Case Report

Cardiac Arrhythmias in Patient with Isolated Persistent Left Superior Vena Cava

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ABSTRACT: We report here the case of a 47-year-old male presented with atypical chest pain, fatigue and arrhythmias that was found to have persistent left superior vena cava. The clinical exam was normal. Twelve leads ECG showed sinus rhythm of 70 beats/min, QRS axis of 90 degree and right bundle branch block. Transthoracic echocardiography revealed left ventricular hypertrophy, a large coronary sinus and mild pulmonary hypertension. Saline contrast echocardiography was performed and micro-bubbles were visible first into the coronary sinus and then passed through the right atrium. Multislice computed tomography confirmed the presence of persistent left superior vein cava and the site of drainage into the right atrium via a large coronary sinus. Right superior vein cava was absent. In this case report, we emphasize the importance of full assessment in patient with persistent left superior vena cava.

KEYWORDS: *persistent left superior vena cava, arrhythmias, embryology*

Introduction

Persistent left superior vena cava (PLSVC) is a rare vascular anomaly; but is considered the most common form of abnormal venous drainage involving the superior vena cava [1], and the most common congenital anomaly of thoracic venous system having an overall prevalence of 0.3-0.5% in the population [*2*]. The few reported cases of PLSVC illustrate the rarity of this pathology. The condition is frequently asymptomatic and is usually detected when cardiovascular imaging is performed for unrelated reasons. PLSVC can be isolated or associated with other cardiovascular abnormalities including bicuspid aortic valve, atrial septal defect, coarctation of aorta, coronary sinus ostial atresia, and cortriatriatum [3]. When symptomatic, invasive approach represents the mainstay of therapy. We herein describe a case of PLSCV in a 47-year-old man, which was successfully treated.

Case presentation

A 47-year-old male with a long history of atypical chest pain, fatigue, arrhytmias and recurrent pulmonary infection, was referred to Cardiac Surgery Department of Institute of Cardiovascular Diseases, Timisoara, for complete cardiac evaluation. The study was performed according to the tenets of the Declaration of Helsinki and was approved by the University Ethics Committee. The patient was fully informed about the possible consequences of the present study, and he filled in an informed consent form to participate.

His vital signs were normal. Physical examination demonstrated a systolic ejection murmur attributed to the mitral and tricuspid valves. Laboratory data and arterial blood gas analyses were within limits. Electrocardiogram revealed sinus rhythm of 70bpm, with right bundle branch block. In the past, the patient had various arrhythmias such as supraventricular tachycardia. Chest X-ray was normal. Transthoracic echocardiography showed left ventricular hypertrophy, ejection fraction of 55%, without dilatation of the right cavities, mild mitral and tricuspid regurgitation, dilated coronary sinus which raised the suspicion of persistent left superior vena cava. Transesophageal echocardiography revealed a dilated coronary sinus of 2.2cm (Fig. 1). Cardiac catheterization showed 50% obstruction of right coronary artery, normal left coronary arteries (Fig. 2). Saline contrast echocardiography and multislice computed tomography confirmed the presence of PLSVC, draining into dilated coronary sinus (Fig. 3). In our patient, the left superior vena cava descends to the left of the aortic arch and main pulmonary artery, traversing under the left atrium, turns medially and inserts into the coronary sinus. Agenesis of right superior vena cava was noted. Jugular internal, subclavian and innominate veins were

normal. Azygos vein drains into persistent left superior vena cava. On appropriate antiarrythmic medication, the course was uneventful. The patient was discharged home, with the recommendation of annual clinical follow up.

