ESC European Society of Cardiology

European Heart Journal - Case Reports (2018) **2**, 1–3
doi:10.1093/ehjcr/yty092

Venous varices of the heart: a case report of spontaneous coronary sinus thrombosis with persistent left superior vena cava

Melissa Y. Y. Moey[1]*, Emanuel Ebin[2], and Constantin B. Marcu[2]

[1]Department of Internal Medicine Vidant Medical Center/East Carolina University, Greenville, NC, USA; and [2]Department of Cardiology, Vidant Medical Center/East Carolina University, Greenville, NC, USA

Received 20 April 2018; accepted 27 July 2018; online publish-ahead-of-print 16 August 2018

Case summary

Absence of coronary sinus (CS) ostium and presence of a persistent superior left vena cava (PLSVC) are rare congenital anomalies.[1] We report a case of spontaneous CS thrombosis with PLSVC associated with new-onset atrial fibrillation (AF) in the absence of recent coronary intervention.

Case description

A 72-year-old African American male patient with a history of alcohol abuse presented with acute encephalopathy and failure to thrive (FTT) with unintentional weight loss. His electrocardiogram showed new-onset AF. As part of his initial work up for FTT, he underwent computed tomography (CT) scan of the chest and abdomen with an incidental finding of congenital absence of the CS ostium resulting in retrograde cardiac venous blood flow through the PLSVC to the left innominate/brachiocephalic vein, into the (right) superior vena cava and back into the right atrium (*Figure 1*). A large thrombus measuring 2.5 cm in its largest dimension and extending into the PLSVC was detected in the coronary sinus (*Figures 1* and *2*). Transthoracic echocardiogram (TTE) confirmed the findings of CS thrombus with engorged cardiac veins (*Figure 3*). Due to high operative risk, the patient was started on anticoagulation instead of thrombectomy

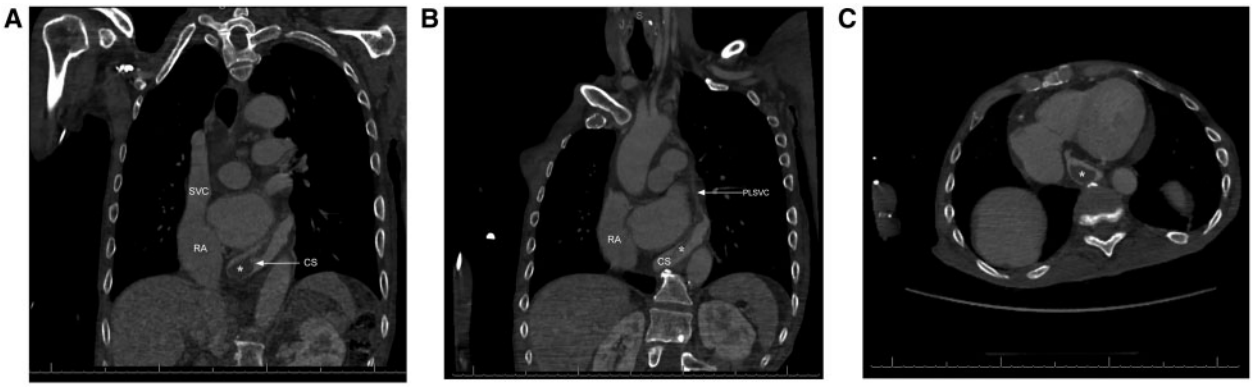

Figure 1 Computed tomography chest, coronal views (*A*,*B*), and axial view (*C*). A large thrombus (asterisk) (measuring up to 2.5 cm) is seen within the cardiac vein in the expected location of the coronary sinus which does not drain into the right atrium. A thin thrombus tract extends into a persistent left superior vena cava. Asterisk indicates the coronary sinus thrombus. CS, coronary sinus PLSVC, persistent left superior vena cava; RA, right atrium.

* Corresponding author. Tel: 252-744-3229, Fax: 252-744-3987, Email: moeym16@ecu.edu. This case report was reviewed by Riccardo Liga and Francesca Musella.

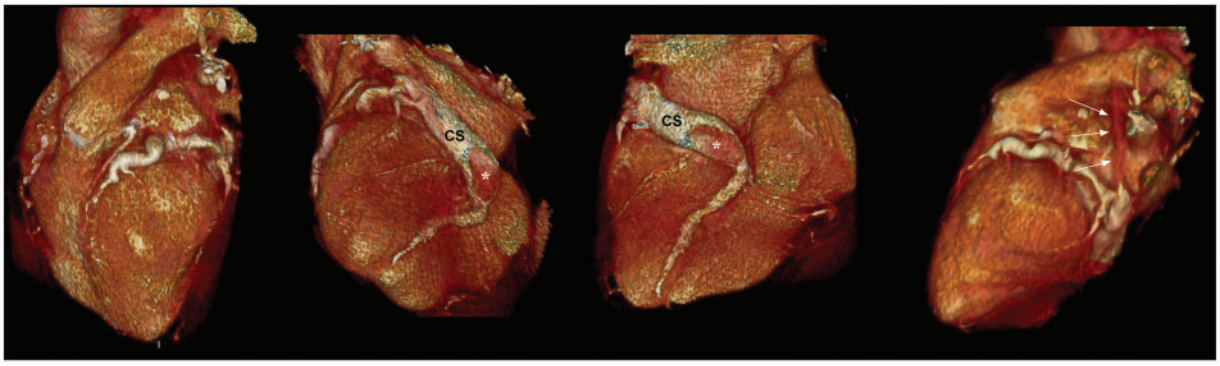

Figure 2 Volume rendering technique images of the heart and cardiac vein anatomy (*A*–*C*). Engorged cardiac veins with presence of a spontaneous large thrombus measuring up to 2.5 cm. Asterisk indicates the coronary sinus thrombus and arrows point to the persistent left superior vena cava. CS, coronary sinus.

