Isolated Persistent Left Superior Vena Cava, Role of Echocardiography Screening and CT angiography

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Abstract

Background: An isolated persistent left SVC with concomittant agenesis of right SVC in adult patients is a very rare abnormality. Physician should consider it particularly in patients, in which venous acces will be performed. Our rare case deals with the importance of detailed echocardiographic examination with screening of coronary sinus dilatation before the electrophysiology study.
Case: A 65-year-old woman came to outpatient clinic for a chief complaint of palpitations. Her ECG showed paroxysmal SVT with WPW syndrome. She underwent echocardiography examination before electrophysiology study and it was found that she had a dilated coronary sinus. Therefore we performed cardiac CT. It was found that she had a persistent left superior vena cava (SVC) and an absence of a right SVC with no other congenital anomaly.
Conclusion: A comprehensive echocardiography examination to look for a dilation of coronary sinus is a first suggestion to screen this anomaly, eventually followed by echocardiography with agitated saline injection and/or computed tomography can help physician to anticipate the anomaly before the invasive procedure involving the thoracic vein.

Keywords: echocardiography, cardiac CT angiography, vascular malformation, superiorvena cava

BACKGROUND

Persistent left superior vena cava (PLSVC) is the most common congenital malformation of the thoracic venous system and it affects about 0.3 to 0.5% of the general population.[1] This incidence increases 10-fold in patients with cardiac malformations.[2] PLSVC with an absent right superior vena cava (RSVC), which is also referred to as isolated PLSVC, is very uncommon, occurring in 0.07 to 0.13% of patients who have congenital heart defects with viscero-atrial situs solitus. Nearly half of the patients with isolated PLSVC have other cardiac malformations, such as atrial septal defect, endocardial cushion defect or tetralogy of Fallot.[3] In this case report we present a patient with isolatedPLSVC with no other cardiac structural abnormalities. The PLSVC was diagnosed by means of echocardiography. The diagnosis was supported by cardiac CT angiography.

CASE

A 65-year old woman went to outpatient clinic with a chief complaint of palpitations. Her ECG showed paroxysmal supraventricular tachycardia (SVT) with Wolf-Parkinson-White (WPW) syndrome. It was decided that she should undergo electrophysiology study and ablation. The echocardiography examination revealed a dilated coronary sinus. A CT angiography was performed because of suspicion of a persistent left superior vena cava (PLSVC). The examination showed a bridging vein draining the right jugular and right subclavian veins; it joined the left brachiocephalic vein and formed thePLSVC, which descended at the left side of the mediastinum, leftward of the pulmonary artery and left atrium (LA) before draining into the right atrium (RA) via a dilated coronary sinus(CS) (Figs 1–3). The RSVC was absent and the PLSVC carriedall venous blood from

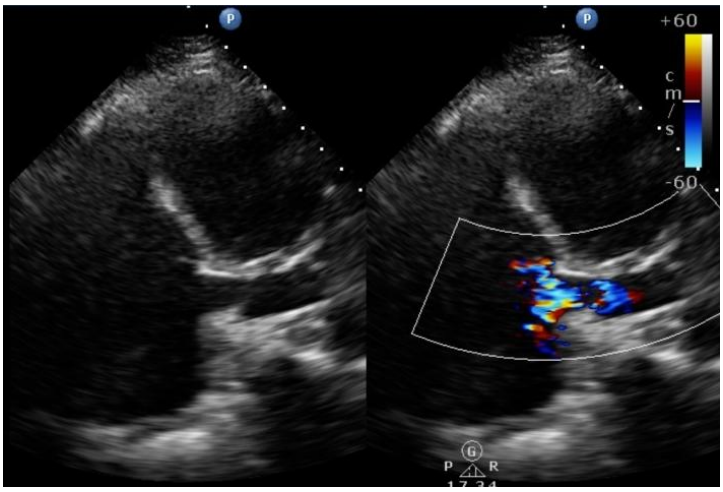

Figure 1. Transthoracal echocardiography showed a dilatation of coronary sinus draining to RA.

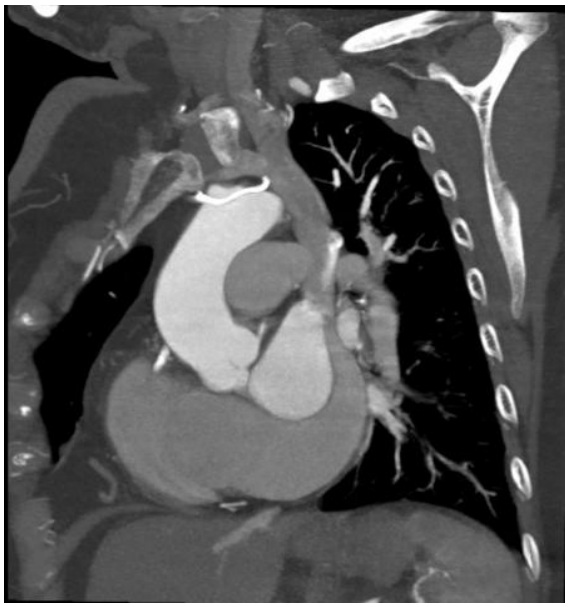

Figure 2. Multiplanar reformatted image demonstrates that the right superior vena cava is absent and there is persistent left superior vena cava (PLSVC).

the head, neck and upper extremities.There was no other intra- or extracardiac pathological structural finding.

