Diagnosis of persistent left superior vena cava: the role of echocardiography

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SOUHRN

Perzistující levostranná horní dutá žíla (persistent left superior vena cava, PLSVC) představuje vzácnou, avšak nejčastější anomálii společných hrudních žil. K jejímu vzniku dochází v případech, kdy nedojde k regresi levé horní kardinální žíly kaudálně k bezejmenné (ramenohlavové) žíle a může souviset s dalšími kardiovaskulárními abnormalitami. I když je PLSVC často asymptomatická, může být během intervenčních výkonů nebo chirurgické léčby srdečních anomálií příčinou závažných komplikací. Náhodné zjištění dilatace koronárního sinu při echokardiografickém vyšetření musí vyvolat podezření na přítomnost PLSVC. Diagnózu je nutno potvrdit kontrastní echokardiografií s agitovaným fyziologickým roztokem.

ABSTRACT

Persistent left superior vena cava (PLSVC) is a rare but the most common thoracic venous anomaly. It results when the left superior cardinal vein caudal to the innominate vein fails to regress and can be associated with other cardiovascular abnormalities. While PLSVC is often asymptomatic, it can cause serious complications during vascular interventional procedures or surgical treatment of cardiac anomalies. Incidental identification of a dilated coronary sinus on echocardiography should raise the suspicion of PLSVC. The diagnosis should be confirmed by agitated saline contrast echocardiography.

Introduction

Although persistent left superior vena cava (PLSVC) is a rare vascular anomaly, it is the most common congenital malformation of the thoracic venous system.[1] Mostly, PLSVC is asymptomatic and detected incidentally in diagnostic and therapeutic examinations due to different reasons.[2] It is most commonly observed in isolation but can be associated with other cardiovascular abnormalities (atrial septal defect, bicuspid aortic valve, coarctation of the aorta, among others) and arrhythmias.[2,3] This condition has implications concerning the placement of catheters in the right heart by the left subclavian route and the administration of retrograde cardioplegia solutions during heart surgery.[1]

PLSVC occurs due to the failure of the left superior cardinal vein to obliterate to form the ligament of Marchall during the early weeks of embryologic development, resulting in a persistent left-sided venous vasculature which, via the coronary sinus, drains into the right atrium.[3] This anomaly causes the dilatation of the coronary sinus due to the increased venous return. Therefore the presence of a dilated coronary sinus detected by an echocardiogram, in a patient without evidence of elevated right-sided filling pressures are an important clue to this diagnosis.[3] PLSVC diagnosis can be confirmed by echocardiography, using agitated saline.[4]

The following echocardiography diagnostic criteria have been proposed: (1) the presence of a dilated coronary sinus on two-dimensional echocardiography in the absence of evidence of elevated right-sided filling pressures; (2) enhancement of the dilated coronary sinus before the right atrium after contrast material infusion into a left arm vein; (3) right atrium opacification before the coronary sinus with contrast injected from the right arm.[2,4]

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Case report

A 34-year-old male was referred to the echocardiography laboratory to perform a transthoracic examination to evaluate cardiac structure and function in the context of frequent ventricular extrasystoles.

The transthoracic echocardiogram (TTE) showed a remarkably dilated coronary sinus in the parasternal long-axis view (Fig. 1). The right cavities were not dilated and there was not any evidence of elevated right-sided filling pressures. The suspicion of a persistent left superior vena cava was proposed.

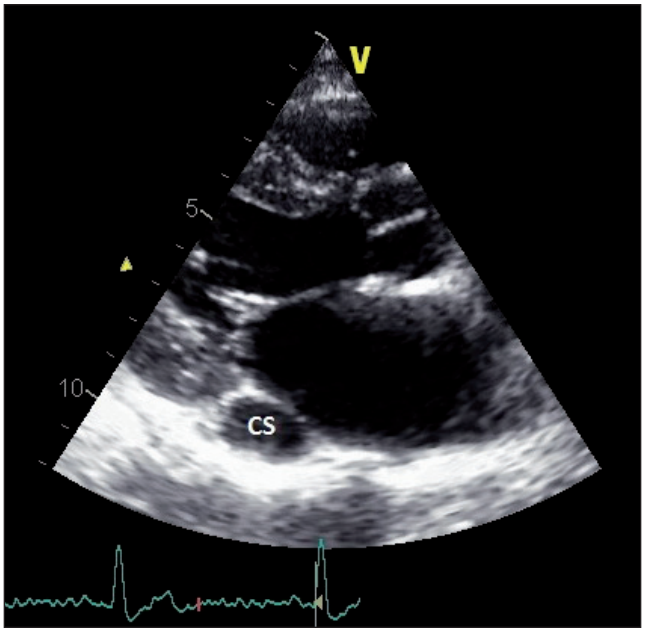

Fig. 1 – Transthoracic echocardiogram, parasternal long axis view, illustrating a dilated coronary sinus.

Then, a TTE with agitated saline was performed using two peripheral venous catheters, one in the left arm and the other in the right arm. After the injection of the agitated saline on the left arm catheter, it was immediately detected by the presence of bubbles in the dilated coronary sinus and only after it filled the right cavities, as observed on a modified 4-chamber view in video 1. Using a tridimensional probe, it was possible to further appreciate these findings as observed on a triplane long-axis view in video 2. In video 3 it is possible to clearly identify the bubbles exiting the coronary sinus and entering the right cavities in a volume-rendered modified 4-chamber view. Performing the same maneuvers but using the right arm catheter, it was evident that the right atrium was filled first, and only then it could be detected in the coronary sinus. Therefore these findings fulfill the criteria for the diagnosis of persistent left superior vena cava.

Conclusion

The presence of a dilated coronary sinus on echocardiography should alert the clinician to the possibility of PLSVC, especially in young patients, without signs of elevated right-sided filling pressures. This diagnosis can be easily achieved using transthoracic echocardiography with agitated saline. Although rare, PLSVC may present severe associated malformations, requiring further cardiologic investigations. This diagnosis is also important due to its implications concerning the placement of catheters in the right heart by the left subclavian route and the administration of retrograde cardioplegia solutions during heart surgery.

Conflict of interest
No conflict of interest.

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Supplementary material
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