DOI: 10.7860/JCDR/2016/20319.7774

Case Report

Internal Medicine Section

Persistent Left Superior Vena Cava: A Rare Case with Clinical Significance

GAUTAM RAWAL[1], RAJ KUMAR[2], SANKALP YADAV[3], DEEPAK VERMA[4]

ABSTRACT

Persistent Left Superior Vena Cava (PLSVC) is a rare congenital vascular anomaly (incidence of 0.3-0.5% of the general population) which being mostly asymptomatic in its presentation, is usually detected incidentally. There are many practical clinical implications associated with it including arrhythmias. We report a rare case of PLSVC with absent Right Superior Vena Cava (RSVC) (isolated PLSVC), in a 55-year-old lady who had complete heart block followed by sepsis and was diagnosed to have this condition during the permanent cardiac pacemaker implantation and central venous catheter insertion showing an abnormal path of the catheter/pacing leads. The authors also give an insight into its clinical relevance.

Keywords: Computed tomography, Coronary sinus, Echocardiogram, Vascular malformation

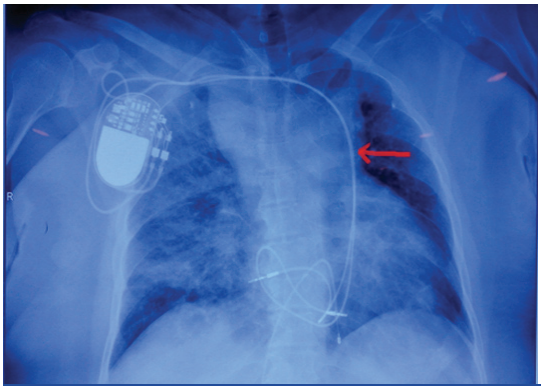
CASE REPORT

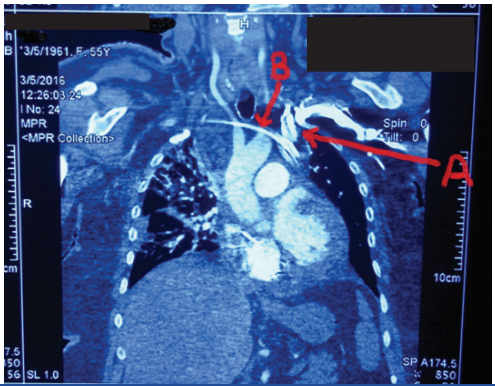
A 55-year-old Indian lady presented to the emergency department with a history of fever and breathing difficulty for the last four days. She had a recent history of admission (four days prior) in a local hospital for recurrent syncope attacks, breathing difficulty, chest heaviness and giddiness and on evaluation was found to have complete heart block on electrocardiogram (ECG) for which permanent dual chamber cardiac pacemaker was implanted through the right subclavian vein. Her chest radiograph (A-P view) done at the local hospital show bilateral chest infiltrates and also the pacemaker leads were seen on the left side of the chest [Table/Fig-1]. Her Computed Tomography (CT) angiogram of the chest done at the local hospital also showed the same with evidence of persistent left Superior Vena Cava (SVC) with absent right SVC [Table/Fig-2a] and cardiac pacing wires seen crossing over to the left hemithorax [Table/Fig-2b]. Her medical history included type 2 diabetes mellitus and hypertension for which she was on regular medicines for past 4 years. She was admitted to the Intensive Care Unit (ICU) for the management of the sepsis due to clinical evidence of pneumonia. On examination, she was drowsy but arousable, moving all limbs, following simple commands with no nuchal rigidity and both pupils equal in size and reacting to light. Her blood pressure was150/80mmHg, pulse 120/minute, random blood sugar 300mg%, no pedal oedema, normal jugular venous pulse, chest-bilaterally clear, heart sounds normal. Her liver function tests, renal function test was normal with leukocytosis. Her echocardiogram showed pacing leads in the right atrium and ventricle with a dilated coronary sinus and left ventricle ejection fraction of 45%. A Central Venous Pressure (CVP) line was inserted in the left Internal Jugular Vein (IJV) for fluid management and monitoring required in a case of severe sepsis. Post CVP line insertion chest radiograph (A-P view) showed abnormal positioning of the central catheter on the left side of the thorax [Table/Fig-3]. The patient was started on intravenous antibiotics, fluids and supportive care. The patient showed initial recovery, but remained critical due to the development of sepsis induced multi-organ dysfunction and her relatives decided to get her discharged against medical advice for further treatment in her home town. A written and informed consent was obtained for using the clinical images and the details of the case.

