

ROLE OF ECHOCARDIOGRAPHY IN SINUS VENOSUS ATRIAL SEPTAL DEFECT COMBINED WITH SYSTEMIC AND PULMONARY VASCULAR DISEASE

JIHUN AHN, MD1, SANG-HO PARK, MD, PhD2, DOHOI KIM, MD, PhD1, TAEHOON KIM, MD1, SEONGSIK JO, MD, PhD3, HYEOKGYU LEE, MD2 and ARA CHO, MD2

1DEPARTMENT OF INTERNAL MEDICINE, SOONCHUNHYANG UNIVERSITY GUMI HOSPITAL, GUMI, KOREA
2DEPARTMENTS OF INTERNAL MEDICINE, 3RADIOLGY, SOONCHUNHYANG UNIVERSITY CHEONAN HOSPITAL, CHEONAN, KOREA

We present a case of persistent left superior vena cava, anomalous right pulmonary venous connection to the right-sided superior vena cava and sinus venosus atrial septal defect detected by computed tomography (CT) pulmonary angiography and echocardiography. These defects were surgically corrected using a double-patch technique. In fact, CT can provide anatomical information about a complex anomaly in the systemic and pulmonary vasculatures. Though modern imaging techniques are useful for detecting complex cardiovascular disease, careful echocardiographic examination should be performed to diagnose complex cardiac anomalies.

KEY WORDS: Atrial septal defect · Pulmonary veins/abnormalities · Computed tomography.
right arm peripheral vein was injected with agitated saline, and normal opacification was noted in the right atrium, followed by opacification of the right ventricle (Fig. 1C). Next, the left arm peripheral vein was injected with agitated saline and abnormal opacification of the coronary sinus was first noted, followed by opacification of the right atrium and then right ventricle (Fig. 1D). On the third day of admission, coronary CT angiography was performed. Coronary CT angiography with 3-dimensional reconstruction demonstrated a more detailed structure of PLSVC drainage into the right atrium via a dilated coronary sinus (Fig. 2D). The patient underwent surgical correction with the double-patch technique. Five

![Fig. 1. Echocardiographic findings in the patient. A: Modified apical 4-chamber view showing markedly dilated right-sided heart chambers and coronary sinus (arrow). B: Transesophageal echocardiography demonstrating sinus venosus atrial septal defect. C: Normal opacification of the right side of the heart following an agitated saline injection through a right brachial vein. D: Early opacification of the abnormally large coronary sinus (arrow) following an agitated saline injection through a left brachial vein. RA: right atrium, SVC: superior vena cava.]

![Fig. 2. Computed tomography (CT) appearance of the patient. A: Coronal reconstruction of the CT pulmonary angiogram (CTA-PA) showing the course of the left superior vena cava (SVC) (arrow). B: Cross-sectional CTA-PA displaying the right superior pulmonary vein draining into the right SVC (arrow). C: CTA-PA showing atrial septal defect (arrow). D: Three-dimensional reconstruction of the coronary CT angiogram demonstrating more detailed structural information of the drainage of persistent left superior vena cava (arrow) into the right atrium via a dilated coronary sinus. CS: coronary sinus, RA: right atrium, RV: right ventricle.]

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months later, follow-up transthoracic echocardiography showed near-normalization of the right ventricular and coronary sinus size.

**DISCUSSION**

PLSVC occurs in approximately 0.3-0.5% of the general population and characteristically drains into the coronary sinus. During and after embryonic development of SVC, SVC develops on the right side from a portion of the right anterior cardinal vein. On the left side, part of the left anterior cardinal vein undergoes normal regression to form the ligament of the left vena cava. PLSVC results from the persistence of the left anterior cardinal vein. Usually, PLSVC is asymptomatic and discovered incidentally during imaging study and pacemaker implantation or central catheterization but sometimes their elucidation is crucial especially during cardiovascular surgery. PLSVC should be considered whenever a dilated coronary sinus is identified at echocardiography and the diagnosis could be confirmed by saline contrast echocardiography. Other modern imaging modalities such as CT or magnetic resonance imaging (MRI) can be used to confirm the diagnosis. In our case, we could not consider the presence of PLSVC before performing CT pulmonary angiography just because of the focus on volume overload of right-sided heart chambers. We performed a contrast echocardiography based on the information obtained from CT pulmonary angiography.

In our case, PAPVC associated with SVD was also found. PAPVC is frequently associated with congenital heart disease such as an ASD. It is estimated that 10-15% of patients have an ASD and approximately 85% of PAPVC are associated with SVD. Usually, the diagnosis of PAPVC can be made by echocardiography, and cardiac catheterization along with angiography is often performed for confirmation of the diagnosis. Nowadays, CT, MRI and TEE with contrast examination are considered as sensitive methods for the detection of PAPVC. In our case, the diagnosis of PAPVC was missed on routine TTE, and a definitive diagnosis of PAPVC could be made on CT pulmonary angiography.

Some authors have previously reported a combined anomaly of systemic and pulmonary venous return associated with SVD. These authors especially emphasized the importance of new imaging modalities in diagnosing complex anomaly of systemic and pulmonary venous return associated with SVD. Also in our case, CT made a definitive diagnosis of the anomaly and provided more detailed structural information. However, this case report also illustrated that careful echocardiographic examination should be performed using several windows and even contrast for diagnosing the anomalies of systemic and pulmonary venous return combined with congenital heart disease. And consideration is required when the case has accompanying cardiac abnormalities besides PLSVC such as an ASD, when coronary sinus is extremely dilated and it is coexisting with enlargement of right ventricle.

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