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GIGANTIC CORONARY SINUS ASSOCIATED WITH CONCURRENT PERSISTENT LEFT SUPERIOR VENA CAVA AND RIGHT VENTRICULAR VOLUME OVERLOAD

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Case Presentation

A 76-year-old woman with known atrial fibrillation and congestive heart failure presented with increasing shortness of breath. A 2-dimensional (2-D) transthoracic echocardiogram was performed to assess left ventricular function. An incidental finding of a very large coronary sinus with a diameter of 4.8 cm was seen, raising a suspicion for the possibility of a persistent left superior vena cava (PLSVC) (Figure 1). Additional pertinent positive findings included a massively dilated right atrium (estimated volume: 538 mL), dilated tricuspid annulus with poor leaflet coaptation, severe tricuspid regurgitation, and pulmonary artery systolic pressure (PASP) of 50 mmHg with an estimated mean right atrial pressure (RAP) of 25 mmHg. After agitated saline administration into the left brachial vein, there was immediate and sequential opacification of the dilated coronary sinus, right atrium, and right ventricle, confirming the presence of a PLSVC (Figure 2). CT angiography provided detailed anatomical and morphological characterization demonstrating drainage of the PLSVC into the gigantic coronary sinus and right-sided cardiac chambers and absence of other vascular or congenital anomaly (Figures 3 and 4).

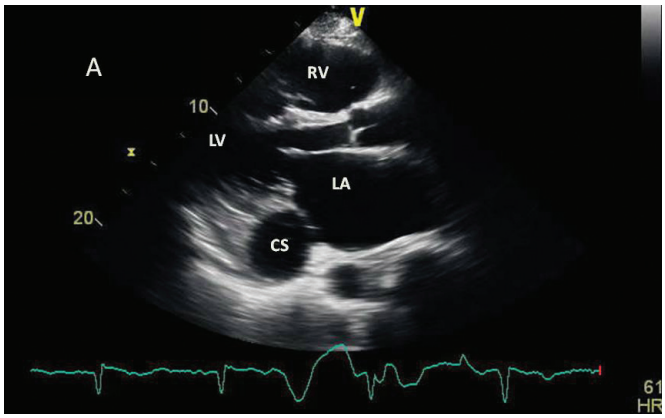


Figure 1. A parasternal long axis view revealing a dilated coronary sinus (CS).

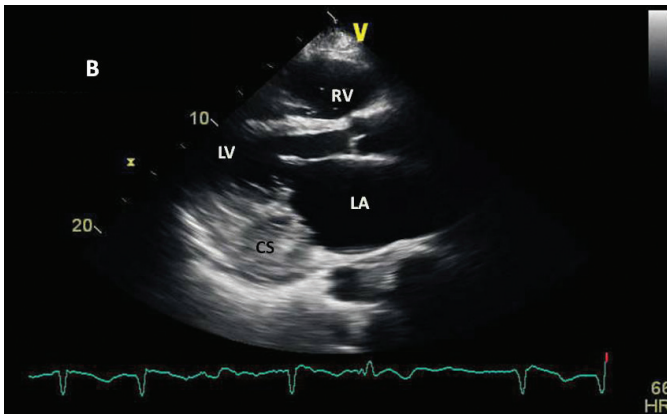
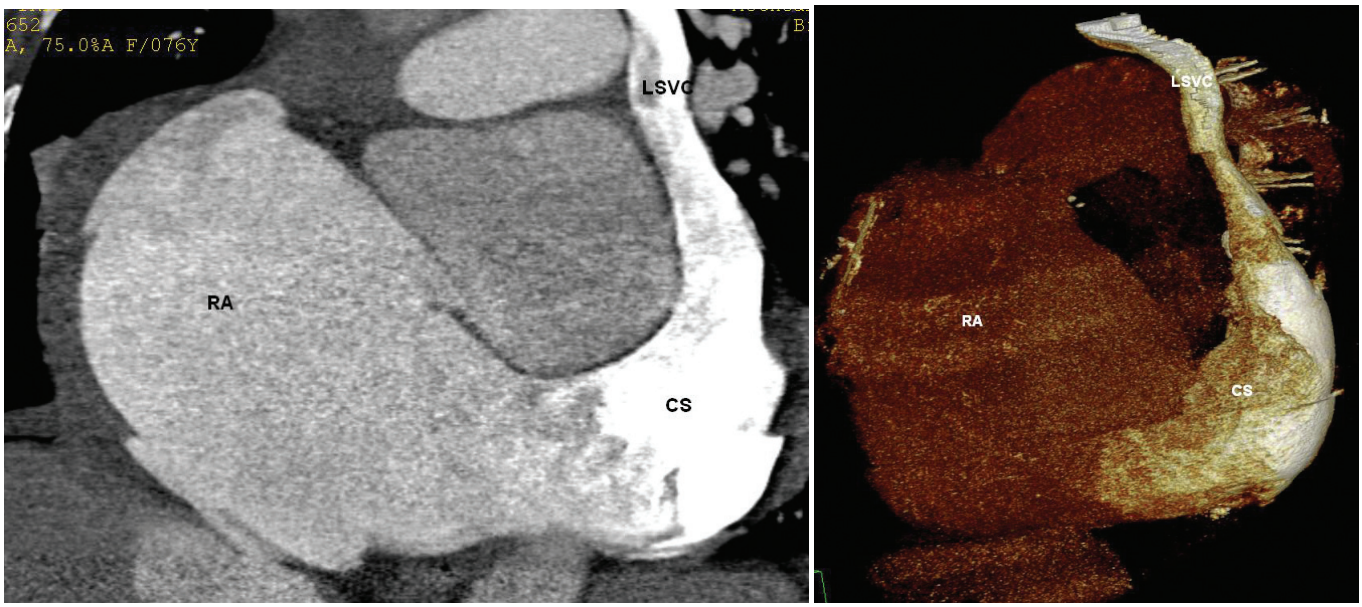


