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Persistent Left Superior Vena Cava: A Case Report

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Abstract: **Presented is a case of left superior vena cava draining into the right atrium through coronary sinus and communicating with right superior vena cava through small left brachiocephalic vein. The right atrium was very large, the right ventricle very small. There was also a defective tricuspid valve. A double superior vena cava is formed due to the persistence of the left anterior cardinal vein and failure of the left brachiocephalic vein to form. In such conditions the left superior vena cava drains into the right atrium through coronary sinus. Such venous abnormalities of thoracic region are commonly associated with cardiac abnormalities such as atrial or ventricular septal defects, Tetralogy of Fallot. When left superior vena cava is present drains the left cranial and upper limb veins to the right atrium, the left brachiocephalic vein may be undeveloped with the right superior vena cava relatively smaller than normal.**

Keywords: **Left superior vena cava, left anterior cardinal vein, coronary sinus, brachiocephalic vein.**

1. INTRODUCTION

Persistence of the left superior vena cava (PLSVC) has an incidence of 0.3–0.5% in the general population (1). In the presence of other congenital heart disease, the incidence of PLSVC increases to 3–10%. The abnormality is often associated with the ventricular septal defect (VSD), atrial septal defect (ASD), coarctation of the aorta and mitral atresia. Precise anatomical knowledge of the great vessels of the neck and thorax is essential for safe anaesthesia, intensive care practice, pacemaker implantation and cardiac surgery. Therefore, this anatomical variant must be recognized to avoid the potential complications such as those associated with placement of central catheters and cardiac surgery.

2. CASE REPORT

This persistent left superior vena cava emptied into the coronary sinus. The coronary sinus itself was enlarged. In this cadaver aged about 50 yrs, we observed the left superior vena cava (PLSVC) with small contracted right ventricle, over-riding of pulmonary artery with a hypertrophied right atrium. The right atrium was acting as one chamber for both, right atrium and right ventricle. Double superior vena cava above the communicating vein showed valves.

3. DISCUSSION

The early development of the heart before the 4 mm. stage includes the formation of the primitive heart tube in the cardiogenic area. The developing liver and systemic veins below the heart are also centrally situated and bilaterally represented. The veins represented 1-5, appropriately labeled, in Fig. 1 & Fig.2, open into the venous sinus, but as this chamber tapers laterally to the right in situs solitus to form the right atrium, the developing liver also increases in size on the right and disappear. The vitelline veins fuse to become a single channel, the hepato-cardiac channel, which connects with the inferior vena cava and thereby forms a single channel to the heart. Failure of disappearance, or reduction in size of the left sinus will permit not only persistent drainage of the anterior cardinal vein (the left superior vena cava) but will permit persistent drainage of the left vitelline vein which continues to drain into this channel. Thus hepatic venous blood from the left lobe of the liver will join the left superior caval and coronary venous blood to enter the right atrium so that no physiological abnormality exists.

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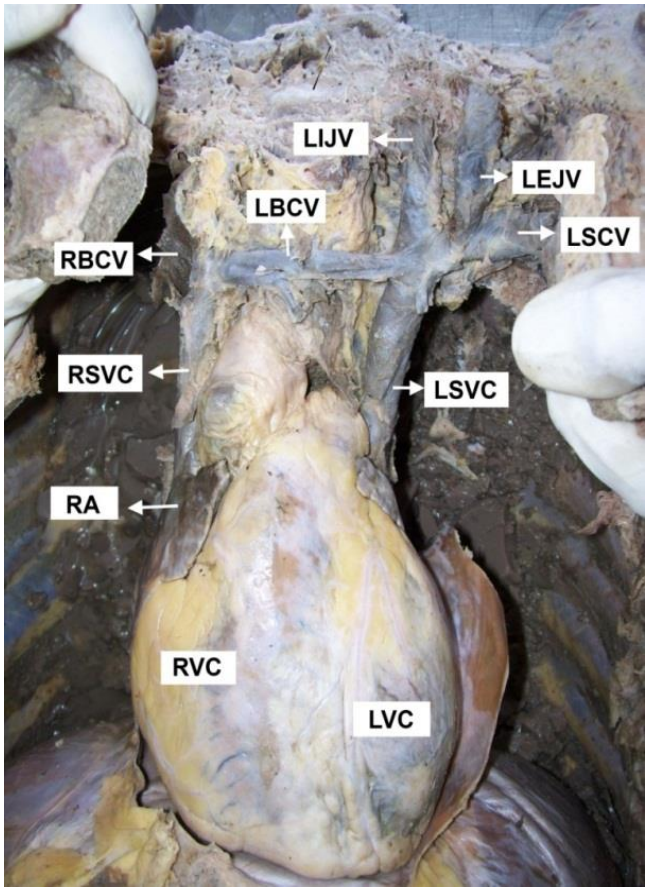

Fig 1: RVC – Right ventricle; LVC-Left ventricle; RA-Right atria; RSVC- Right superior vena cava; LSVC- Left superior vena cava RBCV- Right brachiocephalic vein; LSCV- Left subclavian vein; LEJV-Left external jugular vein; LIJV-Left internal jugular vein; LBCV-Left brachiocephalic vein.

