

CASE REPORT

Persistent left superior vena cava: A disconcerting finding for interventionist

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Abstract

Persistent left superior vena cava (PLSVC) is one of the commonest venous anomalies of thoracic venous system. These cases usually have normal right sided superior vena cava (SVC). Patients with PLSVC are often asymptomatic hence, it is diagnosed as an incidental finding during procedures like central venous line placement or pacemaker implantation. We report here a case that was taken to catheterization laboratory for pacemaker implantation. Due to PLSVC, we struggled to advance pacing lead into right ventricular apex. We thought of right subclavian vein access but contrast medium injection revealed absent right sided SVC. So procedure was abandoned and finally epicardial placement of the pacing lead was done through sub-xiphoid laparotomy incision next morning. This case report highlights a rare variant of PLSVC with absent right sided SVC in structurally normal heart and emphasizes the need of pre-procedure assessment of venous anatomy.

Keywords: Superior vena cava, coronary sinus, cardiac pacemaker

Introduction

The prevalence of PLSVC is about 0.3% to 0.5% in general population and it is the most common form of anomalous venous drainage involving SVC.^{1,2} In majority of cases (82-90%) right sided SVC is present.³ With congenital heart diseases its prevalence rises to 5%. The most commonly associated congenital defect is atrial septal defect. Others include ventricular septal defect, single atrium, tetralogy of Fallot, coarctation of aorta and anomalous pulmonary venous return.⁴ This condition is also associated with various conducting system abnormalities such as tachyarrhythmias (supraventricular tachycardias, atrial fibrillation/flutter) or bradyarrhythmias (atrioventricular and intraventricular conduction blocks).^{5,6} The most easily available modality to diagnose PLSVC is transthoracic echocardiography. A dilated coronary sinus (CS) is the first clue on echocardiography.

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Transthoracic echocardiogram combined with agitated saline injections through left and right antecubital veins provides a simple but elegant study for rapid bedside diagnosis.⁷

Case Report

A 60 years old hypertensive man was presented in outpatient clinic of Tahir Heart Institute with complaint of pre-syncope (vertigo) in April, 2018. The patient was fully informed about the possible consequences of the present study, and he filled in an informed consent form to participate. He underwent percutaneous coronary intervention (PCI) to left anterior descending artery and right coronary artery with drug eluting stents in 2015. His physical examination was unremarkable. ECG showed diffuse conducting system involvement with left bundle branch block, first degree AV block and sinus pauses. Holter recording revealed sinus pause of 2.45 seconds. Echocardiography revealed septal motion abnormality due to conduction defects. Finding of dilated coronary sinus was overlooked. Considering symptomatic bradyarrhythmia and diffuse conducting system disease, decision of permanent pacemaker (PPM) implantation was taken. Patient was taken to catheterization laboratory. Left subclavian vein access was taken with Seldinger's technique. Pacemaker lead was advanced through subclavian vein but its course was unusual. It could not be advanced to right ventricle despite many attempts. Contrast injection into subclavian vein ascertained PLSVC. A Judkin right 4.0 (JR) catheter was advanced on the guide wire through PLSVC. Contrast injection through catheter delineated the hugely dilated CS (Figure 1). The right subclavian vein route was considered but contrast injection in right antecubital vein revealed absent right sided SVC. The procedure was abandoned. The next day, epicardial placement of the pacing lead was done through sub-xiphoid laparotomy incision. Computed tomographic (CT) scan confirmed the presence of PLSVC and the absence of right SVC (Figure 2). It further revealed that at the level of aorto-pulmonary window there is communication of PLSVC with the left hemi-azygous system. Hemi-azygous vein

