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Case Report

Anomalous drainage of left superior venacava into left atrium

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ABSTRACT

The anomalies of drainage of systemic venous communications to the heart are many. But only few cause significant hemodynamic alteration and thus, complications. When they do, they need to be surgically corrected. We report an unusual case of a patient who was found to have left pulmonary veins draining into the left superior venacava (LSVC), which in turn was opening into the roof of the left atrium that had unroofed coronary sinus morphology. Innominate vein was absent. LSVC was divided cranial to where the pulmonary veins were draining into it. Lower end was suture closed. Proximal part of the LSVC was anastamosed to left pulmonary artery.

Keywords: LSVC, Unroofed coronary sinus, Pulmonary venous drainage anomalies

INTRODUCTION

Anomalies of the systemic venous connections and drainage are numerous. Often, they are incidentally detected. They may, at times, behave like right to left shunts causing cyanosis, paradoxical embolism or a stroke.[1] Among these, a persistent LSVC is the most common anomaly occurring in 0.5% of the general population and in 4% of patients with congenital heart disease.[1] Association of an unroofed coronary sinus with a persistent left superior venacava is a rare anamoly, first described by Raghib et al in 1965.[2] Because of nonspecific clinical features, this condition is often incidentally detected on imaging.[3]

CASE REPORT

A 22 year old patient was referred with diagnosis of partial atrio-ventricular canal defect. Blood investigations were normal. ECG showed left axis deviation, right ventricular hypertrophy with a complete heart block. 2D Echocardiography revealed a trifoliate left atrio-ventricular valve with mild regurgitation. On operating table, a good sized right superior venacava was present but no innominate vein. Patient had an LSVC into which all the left pulmonary veins were draining. LSVC was opening onto the roof of the left atrium near the left atrial appendage and had an unroofed coronary sinus. After arresting heart with cold blood cardioplegia, right atrium was opened, anatomy assessed. The left atrioventricular valve was repaired with interrupted 5-0 prolene sutures to abolish regurgitation. Atrial septation was done with native pericardium using 4-0 continuous prolene sutures. LSVC was divided cranial to the site where the pulmonary veins were draining into it. Distal end was suture closed using continuous 5-0 prolene suture. Proximal part of the transected LSVC was anastamosed to the left pulmonary artery. Postoperatively, a permanent pacemaker was implanted as the patient had a complete heart block pre-operatively.

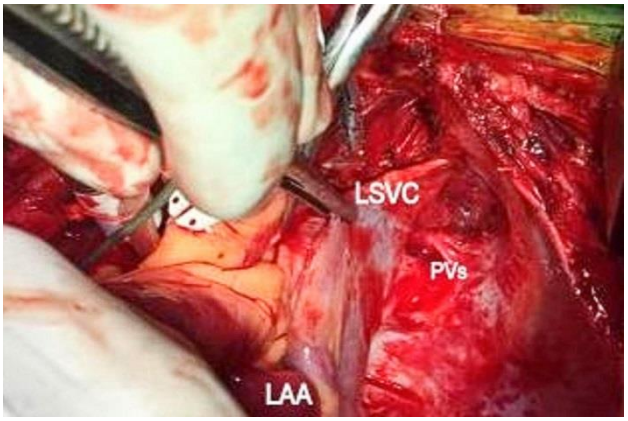

Figure 1: Intraoperative image showing the left pulmonary veins draining into LSVC; LSVC is in turn draining into the roof of the left atrium near the left atrial appendage.
LAA- left atrial appendage; PVs-pulmonary veins.

Figure 2: Transected lower end of the LSVC was closed with continuous 5-0 prolene suture; pulmonary veins draining into this part of the LSVC now drain through it into the left atrium, which is physiological.

