CASE REPORT

Persistent left superior vena cava draining into the left atrium presenting as secondary polycythaemia: A case report

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Abstract

Persistent left superior vena cava (PLSVC) is a rare vascular anomaly which is often asymptomatic yet is potentially life-threatening in cases of abnormal drainage. We present the case of a 46-year-old male experiencing acute dyspnoea attributed to PLSVC. PLSVC, often accompanied by a right superior vena cava, may lead to systemic complications such as hypoxia, cyanosis, and paradoxical embolism. Surgical intervention may be warranted in symptomatic cases. This report underscores the importance of considering PLSVC in differential diagnoses of acute respiratory distress, particularly in individuals with unexplained polycythaemia. Heightened clinical awareness and timely management are crucial in mitigating potential complications associated with this vascular anomaly.

Keywords: Vena cava, superior; Vascular malformations; Tomography, X-ray Computed; Hypoxia; Dyspnoea.

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Introduction

Persistent left superior vena cava (PLSVC) is a rare and generally asymptomatic vascular defect of the systemic venous circulation.[1] It is prevalent in the general population and in patients with congenital heart disease up to 0.3–05% and 12%, respectively. The cranial part of the embryo is drained by anterior cardinal veins. The left and right anterior cardinal veins are connected by an oblique anastomosis which later develops into the left brachiocephalic vein. Furthermore, the left anterior cardinal vein and the left common cardinal vein which are located caudal to the oblique anastomosis eventually degenerate. The former develops into the terminal part of left superior intercostal vein whereas the latter forms the ligament of left vena cava and the oblique vein of the left atrium. However, PLSVC is formed when there is failure of regression of the left anterior cardinal vein.[2] Even though PLSVC is asymptomatic in most individuals, life threatening complications, such as flash pulmonary oedema, can ensue in some individuals.[3] In addition, intracardiac shunting due to drainage of venous blood into the left atrium can lead to complications such as hypoxaemia, cyanosis, and paradoxical embolisation.[4] The presence of a normal right superior vena cava alongside PLSVC often results in the anomaly remaining undetected in the majority of cases. Although PLSVC drains into the left atrium in a minority of patients, this may not always produce significant haemodynamic consequences. However, in some cases, right-to-left shunting can occur, leading to varying degrees of systemic hypoxaemia and clinical manifestations such as cyanosis and dyspnoea.[1,4]

Additionally, PLSVC has important clinical implications during invasive procedures. Cannulation of an unrecognised PLSVC may complicate central venous access.[5]

The objective of this case report is to highlight and analyse the clinical and haemodynamic significance of persistent left superior vena cava draining into the left atrium presenting as secondary polycythaemia.

Case report

The case under consideration involves a 46-year-old male who presented with an episode of acute dyspnoea, rousing him from sleep, during which his roommate observed him gasping for air. Subsequently, he was taken to the Emergency Department of Shaukat Khanum Memorial Hospital, Peshawar, in a wheelchair in March 2024. Initial evaluation revealed mild hypoxia, leading to the initiation of supplemental oxygen at a rate of two litres per minute. His past medical history was significant for similar episodes, with relief noted upon exposure to fresh air. Notably, the patient had a history of chronic smoking but lacked significant comorbidities. Systemic assessment yielded unremarkable findings. Further diagnostic endeavours, such as encompassing baseline investigations, arterial blood gas analysis, electrocardiography, chest radiography, and D-dimer testing, were undertaken. Results revealed type-1 respiratory failure on blood gas analysis and a

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haemoglobin level of 18.6 gm/dl (N- 13.5 to 17.5 gm/dl per on complete blood count, consistent with previously documented polycythaemia since July 2022. Electrocardiography showed non-specific T wave abnormalities. In light of these observations, a Computed Tomography Pulmonary Angiogram was conducted to assess a pulmonary embolus. Although the presence of pulmonary embolism was excluded, an incidental discovery of a separate left superior vena cava draining into the left atrium was noted (Figure 1). As the dyspnoeic episode was transient, subsequent management involved the cessation of oxygen therapy with the patient maintaining satisfactory baseline oxygen saturation ranging between 92–94% without supplemental oxygen. Additionally, the chest radiograph showed lateralisation of vessels. Normally, no opacity should be present lateral to the aorta in a normal chest radiograph, however, in this patient an additional shadow (Figure 2) could be seen representing the presence of PLSVC. Further investigations, including Echocardiography and Pulmonary Function Test, were unremarkable. The patient was managed by the respiratory team, and further evaluation for underlying causes of secondary polycythaemia was planned; however, follow-up data was not available at the time of writing this report.

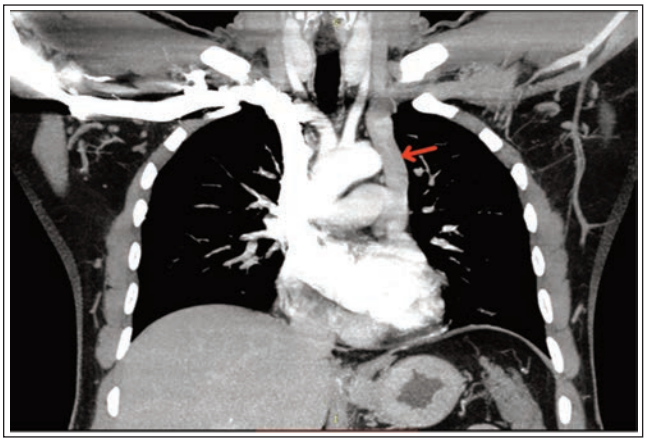

Figure-1: Coronal view of the computed tomography pulmonary angiogram showing persistent left superior vena cava opening into the left atrium as indicated by the arrow.