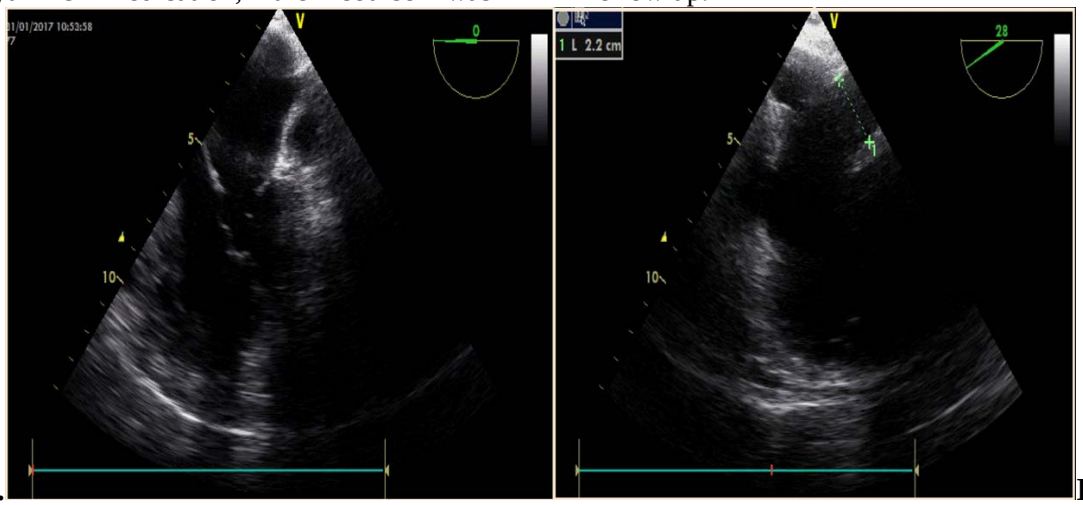

Fig. 1. TE echocardiography. (A)-Mid esophageal four chamber view showing dilated coronary sinus. (B)-Mid esophageal coronary sinus view showing coronary sinus in long axis

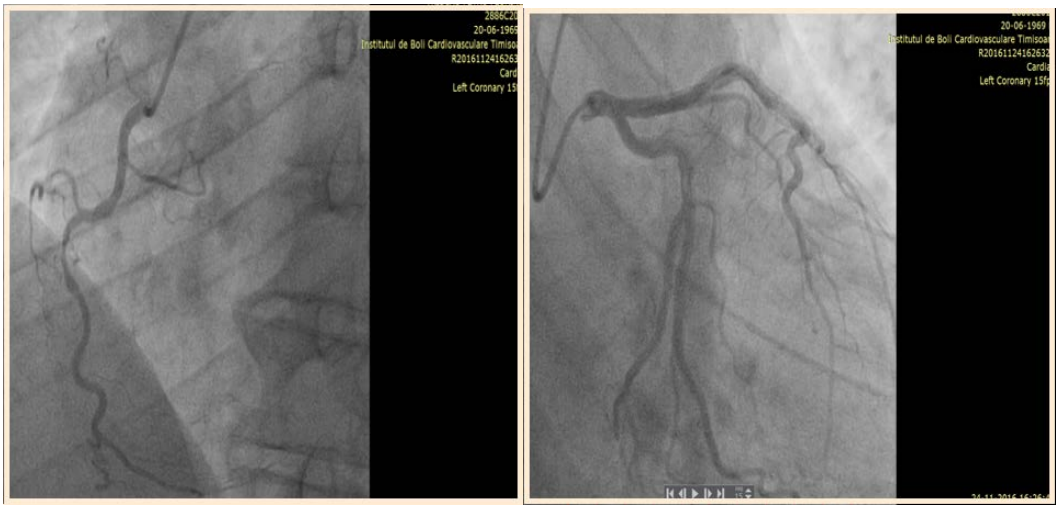

Fig. 2. Coronary angiography revealed 50 % obstruction of right coronary artery. Normal left coronary arteries

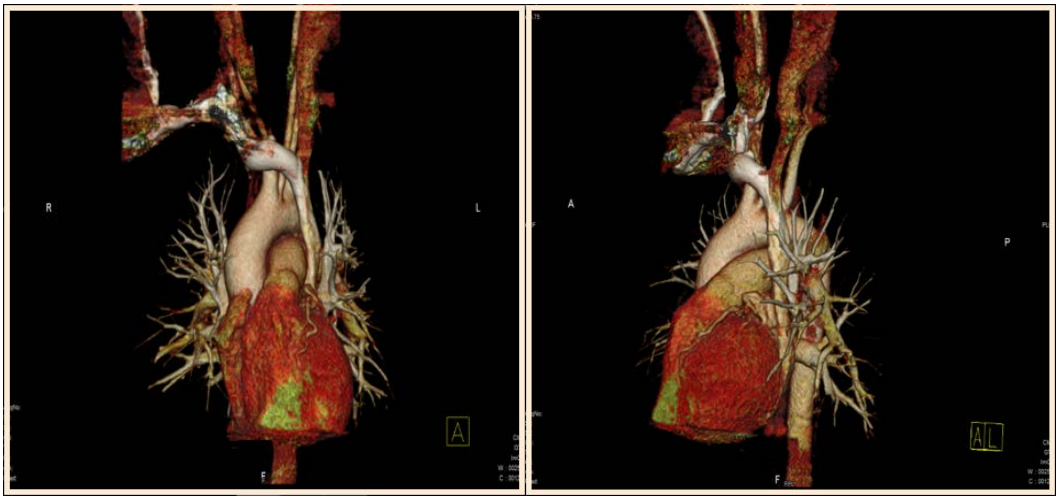

Fig. 3. Multislice computed tomography scan. Chest computed tomography scan demonstrates the persistence of left superior vena cava draining into dilated coronary sinus

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Discussion

Persistent left SVC is the most frequent congenital thoracic venous anomaly showing a general prevalence of 0.3-0.5%, and rising up to 10% in those patients that are suffering from congenital heart diseases [4]. PLSVC can be isolated or associated with cardiovascular abnormalities such as bicuspid aortic valve, atrial septal defect, coarctation of aorta, cortriatriatum and coronary sinus ostial atresia. The condition is mostly asymptomatic and may be an incidental finding. In 80-90 % of individuals, the most common draining site is the right atrium via coronary sinus, but having no hemodynamic consequence. In the remaining cases, it could drain in the left atrium giving rise to a right to left sided shunt, or in inferior vein cava [3]. PLSVC represents the congenital remnant of the left anterior cardiac vein or vein of Marshall, which fails to regress during embryological development [4]. Hence, results a persistent left sided vascular structure draining mostly into coronary sinus, so called persistent left superior vena cava (Fig.4).

