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Double Superior Vena Cava Due to Persistent Left Superior Vena Cava Incidentally Identified on Central Venous Catheterization: A Case Report

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Data Interpretation D
Manuscript Preparation E
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Patient: Male, 40-year-old
Final Diagnosis: Persistent left superior vena cava
Symptoms: None
Medication: —
Clinical Procedure: Central venous catheterization
Specialty: Surgery

Objective: Management of emergency care

Background: Persistent left superior vena cava (PLSVC) results in a double superior vena cava (SVC), and although it is rare, this is the most common venous anomaly of the thorax. PSLVC arises from the junction of the left subclavian and internal jugular veins. It is identified on the left side of the mediastinum adjacent to the aortic arch, and it usually drains into the right atrium through the coronary sinus. This report presents the case of a 40-year-old man with an incidental finding of double SVC due to PSLVC identified on hospital admission following a motor vehicle collision.

Case Report: A 40-year-old man was found to have a double SVC due to PLSVC upon chest radiography during hospital admission for injuries related to motor vehicle trauma. The discovery was made following placement of a central venous catheter (CVC) down the left-sided SVC and into the coronary sinus. The patient suffered no harm as a result. The diagnosis was made by chest radiography and confirmed by computed tomography angiography.

Conclusions: PSLVC is an uncommon condition that can complicate common procedures and therefore must be well-understood by physicians across many medical and surgical specialties. Although PSLVC may be asymptomatic, as in this case, in some patients PLSVC presents as atrial fibrillation or with nonspecific cardiac symptoms. Therefore, all patients identified with PLSVC should be investigated to exclude associated cardiac abnormalities and arrythmias, and before the placement of central venous access devices.

Keywords: Catheterization, Central Venous • Persistent Left Superior Vena Cava

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Background

Double superior vena cava (SVC) due to persistent left superior vena cava (PLSVC) is a rare phenomenon, with an incidence in the general population of 0.3-2.1% [1-3]. In the setting of congenital heart disease, the incidence of PLSVC is 5-11% [1,4]. Anatomical variations in thoracic venous drainage occur during embryologic development and may occur alone or alongside other congenital abnormalities. The SVC drains into the right atrium and is formed from the left and right brachiocephalic veins. The embryologic origin of the SVC begins with the right anterior cardinal veins, which is distinct by the fifth week of embryonic development [5]. The formation of a transverse anastomosis between the left and right anterior cardinal veins leads to eradication of the left anterior cardinal vein proximal to the anastomosis, creating the ligament of Marshall [6]. A PLSVC is established when the left anterior cardinal vein persists, remaining connected to the coronary sinus. In some cases, the right-sided SVC is absent, and in other cases the left brachiocephalic vein is absent [7-9]. Anatomical SVC variation is typically asymptomatic but may complicate clinical intravascular procedures such as central line placement [10], hemodialysis catheterization [11], Swan-Ganz catheterization [12], and pacemaker implantation [13,14]. This report presents the case of a 40-year-old man with an incidental finding of double SVC due to PLSVC identified on hospital admission following a motor vehicle collision.

Case Report

A 40-year-old man presented to the hospital following a motor vehicle collision. He arrived with Glasgow Coma Scale of 8, an open left leg fracture, and clinical signs of shock. He was tachycardic and initial blood pressure was 80/57 mmHg. Rapid sequence intubation was performed for airway protection and emergency uncrossmatched blood was transfused. A right internal jugular (IJ) Cordis introducer was placed and Massive Transfusion Protocol was ordered. The patient was taken to the operating room, where a temporary aortic balloon was placed via a right femoral artery approach followed by splenectomy and small bowel repair. Postoperatively, the patient was admitted to the STICU, where a left internal jugular vein (IJV) central line was placed under ultrasound guidance. No immediate complications were noted. A confirmatory chest radiograph was obtained (**Figure 1**), demonstrating the catheter overlying the left paraspinal region. Pressure tracings demonstrated a central venous pressure wave. CT angiography was performed and showed the catheter to be traversing a PLSVC with the catheter tip resting in the coronary sinus conduit (**Figure 2**). The patient continued to receive care for his multiple injuries and experienced no complications secondary to CVC placement within his left SVC. This patient had no other cardiac or cardiothoracic anatomic abnormalities.

