Atresia of Coronary Sinus Ostium with a Persistent Left Superior Vena Cava Demonstrated on Cardiac Computed Tomography Angiography

Li Zhang[1], Zi-Xian Chen[2], Wei Tian[2], Xin-Ling Yang[3], Shun-Lin Guo[2]

[1]Department of the First Clinical Medical College, Lanzhou University, Lanzhou, Gansu 730000, China
[2]Department of Radiology, The First Hospital of Lanzhou University, Lanzhou, Gansu 730000, China
[3]Department of Magnetic Resonance Imaging, Cardiovascular Imaging and Intervention Center, Fuwai Hospital, State Key Laboratory of Cardiovascular Disease, Beijing 100037, China

To the Editor: A 43-year-old man diagnosed with dilated cardiomyopathy 6 years prior visited our institution recently because of worsened chest congestion with shortness of breath and abdominal distension. The patient had a history of diabetes. His electrocardiography (ECG) demonstrated a sinus rhythm with an intraventricular conduction disturbance, an atypical left bundle branch block, and a QRS interval of 160 ms. Echocardiographic findings were consistent with dilated cardiomyopathy [bicentric type, Figure 1a], in which the left atrial diameter was 44 mm, the left ventricular end-diastolic diameter was 78 mm, and the left ventricular ejection fraction was 30%. The final diagnosis was New York Heart Association Grade III dilated cardiomyopathy. To achieve a good prognosis, improve exercise tolerance, and prevent sudden death, cardiac resynchronization therapy (CRT) with defibrillator implantation was advised. However, coronary sinus (CS) abnormalities were observed during the implantation operation. Therefore, it was forced to stop the operation and a cardiac computed tomography angiography (CTA) evaluation of the abnormalities was arranged.

Cardiac CTA was performed with a dual-source computed tomography (CT) system (SOMATOM Definition Flash; Siemens Healthcare, Forchheim, Germany). Retrospective ECG-gated sequential CT scanning was performed according to the following protocol: detector collimation, 128.0 mm × 0.6 mm; slice thickness, 0.75 mm; reconstruction interval, 0.4 mm; rotation time, 0.28 s; tube voltage, 100 kV; and tube current, 304 mA. A contrast agent (lopamiro® 370; 370 mg iodine/ml, Bracco Sine Pharmaceutical Corp. Ltd., Shanghai, China) was injected intravenously at a flow rate of 4 ml/s through the right cubital vein, followed by a 50-ml saline flush at the same injection rate. The examination was performed with the patient in the supine position and holding his breath. To optimize the scan for venous visualization, the scan started automatically 2 s after the contrast attenuation in the ascending aorta reached 100 Hounsfield units (HU). Three-dimensional volume rendering and maximum intensity projection reconstruction were performed on a workstation (GE ADW 4.6; GE Healthcare, Milwaukee, WI, USA) to obtain general information regarding the anatomy of the cardiac veins.

Cardiac CTA demonstrated an abnormal vessel traversing between the left innominate vein and the CS [Figure 1b and 1d]; then, the blind end of the CS from the right atrium was recognized [Figure 1c and 1d]. The vein between the CS and left brachiocephalic vein is called a persistent left superior vena cava (PLSVC), which finally drained into the right atrium. No other congenital abnormality was identified during this CTA examination such as abnormal coronary vein drainage into the left atrium or an atrial septal defect. Targeted left ventricular lead implantation was performed by atrial transseptal puncture. After this operation, the patient's terminal QRS interval was 110 ms on ECG, the postoperative course was unremarkable, and the patient is currently doing well.

Atresia of the CS ostium (ACSO) with a PLSVC is a rare abnormality.[1] This abnormality can occur without clinical symptoms or significant cardiac dysfunction and is usually ignored by clinicians and radiologists. Current evidence suggests that stimulating the lateral region of the left ventricle provides the greatest benefit in CRT. In CRT, left ventricular pacing is generally achieved by positioning the left ventricular lead in one of the tributaries of the CS, and the CS is a common passageway for this operation. Preoperative identification of the patient's CS anatomy and abnormalities is of critical importance when planning and performing transvenous interventions.

