CASE REPORT

Persistent left superior vena cava causing diagnostic dilemma after central line insertion

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Introduction

Double or duplicated superior vena cava (SVC) is uncommon, occurring in 0.3% to 2.0% of the population. It occurs due to the persistence of the left sided SVC (PLSVC) during embryological development. When it coexists with the normal right sided SVC, the patient develops a double or duplicated SVC. PLSVC is more common in patients with congenital heart disease, occurring at a rate of 10.0% to 11.0% [1],[2].

In cases of double SVC, the right and the left SVCs are formed by the communication of the internal jugular vein (IJV) and the subclavian vein (SCV) of the relevant side. However, the PLSVC drains separately into the atrium. The connection between the right and the left side by the left brachiocephalic vein (LBCV) may not be present.

In 90% of the cases the PLSVC drains into the right atrium through the coronary sinus (CS) [3]. This variation can result in diagnostic dilemmas and complications during the procedures. This case reports a patient with PLSVC who underwent a dialysis access catheter insertion, causing a diagnostic dilemma on postoperative chest x-ray.

Case presentation

A 54-year-old female with end stage renal failure, was admitted for dialysis access line insertion. She had a previous central line insertion on the right side three weeks before, and it was removed following infection. A duplex ultrasound scan of the neck showed that the bilateral internal jugular veins were patent. However, the previous central line insertion site on the right side was not healthy. Therefore, it was decided to insert the dialysis access line to the left internal jugular vein. The procedure was done under the ultrasound guidance, and it was uncomplicated. The dialysis access line was functioning properly after insertion. The post procedure chest X-ray revealed that the dialysis access line descending vertically downwards on the left side of the heart (Figure 1). It was

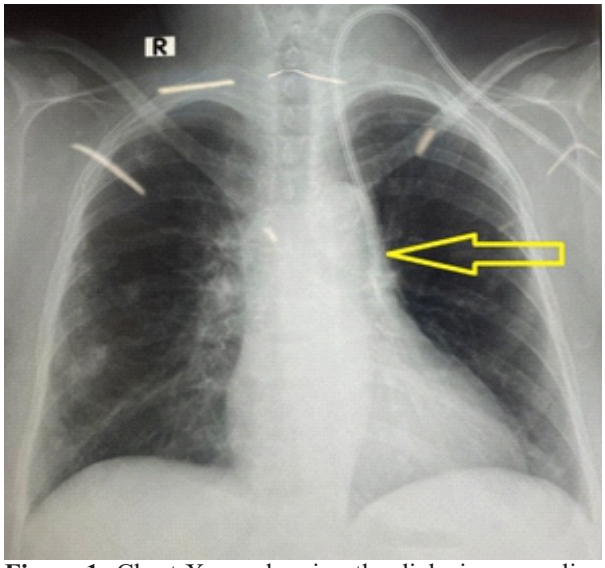

Figure 1: Chest X-ray showing the dialysis access line descending vertically downwards on the left side of the heart (arrow)

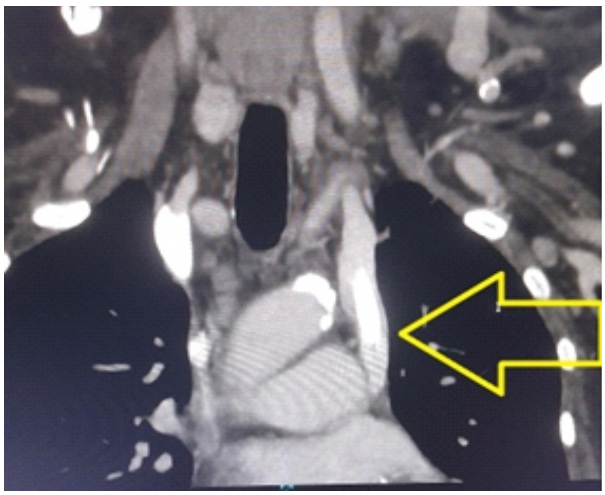

Figure 2: Computerised tomographic (CT) scan showing persistent left sided superior vena cava (PLSVC) and the dialysis access line (arrow) descending vertically downwards on the left side of the heart.

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suspected that the line was not properly positioned or it had accidentally entered the left subclavian artery. Therefore, a computerised tomographic (CT) scan of the lower neck and the chest was planned. The CT scan showed that the dialysis access line was entering the left internal jugular vein and then passing inferiorly along the left side of the heart through the PLSVC (Figure 2). The PLSVC was then draining into the right atrium through the coronary sinus. There was no connection between the left and right IJVs. The right IJV entered the right atrium separately through the normally positioned right SVC. There were no other congenital anomalies. The patient was not cyanosed. A subsequent 2D echocardiogram did not show any cardiac abnormalities.