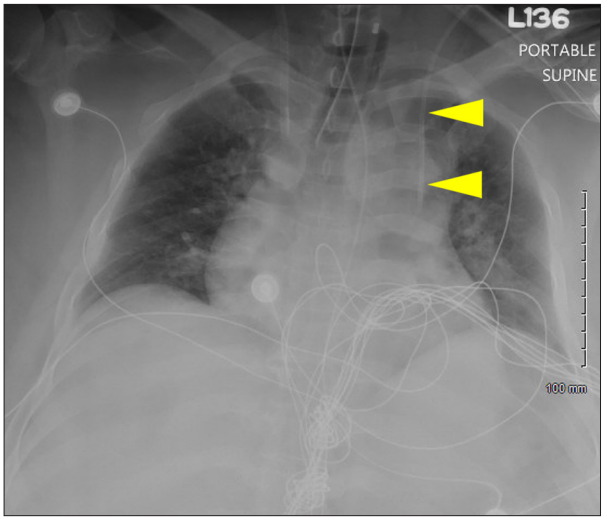

Figure 1. Chest radiography. Chest radiograph confirming placement of the central access line, which did not take its expected course across the midline toward the right atrium.

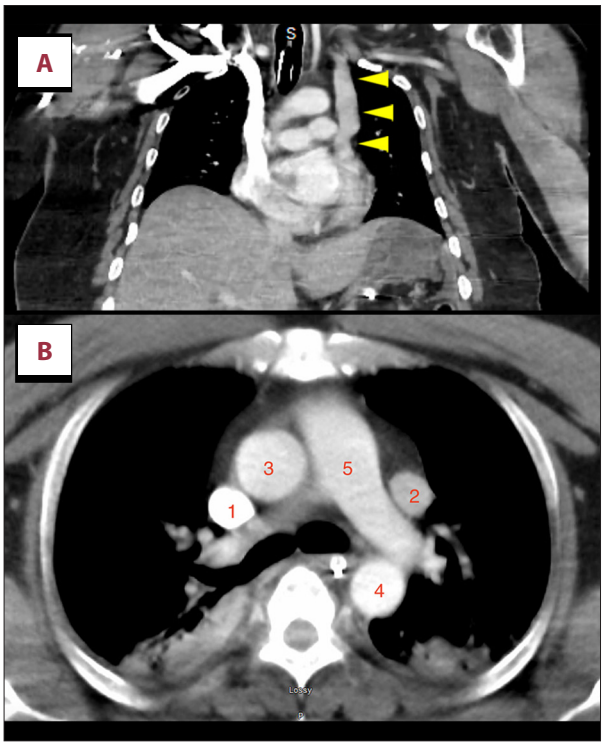

Figure 2. Chest computed tomography. Coronal chest computed tomography with intravenous contrast (**A**) showing a left-sided superior vena cava draining from the left subclavian vein into the coronary sinus. Axial chest computed tomography with intravenous contrast (**B**) demonstrating the right superior vena cava (1), left persistent superior vena cava (2), ascending aorta (3), descending aorta (4), and pulmonary trunk (5).

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Discussion

This case report offers 2 key learning points. First, although this report has shown PLSVC may be asymptomatic, in some patients PLSVC causes anomalous venous return that can result in reduced exercise tolerance, fatigue, cardiac arrythmias, cyanosis, syncope, or chest pain. Therefore, all patients identified with PLSVC should be investigated before the placement of central venous access devices to exclude associated cardiac abnormalities and any associated symptoms. Second, the presence of PLSVC can complicate efforts to obtain central venous access and result in serious complications; this is well-documented in the cardiology literature [4,15]. Our report is unique because it shows PLSVC in an atypical demographic, the surgical trauma patient, which demonstrates the ubiquity of this venous abnormality and the importance for many specialties to understand how to diagnose and recognize complications of PLSVC.

CVC placement is a commonly performed procedure indicated for hemodynamic monitoring, vesicant medicine administration, and when no other intravenous access is available. Described complications include arterial puncture, hematoma, hemothorax, and pneumothorax [16]. The right IJV is the preferred route of access as it provides a direct route to the SVC and is often larger than the left IJV, although the latter can be used if the right IJV is already occupied, as in this case, or is considered unsuitable for any reason [17,18]. The use of ultrasound guidance increases the safety of CVC placement by allowing visualization of local vasculature and confirmation of venous placement [19]. When a PLSVC is discovered, vascular access should be reattempted if the right-sided "true" SVC presents a definite route to the right atrium. The right-sided SVC is actually smaller than the PLSVC in 65% of double SVC cases [7]. The diagnosis can be made via chest radiography when a catheter has already been placed; both anteroposterior and lateral views should be obtained to ensure the line is not located in the pleura or mediastinum [6]. Angiography should then be used to assess the thoracic venous system to confirm the diagnosis and assay for additional cardiovascular anomalies.

