

“Cardiac Stomach”: A Giant Coronary Sinus with Persistent Left Superior Vena Cava and Severe Tricuspid Regurgitation

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Keywords: Giant coronary sinus; Tricuspid regurgitation; Mitral stenosis; Persistent left superior vena cava; Cardiac stomach

Abbreviations:

ASD: Atrial Septal Defect; CS: Coronary Sinus; CT: Computed Tomography; MS: Mitral Stenosis; MVR: Mitral Valve Replacement; PLSVC: Persistent Superior Left Vena Cava; RA: Right Atrium; RV: Right Ventricle; TR: Tricuspid Regurgitation; TEE: Transesophageal Echocardiogram; TTE: Transthoracic Echocardiogram; TVR: Tricuspid Valve Repair

Presentation

A young patient with a history of congenital heart disease may harbor other undiscovered cardiac anomalies, whose consequences

may surface many years later. After correction of the initial defect, if these patients present with further symptoms during chronic follow-up, they should be met with a high clinical index of suspicion and worked up for associated cardiac anomalies. In our case, a 39 year-old female with history of rheumatic heart disease, hypertension, diabetes, and hyperlipidemia presented with progressive dyspnea on exertion and lower extremity edema. Her past surgical history was notable for surgical repair of a congenital atrial septal defect (ASD) 17 years ago. On physical examination, she had a 3/6 holosystolic murmur along the left sternal border, as well as a soft diastolic murmur heard best at the apex. She had crackles in her lungs bilaterally. In addition, she exhibited elevated jugular venous pressure and 2+ pitting edema in her lower extremities bilaterally. Based on this presentation, she was diagnosed with congestive heart failure and admitted for further assessment and treatment.

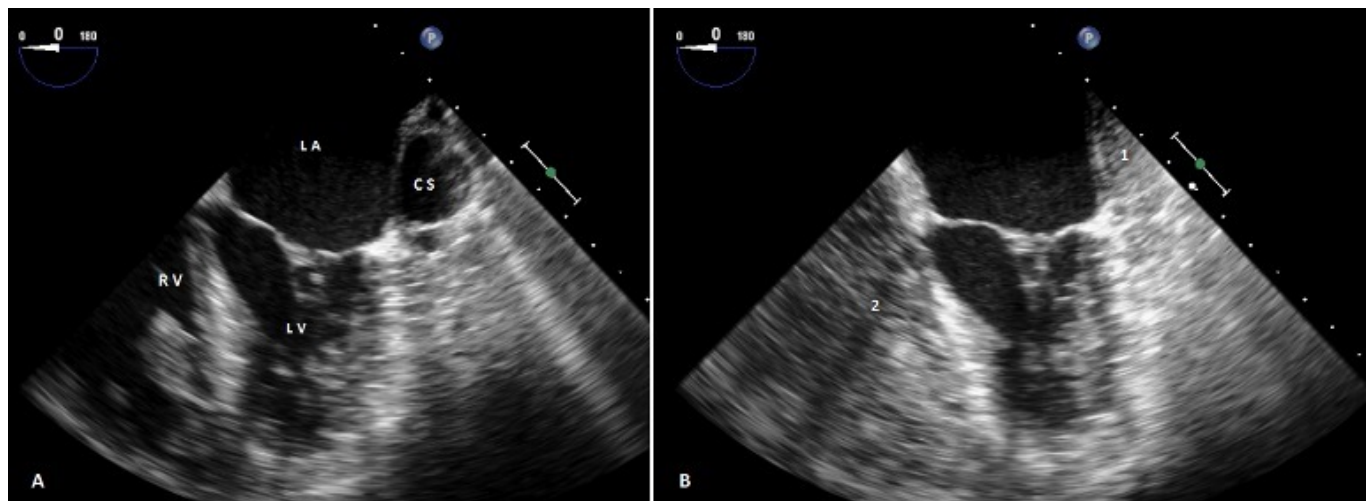


Figure 1: TEE with left-sided venous bubble injection. (A) Four chamber TEE view at zero degrees showing a dilated CS (CS) and its relationship to surrounding cardiac structures at endsystole. (B) Result of left-sided venous bubble injection, showing bubble opacification, first of the CS (1), and then bubble opacification of the right ventricle (2). This is consistent with a PLSVC.

Assessment

During hospitalization, she underwent a transthoracic echocardiogram (TTE) which revealed an ejection fraction of 50%, severe mitral stenosis (MS), severe tricuspid regurgitation (TR), right ventricular (RV) dilatation and RV systolic dysfunction. No other valvular abnormalities were noted. Furthermore, her coronary sinus (CS) was noted to be dilated, with the TR jet directed towards its opening into the right atrium. A left-sided venous bubble study was

conducted, and demonstrated bubble opacification in the CS before the right atrium (RA), consistent with a PLSVC. A transesophageal echocardiogram (TEE) was conducted, and demonstrated a similar finding (Figure 1). A computerd-tomography (CT) scan of her chest (with contrast) significant only for confirmed a persistent left superior vena cava (PLSVC) and a giant CS, draining into the RA (Figure 2). Given the severity of her valvular disease, mitral valve replacement (MVR) and tricuspid valve repair (TVR) surgery were recommended (without concurrent, PLSVC repair). Subsequently, she underwent a

pre-operative cardiac catheterization. During right heart catheterization via the left internal jugular vein, a subtraction venogram confirmed again demonstrated the presence of a PLSVC and a giant coronary sinus (mimicking the shape of a stomach behind the cardiac silhouette), with drainage into the RA (Figure 3, Video 1). No significant coronary stenoses were noted.

