

CASE PRESENTATION

Isolated persistent left superior vein cava unmasked by a simple non-invasive cardiac imaging

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Abstract: This paper presents the case of an asymptomatic persistent left superior vena cava unmasked by a simple non-invasive cardiac imaging, namely microbubble test by transthoracic echocardiography.

A 25 years old man complaining of atypical chest pain at exertion was examined by the cardiologist. The clinical exam was normal. Twelve leads ECG showed sinus rhythm of 55 beats/min, QRS axis of 70 degree and minor right bundle branch block. Transthoracic echocardiography has shown a large coronary sinus of 32 mm, without right cavities dilatation and without any sign of pulmonary hypertension. Transthoracic echocardiogram was repeated during injection of agitated saline solution into the left basilica vein and micro-bubbles unexpectedly filled firstly coronary sinus and then passed through the right atrium. Contrast enhanced computer tomography angiography confirmed the presence of persistent left superior vein cava and the site of drainage into a large coronary sinus. Right superior vein cava was also identified.

It is mandatory to consider persistent left superior vena cava draining into the coronary sinus in the diagnosis of patients presenting with dilated coronary sinus shown by transthoracic echocardiography. Even if persistent left superior vena cava could be diagnosed by a simple, non-invasive imaging test, an associated heart disease must always be excluded in these patients by another imaging method like computer tomography. This case underlines also the importance of multimodality imaging in the evaluation of these abnormalities.

Keywords: coronary sinus, transthoracic echocardiogram, agitated saline, micro-bubbles.

Rezumat: Lucrarea prezintă cazul unui pacient cu persistență de venă cavă superioară stângă asimptomatică demascată printr-o simplă metodă imagistică cardiacă, non-invazivă, și anume testul cu microbule - ecocardiografie transtoracică.

Un bărbat în vârstă de 25 de ani s-a adresat pentru un consult cu acuze de dureri toracice atipice de mai mulți ani. Examenul clinic a fost normal. Electrocardiograma de suprafață a arătat bradicardie sinusală cu 55 bătăi/minut (cu unde P negative în DIII, aVF), axa electrică QRS la 70 de grade și bloc minor de ramură dreaptă. Ecocardiografia transtoracică a fost în limite normale, cu excepția unui sinus coronarian dilatat de 32 mm, fără cavități drepte mărite și fără niciun semn de hipertensiune pulmonară. Ecocardiografia transtoracică cu injectare de soluție salină agitată în vena bazilică stângă a evidențiat opacifierea atriului drept prin sinusul coronarian și nu prin vena cava superioară. Examenul computer-tomografic cu substanță de contrast a confirmat prezența unei vene cavă superioară stângă cu drenaj în sinusul coronarian, care era foarte dilatat. Vena cavă superioară dreaptă era prezentă și nu s-au identificat alte anomalii anatomice.

În cazul unui sinus coronarian dilatat la ecocardiografia transtoracică este obligatoriu să se ia în considerare persistența de vena cavă superioară stângă. Această anomalie poate fi confirmată printr-un test imagistic simplu, non-invaziv, precum testul cu microbule. Întotdeauna trebuie căutată o eventuală anomalie asociată printr-o altă metodă imagistică precum tomografia computerizată cu contrast. Acest caz subliniază importanța utilizării unui test simplu dar și a mai multor metode imagistice în evaluarea acestei anomalii.

Cuvinte cheie: sinus coronarian, ecocardiografie transtoracică, soluție salină agitată, microbule.

BACKGROUND

Persistence of left superior vena cava (PLSVC) is the most common venous anomaly. While affecting 0.5% of the general population, it influences procedures re-

quiring vascular intervention or device implantation and may predispose to atrial fibrillation. Despite being mostly asymptomatic, it can be identified through simple cardiac imaging.

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CASE REPORT

We present you the case of a 25-year old male referred to the clinic for atypical chest pain at exertion. Taking into account the unremarkable medical history and negative family history, we proceeded into investigating the case.

The clinical exam was normal. After performing the ECG, we noted that while being in sinus rhythm with a heart rate of 55 bpm and an AQRS at 60-70 degrees, the patient had a minor right bundle branch block (Figure 1). We also remarked the negative P waves in DIII, aVF.

Twenty four hour ECG monitoring was unremarkable. The fact that the patient's chest pain continued prompted us to perform a transthoracic echocardiography, which showed normal dimensions and functions of the left cavities (Figure 2), and a markedly dilated coronary sinus (CS) of 32 mm in the absence of right cavities dilatation or any signs of pulmonary hypertension (Video 1 still image). The dilated coronary sinus gave rise to the suspicion of a case of PLSVC. The patient refused transoesophageal echocardiography.

