

Persistent Left Superior Vena Cava Associated with Hemiazygos Vein Draining in It and Absence of Left Brachiocephalic Vein, in a Patient with Congenital Heart Defect

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ABSTRACT

Persistent left superior vena cava is an anomalous vein that derives from a malfunction of obliteration of the left common cardinal vein during intrauterine life. The diagnosis can be suggested by a dilated coronary sinus as seen in echocardiography, or other imagistic methods. Due to the lack of hemodynamic impairment, and consequently with few or no symptoms, this vascular anomaly is frequently discovered incidentally. In this brief report we present the case of a 35-year-old male known with a complex congenital cardiovascular malformation that included atrial septum defect, persistent left superior vena cava and anomalous right pulmonary vein drainage in the PLSVC, diagnosed with sinoatrial block that required pacemaker implantation. Due to the patient's medical history, investigations to decide the best approach needed for pacemaker implantation were performed, including a thoracic CT that incidentally found additional anomalies — the hemiazygos vein draining in PLSVC and the lack of the left brachiocephalic vein.

Keywords: persistent left superior vena cava, anomalous hemiazygos vein, pacemaker implantation

INTRODUCTION

Vascular malformations may cause a wide variety of discrepant diagnoses. Imaging techniques might improve the accuracy of the diagnosis. Persistent left superior vena cava (PLSVC) has an incidence of 0.2–0.5% in the general population, and is considered the most frequent thoracic venous anomaly.¹ Most of the times the patients with PLSVC are asymptomatic, and the diagnosis is found incidentally in the course of routine imaging investigations or cardiovas-

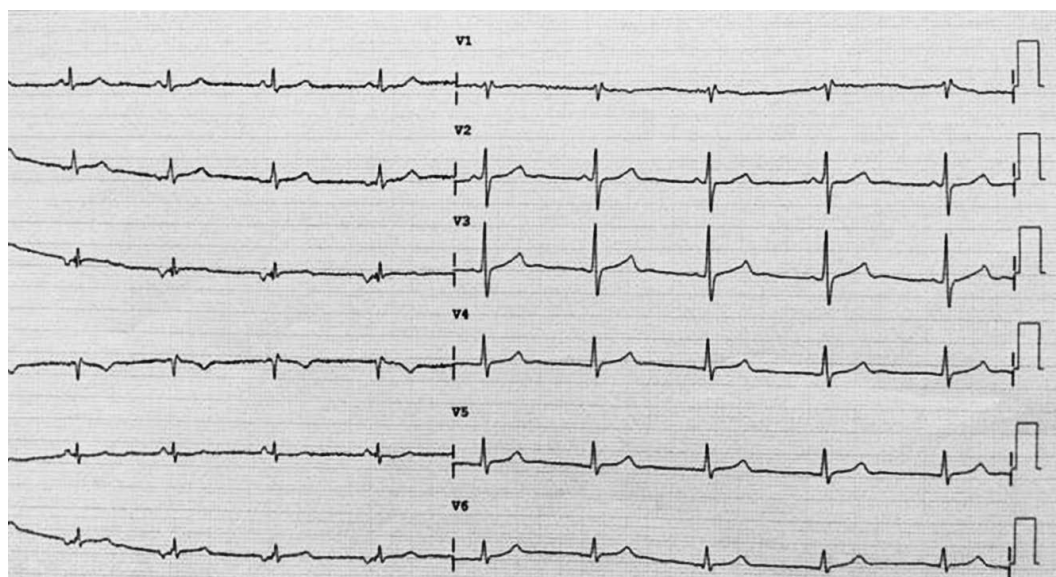


FIGURE 1. Resting ECG – showing lower sinus rhythm, no ischemic modifications

cular surgery. In the majority of cases, the PLSVC drains in the coronary sinus.² Even if these vascular malformations do not cause important hemodynamic changes, it may become a problem when a venous approach is needed, in case of central venous catheter insertions, or cardiovascular device implantations.³ We present the case of a young man with known associated congenital cardiovascular malformations, with multiple admissions for atypical symptoms.