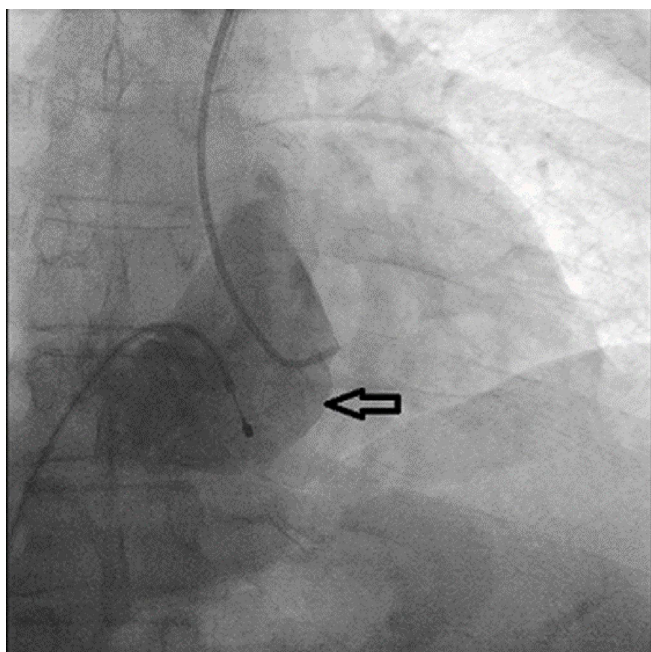


Figure-1: The dilated coronary sinus (Arrow) with JR 4.0 catheter coursing through PLSVC on fluoroscopic antero-posterior projection.

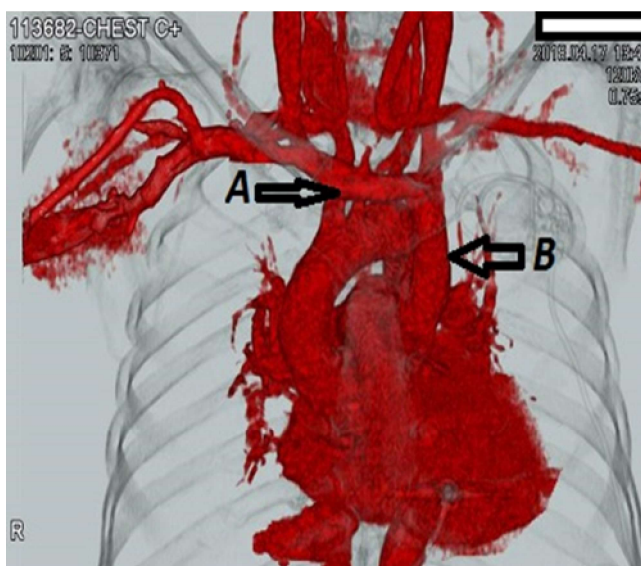


Figure-2: Contrast enhanced CT chest reconstruction image: Arrow A] Expected location of origin of right superior vena cava (absent in this image). Arrow B] Persistent left superior vena cava.

runs in posterior mediastinum with a small horizontal communication with azygous vein at 9th thoracic vertebra.

Patient recovery was uneventful. Cardiac pacing was adequate with PPM in subcutaneous pocket in left infra-

clavicular fossa, attached with the pacing lead tunneled through subcutaneous tissue of anterior chest to epicardial surface of heart. Patient was discharged home with advice of follow-up after one week.

