Persistent left superior vena cava – an anomaly to remember

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Abstract

Persistent left superior vena cava (PLSVC) is the most common anomaly involving central venous return in thorax. Anatomically it is a mirror image of the right superior vena cava and is usually asymptomatic but can cause difficulties during Swan-Ganz catheterisation and insertion of pacing systems. This article presents a comprehensive review of this anomaly and clinical scenarios in which it can prove problematic, illustrated by an example.

Key words: left superior vena cava, congenital anomalies, persistent left superior vena cava (PLSVC).

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Introduction

Persistent left superior vena cava (PLSVC) is the most common anomaly involving central venous return in thorax. The problems that can crop up during Swan-Ganz catheterisation and insertion of pacing systems in patients having PLSVC have been extensively reported. We report the case of a 53-year-old male with PLSVC who underwent coronary revascularisation and closure of atrial septal defect, and also discuss the difficulties that can be faced by cardiologists and cardiac surgeons in such patients.

Case report

A 53-year-old male presented with worsening shortness of breath of one year duration associated with palpitations and limiting angina (CCUS III). He was orthopnoeic and had been having episodes of paroxysmal nocturnal dyspnoea. He was a known hypertensive on treatment. He was on maximal anti-anginal and heart failure medication at the time of presentation.

He had suffered a myocardial infarction (MI) four years previously and had developed severe bradycardia for which he required implantation of a permanent pacemaker. The pacing lead was placed through the left subclavian vein and a check X-ray showed the lead to be following an unusual route along the left border of the cardiac silhouette. A PLSVC was suspected, which was confirmed on transoesophageal echocardiography. A sinus venosus type of atrial septal defect was also identified. Cardiac catheterisation and coronary angiography showed that he had a non-dominant right coronary artery, significant stenosis in the circumflex and in the first and second obtuse marginal arteries. The left ventricle was dilated with mild mitral incompetence. He recovered well from his MI and was managed medically until progression of his symptoms.

Coronary revascularisation and closure of atrial septal defect was planned. A median sternotomy was performed. The ascending aorta, right superior vena cava (SVC) and inferior vena cava were cannulated. The left brachiocephalic vein was absent. The left SVC was a sizeable vessel opening into the coronary sinus, descending along the posterior aspect of the heart. It was not cannulated to avoid disrupting the pacing lead. When cardiopulmonary by-pass surgery was commenced, the venous return was insufficient, signifying the major contribution of the left SVC. The left SVC was cannulated after dissecting it up to its junction with the left subclavian vein, taking care to avoid damage to the pacing lead. The venous return improved remarkably. The right coronary artery and the obtuse marginal branch of the circumflex artery were grafted using saphenous vein grafts. A right atriotomy revealed a large coronary sinus opening and a small secundum type of atrial septal defect close to the right superior vena caval orifice. The septal defect was repaired by direct closure using 2'0 prolene. There were no post-operative complications and the patient was discharged on the seventh post-operative day. Pre-discharge echocardiography revealed good left ventricular function and an intact inter atrial septum.

Discussion

PLSVC is the most common congenital anomaly involving central venous return in the thorax.[1-3] Anatomically it represents the counterpart of right SVC, formed by the union of the left internal jugular vein and left subclavian vein. Normally it disappears in the embryological stage. Its incidence has been reported to be 0.3–0.5% in otherwise normal individuals and about 3–10% in patients with other congenital heart anomalies.[4-6] It remains asymptomatic in the majority of patients until they reach adulthood. It is most commonly detected coincidentally on a chest X-ray, following central venous catheter insertion through the left subclavian vein, where the catheter is seen to take an unusual route. PLSVC usually drains the systemic venous blood from the

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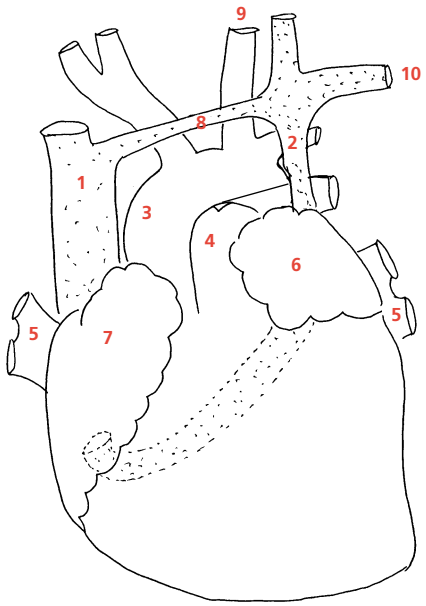
Figure 1. Anatomical relationships of the persistent left superior vena cava (SVC) (anterior view). Note the relation of the left SVC passing anteriorly to the left pulmonary artery and pulmonary veins. It then enters the atrioventricular groove posterior to the left auricular appendage to join the coronary sinus and proceeds to end in the right atrium on the diaphragmatic surface of the heart



