CASE REPORT

Persistent Left Superior Vena Cava Drain Into the Left Atrium: A Rare Cause of Hypoxia in Childhood

Anis Munirah Mohd Kori[1,2]

[1.] Department of Paediatrics, School of Medical Sciences,Universiti Sains Malaysia, 16150 Kota Bharu, Kelantan,Malaysia
[2.] Hospital Universiti Sains Malaysia (HUSM), Health Campus,Kubang Kerian, 16150 Kota Bharu, Kelantan,Malaysia

ABSTRACT

Persistent left superior vena cava drain into the left atrium is frequently overlooked since it rarely has a major influence on haemodynamic in children. Therefore, it is frequently asymptomatic and accidentally found during catheterization and imaging modalities. It has the potential to mimic pulmonary disease, especially in individuals who do not have a congenital cardiac anomaly. We report the case of a five-year-old boy who had recurrent pneumonia with persistent hypoxaemia and was discovered to have persistent left superior vena cava into the left atrium by computer tomography pulmonary angiography. Due to the lifetime risk of emboli and bacteraemia from right to left shunting, the left superior vena cava must be terminated either percutaneously or surgically. *Malaysian Journal of Medicine and Health Sciences (*2024) 20(4): 400-402. doi:10.47836/mjmhs20.4.50

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Corresponding Author:
Anis Munirah Mohd Kori,Master of Medicine (Paediatrics)
Email: anismun@usm.my
Tel: +603-79760000

INTRODUCTION

Persistent left superior vena cava (PLSVC) is the most common thoracic venous abnormality, accounting for 0.3% of all cases [1]. It is often asymptomatic and is found accidentally on thoracic imaging modalities or during vascular catheterization. In general, 90% of PLSVC drain into the right atrium via the coronary sinus (CS) and have no haemodynamic effect [2]. Even though it is associated with a ten-fold increase in congenital heart defect (CHD), its solitary finding is usually benign, unless it drains into the left atrium (LA), which increases the risk of bacteraemia and septic emboli due to right to left shunting [1]. It causes mild hypoxaemia in children without causing haemodynamic abnormalities, therefore it may mimics the presence of lung illness in those who do not have CHD. Nevertheless, patients are frequently asymptomatic because a greater volume of venous blood returns via the inferior vena cava, empties into the right atrium, and is normally oxygenated. Cyanosis is typically a significant finding; however, it may be undetected in these patients when transthoracic echocardiography is performed on the suspicion of congenital anomalies of the heart chambers, the results are typically normal causing the right-to-left shunt to go undetected[2]. We report the case of a five-year-old boy who had recurrent pneumonia with persistent hypoxaemia and was discovered to have PLSVC into the LA by computer tomography pulmonary angiography (CTPA).

CASE REPORT

A five-year-old boy with no known medical illness had a history of recurrent pneumonia since the age of three. Within a year, he experienced a cumulative occurrence of three episodes of pneumonia. During his first and second admissions, he required supplementary oxygen with no evidence of persistent hypoxemia. Serial chest radiographs revealed interstitial lung changes which, in conjunction with clinical symptoms and biochemical parameters, were most likely caused by a viral infection of the lungs. Recently, he presented with fever and fast breathing for three days and was diagnosed with viral pneumonia. Although his lung pathology was resolved, he had low oxygen saturation under room air which ranging between 88% and 92%.

Upon examination, the patient appeared to be in good general condition, with finger and foot clubbing. His vital signs: blood pressure of 85/60 mmHg, respiratory rate of 28 breaths per minute, oxygen saturation of 89% on room air, heart rate of 100 beats per minute and temperature of 37.3 °C. Auscultation of the lungs and heart were

normal. Laboratory analysis revealed haemoglobin concentration of 14.5 g/dL. Electrocardiography (ECG) with 12 leads revealed sinus rhythm. Chest radiography showed clear lung field and normal cardiothoracic index. Transthoracic echocardiography revealed that the pressure in the pulmonary artery, ejection fraction, and cardiac chambers were all within normal limits.

He was scheduled for CTPA because of the unexplained hypoxemia. The presence of PLSVC draining into the LA was confirmed (Figure 1a, b, c). A thin bridging vein linking the right superior vena cava and LSVC was observed (Figure 2a, b). The remaining pulmonary and systemic venous drainage is normal. Persistent right-to-left shunting posed a lifetime risk of infection and thromboembolic events; therefore, he was referred to a cardiac surgeon for surgical intervention.

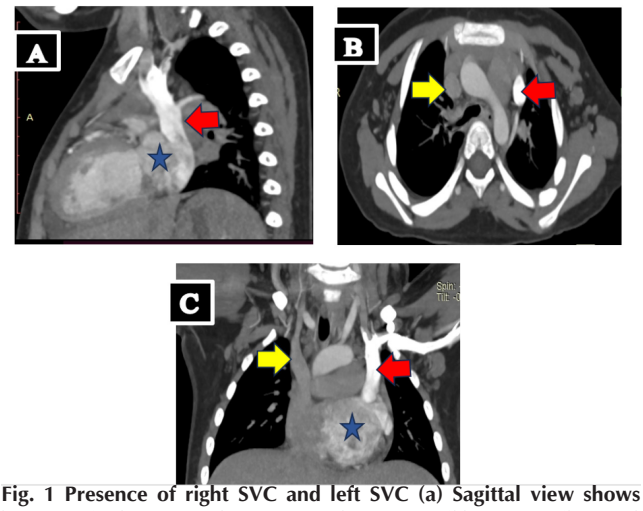

Fig. 1 Presence of right SVC and left SVC (a) Sagittal view shows left SVC (red arrows) drain inferiorly into LA (blue stars) (b) Axial view (c) Coronal view show left SVC having similar sizes to right SVC (yellow arrows)