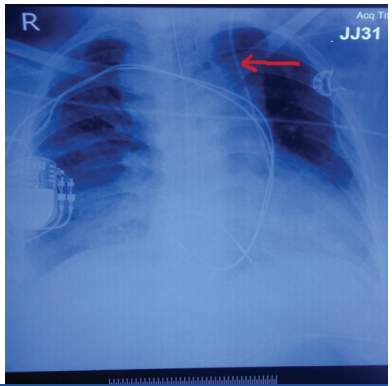
DISCUSSION

Persistent Left Superior Vena Cava (PLSVC) is a thoracic vascular anomaly, usually diagnosed incidentally while performing imaging for unrelated invasive or non-invasive procedures. However, although rare, it is considered the most common congenital anomaly of the thoracic venous system with a prevalence of 0.3% to 0.5% in the general population and 10% of those with congenital heart disease [1-3]. The authors present a rare case of isolated PLSVC with absent right SVC, with its relevant implications.

PLSVC occurs due to the failure of the left superior cardinal vein to regress/obliterate to form the 'ligament of Marshall' during the early weeks of embryologic development, resulting in a persistent left-sided venous vasculature which, via the coronary sinus, drains into the right atrium [1-3]. This causes the dilatation of coronary sinus due to the increased venous return [1,2]. In 82-90% of







[Table/Fig-1]: Chest radiograph (A-P view) showing cardiac pacemaker leads (arrow) on the left side of the thorax. **[Table/Fig-2]:** Coronal section of CT angiogram chest: - A: Left superior vena cava draining into right atrium. B: Cardiac pace maker leads in right subclavian vein seen crossing the mid line and entering into left superior vena cava. **[Table/Fig-3]:** Chest radiograph (A-P view) showing abnormal path of left IJV central catheter (arrow) in the left hemi thorax.

the PLSVC cases superior vena cava is bilateral (both right and left) and the concomitant agenesis of the right-sided SVC is rare (known as isolated PLSVC) [1,4]. In 90% of individuals, the PLSVC drains into the right atrium via the coronary sinus and causes no haemodynamic consequence [1-4]. In the remaining cases, it may drain into the left atrium resulting in a right to left sided shunt, or into the inferior vena cava or hepatic vein [1-4]. PLSVC is usually asymptomatic and haemodynamically of no significance, but may be associated with disturbances of cardiac rhythm. The diagnosis is usually incidental during cardiovascular imaging (echo cardiogram showing dilated coronary sinus), cardiac pacemaker implantation or chest imaging after placement of a central venous catheter [1,2,4-8].

Clinical and practical relevance of PLSVC: In about 40% of the patients with PLSVC, congenital cardiac anomalies are associated and they are more commonly seen in cases with concomitant absence of right SVC (isolated PLSVC) [1,4]. These anomalies include atrial septal defect, bicuspid aortic valve, coarctation of the aorta, coronary sinus ostial atresia, and cor triatriatum [4-9].