To identify CS abnormalities, interventional coronary angiography (ICA), CTA, and transthoracic echocardiography (TTE)[2] can be performed. TTE assesses a lesion after the injection of agitated

Access this article online
Quick Response Code:
Website: www.cmj.org
DOI: 10.4103/0366-6999.237407

Address for correspondence: Prof. Shun-Lin Guo, Department of Radiology, The First Hospital of Lanzhou University, Lanzhou, Gansu 730000, China
E-Mail: guoshunlin@msn.com

Received: 11-04-2018 **Edited by:** Yuan-Yuan Ji
How to cite this article: Zhang L, Chen ZX, Tian W, Yang XL, Guo SL. Atresia of Coronary Sinus Ostium with a Persistent Left Superior Vena Cava Demonstrated on Cardiac Computed Tomography Angiography. Chin Med J 2018;131:1882-3.

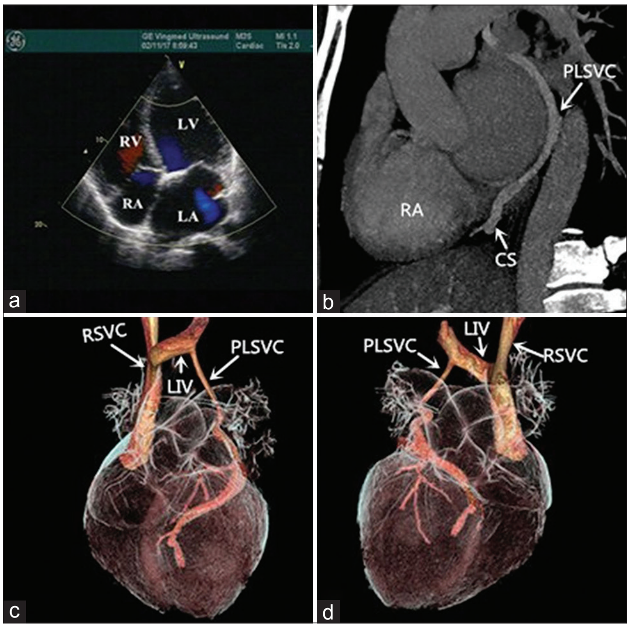

Figure 1: (a) Ultrasonography: Four-chamber view showing global enlargement of heart. (The length/transverse diameter of right atrium, left atrium, right ventricle, and left ventricle are 63/46 mm, 60/49 mm, 75/52 mm, and 85/61 mm, respectively.) (b) Maximum intensity projection images of CT show blind end of the CS from RA and the PLSVC runs on the lateral edge of the left ventricle. (c and d) 3D VR of CT shows an abnormal communicating vessel from the LIV to CS. CT: Computed tomography; CS: Coronary sinus; RA: Right atrium; PLSVC: Persistent left superior vena cava; 3D: Three dimensional; VR: Volume rendering; LIV: Left innominate vein.

saline (air-filled microbubbles are created by shaking the saline solution in a syringe), but the accuracy of the ultrasonography diagnosis depends on the operator's experience and examination skills. Moreover, ultrasonography cannot visualize the entire course of a PLSVC because of acoustic window limitations and obstructions caused by bones and gases. Compared with ICA, CT is a noninvasive technique that conveys no risk of damage to the CS, requires less contrast agent, and the examination duration is shorter. However, this shorter duration necessitates tighter contrast enhancement protocols, and venous opacification may be insufficient using scanning protocols optimized for imaging the coronary artery system. Therefore, a longer duration may be required between contrast injection and scanning initiation to visualize the cardiac veins.

The scanning protocol in our case was designed such that image acquisition commenced 2 s after reaching the threshold of 100 HU, and we acquired the best images of the coronary venous system during the systolic phase. Despite this adjustment, veins other than the proximal coronary veins (the CS, middle cardiac vein, small cardiac vein, and posterior vein) were not visualized optimally. It has been reported in the literature that contrast injection via the left arm would have demonstrated ACSO and the anomalous left superior vena cava more clearly.[3]

In summary, CTA is a noninvasive examination for CS abnormalities, but the design of the scanning protocol is crucial; a successful examination also depends on the experience of the radiologist.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understand that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Acknowledgments

The authors would like to thank the Department of Magnetic Resonance Imaging, Fuwai Hospital, State Key Laboratory of Cardiovascular Disease for providing the reconstruction workstation (GE ADW 4.6) for volume rendering and maximum intensity projection CT images.

Financial support and sponsorship

This work was partly supported by a grant of the First Hospital of Lanzhou University Science Foundation (No. ldyyyn 2015-06).

Conflicts of interest

There are no conflicts of interest.

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