Cardiology in the Young

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Persistent left superior vena cava without right superior vena cava during fetal life

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Brief Report

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Abstract

Enlarged coronary sinus detected during cardiac examination in the prenatal period is the first finding that raises suspicion for persistent left superior vena cava. In this report, a patient was presented who was referred with the prediagnosis of cor triatriatum sinister in the antenatal period and diagnosed with isolated persistent left superior vena cava (SVC).

Persistent left superior vena cava (SVC) is the most common variation of the thoracic venous system.[1] It is found in 0.3–0.5% of the general population and 4–8% in those with CHD.[2–5] In most cases, the right and left SVC coexist. Persistent left SVC without the right SVC, also called isolated persistent left SVC, is very rare and occurs in 0.07–0.13% of patients with CHDs with visceroatrial situs solitus. An enlarged coronary sinus detected during cardiac examination in the prenatal period is the first finding that raises suspicion for this anomaly.[6] Diagnosis is made with three vessels view, which is one of the standard cardiac examination sections, and an additional vessel to the left of the pulmonary artery and ductus arteriosus in the tracheal image or indirectly draining into the left or right atrium. The wideness of the coronary sinus may confuse the diagnosis with cor triatriatum sinister during fetal cardiac evaluation. Isolated persistent left SVC is generally considered a benign condition. When present with cardiac and extracardiac anomalies and chromosomal anomalies, perinatal and neonatal outcomes are adversely affected.[7]

Case

In the fetal echocardiographic evaluation performed during the 33rd week of pregnancy in a female patient who was sent from an external centre with the prediagnosis of cor triatriatum sinister, it was observed that the coronary sinus was wide. In the evaluation of the region where the coronary sinus ostium is located, it was observed that the coronary sinus was wide and there was an additional structure to the left of the pulmonary artery in three-vessel view (Video S1). As a preliminary diagnosis, persistent left SVC was suspected alongside cor triatriatum sinister. The diagnosis was confirmed as isolated persistent left SVC which is drained to the coronary sinus by echocardiography performed in the postnatal period and contrast echocardiography performed with agitated saline (Video S2).

Conclusion-discussion

We presented a patient with isolated persistent left SVC diagnosed by fetal echocardiography. Persistent left SVC without right SVC (isolated persistent left SVC) is a very rare venous malformation. During normal fetal development, the left anterior venous cardinal system regresses and lags the coronary sinus and Marshall ligament. Failure of the left anterior cardinal vein to close results in persistent left SVC. Generally, persistent left SVC is associated with right SVC and drains into the right atrium through a dilated coronary sinus. When developmental arrest occurs at an earlier stage, the absence of coronary sinus is seen and the persistent left SVC drains into the left atrium. In the embryonic period, the anterior cardinal veins provide venous drainage of the cephalic region and upper extremity. Except for a small portion forming the left superior intercostal vein, the left anterior cardinal vein regresses by the eighth week of embryogenesis. Failure of this regression results in persistent left SVC as our case.[8]

This venous malformation, either isolated or associated with right SVC, does not itself cause haemodynamic disturbance and is usually diagnosed incidentally. In the study of Ari et al., after the retrospective evaluation of 2663 patients with CHD, between 2005 and 2012, persistent left SVC was found in 88 (3.3%) patients, while isolated persistent left SVC was found in 5 patients (0.001%).[5]

Contrary to most reported cases of isolated persistent left SVC,[9] the case presented herein was a rare condition due to the presence of situs solitus and right SVC agenesis and no associated CHD, and it was diagnosed by differential diagnosis from cor triatriatum in the fetal period.

Persistent left SVC is typically asymptomatic if there is not associated other heart defects. On the other hand, persistent left SVC may cause difficulty in introducing central venous catheters, pacemaker, or defibrillator leads. Also, the use of retrograde cardioplegia, for coronary artery bypass grafting, may cause ineffective.[10] In adition to these, arrhythmias may result from dilatation of the coronary sinus opening, causing stretching of the atrioventricular (AV) node and bundle of His, especially in cases with absent SVC, wherein the coronary sinus acts as the only drainage source for flow from the entire SVC.[11,12] We have been following our patient asymptomatically for 2 years.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951123001014.

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Conflicts of interest. None.

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