Case Report

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Persistent Left Superior Vena Cava and its Clinical Correlation: A Cadaveric Case Report

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Abstract

Presented is a case of Persistent left superior vena cava draining into the right atrium through coronary sinus and finally opens into Right Atrium. Abnormalities of the vascular system are more commonly seen due to its importance in circulation. Persistent left superior vena cava is rare but important congenital vascular anomaly. It results when the left superior cardinal vein caudal to the in nominate vein fails to regress. The venous anomaly of a persistent left superior vena cava (PLSVC) affects 0.3%–0.5% of the general population. Normally the superior vena cava is a single vascular structure formed by the union of right and left brachio-cephalic veins which are in turn formed by the union of corresponding internal jugular and subclavian veins of corresponding side, draining the head and neck as well as the superior extremity. During routine dissection in the Department of Anatomy, Jaipur National University Institute for Medical Sciences And Research Centre (JNU IMSRC) Jaipur. We found persistent left superior vena cava in a 64-year-old male cadaver. Both the vena cavae were formed as continuations of brachiocephalic veins of the corresponding side. The persistent left superior vena cava opened into the enlarged coronary sinus that drained into the right atrium between the opening of inferior vena cava and right atrio-ventricular orifice it has important clinical implications in certain clinical interventions. It may complicate placement of cardiac catheters or pacemaker leads.

Keywords: Persistent Left Superior Vena Cava; Coronary Sinus; Superior Vena Cava; Right Atrium.

Introduction

Persistent left superior vena cava (PLSVC) is a rare venous abnormality. It is, however, the most common congenital anomaly of thoracic venous system with a frequency of less than 0.5% of the general population and up to 10% of patients with congenital heart disease [1-3]. Normal anatomy describes the formation of a single superior vena cava by the union of right and left brachi-cephalic veins which are in turn is formed by the union of corresponding internal jugular and sub-clavian veins, draining the head and neck as well as the superior extremity [4]. Double superior vena cava (SVC) with the persistent left superior vena cava (PLSVC) is a rare venous malformation. Patients with PLSVC may have other associated cardiac malformations such as atrial septal defect, ventricular septal defect or endo-cardial cushion defect [5,6]. Presence of PLSVC may also interfere and cause problems during various invasive procedures such as pacemaker implantation, central venous catheterization, retrograde delivery of cardioplegia and retrograde left ventricular pacing [7,8].

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Case Report

During routine dissection in the department of Anatomy, JNU IMSRC, Jaipur, we found double superior vena cava with persistent left superior vena cava (PLSVC) in a 64-year-old male cadaver there is no joining between left brachio-cephalic vein and right brachio-cephalic to forming a single superior vena cava , Both the vena cavae were formed as continuations of brachio-cephalic veins of the corresponding sides (Figure 3). The PLSVC had the larger length compared to the superior vena cava (Figure 1). When we traced, it opened into the enlarged

coronary sinus that further drained into the right atrium between the opening of inferior vena cava and right atrio-ventricular orifice (Figure 2). There was no communication between the two vena cavae (Figure 1). The hemiazygos and accessory hemiazygos veins drained normally into the azygos vein which in turn drained into the right superior vena cava. No other associated variations were observed.

