Ginormous Coronary Sinus with Persistent Left Superior Vena Cava

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Abstract

An echocardiogram is the most common diagnostic imaging done in chest pain evaluation workup. Persistent left superior vena cava is a rare thoracic congenital anomaly but commonly reported one in the literature. Dilated coronary sinus on echocardiogram is a signal to think of persistent left superior vena cava which can be presented with atypical chest pain and palpitation but is often asymptomatic. As a catheter from left subclavian approach can incidentally advance to the right side of the heart, early awareness of PLSVC is crucial to prevent unwanted procedure related complications.

Keywords: Chest pain; Persistent Left Superior Vena Cava; Congenital venous anomaly

Case Report

A 54-year-old man with a history of hypertension was referred to cardiology clinic for the evaluation of left sided atypical chest pain and palpitation. On examination, no significant physical findings were found with a pulse rate 65/min and blood pressure of 160/90 mmHg. Further investigation showed normal results of complete blood count, basal metabolic profile, thyroid and liver function test. Electrocardiogram (EKG) showed normal sinus rhythm with left atrial enlargement (Figure 1). Transthoracic echocardiography (TTE) demonstrated mild concentric left ventricular hypertrophy with low-normal systolic function (EF ~ 54%) and dilated coronary sinus (CS), approximately 2.3 cm x 2.4 cm in the posterior atrioventricular groove in left parasternal long axis and apical 4-chamber views (Figure 2). Since dilated CS is a relatively rare finding, the differential diagnosis should include right-sided heart disease with pressure and volume overload, unroofed coronary sinus, and anomalous venous drainage into the coronary sinus. A TTE with agitated normal saline contrast injected into the left antecubital vein verified the presence of air bubbles entering the coronary sinus first and then in the right atrium, suggestive of a persistent left superior vena cava (PLSVC) draining directly into the coronary sinus (Figure 3).

Discussion

PLSVC has been recognized as a rare venous anomaly but the most commonly reported congenital thoracic venous anomaly with a prevalence of 0.3% to 0.5% of general population [1]. During the early stage of fetal life, the thoracic venous system is mainly composed of anterior and posterior cardinal veins. While the proximal part of left cardinal vein combines with right anterior cardinal vein remaining as the SVC, the distal portion usually dissipates, forming a ligament of Marshall. However, a failure of the left cardinal vein degeneration leads to the persistent left SVC. Saline agitated TTE (bubble study) is the gold standard to confirm the diagnosis. The following TTE findings are part of diagnostic criteria: (1) the presence of a dilated coronary sinus in the absence of evidence of elevated right-sided filling pressures; (2) enhancement of the dilated coronary sinus before the right atrium

Figure 1: Electrocardiogram shows normal sinus rhythm with broad and bifid P wave in all leads.
As PLSVC is a congenital disorder, up to 40% of PLSVC can be associated with other anomalies including atrial septal defect, coarctation of aorta, bicuspid aortic valve, unroofed coronary sinus, coronary sinusostial atresia and cor triatriatum [3,4]. It is often asymptomatic and discovered incidentally during central venous cannulation, devices placement such as pacemaker and implantable cardioverter defibrillator, and cardiac catheterization. However, PLSVC has several practical implications. A thorough understanding of venous drainage system is imperative especially in PLSVC cases with a planned central line insertion or catheterization because the catheter might be inserted unexpectedly into PLSVC leading to unintended consequences such as arrhythmia, cardiogenic shock, and coronary sinus thrombosis.

The clinical implication of PLSVC is different based on venous drainage systems, and it usually drains into the right atrium via the coronary sinus without significant complications, which represent almost 80–92% of cases [5]. In remaining 10% of cases, the PLSVC can drain directly into the left atrium resulting right-to-left shunting which can lead to increased risk of paradoxical thromboembolic complications [6]. Cardiac arrhythmia especially atrial fibrillation (AF) could be a part of PLSVC presentation, which sometimes might be an arrhythmogenic source. Thus, LSVC isolation rather than pulmonary vein isolation will be required to suppress AF recurrence [7].

Fortunately, there was no evidence of AF and other congenital anomalies in our patient and treated with a low dose beta blocker. In the subsequent follow-up visit, he had a better control of blood pressure without the requirement of cardiac catheterization.

Conflict of interest
The authors have no conflicts of interest or financial relationships to disclose.

References


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