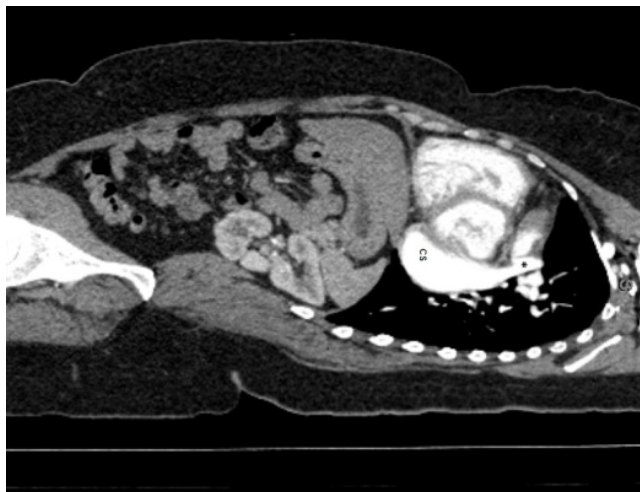


Figure 2: Computed Tomography of chest and abdomen with contrast. Sagittal view showing persistent left superior vena cava (*) draining into a dilated coronary sinus (CS).

Diagnosis

Our patient had a known ASD, and a PLSVC that was discovered after our evaluation. Her valvular dysfunction was felt to be the result of long-standing rheumatic heart disease. The combination of severe MS, severe TR and PLSVC resulted in a giant CS with the unique angiographic appearance of a stomach ("cardiac stomach"). A PLSVC is a rare anomaly, with an incidence is 0.3% in the general population and 4.4% in patients with congenital heart disease [1-3]. In nearly 40% of patients, a PLSVC is accompanied by other cardiac anomalies, such as atrial septal defect, bicuspid aortic valve, coarctation of aorta, coronary sinus ostial atresia, and cor triatriatum [4,5]. If clinically suspected, a TTE with a left-sided venous bubble injection can be used to pursue the diagnosis of a PLSVC (Figure 1, Video 1). The appearance of bubbles in the CS prior to the RA is consistent with a PLSVC. The diagnosis may also be made using other imaging modalities such as CT scan or cardiac catheterization (as demonstrated above).

Management

The majority of PLSVC are found to drain into the CS (and subsequently into the RA) and have no hemodynamic consequences [6]. The long term clinical course of such patients is usually uneventful in the absence of other congenital heart defects. In patients with accompanying congenital heart disease, their long term course is mostly influenced by particular congenital defect(s) [7]. The development of symptoms related to cardiac arrhythmia via secondary stretching of the conduction tissue surrounding the dilated CS has also been reported [8,9]. However, Should a PLSVC drain directly into the

left atrium (LA), or into an unroofed CS (such that there anomalous connection with the LA), a hemodynamically significant right-to left shunt may form. Such a shunt may cause significant systemic desaturation and place the patient at risk for a paradoxical embolic stroke. Surgical repair of the PLSVC may be offered if a patient suffers complications due to a right-to-left shunt. The surgical techniques used to repair a PLSVC are well described, and are specific to the patient's particular anatomical defect [10]. More recently, some authors have described successful percutaneous closure of PLSVC using the Amplatzer vascular plug device as well [11]. Our patient underwent a successful MVR with a 29 mm Medtronic ATS mechanical valve and TVR with a 27 mm flexible band. The PLSVC was not surgically repaired. Following discharge from the hospital, she demonstrated marked improvement in symptoms on postoperative follow-up.

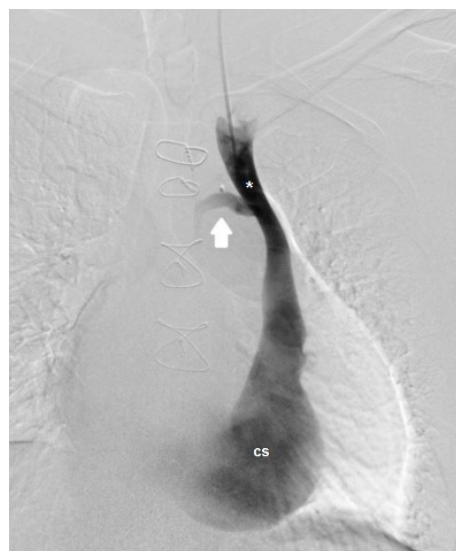


Figure 3: Cardiac Catheterization. Angiographic image (AP view) demonstrating a persistent left superior vena cava (*) draining into a dilated coronary sinus (CS). Here the left brachiocephalic vein is also seen connected to the persistent left superior vena cava (arrow).

Conclusion

Our patient had a known ASD, and a PLSVC that was discovered after our evaluation. Her valvular dysfunction was felt to be the result of long-standing rheumatic heart disease. The combination of severe MS, severe TR and PLSVC resulted in a giant CS with the unique angiographic appearance of a stomach ("cardiac stomach").

This case highlights that patients with congenital heart defects, may harbor other undiscovered defects. In our case, a finding of a dilated coronary sinus in patient with a prior ASD prompted further work up and discovery of a PLSVC. Although her clinical presentation on this occasion was not directly related to the giant CS or the PLSVC, their discovery may have practical implications for her in future clinical settings, especially if she were ever to require a left-sided central venous line, a swan-ganz catheter, an electronic pacemaker, or an implantable defibrillator.

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