CASE REPORT

We report the case of a 35-year-old male, admitted to the Clinic of Cardiology of the County Emergency Clinical Hospital of Tîrgu Mureș. He was admitted with the following symptoms: shortness of breath, constrictive chest pain, headache and palpitations that occurred during moderate physical exertion.

The patient agreed with the publication of his data and the publication of the case was approved by the ethics committee of the Cardio Med Medical Center, where the investigations were performed.

The patient was diagnosed with a complex cardiovascular malformation that included sinus venosus atrial septum defect, persistent left superior vena cava and anomalous right pulmonary vein insertion. The right pulmonary veins were draining in the persistent left superior vena cava (PLSVC), which in turn was draining in the coronary sinus. Surgical correction of the malformation included the closing of the atrial septum defect using a Dacron Velour patch, as well as the reinsertion of the right pulmonary

veins at the level of the left atrium by performing an atrio-caval plasty with a Gore-Tex patch.

After the surgical intervention, the patient's condition was stable, and he had been constantly followed-up.

After 20 years, the patient was admitted to our clinic for constrictive chest pain with duration of 5–10 minutes, associated with dyspnea and fatigue. The laboratory results did not show any pathological findings, and the echocardiographical examination revealed mild mitral regurgita-

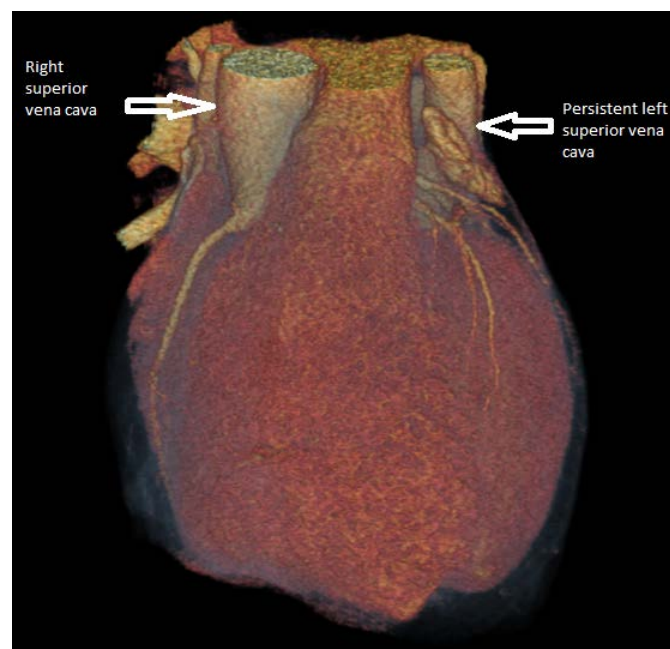


FIGURE 2. Cardiac CT with the presence of the persistent superior vena cava

tion, third degree tricuspid regurgitation, a pulmonary artery pressure of 57 mmHg (moderate arterial hypertension), a dilated coronary sinus (20/12 mm), and no evidence of left-to-right atrial shunt. The electrocardiogram showed lower atrial rhythm (Figure 1). For the evaluation of heart anatomy, a cardiac computed tomography was performed that showed absent coronary atherosclerotic lesions, and the presence of the PLSVC (Figure 2). In order to assess the origin of the palpitations, a 24 h Holter electrocardiogram was conducted, which showed sinus bradycardia, with a minimum heart rate of 45 beats per

minute. The patient had received treatment with angiotensin converting enzyme inhibitor, and was discharged with improved symptoms.