Discussion

PLSVC represents the congenital remnant of the vein of Marshall, which fails to regress during embryological development.⁵ The condition is mostly asymptomatic and may be an incidental finding. In 92% of individuals, the draining site is the right atrium via CS, while in others (8%) it drains into left atrium giving rise to right to left shunt which is not large enough to cause symptoms or cyanosis. As described earlier, in about 90% cases right sided SVC is present which from interventionist perspective is a blessing. In 25-35% cases a persistent bridging vein communicating the two SVC is also present.^{4,8} PLSVC is associated with various tachy & brady arrhythmias. This could be due to the direct effect of dilated CS pressing on conducting tissues like atrio-ventricular node and HIS-bundle. This association of PLSVC with arrhythmia could explain the higher prevalence (4% versus 0.5% in general population) of PLSVC detection during PPM implantation.⁹ Transthoracic echocardiography is very helpful in diagnosis. In this case, patient's echocardiography was done at the time of PCI and echocardiographer was probably more concerned in segmental motion wall abnormalities and finding of dilated coronary sinus was missed. Fresh echocardiography was not requested. The abnormal left sided course of guide wire through the subclavian sheath was the first indication of PLSVC. In the majority of the cases in which PLSVC is discovered during or before the procedure, access is switched to the right subclavian vein, allowing for an easier route for lead navigation.¹⁰ This was not a viable option in this case as right sided SVC was absent. Though there are some reports of successful navigation of pacing lead through PLSVC into right ventricular apex.¹¹ But it depends on patient's anatomy. With absent right sided SVC, all venous circulation from upper extremities, is through PLSVC and then via CS to right atrium, resulting in more dilatation of CS and more awkward course for lead navigation. It is our practice to prepare subcutaneous pocket for PPM after vein access. The abnormal course of wire through the arterial sheath into PLSVC enabled us to diagnose PLSVC before pocket preparation and

unnecessary incision. A pre-procedure contrast injection through antecubital vein cannula on both sides under fluoroscope could serve the purpose. It could delineate the venous anatomy there and then.

Conclusion

This case suggests that the diagnosis of PLSVC can be missed due to benign nature of this anomaly. But if such a patient undergoes procedure like PPM implantation then it could be disconcerting for interventionist. Failure of device implantation would add to patient misery. A pre-procedure confirmation of venous anatomy by contrast injection through antecubital veins of both sides under fluoroscope, irrespective of echocardiographic findings would be helpful for interventionist.

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References

1. Paval J, Nayak S. A persistent left superior vena cava. Singapore Med J 2007; 48(3): e90.
2. Mohapatra R, Pattnaik NK, Sahu S, Nayak D. Isolated Persistent Left Superior Vena Cava: A Case Report and its Clinical Implications. Ann Card Anaesth; 2017; 20:104-7.
3. Cormier MG, Yedlicka JW, Gray RJ, Moncada R. Congenital anomalies of the superior vena cava: a CT study. SeminRoentgenol1989;24:77-83.
4. Pretorius PM, Gleeson FV. Case 74: right-sided superior vena cava draining into left atrium in a patient with persistent left-sided superior vena cava. Radiology 2004; 232: 730-4.
5. Morgan LG, Gardner J, Calkins J. The incidental finding of a persistent left superior vena cava: implications for primary care providers-case and review. Case Rep Med 2015; 2015:198754.
6. Morgan DR, Hanratty CG, Dixon LJ, Trimble M, and O'Keeffe DB. Anomalies of cardiac venous drainage associated with abnormalities of cardiac conduction system. Europace 2002;4: 281-7.
7. Recupero A, Pugliatti P, Rizzo F, Carerj S, Cavalli G, de Gregorio C, et al. Persistent left-sided superior vena cava: integrated noninvasive diagnosis. Echocardiography 2007; 24: 982-6.
8. Atwell WJ, Zoltowski P. A case of left superior vena cava without a corresponding vessel on the right side. Anat Rec 1938; 70: 525-32.
9. Hsu LF, Ja'is P, Keane D, Wharton JM, Deisenhofer I, Hocini M, et al. Atrial fibrillation originating from persistent left superior vena cava. Circulation 2004;109: 828-32.
10. Jovic Z, Mijailović Z, Obradović S, Tavciovski D, Matunović R, Rusović S, et al. Successful implantation of a permanent pacemaker through a persistent left superior vena cava by using a right subclavian approach. Vojnosanitetski Pregled 2011; 68: 792-4.
11. Sankhla V, Vajifdar B, Shah M, and Lokhandwala Y. Left sided biventricular pacemaker implantation in the presence of persistent left superior vena cava. Ind Heart J. 2010;62(4):344-5.