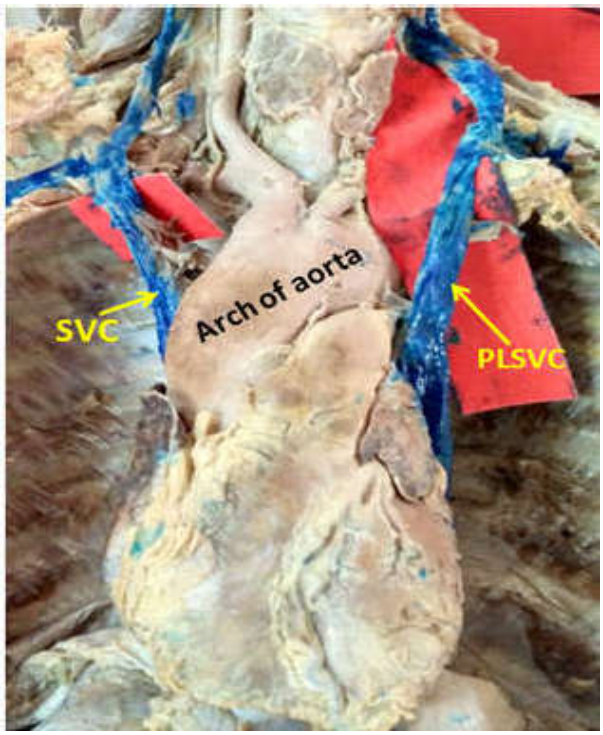

Fig. 1: Showing persistent left superior vena cava (PLSVC). Both the vena cavae (SVC & PLSVC) were formed as continuations of brachio-cephalic veins of the corresponding side. There is no communication between the two veins.

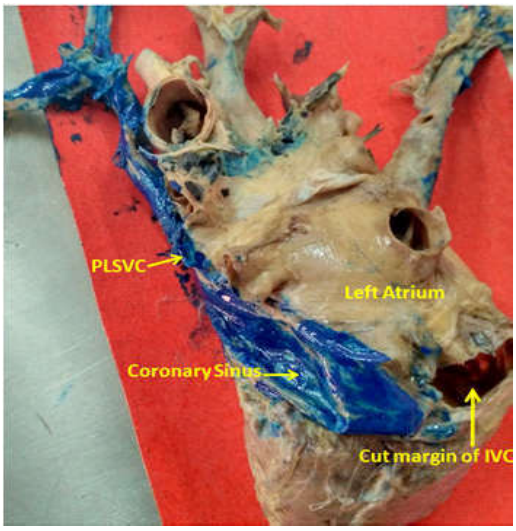

Fig. 2: Showing the persistent left superior vena cava (PLSVC) opening into the enlarged coronary sinus that drains into the right atrium between the opening of inferior vena cava and right Atrio-ventricular orifice.

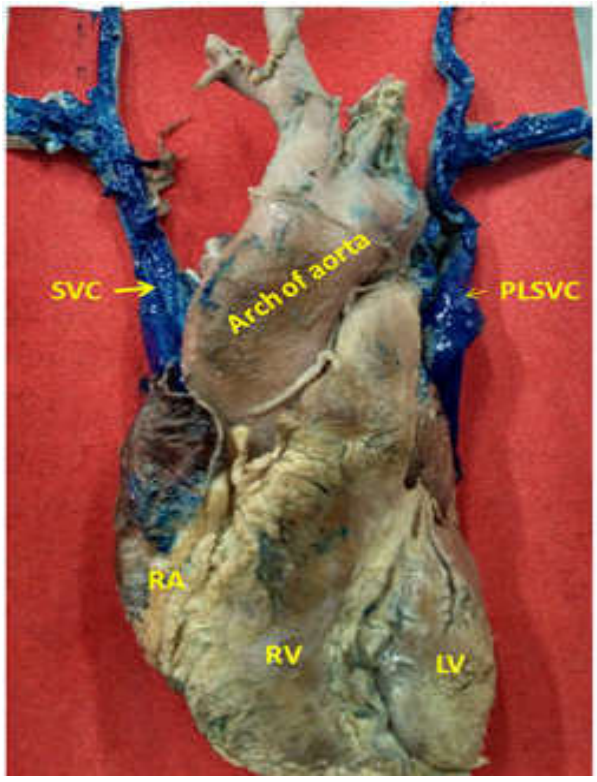

Fig. 3: Showing persistent left superior vena cava (PLSVC) and superior vena cava (SVC) on Both side.the There is no communication between the two veins. **SVC**- Superior vena cava
PLSVC- persistent left superior vena cava **RA**- Right Atrium, **RV** –Right ventricle, **LV**- left ventricle

