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Persistent Left Superior Vena Cava: About a Case and Review of Literature

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ABSTRACT

Persistent left superior vena cava (PLSVC) is a rare and benign congenital malformation. It results when the left superior cardinal vein caudal to the innominate vein fails to regress. It is often asymptomatic and usually diagnosed incidentally while performing imaging. The notation of a dilated coronary sinus on echocardiography should raise the suspicion of PLSVC. We report a rare case of a 56 year-old lady in whom we discover this anomaly because of dyspnea.

Introduction

Persistent of left superior vena cava is the most common congenital thoracic venous anomaly with a prevalence of 0.3-0.5% in general population and 10% of those with congenital heart disease **(1)**. It is most commonly observed in isolation but can be associated with other cardiovascular abnormalities including atrial septal defect, coarctation of aorta, bicuspid aortic valve, cor triatriatum, and coronary sinus ostial atresia. Dilation of the coronary sinus constitutes the main echocardiographic sign leading to suspicion of its presence.

Case report

A 56 year-old-lady, with no cardiac risk factors and had no history of disease, was admitted to the hospital for four weeks history of dyspnea stade III of NHYA and increased volumes of both lower limbs. The cardiovascular examination revealed a systolic ejection murmur in the pulmunary area.

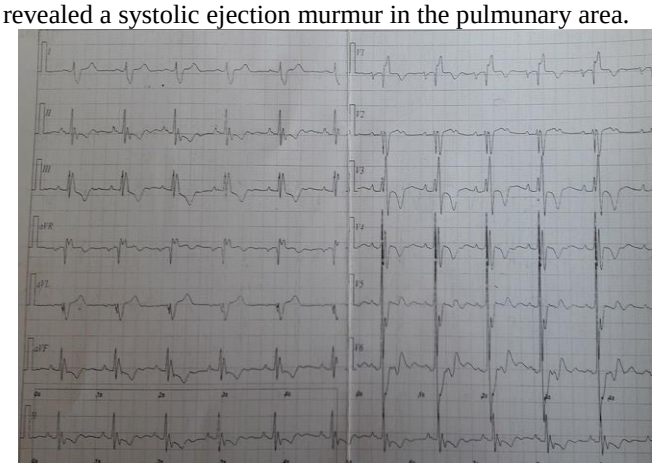

Figure 1. Electrocardiogram.

The electrocardiogram showed complete left brundle branch block type left ventricular hypertrophy and negatives T-waves in V3-V4-V5-V6 leads **(Figure1)**. The echocardio graphy revealed a persistent of left superior vena cava **(Figure 2)** with a dilated coronary sinus and a large ostuim secundum atrial septal defect (19 mm) which continues till the roof of the coronary sinus **(Figure 3 and 4)**.

The left cavities was dilated with pulmonary arterial hypertension of debt (SPAP: 51mmhg).The pumonary venous drainage was normal. Her Computed Tomography (CT) angiogram of chest showed the same with evidence persistent left superior vena cava.

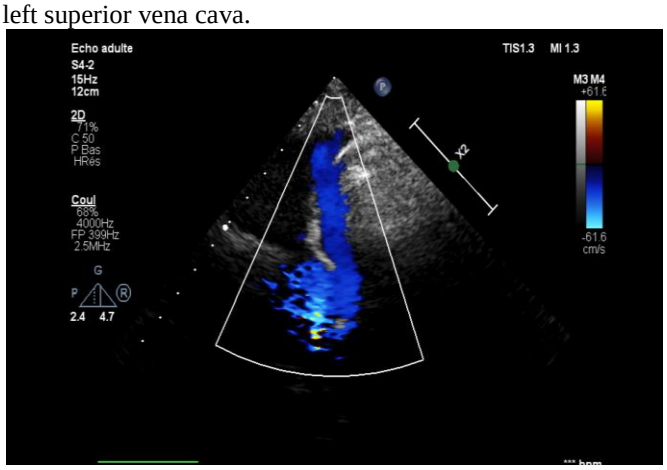

Figure 2. Transthoracic echocardiogram showing the persistent of left superior vena cava.

