. **a, b)** a markedly dilated coronary sinus – 26 mm in diameter (yellow arrow); **c)** Persistent left superior vena cava (white arrow), draining in the coronary sinus (yellow arrow); **d)** Persistent left superior vena cava draining into a dilated coronary sinus (yellow arrow), which compresses the posterior wall of the left atrium (LA); **e, f)** Isolated persistent left superior vena cava (yellow arrow) with absent right superior vena cava (white arrow)

The identified vascular anomaly required no specific therapeutic intervention, as it had no impact on the patient's physical activity. In view of the known ischemic heart disease and existing cardiovascular risk factors, the patient was considered suitable for optimal medical management and risk factor control.

DISCUSSION

Thoracic venous anomalies are uncommon findings that may involve the systemic or pulmonary venous systems. Awareness of these variants is essential for accurate diagnosis, appropriate patient management, and to prevent unnecessary additional investigations when such findings mimic pathological conditions and pose differential diagnostic challenges. PLSVC, although it is a rare anomaly, represents the most common congenital variation of the thoracic venous system. The true prevalence is likely higher than reported, since the anomaly is asymptomatic and often detected incidentally. Cardiac defects are more frequently observed in fetuses than among the general population, likely because severe malformations can lead to miscarriage or early fetal death. Nonetheless, in patients with congenital heart diseases, the incidence has been reported to be as high as 12% [7].

In the majority of cases – approximately 92% – the PLSVC collects deoxygenated blood from the left side of the head, neck and left arm, drains into the right atrium via the CS, leading to its characteristic dilatation and it is a benign condition with no apparent effect on cardiac hemodynamics, while in 8% of cases, it drains directly or via the pulmonary veins into the left atrium, producing a right-to-left shunt that may result in systemic desaturation and paradoxical embolism [8]. There are two main anatomical variants of PLSVC. The double (or bilateral) SVC, which accounts for approximately 90% of cases, is characterized by the presence of both right and left SVC, with the right SVC draining into its normal location in the right atrium. In such cases, a connecting innominate (brachiocephalic) vein is observed in about 30% of patients. When the right SVC is absent – as in the present case – the isolated persistent left superior vena cava (IPLSVC) carries venous return from the right upper body via the brachiocephalic vein to the left SVC, which then becomes the sole pathway for upper body venous return, leading to marked dilatation of the CS [1, 9].

In early fetal development, the venous drainage of the thorax is performed by two pairs of cardinal veins – anterior veins, draining the upper part of the embryo, and posterior veins, draining the lower part. Each pair converges to form common cardinal veins that enter the primitive heart. Around the eighth week of gestation, a connection forms between the right and left anterior cardinal veins and later develops into the brachiocephalic (innominate) vein. Normally, the lower segment of the left anterior cardinal vein regresses and becomes the ligament of Marshall, while the corresponding segment on the right side persists as the right SVC. If this regression on the left side does not occur, a PLSVC remains, typically draining into the CS. The brachiocephalic vein may or may not be present, leading to different anatomic patterns [10, 11].

This venous anomaly has been reported to occur more frequently in association with other congenital cardiac defects. Notably, almost 40% of individuals with a PLSVC present with associated cardiac anomalies, underscoring the strong relationship between this venous variation and other coexisting cardiac defects. Although an isolated PLSVC as in the case described is usually asymptomatic and has no hemodynamic consequence, its presence in association with congenital cardiovascular malformations is clinically significant, as these defects may be responsible for the development of symptoms [2].