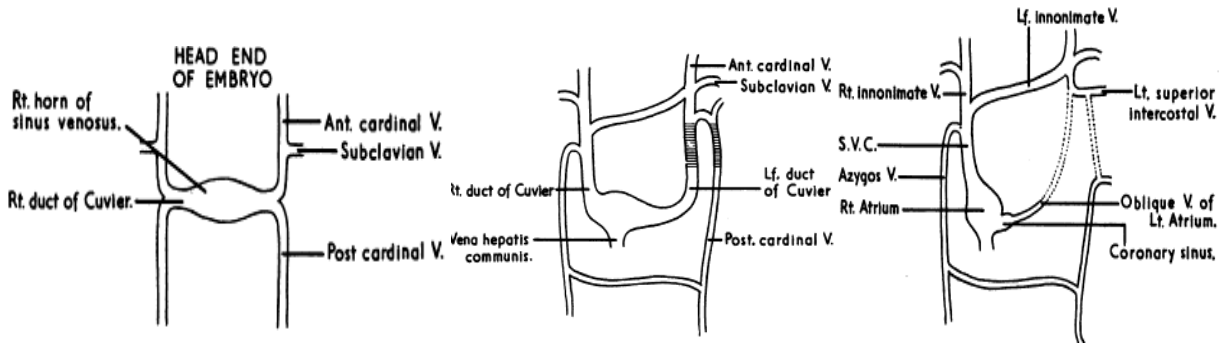

Fig. 4. Embryological development of left sided superior vena cava (SVC). Initially, the anterior cardinal vein drains into the sino-atrial chamber (left image). The left cardinal vein obliterates (center image), leaving only the oblique vein of Marshall as a remnant (right image). The failure of obliteration leads to a persistent left superior vena cava (adapted from Campbell and Deuchar) [5]

This condition is frequently associated with other anomalies of the intrinsic heart's conduction system, leading to arrhythmias such as tachyarrhytmias (supraventricular tachycardias, atrial fibrillation/flutter or Wolff Parkinson White) or bradyarrhytmias (atrioventricular and intraventricular conduction blocks) [6].

In a study of 300 patients with arrhythmias who underwent an electrophysiologic study prior to pacemaker or implantable cardioverter defibrillator placement, demonstrated that approximately 4% had an anomaly of venous drainage [6].

The pathophisyology of arrhythmias has its explanation in early development of the conductive tissue.

The right sided area (located in the sinus venous) usually forms the sinoatrial node while the left sided (located in the posterior cardinal vein) migrates downward to an area near the coronary sinus.

The left sided tissue in normal development loses its conduction ability. Contrary, in case of persistence of left superior vena cava, abnormal electrophysiologic function can arise from this site [6].

Cardiac arrhythmias may have secondary causes such as stretching of atrioventricular node and bundle of His due to coronary sinus dilatation [1].

Amongst other important clinical implications of PLSVC, we mention difficult placement of cardiac catheters or pacemaker leads, or ineffectiveness of retrograde cardioplegia during cardiac surgery, in patients with dilated coronary sinus [1].

Being mostly asymptomatic, persistent left SVC is an incidental finding during cardiovascular imaging or surgery. An incidental finding of dilated coronary sinus on an echocardiography examination should also raise the suspicion of PLSVC. The diagnosis must therefore be confirmed by saline contrast echocardiography and multislice computed tomography [3].

In addition, in patients with abnormal venous return it is very important to perform an overall assessment, complete imaging and electrophysiological studies. Annual clinical followup, including an electrocardiogram, is mandatory and any deviations from the patient's baseline should be a criteria for cardiac evaluation [6].

Conclusion

Our patient was investigated for atypical angina and arrhythmias. Transthoracic echocardiography had raised the suspicion of PLSVC, the diagnosis being confirmed by contrast echocardiography examination and multislice computed tomography. Cardiac catheterism revealed 50% obstruction of right coronary artery, normal left coronary arteries. On proper medication, the patient had an uneventful recovery, being discharged to a rehabilitation center. In conclusion, a complete assessment and appropriate understanding of the embryology and pathophysiology of PLSVC, as well, will decrease useless and potentially damaging examinations, while providing optimal care for those few patients who truly need complete cardiac evaluation and treatment.

References

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