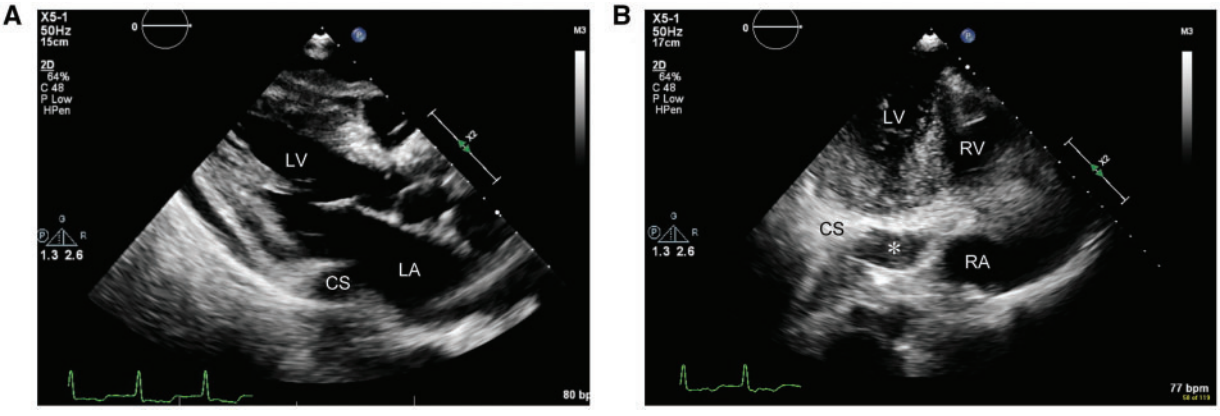

Figure 3 Two-dimensional echocardiography of (*A*) parasternal long view and (*B*) apical four chamber view. (*A*) Parasternal long axis view demonstrates dilated coronary sinus and small <1 cm pericardial effusion. (*B*) Targeted apical four chamber view demonstrating dilated coronary sinus occupied by thrombus (*). CS, coronary sinus; LA, left atrium; LV, left ventricle; RA, right atrium.

and eventually discharged to a nursing facility. Six months following his initial diagnosis, he presented with right upper quadrant abdominal pain where he was found to have a large pericardial effusion on CT chest, abdomen, and pelvis. As the CT image was performed without contrast, the thrombus was not visualized, though CS dilatation was still observed. A repeat echocardiogram noted the dilated coronary sinus and persistent thrombus. The patient had features of cardiac tamponade requiring pericardiocentesis with fluid studies suggestive of an effusive-constrictive disease. He continued to decompensate and his family opted for comfort measures.

Persistent superior left vena cava results as failure of obliteration of the left common cardinal vein during embryological development of the coronary venous system resulting in connection with the CS via the vein of Marshall. Dilation of the CS seen on routine TTE leads to suspicion for PLSVC.[1] To confirm this finding on TTE, criteria include (i) the presence of CS dilatation on 2D echocardiography in the absence of right-sided elevated filling pressures, (ii) enhancement of the CS before the right atrium (RA) after contrast infusion into a left arm vein, and (iii) normal transit of contrast with RA opacification before the CS with contrast injection in the right arm vein.[1] Other imaging modalities to rule out additional anomalous findings and delineate coronary anatomy include CT and magnetic resonance venography.

While this congenital anomaly is usually asymptomatic, it is possible that our patient's AF may have been due to the presence of a PLSVC. In a small study of five patients with symptomatic medication-resistant AF, catheter ablation of the PLSVC resulted in successful conversion and maintenance of sinus rhythm at 15-month follow-up.[2] The presence of spontaneous CS thrombosis in our patient without any prior intervention is unique. In a histological analysis from necropsied hearts, a cuff of striated muscle from the ostium was seen in the wall of the CS that inserted distally in the left atrium.[3]

As such, it is plausible that in the setting of AF with an absent CS ostium and contraction, venous stasis led to formation of thrombus at the terminal end of the CS.

Treatment for CS thrombosis may include thrombectomy to prevent embolization into the pulmonary circulation or alternatively medical management with anticoagulation alone. Despite significant clot burden with extension ascending into the PLSVC, our patient was considered a very poor surgical and interventional candidate. There are no current studies outlining anticoagulation choice for CS thrombosis. The patient was initially started on warfarin for CS thrombosis and new-onset AF. He was discharged to a nursing home where he was transitioned to rivaroxaban. We suspect this was for ease of administration and monitoring in the setting of normal kidney and liver function.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

References

1. Goyal SK, Punnam SR, Verma G, Ruberg FL. Persistent left superior vena cava: a case report and review of literature. *Cardiovasc Ultrasound* 2008;**6**:50.
2. Hsu LF, Jais P, Keane D, Wharton M, Deisenhofer I, Hocini M, Shah DC, Sanders P, Scavee C, Weerasooriya R, Clementy J, Haissaguerre M. Atrial fibrillation originating from persistent left superior vena cava. *Circulation* 2004;**109**:828–832.
3. Chauvin M, Shah DC, Haissaguerre M, Marcellin L, Brechenmacher C. The anatomic basis of connections between the coronary sinus musculature and the left atrium in humans. *Circulation* 2000;**101**:647–652.