A broad spectrum of congenital cardiac defects can be associated with PSVC and various authors have proposed subgroup classifications of these anomalies: shunt lesions are common and include atrial septal defects, ventricular septal defects (VSD), atrioventricular septal defects, patent ductus arteriosus and anomalous pulmonary venous drainage; conotruncal malformations – tetralogy of Fallot, pulmonary atresia with VSD, levo- or dextro-transposition of the great arteries, truncus arteriosus; left-sided obstructive lesions – coarctation of the aorta, cor triatriatum, mitral stenosis and bicuspid aortic valve; right-sided lesions-pulmonary stenosis, pulmonary atresia, tricuspid atresia, bicuspid pulmonary valve, and Ebstein anomaly; single ventricular anomalies and aortic arch anomalies – cervical aortic arch, right aortic arch, aberrant right subclavian artery and right aortic arch with aberrant left subclavian artery [1, 12, 13]. The most frequent cardiac anomalies coexisting with PSLVC are VSD, atrial septal defect; atrioventricular septal defect, tetralogy of Fallot; transposition of the great vessels [7, 14, 15]. Among extracardiac anomalies, esophageal atresia is reported most frequently in association with PSLVC [7]. Patients with IPLSVC, particularly those with drainage into the left atrium, exhibit a higher rate of coexisting congenital cardiac abnormalities [16].

Coexistence of PLSVC and heterotaxy is relatively frequent. Heterotaxy, which includes situs inversus and situs ambiguus (right or left isomerism), is found in up to 45% of fetuses with PLSVC detected prenatally. According to some authors, the coexistence of isolated PLSVC and situs inversus is regarded as a normal mirror-image configuration rather than a true anomaly. Among patients with heterotaxy and SVC anomaly, double SVC occurs in about 70% of cases, while others present with an IPLSVC. The most common associated cardiac defects in these individuals include complete atrioventricular septal defect, right ventricular outflow tract obstruction, and double outlet right ventricle. Absence of CS dilatation, often due to an unroofed CS, is a characteristic feature in heterotaxy and does not exclude the presence of PLSVC [13, 17, 18].

Most individuals with PLSVC are asymptomatic, and the anomaly is frequently detected incidentally during imaging, cardiac catheterization, or device implantation. Several diagnostic tools exist for the evaluation of patients with a PLSVC, each with its own advantages and limitations. Basic diagnostic modalities include transthoracic and transesophageal echocardiography, which allow non-invasive visualization of the CS and venous drainage, conventional contrast venography, CT and magnetic resonance (MR) venography, providing more detailed anatomical information and aiding in differentiation from other mediastinal or atrial structures [1].

Echocardiography is a widely available, first-line imaging modality in the evaluation of PLSVC. It has the advantages of being inexpensive, free from ionizing radiation and unaffected by cardiac rhythm, making it a safe and reliable imaging method for repeated or bedside evaluation. However, it is operator- and acoustic-window dependent, and its spatial resolution is lower than that of CT or MRI [1]. On TTE, a dilated CS is often the first clue. However, a dilated CS can arise from several conditions and is not exclusively indicative of a PLSVC. While the most frequent cause is increased right atrial pressure, other possible explanations include a coronary arteriovenous fistula, partial anomalous pulmonary venous return, or an unroofed CS. Confirmation of the existence of PSLVC is achieved with a contrast (bubble) study: injection of agitated saline into a left arm vein results in early opacification of the CS before the right atrium, whereas injection into a right arm vein produces normal right atrial opacification before the CS, confirming a normal right SVC. In cases of IPLSVC, a bubble study shows early opacification of the CS after injection into

a right arm vein [2]. TTE is useful for identifying coexisting cardiac anomalies, while contrast-enhanced and transesophageal echocardiography provide additional value in detecting PLSVC. Fetal echocardiography should also be mentioned, as it plays a crucial role in the prenatal detection of vascular anomalies, cardiac malformations, and cardiac tumors [19].

However, as illustrated in this case, the echocardiographic appearance may mimic a cystic or mass-like structure adjacent to the left atrium, posing a diagnostic challenge. Echocardiographic features suggesting a vascular nature include a well-defined, echo-lucent structure located posterior to the left atrium and the demonstration of venous flow directed toward the right atrium on color Doppler imaging.

Multidetector CT is a rapid and widely available imaging technique that offers excellent spatial resolution and multiplanar reconstruction, making it highly effective for detailed evaluation of PLSVC and associated anomalies. Its main limitations include radiation exposure, contrast-related risks, and possible motion artifacts from cardiac rhythm changes. Optimal assessment is achieved with ECG-gated cardiac CT angiography and delayed venous phase imaging [1].