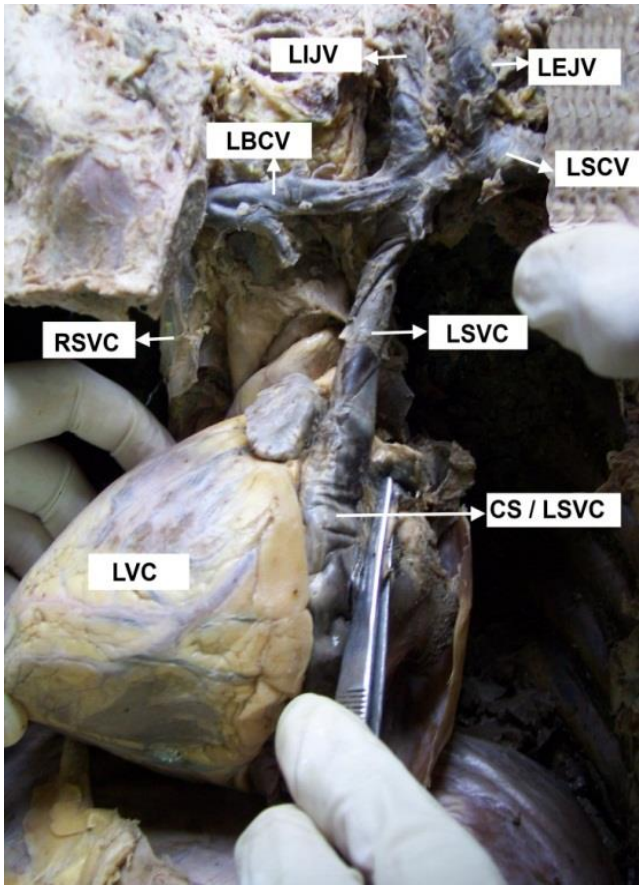

Fig 2: LVC-Left ventricle; RSVC- Right superior vena cava; LSVC- Left superior vena cava; RBCV- Right brachiocephalic vein; LSCV- Left subclavian vein; LEJV-Left external jugular vein; LIJV-Left internal jugular vein; LBCV-Left brachiocephalic vein; CS / LSVC-Coronary sinus/Left superior vena cava.

The persistence of the left superior vena cava (PLSVC) is a congenital anomaly resulting from failure of degeneration of the left cardinal vein. While the embryo grows, new organs appear and persist and others are transient and disappear. The development of the great systemic veins is a complex process and of clinical importance. In a 4 mm embryo, three main groups of veins are seen. These include the omphalo-mesenteric veins, the umbilical veins and the common cardinal veins. The anterior and posterior cardinal veins join to form common cardinal veins and enter the right and left horns of the sinus venosus. Due to the rightward direction of blood flow, the right horn of the sinus venosus develops. Additionally, the left common cardinal veins and the distal part of the left horn become atretic and is designated the ligament of Marshall or ligament of the left superior vena cava. The failure of the left anterior cardinal vein to obliterate results in PLSVC This vein generally drains to the coronary sinus and then into the right atrium. (Paval). The incidence of the persistence of the left superior vena cava is approximately 0.3% in the general population and in 3% to 10% of patients have additional cardiac defects.

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. (1, 2)

A PLSVC draining into the coronary sinus is not only the most common thoracic venous anomaly but also the most frequent cause of enlargement of the coronary sinus (4). In a majority of cases, there is a right superior vena cava also, but frequently there is hypoplasia or agenesis of the left innominate vein. In general, the sizes of the two venae cavae is

complementary; the larger the left superior vena cava, the smaller the right. Sometimes there is atresia of the right superior vena cava. In this instance, the anomaly is of major importance as the PLSVC is the sole route of venous return from the upper body. Not so harmless are the congenital atrioventricular conduction defects and arrhythmias that may occur in this setting Moreover, PLSVC with absence of the right superior vena cava complicates pacemaker lead implantation via the transvenous approach Persistent left superior vena cava is joined to the right superior caval vein by an innominate vein in about sixty per cent of cases). In this arrangement, the PLSVC can simply be clamped or ligated to avoid flooding the field when the heart is opened.

A prevalence of 3.3% has been recorded in the general population (3). A far less incidence, 0.5, has been recorded by Sharma (4).

The anatomy of the upper caval veins is important for pacemaker implantation and for cardiac surgery. The PLSVC is often found during surgery or catheterization due to the low frequency of presence of some diagnostic signs on the conventional chest X-rays. Some researchers have proposed some diagnostic features which include the widening of the aortic shadow, paramedian bulging and a paramedian strip or crescent along the left heart border on chest X-ray. The shadow of the PLSVC may also be seen along the left upper border of the mediastinum. Moreover, it was reported that when the chest radiograph shows the central venous catheter passing along the border of the left heart and good blood return through the catheter then a PLSVC would be suspected. It is also possible to diagnose PLSVC by angiography, echocardiography, computed tomography and magnetic resonance imaging. Diagnosis of PLSVC by echocardiography has 100% specificity and 96% sensitivity.

This PLSVC drains into the right atrium *via* the coronary sinus in 92% of cases (4, 5). 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.

The cyanotic cases have the risk of paradoxical embolus. Cerebral emboli and abscess have been reported in these patients. Detailed study of the anatomy of the venous system is essential before cavo pulmonary procedures such as Glenn anastomosis, bidirectional cavo pulmonary connection or Fontan-type procedure to determine the anatomical variants of the venous system. Intensive care clinicians should be aware of the presence of a left superior vena cava in order not to place catheters outside the venous circulation and to avoid complications like perforation, shock, cardiac arrest, cardiac tamponade and thrombosis.

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