Discussion and conclusions

During the development, a pair of (right and left) anterior (ACV) and posterior cardinal veins (PCV) develops on either side of the midline [4]. The right and left ACV drain the anterior (head and upper limb) part of the developing embryo. The ACV and the PCV on either side join to form the common cardinal veins (CCV) (ducts of Cuvier). The CCV drains into the horn of the sinus venosus (SV). The right and the left horns of the SV drain into the sinus venosus. Later in the development, a connecting transverse vein connects the right and the left ACVs. This transverse connecting vein develops into the left brachiocephalic vein (LBCV). The part of the ACV distal to the BCV, the CCV and the right horn of the SV develop into the superior vena cava (SVC). The PCV develops into the azygous vein (AV) in adults. On the left side the ACV distal to the BCV degenerates (Figure 3).

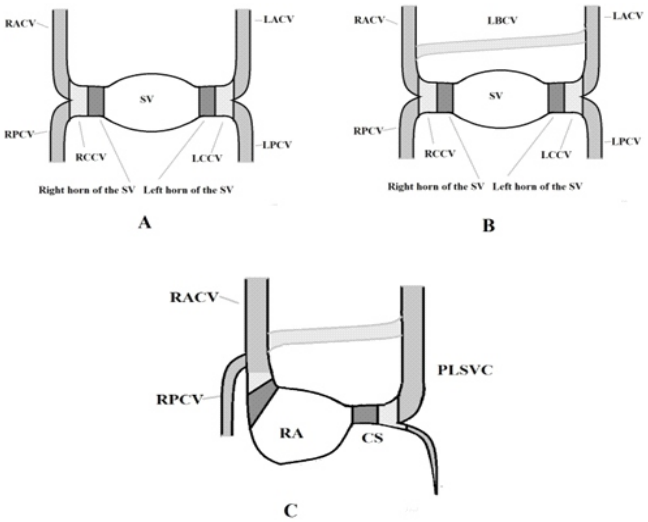

Figure 3: Development of the persistent left sided superior vena cava (PLSVC). Right (R), left (L), anterior cardinal vein (ACV), posterior CV (PCV), common CV (CCV), sinus venosus (SV), coronary sinus (CS), left brachiocephalic vein (LBCV), right atrium (RA)

The distal part of the left CCV and the left horn of the SV form the coronary sinus in adults. When the left ACV remains patent in between the left BCV and the coronary sinus, it develops into the PLSVC in adults. If the LBCV does not develop or degenerates, separate right and left SVC develops in adults. PLSVC with absent right SVC is a rarer abnormality occurring in 0.09% to 0.13% [5]. In 90% of the cases the PLSVC drains into the right atrium through the coronary sinus (CS). Other sites of drainage include the left atrium, inferior vena cava, and the hepatic veins [3]. The PLSVC is associated with congenital heart diseases, especially when the PLSVC drains into sites other than the right atrium. The above described case did not have any evidence of congenital heart disease, either clinically or on the echocardiography.

Most patients with PLSVC are asymptomatic. However, the patients can present with arrhythmia and with cyanosis. The increased risk of arrhythmias in patients with PLSVC occurs due to the widening of the CS opening, resulting in expansion of the atrio-ventricular node. Cyanosis can occur when the PLSVC drains into the left atrium. PLSVC can also lead to serious complications during procedures like central line insertion and during cardiac surgeries. After central line insertion, thrombosis of the coronary sinus can occur, and also there can be arrhythmias.

The diagnosis is suspected on the chest x-ray following central line insertion, as in the case described above. In cases of PLSVC, the central line descends inferiorly on the left side of the heart, rather than crossing to the right side (via the LBCV) as in normal cases. In the 2D echocardiography, the feature suggesting PLSVC is the presence of a dilated coronary sinus in the absence of raised right atrial pressure. In addition, echocardiography is useful in diagnosing the associated congenital cardiac abnormalities. In the above described case, there were no congenital cardiac abnormalities. A contrast enhanced computerised tomographic (CT) scan with CT venography is needed to confirm the exact anatomy of the abnormalities.

During central venous cannulation, the cannula and the guide wire can accidentally injure the left SVC due to its anatomical orientation. In addition, PLSVC is known to have stenosis and a dilated coronary sinus which can be aneurysmal. These abnormalities can also lead to injury to the PLSVC or the coronary sinus, resulting in haemothorax, haemoperi cardium, and cardiac tamponade [6]. In addition the central venous lines can cause arrhythmias and coronary sinus thrombosis [7]. Therefore, this case illustrates that all central venous line insertions should be done with ultrasound and

fluoroscopic guidance. When using fluoroscopy, a contrast agent can be injected directly into the central line, and the anatomy can be determined. In case of any doubts about congenital variations, the procedure should be abandoned and the patient should be further investigated (with CT scan) to demonstrate the exact anatomy.

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Learning Points:

- A persistent left superior vena cava is a relatively uncommon congenital venous anomaly, but it is clinically important.
- This variation can lead to serious complications during procedures such as central venous catheter insertion and cardiac surgeries due to the abnormal venous pathway.
- If there is any suspicion of this anatomical variation during a procedure, the procedure should be stopped and the patient should be further evaluated, usually with imaging such as a CT scan to confirm the anatomy.