Upon discovery, the presence of a PLSVC merits exploration of patient history for the presence or absence of clinical symptoms, including tachycardia, palpitations, syncope, and dyspnea. Importantly, PLSVC can be an arrhythmogenic focus in atrial fibrillation [20]. Isolation of the PLSVC is a catheter ablation strategy that has shown success in treating atrial fibrillation [20,21]. A rarer phenomenon is drainage of the PLSVC into the left atrium rather than into the right atrium; this can present with symptoms of shunting including dyspnea, hypoxia, cyanosis, and paradoxical emboli [22-24]. Echocardiography should be obtained to characterize function when patients with PLSVC endorse any of the above symptoms or when CT angiography demonstrates further atypical cardiovascular anatomy. Finally, it is important to document the presence of a PLSVC in medical records so that fellow clinicians are aware of the relevant anatomy before obtaining central venous access.

The routine use of chest radiography to confirm correct CVC placement is debatable, with some clinicians considering radiography unnecessary in the event of uncomplicated insertion [25,26]. While imaging may be unnecessary in cases of uncomplicated right-sided CVC placement, we believe that confirmatory chest radiography should be used whenever a CVC is placed through the left IJV owing to the frequency of left-sided complications and the not insignificant rate of PLSVC in the general population. A randomized study of 120 patients concluded left-sided versus right-sided IJV cannulation is more time consuming and is associated with more complications (20% vs 10%, $P<0.05$) [27]. Additionally, the left IJV is significantly smaller than the right IJV, with a mean difference in diameter of 1.44 mm (95% CI 0.79-2.09 mm, $P<0.0001$) according to an evaluation of 100 computed tomography scans [8]. Radiography alone in this case was unable to confirm placement in the vena cava, which led to concern for placement in the aorta or pleural space. Subsequently, measuring pressure wave form valuation at bedside was a helpful means of affirming central venous placement.

The treatment of complications related to central venous catheterization has previously been reviewed in great detail [28]. Radiography can detect procedural complications of pneumothorax and hemothorax. Angiography is used to diagnose suspected intravascular perforation or thrombosis. Symptoms associated with all the aforementioned complications include new-onset dyspnea, hypotension, and jugular venous distention. The onset of such symptoms may be delayed for several days following catheterization. Tube thoracostomy is the preferred treatment for CVC-associated pneumothorax and hemothorax. Clinically insignificant small pneumothoraces do not require treatment. Cases of large, symptomatic, or tension pneumothorax, as well as hemothorax, should be treated immediately. Both clotting disorders and therapeutic anticoagulation can be seen among patients requiring central lines. Bleeding or thrombosis related to coagulopathy and line insertion should be addressed before a decision to remove a catheter is made. Resuscitation and inotropic cardiac support should be provided by another route if the line is suspected of incorrect placement.

Stenosis is common within PLSVC, increasing the risk of vascular perforation with guidewire placement or catheter advancement. Serious complications such as hemothorax, cardiac tamponade, phrenic nerve damage, or type 2 myocardial infarction can occur due to perforation [7,10]. If the line is advanced into the wall of the right atrium or the coronary sinus, there

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is risk for arrythmia and coronary sinus thrombosis [29,30]. Additionally, procedural aids such as ultrasound and contrast dye should be utilized whenever uncertainty arises to help obtain proper intravascular access. Most commercially available central access kits have insertion directions requiring use of fluoroscopy for insertion except for cases of emergent placement, but fluoroscopy is rarely used in clinical practice. A joint guideline from the American Society of Echocardiography and the Society of Cardiovascular Anesthesiologists recommends that properly trained clinicians use real-time ultrasound during IJ cannulation whenever possible to improve cannulation success and reduce the incidence of complications (Category A, level 1); use of fluoroscopy is not routinely recommended [31].

Conclusions

PSLVC is an uncommon condition that can complicate common procedures and therefore needs to be well-understood by physicians across many medical and surgical specialties. Although PLSVC may be asymptomatic, as in this case, in some patients PLSVC presents as atrial fibrillation or with nonspecific cardiac symptoms. Therefore, all patients identified as having PLSVC should be investigated to exclude associated cardiac abnormalities and arrythmias, and before the placement of central venous access devices.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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