The patient presented repeated chest pain, dyspnea, coughing and lower limb paresthesia. The 24 h Holter ECG was repeated, and the result concurred with the previous one, therefore, a ECG cicloergometric stress test was carried out. The stress test was clinically positive, and was discontinued at 75w due to marked shortness of breath and constrictive chest pain, but the ECG revealed no ischemic electrocardiographical changes. The patient was treated

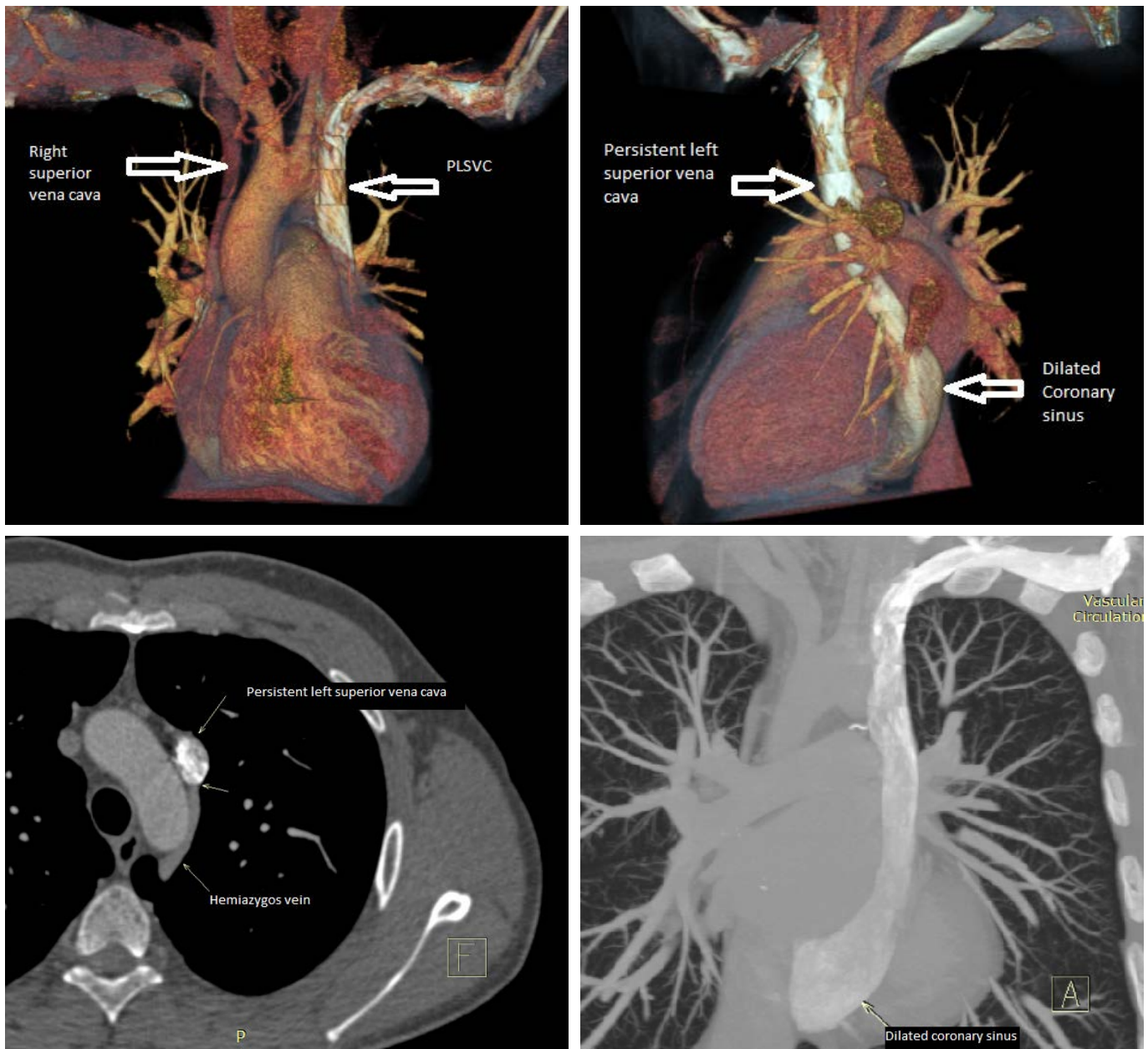


FIGURE 3. Contrast enhanced thoracic angio-CT showing the absence of the left brachiocephalic vein, the PLSVC, dilated coronary sinus, and the hemiazygos vein draining in the PLSVC

with ACEI and nitrates, and was discharged, being scheduled for an invasive exploration.