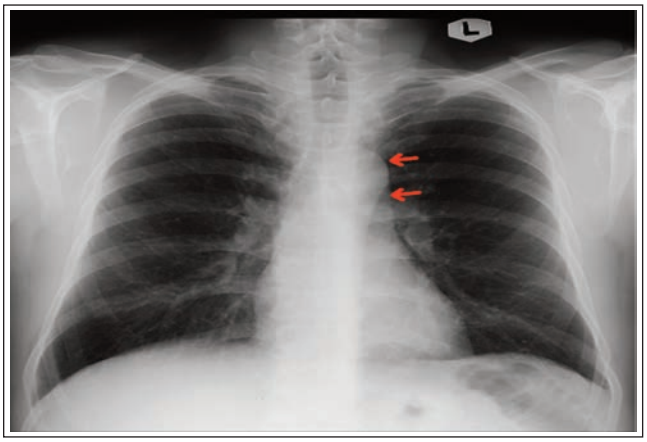

Figure-2: Chest radiograph showing an additional opacity lateral to the aortic knuckle indicating persistent left superior vena cava as shown by the arrows.

Discussion

PLSVC drains into the coronary sinus in close proximity to the atrioventricular valves in approximately 90 percent of the cases and in 10 percent of the cases it opens directly into the left atrium or one of the pulmonary veins which can lead to a right to left shunt. The PLSVC occurs in conjunction with a right superior vena cava in 82-90 percent of cases.

On echocardiography, a dilated coronary sinus may be observed, and the presence of PLSVC can be confirmed by injection of agitated saline into both arms, demonstrating sequential opacification of the coronary sinus and the right atrium. Direct visualisation of PLSVC and its anatomical variations can be achieved using multi-slice computed tomography or magnetic resonance imaging.[5]

In the majority of individuals, there are no significant clinical consequences of PLSVC as venous blood can return to the right atrium through a normal right superior vena cava.[1] Due to variations in its course and drainage, PLSVC may occasionally result in blood shunting. Drainage into the right atrium via the coronary sinus is typically haemodynamically insignificant; however, un-roofing of the coronary sinus may create a communication with the left atrium. In cases where PLSVC drains directly into the left atrium, a functional right-to-left shunt can occur, allowing deoxygenated blood to bypass pulmonary circulation and enter the systemic circulation, leading to hypoxaemia, cyanosis, and, in some cases, paradoxical embolism.[5]

The current case demonstrated anatomy similar to the latter senario with the left superior vena cava (LSVC) directly opening into the left atrium. This led to the introduction of deoxygenated blood from the venous circulation directly into the arterial circulation. The formation of the functional right-to-left shunt resulted in chronic mild hypoxia, occasional episodes of short-lived nocturnal dyspnoea and compensatory secondary polycythaemia to balance the oxygen requirements of the tissues. In contrast to the usual drainage of PLSVC into the coronary sinus, the current patient demonstrated direct drainage into the left atrium, resulting in a functional right-to-left shunt. This led to the admixture of deoxygenated venous blood with oxygenated arterial blood, causing chronic hypoxaemia. The resultant compensatory erythrocytosis explains the patient's persistent secondary

polycythaemia. This highlights an important clinical implication of this anatomical variant, where unexplained polycythaemia may serve as a diagnostic clue to underlying intra-cardiac shunting. Similar cases reported in recent literature demonstrate that although PLSVC is often asymptomatic, anomalous drainage into the left atrium can result in clinically significant right-to-left shunting and complications such as hypoxaemia.[4] A recent case report from Pakistan has also highlighted the clinical importance of recognising persistent left superior vena cava, particularly during central venous catheterisation.[6]

Surgical intervention may be considered in symptomatic patients, particularly in those with significant right-to-left shunting or cyanosis.[7]

This presentation may represent a variant of Raghib syndrome, which is characterised by persistent left superior vena cava draining into the left atrium, typically in association with an atrial septal defect; however, complete diagnostic criteria were not fully evaluated in this case.[8]

Conclusion

Persistent left superior vena cava is a rare anatomical defect which is usually asymptomatic in most individuals. Occasionally, individuals can present with clinical consequences of PLSVC due to right to left cardiac shunting. These can be dyspnoea, hypoxaemia, cyanosis, secondary polycythaemia, and rarely acute ischaemic events due to paradoxical embolism. PLSVC should be considered an essential differential in individuals presenting with dyspnoea, hypoxia, and a background of secondary polycythaemia when other causes have been excluded.

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Author Contribution:

HZ, FUH, FM & SSH: Concept, design, data acquisition, analysis, interpretation, drafting, revision, final approval and agreement to be accountable for all aspects of the work.