Figure 2. Injection of agitated saline via the left brachial vein shows contrast appearing first in the coronary sinus (CS), then right atrium and right ventricle consistent with the diagnosis of PLSVC.



Figures 3 and 4. Multiplanar reformat and 3-dimensional reconstruction CT angiography confirming the presence of PLSVC after contrast injection through the left brachial vein.

Comment

PLSVC is a very rare congenital remnant with a prevalence of 0.05-0.5% in the general population.[1, 2] Nevertheless, it is considered the most common thoracic vein anomaly and is more frequently encountered in conjunction with other congenital heart diseases such as Tetralogy of Fallot, with prevalence rates approaching 2.1% to 4.3%.[3, 4] It results when the caudal portion of the left anterior cardinal vein fails to regress. A PLSVC usually drains into the right atrium through a dilated coronary sinus and does not alter the normal systemic venous circulation.

Although PLSVC is the most common finding associated with a dilated coronary sinus, other differential diagnoses include a partially unroofed coronary sinus; pulmonary venous connection to the coronary sinus; coronary artery fistula; partial anomalous hepatic venous connection to the coronary sinus; and continuity of IVC with SVC through a hemiazygous vein.[3, 5] Furthermore, coronary sinus dilatation is commonly observed in pulmonary hypertension and is directly associated with elevated PASP and RAP, as well as with dilated right-sided chamber size and IVC diameter.[6–8] Our case illustrates that the presence of concurrent PLSVC and pulmonary hypertension likely contributed to the marked dilatation of the coronary sinus.

References

1. Pahwa R, Kumar A. Persistent left superior vena cava: an intensivist's experience and review of the literature. South Med J. 2003 May;96(5):528-9.
2. Snider AR, Ports TA, Silverman NH. Venous anomalies of the coronary sinus: detection by M-mode, two-dimensional and contrast echocardiography. Circulation. 1979 Oct;60(4):721-7.
3. Tak T, Crouch E, Drake GB. Persistent left superior vena cava: incidence, significance and clinical correlates. lnt J CardioI. 2002 Jan;82(1):91-3.
4. Nsah EN, Moore GW, Hutchins GM. Pathogenesis of persistent left superior vena cava with coronary sinus connection. Pediatr Pathol. 1991 Mar-Apr;11(2):261-9.
5. Chen MC, Hung JS, Chang KC, Lo PH, Chen YC, Fu M. Partially unroofed coronary sinus and persistent left superior vena cava: intracardiac echocardiographic observation. J Ultrasound Med. 1996 Dec; 15(12):875-9.
6. Mahmud E, Raisinghani A, Keramati S, Auger W, Blanchard DG, DeMaria AN. Dilation of the coronary sinus on echocardiograrn: prevalence and significance in patients with chronic pulmonary hypertension. J Am Soc Echocardiogr. 2001 Jan;14(1):44-9.
7. Gunes Y, Guntekin U, Tuncer M, Kaya Y, Akyol A. Association of coronary sinus diameter with pulmonary hypertension. Echocardiography. 2008 Oct;25(9):935-40. Epub 2008 Sep 2.
8. Isaacs D, Hazany S, Gamst A, Stark P, Mahmud E. Evaluation of the coronary sinus on chest computed tomography in patients with and without pulmonary artery hypertension. J Comput Assist Tomogr. 2009 Jul-Aug;33(4):513-6.