After several months, the patient presented again with the previously mentioned symptoms, relating also an episode of loss of consciousness. He has undergone a third Holter 24 h ECG examination, but this time the results showed an intermittent sinoatrial block, with a minimum heart rate of 30 beats per minute, associated with syncope.

To investigate possible anatomical particularities for the venous approach during the implantation of a pacemaker, a thoracic computed tomography angiography was obtained. The imaging results showed that the atrial septum defect was successfully closed, that the pulmonary arteries were properly draining into the left atrium, and the PLSVC was draining into a dilated coronary sinus. In addition, we detected another, previously missed venous anomaly: the fact that the hemiazygos vein was draining into the persistent left superior vena cava, and also, the absence of the left brachiocephalic vein (Figure 3).

After finalizing the paraclinical investigations, the patient had received a permanent DDDR pacemaker, using right subclavian approach.

DISCUSSIONS

The real incidence of PLSVC is only predictable, because most of the patients are asymptomatic.¹

The embryological origin of the PLSVC derives from the lack of involution of the lower segment of the left cardinal vein. The embryological superior venous system consists of two cardinal veins, the right, which will become the superior vena cava, and the left cardinal vein. The superior segment of the left cardinal vein will become the internal jugular, while from the inferior part the coronary sinus and the left atrial vein will form, and the remaining left cardinal vein will degenerate into a fibrous ligament. In PLSVC anomaly, the regression of the left cardinal vein fails to occur, and the left cranial and upper limb venous system will drain through the PLSVC into the coronary sinus, which becomes dilated.⁴

Other congenital defects that are associated with PLSVC include the absence of the left brachiocephalic vein, atrial septum defect, pulmonary vein drainage anomalies, absence of the right superior vena cava, and cor triatriatum.^{5,6}

Many related studies showed that the malformations of the great veins and the atypical drainage into the cardiac cavities can be associated with arrhythmias. This fact can be explained due to the close position of the coronary sinus, which is dilated, with the left sided pacemaker tissue.⁷

The noninvasive diagnosis of the PLSVC is usually obtained by performing a "bubble echocardiographic" study, in which the injection of agitated saline through the veins of the left upper limb will show the coronary sinus filling before the right atrial one, and a dilated coronary sinus.⁸ Other diagnostic methods include thoracic multislice computed tomography, magnetic resonance imaging and radionuclide angiography.⁹

The particularity of this patient was that he was previously diagnosed with a complex cardiovascular congenital disorder that included the PLSVC, atrial septum defect and abnormal draining of the pulmonary veins, which had been surgically corrected when the patient was 12 years old. The current symptoms, including the presence of the syncope, led to the decision to perform a permanent pacemaker implantation, with class IA indication according to the European Society of Cardiology guidelines.¹⁰ Incidentally, associated malformations were discovered during a repeated thoracic CT that was performed in order to evaluate the anatomical particularities of the pacemaker implantation approach, including the absence of the left brachiocephalic vein and the anomalous hemiazygos venous return in the PLSVC, leading to an even more enhanced dilation of the coronary sinus.

The presence of the PLSVC and the lack of the brachiocephalic vein led to the decision to implant the permanent pacemaker by a right subclavian approach.

In case of patients with PLSVC, this anomaly may provoke difficulties in performing other invasive procedures that require a superior venous approach, besides implantable cardiovascular devices, such as insertion of central venous catheters, and also the administration of chemotherapy through an implantable catheter in oncological patients.¹¹ Therefore, in patients with known congenital malformations, the physicians ought to investigate these possible associated anomalies.

CONCLUSIONS

The persistence of the left superior vena cava has no hemodynamic consequences, it may evolve without any symptoms, therefore it is usually incidentally diagnosed. It is frequently associated with other congenital defects, hence it would be strongly recommended to search for additional malformations in a patient diagnosed with PLSVC.

This anomaly may influence the conduction system of the heart due its proximity to the extremely dilated coronary sinus. The anatomical particularity of this malformation may lead to difficulties in the approach used during pacemaker implantation procedures.

CONFLICT OF INTEREST

Nothing to declare.

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