Key: 1 = right superior vena cava; 2 = persistent left superior vena cava; 3 = ascending aorta; 4 = pulmonary artery; 5 = pulmonary veins; 6 = left auricular appendage; 7 = right auricular appendage; 8 = brachiocephalic vein (can be rudimentary); 9 = left internal jugular vein; 10 = subclavian vein

left upper half of the body into the coronary sinus. However, in some it drains into the left atrium. It could be a part of complex congenital anomalies like unroofed coronary sinus, where the left SVC opens into the left atrium and is associated with atrial septal defect.[7] In such cases it causes problems when floating Swan-Ganz catheters through the left subclavian vein. Sometimes it is only identified at the time of cardiac surgery, when it can cause problems with the venous return for cardiopulmonary bypass.

Anatomy

Anatomically the PLSVC can be imagined as a mirror image of the right SVC originating with the confluence of the left subclavian vein and internal jugular vein behind the left sternoclavicular joint. It descends down the left border of superior mediastinum, anterior to the left pulmonary artery and posterior to the left atrial appendage, where it enters the atrioventricular groove and joins the coronary sinus. It terminates variably into the left or right atrium. (figure 1) Variations in the presence and absence of both vena cava can occur ranging from complete absence of the right SVC to presence of bilateral SVC and the presence of a rudimentary SVC, either right or left. In one study the PLSVC was found to be present in 2.6% of the patients with congenital anomalies and 67% of these had two vena cavae.[8] In another study, PLSVC with absent right SVC was found in 0.15% of patients (six of 4,100) at cardiac catheterisation for suspected congenital cardiac disease.[9]

Embryology

The cardinal veins constitute the main venous return to the embryonic heart (figure 2a–c), with the anterior cardinal veins draining the cranial part of the body and the posterior cardinal veins draining the caudal part. The left brachiocephalic vein is formed in the eighth week of gestation connecting both the anterior cardinal veins. The caudal part of the right anterior cardinal vein distal to the junction to left brachiocephalic vein forms the right SVC. The portion of the left anterior cardinal vein distal to the left brachiocephalic vein usually degenerates, but is represented distally as the coronary sinus in the atrioventricular groove. Persistence of this middle portion results in PLSVC (figure 2d). This is commonly associated with a small or absent anastamosis between the SVCs that form the left brachiocephalic vein.[10]

Investigations

A routine chest X-ray shows PLSVC as a widening of the aortic shadow with a mediastinal bulge under the aortic arch, or as a strip along the left margin of the mediastinum in the upper part of cardiac silhouette.[11] Following insertion of the central venous catheter through the left subclavian vein, a check X-ray would show the catheter to take a course along the left border of the cardiac silhouette.

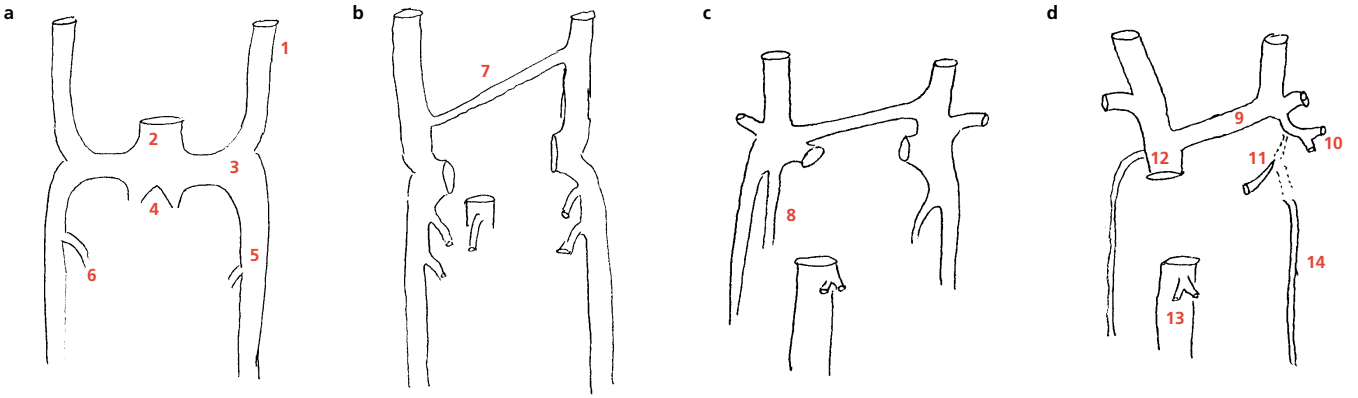
Two dimensional transthoracic echocardiography (TTE)[4] shows dilation of the coronary sinus on parasternal long- and short-axis views and apical four-chamber view angulated posteriorly. PLSVC can be confirmed by injection of echo contrast material into the left arm vein, which opacifies the coronary sinus and the right atrium. Injections of the echo contrast material into the right arm will opacify only the right atrium. At Doppler echocardiography the abnormal contour of the left atrium may be mistaken for a mass.