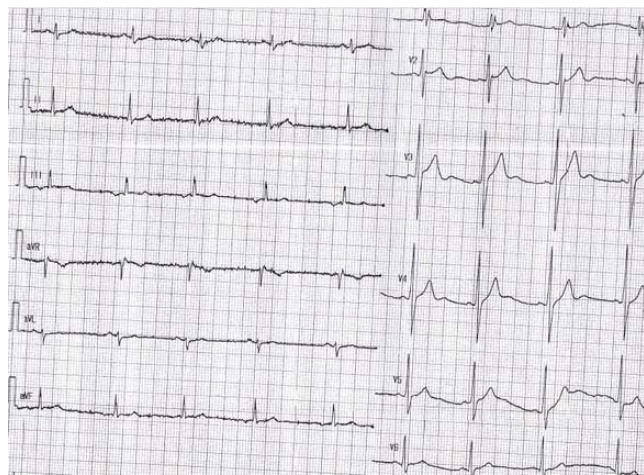


Figure 1. Standard electrocardiogram in 12 leads.



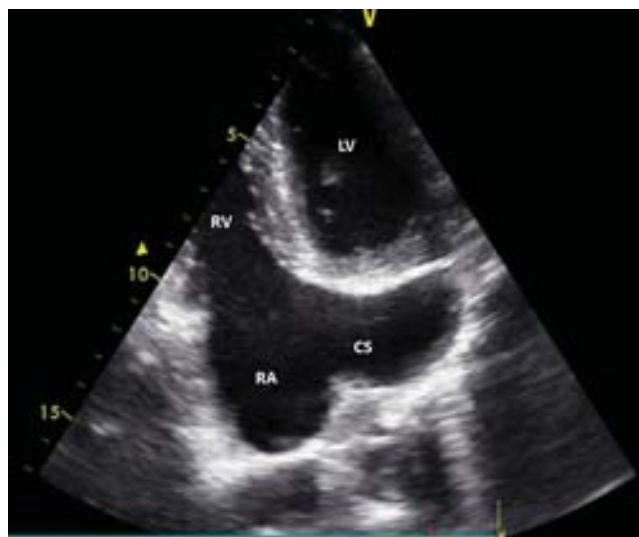
Figure 2. Left image: Bidimensional transthoracic echocardiography in parasternal long axis view showed a markedly dilated coronary sinus. Right image: Bidimensional transthoracic echocardiography in apical 4 chambers view showed normal right cavities, without signs of pulmonary hypertension and dilated coronary sinus. Ao: aorta; CS: coronary sinus; LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle.

We decided to perform an agitated saline study, injecting the solution in the left arm. Filling of the right atrium with microbubbles not via the superior vein cava, but through the coronary sinus testified for the presence of the aforementioned anomaly (Video 2 still image).

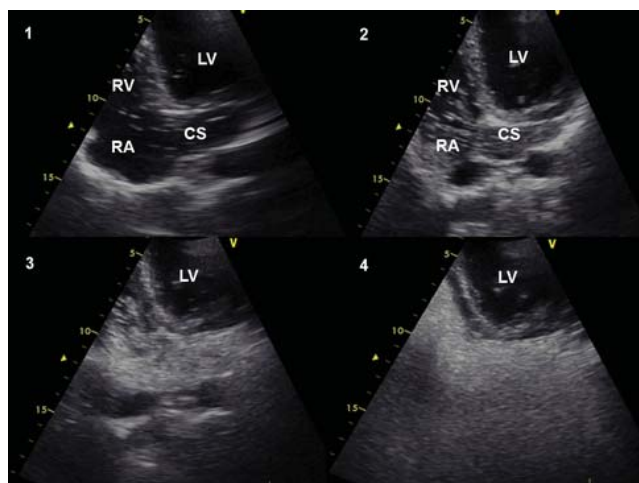
Contrast enhanced computer tomography (CT) angiography confirmed the anomaly, showing the PLSVC draining into the coronary sinus in the presence of normal superior and inferior vein cava (Figure 3).

DISCUSSION

PLSVC is the most common congenital anomaly involving thoracic central venous return¹. While ~0.5% of the general population is affected, the percentage rises up



Video 1 still image. Bidimensional transthoracic echocardiography in apical 4 chambers view showed a severely dilated coronary sinus. CS: coronary sinus; LV: left ventricle; RA: right atrium; RV: right ventricle.



Video 2 still image. Microbubble test by transthoracic echocardiography in apical 4 chambers view showed filling of the right atrium through the coronary sinus not through the left superior vein cava. CS: coronary sinus; LV: left ventricle; RA: right atrium; RV: right ventricle.