DISCUSSION

PLSVC with absent RSVC (isolated PLSVC) is a very rarevenous malformation. During normal fetal development, theleft-sided anterior venous cardinal system regresses, leaving theCS and the ligament of Marshall. Failure of the closure of the leftanterior cardinal vein results in PLSVC.[4] In general, PLSVC isassociated with RSVC and drains into the RA via a dilated CS.When developmental arrest occurs at an earlier stage, the CSis absent and the PLSVC drains

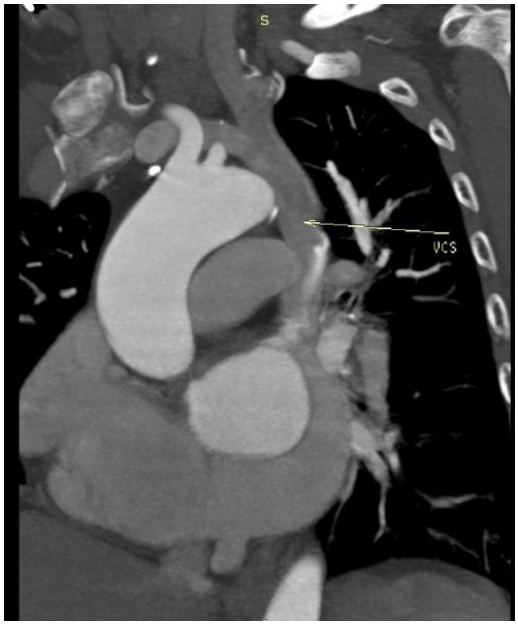

Figure 3. Multiplanar reformatted image reveals the persistent left superior vena cava (PLSVC) draining into a dilated coronary sinus (CS)

into the LA. Either isolated orassociated with RSVC, this venous malformation itself causes no haemodynamic disturbance and is usually diagnosed incidentally. [5-7]

However, it has several clinical implications. A PLSVC cancause problems during central venous catheterisation (access tothe CS can cause hypotension, angina, perforation of the heart,tamponade and arrest),[8] pacemaker implantation (due to thecircuitous path taken by the electrode, it can be difficult to obtaina stable electrode position and sustained capture),[9] or cardiopulcardiopulmonarybypass (isolated PLSVC impairs the use of retrogradecardioplegia).

In addition, a higher incidence of arrhythmias and conductionsystem abnormalities has been described in patients withPLSVC. There are two proposed mechanisms for this association:a dilated CS stretches the atrioventricular nodal tissue,which prepares a substrate for re-entrant tachycardias; or, theearly conduction tissue has close proximity to the cardinalvenous tissue and this leads to sinus node dysfunction. Lenox*et al*. found sino-atrial node abnormalities in some patients withabsent RSVC and this condition may predispose to sick sinussyndrome.[10-12]

In 10% of patients, a PLSVC may drain into the LA eitherdirectly or via an unroofed CS. This creates a right-to-left shuntand the risk of paradoxical embolism is markedly increased. Inaddition, drugs directly enter the systemic circulation when theyare applied from the left brachiocephalic vein.A final clinical implication of PLSVC (especially whenisolated) is a high incidence of accompanying congenital heartdefects, for example ventricular septal defect, atrial septal defect,endocardial cushion defect or tetralogy of Fallot.[3,13] Thereforeassociated congenital heart disease should be meticulouslysearched for.

When PLSVC is present, the ECG often shows an abnormalP-wave axis and a normal or shortened PR interval. A geometricchange in the LA may be a possible mechanism for the

left-axisdeviation of the P wave.[14] On chest X-ray, a crescent-shapedshadow of the PLSVC can be seen at the aortic knob or left uppermediastinum. After insertion of a pulmonary artery catheter intothe left subclavian or jugular vein, a control chest X-ray gives thefalse appearance that the catheter has passed through the vessel. The diagnosis can be confirmed by TTE, transoesophagealechocardiography (TEE), venous angiography, computed tomography(CT) or magnetic resonance imaging (MRI).

On two-dimensional B-mode TTE, the characteristic finding is a dilated CS on parasternal long-axis view. The normal diameter of the CS is smaller than 1 cm and in the case of isolated PLSVC, severely increased flow can cause a truly giant CS.[15,16] Other causes of dilated CS are: increased RA pressure, an anomalous systemic or pulmonary venous connection or a fistulous connection with the coronary arteries.[17]

The next step in the echocardiographic evaluation should becontrast application with agitated saline. In normal individuals, agitated saline injection from the left or right antecubital vein results in opacification of the RA. In isolated PLSVC, as in ourcase, contrast given from the left or right arm opacifies the CS. When PLSVC is associated with an unroofed CS, contrast injection from either arm results in opacification of the LA. If RSVC accompanies the PLSVC, contrast given from the left arm first appears in the CS, whereas contrast given from the right arm first appears in the RA. On TEE, the anomalous PLSVC and absence of RSVC can be well visualised. In mid-oesophageal views, the PLSVC can be seen near to the left atrial appendage and left upper pulmonary vein. In the bicaval view, the absence of RSVC can be demonstrated.

Other techniques (venous angiography, CT, MRI) directly visualise the venous anatomy and confirm the diagnosis. In the absence of an RSVC, central venous access should bemade from the femoral vein in patients with PLSVC. During right-sided open-heart surgical procedures, a PLSVC has to be drained by inserting a separate cannula into it. If the PLSVC drains into the LA and creates a large right-to-left shunt, surgical correction should be made. Again, central venous access via the femoral vein is a safer choice in this variation. When implanting permanent pacemakers, the left subclavian vein is preferred, as lead manipulation is easier. There is an acute angle between the CS ostium and the tricuspid valve, therefore the lead should belooped in the RA in order to enter the right ventricle.[18] Handshapedstylets and active fixation leads are also helpful to overcometechnical difficulties.[19]

Finally, a wide spectrum of clinicians (radiologists, sonographers,intervenists, intensivists, anaesthesiologists, cardiothoracic-surgeons) should be aware of PLSVC and its variations inorder to avoid possible complications.

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