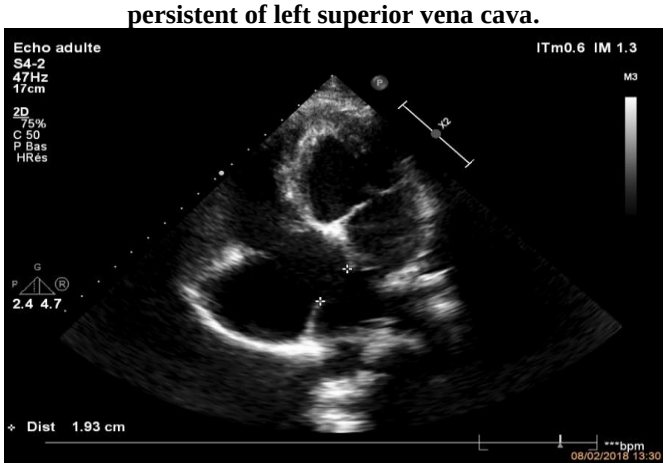

Figure 3. Transthoracic echocardiogram, four cavities view, illustrating a large ostuim secundum atrial septal defect and a dilated coronary sinus.

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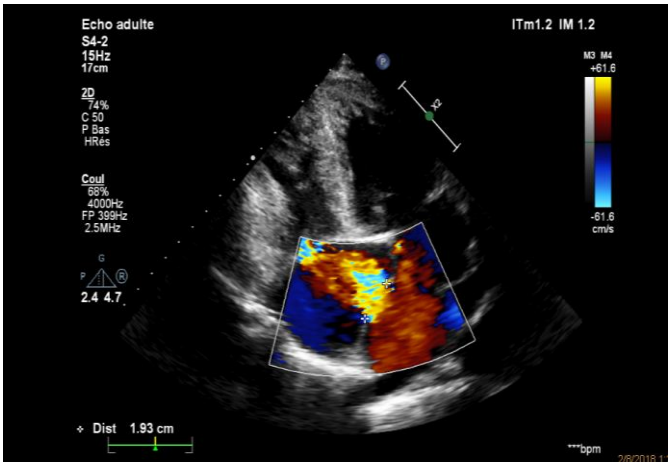

Figure 4. Transthoracic echocardiogram, four cavities view, illustrating a large ostuim secundum atrial septal defect (color doppler).

Discussion

Persistent left superior vena cava is the most common form of anomalous venous drainage involving the superior vena cava. The fetal development of systemic and pulmonary veins is complex and variable. In normal cardiac development, the anterior cardinal veins draining the cephalic portion of the embryo combine with the posterior cardinal veins to form common cardinal veins that drain into the sinus venosus. During the eighth week, an anastomosis develops between the right and left anterior cardinal veins, creating the left brachiocephalic vein. Portions of the right anterior cardinal vein and the right common cardinal vein develop into the RSVC. Part of the left anterior cardinal vein caudal to the brachiocephalic vein regresses, leaving the ligament of Marshall and coronary sinus. In rare cases in which the left anterior cardinal vein fails to regress, a PLSVC forms and usually drains into the right atrium via the coronary sinus **(2,3)**.

This anomaly may be isolated or most often associated cardiac and extracardiac anomalies, as well as with multiple malformation syndromes. The most common associated congential heart abnormalities are atrial septal defect and ventricular septal defect, followed by aortic coarctation, transposition of the great vessels, Tetralogy of Fallot, and anomalous connections of the pulmonary veins **(4,5,6)**. Conversely, the most frequently associated extra-cardiac anomaly is esophageal atresia **(4)**.In our case,the patient has a large ostuim secundum atrial septal defect.

Diagnosis is usually suspected when dilated coronary sinus is noted on transthoracic echocardiography or through an intravenous catheter in the left arm.Other imaging modalities like magnetic resonance imaging, directly visualize the venous anatomy and confirm the diagnostic.

Conclusion

Persistent left superior vena cava is a rare anomaly of systemic venous return. His diagnosis should be suspected if a dilated coronary sinus is visualized by echocardiography and confirmed by cardiac angio-magnetic resonance imaging.

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