Magnetic resonance imaging is a radiation-free modality that provides high spatial resolution and multiplanar imaging, enabling detailed assessment of venous anatomy and flow direction. It can visualize PLSVC even without contrast, though contrast-enhanced and phase-contrast MR angiography may offer additional detail. Limitations include higher cost, low scanning speed, and contraindications, such as magnetic implants or claustrophobia [1].

Invasive angiography remains the gold standard for defining venous anatomy, providing excellent morphological detail and the possibility for intervention when necessary. However, its use is limited by its invasive nature, radiation exposure, and the need for iodinated contrast, which carries risks of allergy or nephrotoxicity. Although catheter venography with water-soluble contrast can precisely delineate PLSVC, it is not routinely performed for diagnosis and is often identified incidentally during procedures such as central venous catheterization or pacemaker implantation [1].

The clinical significance of a PLSVC depends primarily on its drainage site and the presence of associated cardiac anomalies. In patients with concomitant congenital heart defects, the clinical manifestations are usually attributable to those associated anomalies rather than to the PLSVC itself [5]. The PLSVC is often hemodynamically insignificant and asymptomatic when it drains into the right atrium via the CS. However, a markedly enlarged CS, even to the point of aneurysmal dilation, can occasionally compress adjacent cardiac structures, such as the atrioventricular node or bundle of His (BH), predisposing patients to conduction disturbances or arrhythmias [1]. On the other hand, compression of the left atrium and a consequent decrease in cardiac output may occur due to this enlargement. A markedly dilated CS has even been reported to cause left ventricular inflow obstruction, potentially leading to the development of heart failure [20].

When the PLSVC drains into the left atrium, right-to-left shunt occurs, which may lead to systemic desaturation, cyanosis, and an increased risk of paradoxical embolism or brain abscess, requiring surgical correction [16].

The presence of a PLSVC can complicate procedures, such as central venous catheterization, pacemaker- or defibrillator- lead placement, or cardiopulmonary bypass cannulation. Performing Swan-Ganz catheterization at the bedside can be challenging without imaging guidance. During permanent pacemaker implantation, leads introduced from the left side may follow an unusual course through the CS and can be a reason for ineffective fixation. Since the procedure is always performed under fluoroscopic guidance, a PLSVC is typically detected at that time. Insertion of pacemaker leads or catheters through a PLSVC has been associated with serious complications, such as arrhythmias, cardiogenic shock, cardiac tamponade, and thrombosis of the CS. However, these events are relatively uncommon, with technique and device improvement over time [1, 2].

In patients undergoing cardiac surgery, a PLSVC is a relative contraindication for retrograde cardioplegia, as its administration is hampered by this venous anomaly. The PLSVC can cause excessive runoff of the solution into itself and the right atrium, potentially resulting in inadequate myocardial protection and rendering the cardioplegia ineffective. Therefore, preoperative identification of a PLSVC is essential [21].

In asymptomatic individuals with PLSVC draining into the right atrium and without associated cardiac malformations, no specific treatment is required [22]. However, documentation of the anomaly is important for future reference, particularly prior to invasive cardiac or venous procedures. Periodic follow-up with echocardiography may be warranted in cases with marked CS dilatation to ensure no compression or hemodynamic effects develop over time.

CONCLUSION

This case highlights the need to consider vascular anomalies, such as a PLSVC, when evaluating

cystic or echo-lucent structures near the left atrium on echocardiography. The use of multiple imaging modalities, including echocardiography and CT, was key in establishing the correct diagnosis and preventing misidentification as a pathological mass. Our case represents a rare, asymptomatic instance of IPLSVC occurring without any other cardiac or structural anomalies, which is unusual compared to most previously reported cases. Familiarity with this congenital variant is important for cardiologists, radiologists, and cardiac surgeons in order to minimize the risk of diagnostic mistakes and procedural complications.

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Consent for publication: *Consent form for publication was signed by the patient and collected.*

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