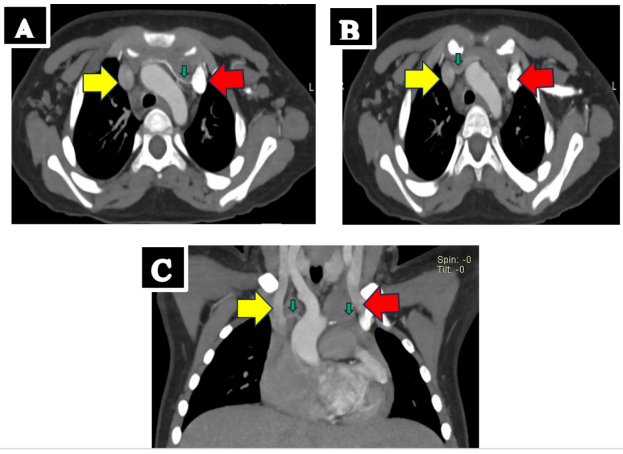

Fig. 2 (a,b) Axial view (c) Coronal view presence of hypoplastic left brachiocephalic vein / bridging vein (green arrows) connecting right SVC (yellow arrows) and left SVC (red arrows)

DISCUSSION

PLSVC is characterised by the elimination of the common cardinal vein and the proximal portion of the right anterior cardinal vein, in addition to the persistence of the left anterior cardinal vein [1]. When the PLSVC drains into the left atrium, as in our patient, venous drainage destined for the right side of the heart (RA) is rerouted to the left side of the heart (LA). The potential cause of intermittent or absent cyanosis is the existence of a bridging vein, which functions as an interatrial conduit [2]. This explains why the cyanosis was detected late, despite the patient had been admitted twice previously for pneumonia.

PLSVC is predominantly asymptomatic, the majority of diagnoses were coincidental. In this case, it is also challenging to detect this anomaly via echocardiography, as the absence of dilated CS would preclude suspicion of PLSVC. However, considering the patient's medical history of unexplained chronic hypoxia, it is recommended to perform an agitated saline contrast study using echocardiography through the left arm in order to detect immediate air microbubbles in the left atrium. Several case reports have documented the use of this technique for confirming the diagnosis without the need for additional detailed imaging modalities such as CT and magnetic resonance imaging (MRI) [1]. The differential diagnosis for asymptomatic cyanosis with a normal echocardiography includes pulmonary arteriovenous malformation or congenital methemoglobinemia [3]. It is crucial to elucidate the duration of cyanosis and any symptoms associated with the cardiovascular and pulmonary systems during the medical history.

PLSVC can cause problems during establishing central venous access, pacemaker implantation (due to the tortuous course of the electrode), or cardiopulmonary bypass (isolated PLSVC impairs the use of retrograde cardioplegia). In the presence of PLSVC, central venous access via the femoral vein is a safer choice in case of this anatomical variation. When implanting permanent pacemakers, the left subclavian vein is preferred, as lead manipulation is easier [4]. Anatomy variation awareness can provide a valuable opportunity to enhance our understanding of cardiovascular anatomy and physiology, and this case is an example of the importance of thorough anatomical knowledge and clinical awareness in our clinical practice.

Failure to correct the problem prior to old age increases the likelihood of developing septic emboli and bacteraemia. Few case reports from the adult

population revealed that the majority of complications involved stroke and abscess [2]. The lungs function as a mechanical filter for the blood, and the existence of a right-to-left shunt may permit septic emboli to enter the arterial circulation[4].In order to eliminate these lifetime risks, it is recommended to terminate the connection between LSVC and LA. Although the guidelines for when to treat PLSVC are uncertain, expert opinion indicates that correction should be considered when significant shunting and symptomatic hypoxemia are present[1]. Multiple surgical options are available, there are include ligation of PLSVC, intra-atrial baffle formation, transection of PLSVC with left atrial tissue and implantation to the right atrium, end-to-side anastomosis of PLSVC to the left pulmonary artery and performing end-to-side anastomosis to the right SVC under or over the aortic arch [4]. The outcome of surgical correction is good with low mortality and morbidity. Despite that, percutaneous closure could prevent the need for a sternotomy and shorten the length of hospital stay[4]. Percutaneous closure with an Amplatzer device of a PLSVC draining into the left atrium is feasible. A thorough evaluation of collateral vessels that can run off venous blood during PLSVC occlusion is essential to prevent venous hypertension. Furthermore, it is crucial to determine the precise location of azygous vein drainage to the PLSVC, as the device must be positioned proximate to the junction of the azygous vein and PLSVC to prevent any residual right-to-left shunting [5]. It is a relatively new procedure in our country; therefore, the patient has to be referred to a surgeon for correction.

CONCLUSION

PLSVC draining into LA is uncommon and typically results in low oxygen saturation, which is discovered incidentally in children. For the detection of this anomaly, which is frequently overlooked, particularly in patients without CHD, contrast transthoracic echocardiography is of the utmost importance. Early detection and appropriate intervention are essential in order to prevent emboli and abscesses in adults

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