The presence of PLSVC has been found to be associated with the abnormal electrophysiological functioning of the heart due to the anatomical and architectural abnormalities of the sinus node and the conduction tissues (due to dilation of coronary sinus) [4-10]. This can result in both tachyarrhythmias (supraventricular tachycardias, atrial fibrillation syndrome) and bradyarrhythmias (due to atrioventricular conduction blocks) [4,5,9,10]. PLSVC can also complicate invasive procedures like the placement of permanent pacemaker or implantable cardioverter defibrillator (ICD), CVP catheter or haemodialysis catheter placement (where the catheter follows an unusual course on the chest radiographs) and surgical procedures like cardiopulmonary bypass surgery or heart transplantation [4,5,9,10]. PLSVC draining into the left atrium may result in incidences of cerebral abscess and thromboembolic events [3].

CONCLUSION

Critical care physicians and cardiologists should be aware of this anomaly of PLSVC, which though rare, is clinically relevant due to its association with congenital heart disease and cardiac electrophysiological conduction abnormalities. It should be considered whenever a catheter or guide wire inserted in the upper major venous system takes an unusual left-sided downward path or if there is presence of dilated coronary sinus on echocardiogram.

REFERENCES

[1] Irwin RB, Greaves M, Schmitt M. Left superior vena cava: Revisited. *Eur Heart J Cardiovasc Imaging*. 2012;13:284–91.
[2] Gupta R, Pearson A. Diagnosis of persistent left superior vena cava draining directly into the left atrium. *N Am J Med Sci*. 2013;5(8):496–97.
[3] Lee MS, Pande RL, Rao B, Landzberg MJ, Kwong RY. Cerebral abscess due to persistent left superior vena cava draining into the left atrium. *Circulation*. 2011;124(21):2362-64.
[4] Uçar O, Pasaoglu L, Ciçekçioglu H, Vural M, Kocaoglu I, Aydogdu S. Persistent left superior vena cava with absent right superior vena cava: a case report and review of the literature. *Cardiovasc J Afr*. 2010;21(3):164-66.
[5] Sheikh AS, Mazhar S. Persistent left superior vena cava with absent right superior vena cava: review of the literature and clinical implications. *Echocardiography*. 2014;31(5):674-79.
[6] Duymus M, Yesilkaya Y, Orman G, Bozkurt A, Yilmaz O. Persistent left superior vena cava draining to the left atrium: A case report and review of the literature. *Pol J Radiol*. 2012; 77(4):65-66.
[7] Schreve-Steensma AM, van der Valk PHM, ten Kate JBL, Kofflard MJM. Discovery of a persistent left superior vena cava during pacemaker implantation. *Neth Heart J*. 2008;16(7-8):272-74.
[8] Parreira LF, Lucas CC, Gil CC, Barata JD. Catheterization of a persistent left superior vena cava. *J Vasc Access*. 2009;10(3):214-15.
[9] Eldin GS, El-Segaier M, Galal MO. High prevalence rate of left superior vena cava determined by echocardiography in patients with congenital heart disease in Saudi Arabia. *Libyan J Med*. 2013;8(1):21679.
[10] Sarodia BD, Stoller JK. Persistent left superior vena cava: case report and literature review. *Respir Care*. 2000;45(4):411-16.

PARTICULARS OF CONTRIBUTORS:
1. Attending Consultant, Department of Respiratory Intensive Care, Max Super Specialty Hospital, Saket, New Delhi, India.
2. Senior Consultant and Incharge, Department of Respiratory Intensive Care, Max Super Specialty Hospital, Saket, New Delhi, India.
3. General Duty Medical Officer-II, Department of Medicine & TB, Chest Clinic Moti Nagar, North Delhi Municipal Corporation, New Delhi, India.
4. Attending Consultant, Department of Respiratory Intensive Care, Max Super Specialty Hospital, Saket, New Delhi, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Gautam Rawal,
Flat No. 417, Dhruva Apartments, Plot no. 4, I P Extension, Patparganj, Delhi-110092, India.
E-mail: drgautamrawal@hotmail.com

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Mar 25, 2016**
Date of Peer Review: **Apr 04, 2016**
Date of Acceptance: **Apr 05, 2016**
Date of Publishing: **May 01, 2016**