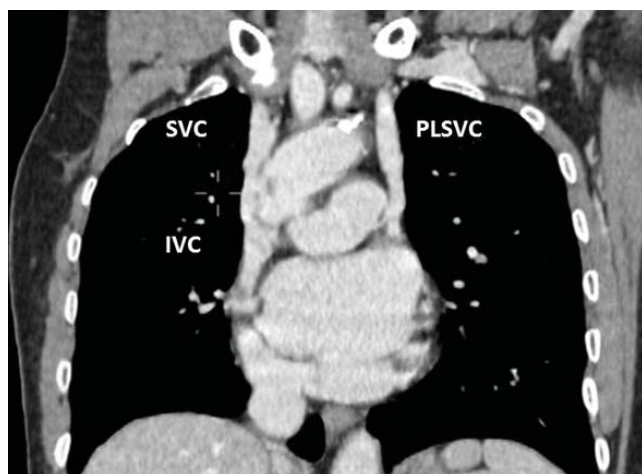


Figure 3. CT angiography (frontal view) confirms the presence of PLSVC. IVC: inferior vein cava; PLSVC: persistent left superior vein cava; SVC: superior vein cava.

to 10% in those suffering from congenital heart diseases^{1,2}. A PLSVC is an embryological remnant representing persistence of the left anterior cardinal vein that fails to obliterate during embryologic development of right atrium^{1,3}. The right and left anterior cardinal veins drain the cranial regions into the respective ducts of Cuvier, which in turn drain into the sinus venosus. The right cardinal vein and right duct of Cuvier form the superior vein cava. Bridging connection between the anterior cardinal veins forms the left innominate vein. The sinus venosus could be absorbed into the structure of the right atrium.

It has multiple possible draining sites including inferior vein cava, hepatic vein or left atrium, the most common being right atrium via the CS (about 90% of cases)¹. Another possibility is drainage into the left atrium through an unroofed CS, thus giving rise to a right-to-left shunt. Most of the times it remains an incidental finding, as this anomaly is clinically silent in its most common form. In rare cases, PLSVC can determine severe right heart dilatation by draining into left atrium in association with an unroofed CS and CS atrial septal defect⁴. However PLSVC typically drains into the coronary sinus via the vein of Marshall.

Usually the LSVC descends vertically, anterior, and to the left of the aortic arch and main pulmonary artery and enters the left atrium between the pulmonary veins and the atrial appendage, draining, most frequently into the CS. In patients with suspected PLSVC while left arm injection during microbubble test determines fills CS first, followed by right atrium, right arm injection will determine fills right atrium first. Bilateral arm contrast injections should be performed if suspecting a

PLSVC, such as an enlarged CS, but do not expand on the reasoning. Bilateral injections can determine right- and left-sided anomalous drainage.

Though drainage into the right atrium via the CS holds no hemodynamic consequences, it could translate into a difficulty when undergoing device implantation or other procedures requiring vascular access. Another risk cited in literature is developing cardiac arrhythmias, particularly atrial fibrillation due to the ligament of Marshall. Given the right-to-left shunt that drainage into the left atrium creates, patients most typically present with cyanosis at this point. Other cardiovascular anomalies have been associated with PLSVC, such as atrial or ventricular septum defects, tetralogy of Fallot, and CS ostium atresia¹.

The differential diagnosis of a dilated CS includes partial anomalous hepatic venous drainage and continuity of the inferior vein cava with an LSVC via the hemiazygos vein. Various shunts may result in CS dilatation, either low pressure in the case of unroofed CS and anomalous pulmonary venous drainage, or high pressure in the form of a coronary artery to CS fistula⁵. Finally, any cause of raised right atrial pressure may cause dilatation.

Despite being mostly asymptomatic, the fact that this anomaly is easily detected through cardiac imaging such CT, magnetic resonance imaging and transthoracic echocardiography should prompt clinicians for a throughout set of investigations when suspecting such an anomaly. We also emphasize the importance of the microbubble test that, while being easily reproduced with a minimum set of risks for the patient, it enables early detection of abnormal intracardiac blood flow. Even if PLSVC could be diagnosed by a simple, non-invasive imaging test, an associated heart disease must always be excluded in these patients by another imaging method like CT. However multimodality imaging in the evaluation of these abnormalities is very important.

The authors presented the case of a young asymptomatic patient with a benign congenital anomaly unmasked by a simple non-invasive cardiac imaging, namely microbubble test (very useful but perhaps underused in clinical practice) and confirmed by another cardiac imaging technique like CT angiography.

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Video legends

Video 1: Bidimensional transthoracic echocardiography in apical 4 chambers view show markedly dilated coronary sinus in the absence of right cavities dilatation or any signs of pulmonary hypertension.

Video 2: Bidimensional transthoracic echocardiography in apical 4 chambers and agitated saline study: filling of the right atrium with microbubbles not via the superior vein cava, but through the coronary sinus.