Case Report

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Persistent left superior vena cava and its clinical implications: case report and literature review

Abstract

The persistent left superior vena cava venous anomaly is the most common congenital malformation of the thoracic venous system with an estimated prevalence of 0.3 to 0.5%. After dissection of a 30-week-old male fetus, was observed a persistent left superior vena cava that drained into the left atrium. Understanding this anomaly is crucial, given the widespread use of invasive cardiac and electrophysiological procedures.

Keywords: anatomical variation, left superior vena cava, left atrium, congenital heart disease, fetal echocardiography, embryology

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Abbreviations: AA, aortic arch; BA, brachiocephalic artery; LA, left atrium; LCCA, left common carotid artery; LIJV, left internal jugular vein; LIPV, left inferior pulmonary vein; LL, left lung; LSA, left subclavian artery; LSV, left subclavian vein; LSVC, left superior vena cava; LUPV, left upper pulmonary vein; LV, left ventricle; PAT, pulmonary artery trunk; RA, right atrium; RCCA, right common carotid artery; RIJV, right internal jugular vein; RL, right lung; RSA, right subclavian artery; RSV, right subclavian vein; RSVC, right superior vena cava; RV, right ventricle; PLSVC, persistent left superior vena cava

Introduction

Persistent left superior vena cava (PLSVC) is the most common systemic venous return anomaly, with an estimated prevalence of 0.3-0.5% in the general population, of 4-8% in patients with congenital heart disease[1–4] and 0.9% in the fetal population.[5]

The origin of the PLSVC is not yet completely clear, it is believed to be the result of an intrauterine failure of the development of the left anterior cardinal vein, resulting in the bilateral presence of superior vena cava,[6] but in the last years the prenatal diagnosis of PLSVC has been done based on an abnormal view of three vessels in the upper mediastinum, showing a supernumerary vessel to the left of the pulmonary trunk and arterial duct.[7]

Although there are many divergences regarding the true prevalence, the PLSVC does not have major clinical impacts, as systemic venous blood continues to return to the right atrium through the coronary sinus, and there is usually a normal right superior vena cava.[8,9]

The recognition of PLSVC is of great importance for anatomists, clinicians and surgeons, especially in the implantation of long-term central venous catheter.[10–12] Thus, the present article aimed to report a finding of a persistent left superior vena cava in a human fetus corpse and to review the literature.

Case report

During a routine dissection at the anatomy laboratory of the Federal University of Sergipe, São Cristóvão, SE, Brazil, it was observed in a male human fetus, with an estimated age of 30 weeks (hallux-calcaneus measurement), a persistent left superior vena cava (Figure 1). The left superior vena cava was lateral to the left common carotid artery and anterior to the left subclavian artery, had a caudal direction, passed lateral to the pulmonary artery trunk and aortic arch, ventral to the left lung hilum and flowed into the left atrium along with the pulmonary veins (Figure 2). The right superior vena cava also followed a descending path along the right upper part of the mediastinum and ended into the right atrium.

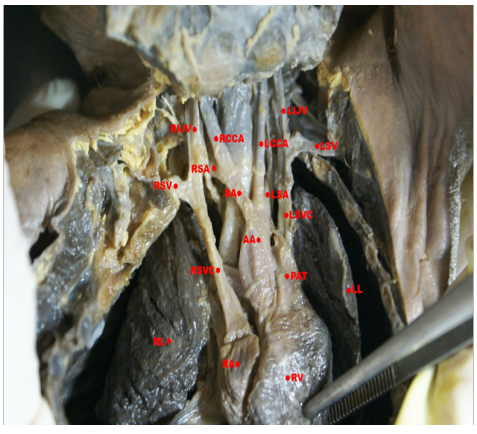

Figure 1 Persistent left superior vena cava.