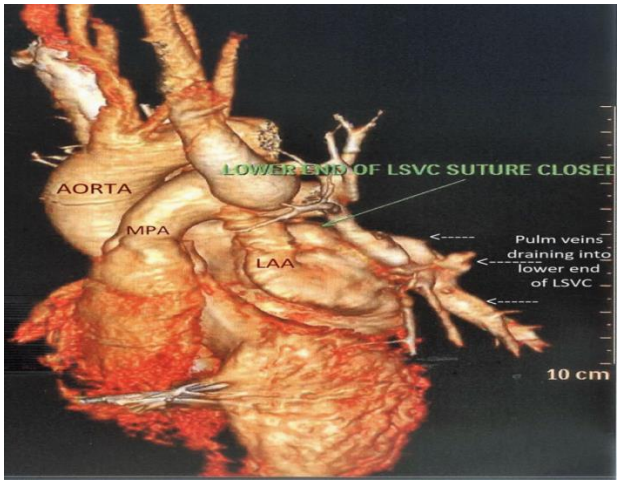

Figure 3: Postoperative computer tomography scan imaging showing the pulmonary veins draining into the lower end of the transected left superior venacava and through it drain into the left atrium.
MPA- main pulmonary artery; LAA- left atrial appendage.

DISCUSSION

Pulmonary veins may drain anomalously into the innominate vein, right atrium or coronary sinus.[4] When right pulmonary veins drain through sinus venosus defects into the SVC or IVC, they are re-routed into left atrium by intra-atrial baffling using native pericardial patch with closure of the ASD. If the anomalous vein joins IVC too deep below the diaphragm, it can be ligated, divided and anastamosed to the left atrium.[5] Anomalous drainage of the left pulmonary veins constitute only 10% of all PAPVC and drainage may be into the innominate vein through a vertical vein or to the coronary sinus. The vertical vein is ligated high up, transected and the transected end is anastamosed to the left atrium.[5] When they drain into coronary sinus, defect is repaired by excision of the septum between the coronary sinus and the ASD, and by a pericardial patch closure of the defect.[5]

LSVC results due to persistence of left anterior and left common cardinal veins was first described by McCotter in 1916. It presents as an isolated anomaly or as part of complex congenital cardiac defects. Often, LSVC drains into coronary sinus; rarely, into the IVC or hepatic veins. It can drain into roof of the left atrium, either through an unroofed coronary sinus or as a direct communication into the roof of left atrium or to the left superior pulmonary vein.[1,6] When it drains into the left atrium, it causes a right to left shunt predisposing to paradoxical embolism, brain abscess or stroke. In the majority of these cases, the right SVC is also present and drains normally. There is absence of innominate vein in 65% of such cases.[1,6] Diagnosis of LSVC is made on transthoracic echocardiography showing a dilated coronary sinus and can be confirmed with agitated saline contrast echochardiography. It is most often an incidental finding in cardiovascular imaging or surgery.[6]

LSVC without unroofed coronary sinus can be dealt with by ligating it when there is an adequate sized innominate vein and RSVC. Alternatively, it can be divided and re-implanted into the right atrium, right superior venacava or pulmonary artery. In case of a short LSVC, an interposition conduit may be necessary to reach the appropriate re-implantation site.[7] Its technically difficult to place pacemaker leads, hemodialysis catheters or Swan-Ganz catheters by catheterizing the LSVC. Supraventricular tachycardia, atrial fibrillation and sudden death may occur as there is a possibility to irritate the coronary sinus, where an ectopic pacemaker can be present.[8] Administering retrograde cardioplegia is difficult in the presence of an LSVC.[8] LSVC has been associated with architectural abnormalities of the sinus node and conduction tissue.[9]

We found no case reports of persistent LSVC draining the pulmonary veins into left atrium with unroofed coronary sinus morphology. The closest reference, we found was that of an isolated right PAPVC with

concomitant anomalous connection of the RSVC to left atrium.[10]

CONCLUSION

A persistent LSVC is a very common anomaly present in about 0.5% of the general population and often associated with numerous complex congenital heart defects. However, drainage of left pulmonary veins into the right atrium through the LSVC is a very rare anomaly. Rarer still, is the association of unroofed coronary sinus morphology with these defects. When an anomaly causing complications such as our case is found, it should be corrected surgically.

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