Transoesophageal echocardiography (TOE)[12] is a more sensitive modality for the diagnosis of PLSVC and the associated anomalies than transthoracic studies. First pass radionuclide angiography in the right or left anterior oblique projection is another important modality for recognising PLSVC.[13] Computerised tomography[14] can also be used to visualise the PLSVC, especially with contrast. Magnetic resonance imaging[14] shows more accurate images of the anomaly.

Clinical implications

Clinically, most of the time, a PLSVC is asymptomatic and is discovered incidentally. But it can cause problems while floating Swan-Ganz catheters through the left subclavian vein, especially if the PLSVC opens into the left atrium. It might show damped central venous tracing while inserting central venous lines for monitoring central venous pressure. A longer length of pacemaker wires may be needed to start pacing the right chambers

Figure 2. Drawings showing the normal embryology of the central venous system. **a–c** show the primitive venous system in the early embryo, **d** shows normal adult central venous system (dotted line represents PLSVC)



Key: 1 = precardinal veins; 2 = sinus venosus; 3 = common cardinal vein; 4 = hepatico-cardinal vein; 5 = post-cardinal vein; 6 = subcardinal vein; 7 = oblique intercardinal anastamosis; 8 = azygous vein; 9 = brachiocephalic vein; 10 = superior intercostal vein; 11 = oblique vein and ligament of left atrium; 12 = right superior vena cava; 13 = inferior vena cava; 14 = hemi-azygous vein

because the wires have to course through the coronary sinus before reaching the right atrium.

The incidence of post-operative supra ventricular arrhythmias is high in patients with PLSVC. It has been hypothesised that a dilated coronary sinus may stretch the atrioventricular node and bundle of His, hence predisposing these patients to arrhythmias.[15]

Surgical implications

PLSVC may be found at the time of cardiac surgery. Cardiac surgery in patients with known persistent left superior vena cava should always be planned. Having the following information pre-operatively is helpful:

- The patency of the right superior vena cava
- The presence of a left brachiocephalic vein that allows snaring of left superior vena cava and therefore avoids cannulation or allows ligation of the anomalous vessel
- The size of the anomalous vessel. This gives an estimate of the venous drainage and the need for cannulation. This can be assessed using transoesophageal echocardiography.
- The site of drainage of the anomalous vessel.
- Presence of other congenital cardiac anomalies.

Intra-operatively the following difficulties could be faced:

- Inadequate venous return may result if the PLSVC is a significant sized vessel.
- If retrograde cardioplegia is planned, there will be loss of cardioplegic solution into the systemic circulation and inadequate myocardial protection,[16] so an alternative method of myocardial protection should be planned or the PLSVC either clamped temporarily or ligated.
- An existing pacing system through the anomalous left vein may cause problems during cannulation.
- Inadvertent ligation of the PLSVC should be avoided because there may be associated coronary sinus ostial stenosis; this may be the only route of drainage for the cardiac venous blood. Ligation in such circumstances can lead to myocardial ischaemia and necrosis.[17]

Key messages

- Persistent left superior vena cava is an anomaly to remember
- It is usually asymptomatic and associated with other anomalies
- An abnormal course of central catheter should lead to suspicion
- Further investigation is needed to rule out other anomalies
- Operative strategy should be planned; additional procedures may be necessary

Treatment

Most of the patients with PLSVC are asymptomatic. In case of a left to right shunt, the PLSVC can be ligated just above the level of left atrium if there is an alternative means of drainage into the right atrium. If the PLSVC is the only vein draining the upper body, then the ASD or the intra-cardiac lesion needs to be corrected.

Conclusion

PLSVC is a common anomaly that can pose challenges on occasions to both cardiologists and cardiac surgeons, hence awareness of this 'stranger' in the chest is essential.

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