Discussion

During the fifth week of intrauterine life, in the human fetus, three pairs of major veins can be distinguished: the vitelline veins, carrying blood from the yolk sac to the sinus venosus; the umbilical veins, originating in the chorionic villi and carrying oxygenated blood to the embryo; and the cardinal veins, draining the body of the embryo proper. The cardinal veins form the main venous drainage system of the embryo. This system consists of the anterior cardinal veins, which drain the cephalic part of the embryo, and the posterior cardinal veins, draining the remaining part of the body of the embryo. The anterior and posterior veins join to form common cardinal veins and enter the right and left horns of the sinus venosus. Formation of the vena cava system is characterized by the appearance of anastomoses between the left and right sides in such a manner that the blood from the left side is directed to the right side. The anastomosis between the anterior cardinal veins develops into the left brachiocephalic vein. Most of the blood from the left side of the head and the left

upper extremity is thus directed to the right. The terminal portion of the leftanterior cardinal vein entering into the left brachiocephalic vein is retained as the left superior intercostal vein. The superior vena cava is thus formed by the right common cardinal vein and the proximal portion of the right anterior cardinal vein. On the other hand, the left common cardinal vein and the distal part of the left horn become atretic and forms the ligament of Marshall or ligament of the left superior vena cava. If this normal regression of the left cardinal vein fails to occur, it results in a PLSVC [9].

The PLSVC is normal in some mammals but it is rare in man. Congenital abnormalities of the superior vena cava generally fall into one of two categories: anomalies of position or anomalies of drainage. Anomalies of position, especially a PLSVC are far more frequent than those of drainage. A PLSVC in itself causes no haemodynamic disturbance [10].

PLSVC is an uncommon and yet the most commonly reported thoracic venous abnormality. The frequency of a PLSVC is 0.3-0.5% among healthy individuals and as many as 10% of patients with congenital heart diseases [1-3]. There are two types of PLSVC described in the literature. In 92% of cases, PLSVC connects to the right atrium via coronary sinus with no hemodinamically significant consequence and in 8% of cases, PLSVC connects directly or through the pulmonary veins to the left atrium causing a right to left shunt [11].

The most common thoracic venous abnormality is the LPSVC draining into the coronary sinus in the presence of both left and right superior vena cavae. This anomaly is usually asymptomatic and does not require treatment unless accompanied by other cardiac anomalies [12].

This PLSVC drains into the right atrium *via* the coronary sinus in 92% of cases . But in the remainder of cases, it connects to the left atrium in such variants with absent or unroofed coronary sinus or normal coronary sinus and so creates a right-to-left shunt. Although the anomalies of systemic venous connection to the right atrium require no treatment when they occur alone, the PLSVC assumes particular significance when it communicates with the left atrium. Such patients usually present with cyanosis, polycythaemia or clubbing, although some have no clinical findings [13].

In the present case, a double SVC with a PLSVC was observed and there was no communication between the two superior vena cavae unlike as reported previously. PLSVC may also give rise to rhythm disturbances such as sinus node dysfunction and atrioventricular block. These rhythm problems may be related to the stretching of the conduction tissue caused by the enlargement of the coronary sinus [3]. It may also be associated with other malformations such as situsinversus or tetralogy of Fallot [9].

During cardiac surgery, the presence of PLSVC would be a relative contraindication to the administration of retrograde cardioplegia. It may be possible to clamp the PLSVC to avoid the cardioplegia solution from per fusing retrograde up the PLSVC and its tributaries. However, there is a possibility that there may be some steal of cardioplegia solution through an accessory vein. Further, the coronary sinus catheter balloon may not be able to occlude the dilated coronary sinus. This may result in the failure of flow of cardioplegia solution to the myocardium. Thus, the cardioplegia solution administered would largely be distributed to the left internal jugular and left subclavian veins, rather than the myocardium [14].

Conclusion

Persistent left superior vena cava PLSVC is a rare congenital anomaly and is separately available in the medical literatures. Therefore for the clinicians should be alerted about the possible existence of these venous anomalies, other cardiac abnormities associated with it and their clinical consequences so as to prevent possible complications in routine clinical practice and during cardiopulmonary bypass surgeries. The present case report adds to the existing knowledge of these congenital abnormalities and stresses on the use of different diagnostic techniques for its accurate diagnosis thereby avoiding further complications while planning different interventions.

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Conflict of Interest: No conflict.

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