AA, aortic arch; BA, brachiocephalic artery; LCCA, left common carotid artery; LIJV, left internal jugular vein; LL, left lung; LSA, left subclavian artery; LSV, left subclavian vein; LSVC, left superior vena cava; PAT, pulmonary artery trunk; RA, right atrium; RCCA, right common carotid artery; RIJV, right internal jugular vein; RL, right lung; RSA, right subclavian artery; RSV, right subclavian vein; RSVC, right superior vena cava; RV, right ventricle.

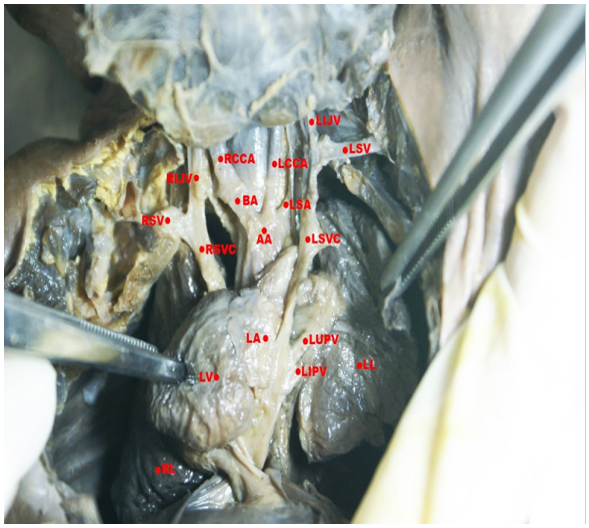

Figure 2 Persistent left superior vena cava draining directly into the left atrium.

AA, aortic arch; BA, brachiocephalic artery; LA, left atrium; LCCA, left common carotid artery; LIJV, left internal jugular vein; LIPV, left inferior pulmonary vein; LL, left lung; LSA, left subclavian artery; LSV, left subclavian vein; LSVC, left superior vena cava; LUPV, left upper pulmonary vein; LV, left ventricle; RCCA, right common carotid artery; RIJV, right internal jugular vein; RL, right lung; RSV, right subclavian vein; RSVC, right superior vena cava.

Discussion

The persistence of the left superior vena cava has been observed in the general population, being the most common abnormality of intrathoracic vessels,[8] caused by variations in the embryological development of the thoracic venous system.[13] This abnormal vein originates from the left anterior cardinal and common cardinal veins, and normally flows into the right atrium through the coronary sinus.[14,15]

The anomalous systemic venous connection is a rare abnormality, which can be completely asymptomatic, as the persistent left superior vena cava drains in more than 90% into the coronary sinus, but can also drain directly into the left atrium in just 0.03% of the population, and in this case causing cyanosis.[16,17] In our case, the persistent left superior vena cava drained directly into the left atrium.

Drainage to the left atrium is often associated with atrial septal defect and can be seen in the absence of a coronary sinus,[18] with the coronary sinus orifice persisting as an interatrial communication.[19] The persistent left superior vena cava is associated with varied cardiac structural abnormalities, including bicuspid aortic valve, atrial septal defect, coronary sinus ostia atresia and aortic coarctation.[3]

In most cases, these variations are clinically silent and can be discovered accidentally in radiological studies done for other reasons. However, they should be kept in mind when these patients are candidates for insertion of central venous catheters or cardiothoracic surgery.[20,21]

In patients with known congenital anomalies of the superior vena cava undergoing insertion of central venous catheter insertion, several important aspects must be considered for a safe and effective procedure: accurate preoperative planning with image description of the anomalies of the veins, careful selection of the most appropriate vascular access and the use of ultrasound and fluoroscopic guidance by qualified professionals,[22] which is of great importance in all invasive and non-invasive procedures, such as transthoracic echocardiography and cardiac tomography.

Conclusion

Persistent left superior vena cava is a rare abnormality of systemic venous return often associated with other congenital cardiac malformations and physicians should be aware of such anomaly and its clinical implications in order to avoid possible complications.

Acknowledgments

None.

Conflicts of interest

The authors declare there are no conflicts of interest.

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