Persistent Left Superior Vena Cava Identified After Hemodialysis Catheter Insertion: A Case Report

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Introduction: Central venous catheter (CVC) insertion is the most commonly performed clinical procedure when a patient initiates hemodialysis. Despite its clinical benefits, CVC insertion has several risks of complications. Thrombosis, venous stenosis, infection, arrhythmia, pneumothorax, and bleeding are among these complications. Malposition of the tip of the CVC can also occur with an incidence of up to 7%. One of several factors that could contribute to malposition is venous anatomy variation. Persistent left superior vena cava (PLSVC) is an extremely rare venous anatomical disorder but might have a significant clinical impact.

Case Presentation: Here we report a PLSVC case that was identified in chest radiography after the insertion of a CVC catheter in a patient with end-stage renal disease (ESRD). A 40-year-old woman with a history of type 2 diabetes mellitus, hypertension, dyslipidemia, and obesity was presented in the emergency room with dyspnea for 1 week. Acute hemodialysis was required because of the ESRD and pulmonary edema. The PLSVC condition accompanied by various complications that occurred in this patient became a dilemma for the nephrologist in determining the diagnosis and proper CVC management.

Discussion: PLSVC is the most common congenital abnormality of the vena cava, even though it has a very small incidence. PLSVC occurs in about 0.1–0.5% of the total population and reaches 10% in individuals with congenital heart abnormalities. Most PLSVC presents along with normal superior vena cava and drains into the right atrium, which makes it very difficult to see the clinical signs and symptoms. Almost all PLSVC conditions are found incidentally during or after invasive procedures such as CVC insertion. CVC insertion in the PLSVC condition needs proper management to minimize the risk of complications.

Conclusion: This case shows the importance of understanding the PLSVC condition, which, although very rare, is expected to increase the awareness of the nephrologist in making the diagnosis, determining appropriate management, and preventing complications, thereby improving patient safety.

Keywords: persistent left superior vena cava, PLSVC, central venous catheter, CVC, malposition, hemodialysis, end-stage renal disease, ESRD

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Introduction

Central venous catheter (CVC) insertion is the most commonly performed clinical procedure when a patient initiates hemodialysis.¹ Although the National Kidney Foundation recommends using AV access (fistula or graft) for hemodialysis initiation, the use of CVC still dominates with an incidence reaching 80%.^{1,2} Its ability to

be used immediately after insertion makes it preferred for use in urgent or emergent conditions.³

Despite its clinical benefits, CVC insertion has several risks of complications. Thrombosis, venous stenosis, infection, arrhythmia, pneumothorax, and bleeding are among these complications.^{4,5} Malposition of the tip of the CVC can also occur with an incidence of up to 7%.⁶ Several factors could contribute to malposition, such as bevel orientation upon needle insertion, the patient's body habitus, and venous anatomy variation.⁶ Persistent left superior vena cava (PLSVC) is an extremely rare venous anatomical disorder but might have a significant clinical impact.⁷

Here we report a PLSVC case that was identified in chest radiography after the insertion of a CVC catheter in a patient with end-stage renal disease (ESRD).

Case Presentation

A 40-year-old woman with a history of type 2 diabetes mellitus, hypertension, dyslipidemia, and obesity, presented in the emergency room with dyspnea for 1 week. Patient was hospitalized with the diagnosis of community-acquired pneumonia (CAP) and diabetic foot ulcer with secondary sepsis. Acute hemodialysis was required because of the ESRD and pulmonary edema.

The first CVC for hemodialysis was immediately inserted to the right subclavian venous using ultrasound guidance. In the chest X-ray evaluation, the tip of the catheter appeared to be at the projection of superior vena cava and right atrium border. After 1 week of usage, the catheter was immediately removed due to thrombus and bloodstream infection.

The second CVC insertion was delayed until the 13th day of hospitalization due to the patient's unstable hemodynamics. The catheter was inserted using ultrasound guidance through the left jugular vein because of thrombus in the right side. There were no signs of hypotension, cyanosis, or bleeding during and after the insertion. The electrocardiography showed normal sinus rhythm. In the post-insertion chest x-ray evaluation, the tip of the catheter appeared to be at the left mediastinum (Figure 1).

The left internal jugular or subclavian catheter insertion should cross the midline to reach the right atrium. The inadvertent cannulation of the carotid artery was ruled out by blood gas analysis. An echocardiography was performed in the following day to confirm whether there was PLSVC. The result showed that there was definitely PLSVC (Figure 2).

Figure 1 Anteroposterior chest x-ray shows the tip of the catheter inserted through the left jugular vein was located in the left mediastinum (black arrow).

The patient's condition worsened despite the optimal treatment. She died on the 16th day of hospitalization due to septic and hemorrhagic shock. The use of PLSVC as hemodialysis access could not be evaluated because the hemodialysis procedure was not done.

Discussion

PLSVC is the most common congenital abnormality of the vena cava, even though it has a very small incidence. PLSVC occurs in about 0.1–0.5% of the total population and reaches 10% in individuals with congenital heart abnormalities.⁷ The abnormalities that are most commonly found together with PLSVC are atrial septal defect, ventricular septal defect, and aortic coarctation.⁵ In clinical practice, venous system imaging is not routinely performed on patients who will undergo invasive procedures such as CVC insertion, so it is almost certain PLSVC will be found incidentally during or after the procedure.⁸

In normal embryological development, the venous system consists of two pairs of cardinal veins symmetrically located. The cranial portions of both anterior cardinal veins form the internal jugular veins. The caudal portion of the right anterior cardinal vein develops into the superior vena cava while the left regresses into the ligament of Marshall. The failure of this regression will cause PLSVC (Figure 3).^{9,10}

Figure 2 Echocardiography examination shows: (A) Coronary sinus enlargement due to the blood flow from PLSVC to right atrium (RA); (B) PLAX view shows that there is an enlargement of the coronary sinus (CS) adjacent to the descendant aorta (DA); and (C) Bubble test injected through the left cubital vein shows air bubbles filling the right atrium through the coronary sinus.

There are four types of superior vena cava based on Schummer's classification (Table 1).¹¹ About 80% of the PLSVC present along with the normal superior vena cava and most of the PLSVC (92%) drains into the right atrium. Both of these make most PLSVC cases asymptomatic.^{9,12}

Based on a guideline for determining the location of catheter malposition by Gibson and Bodenham,¹³ the position of the hemodialysis catheter in this patient was consistent with the PLSVC condition. The criteria are that blood flow can pass through all lumens, with venous blood characteristics (low flow, non-pulsatile, dark color), and is located in the left mediastinum when evaluated by chest x-ray. Echocardiography can be used to diagnose PLSVC quickly and non-invasively,¹⁴ as was done in this patient. Despite its small proportion (8%), PLSVC which drains into the left atrium can cause serious complications, such as embolization.¹² Additional diagnostic studies can be used to detect the

presence and ascertain the drainage of PLSVC, such as radionuclide angiocardiology, computerized tomography (CT) scan, or magnetic resonance venography.⁹

Some nephrologists believed that PLSVC has relatively thin walls and low blood flow, making it unsuitable for long-term access to hemodialysis.⁷ Use of PLSVC as hemodialysis access was also associated with some complications, including arrhythmia, pericardial effusion, thrombus formation, and reduced blood flow to the heart.^{7,15-17} However, literature reviews of the same case show the possibility of PLSVC being used as hemodialysis access in short- and long-term hemodialysis patients, certainly with tighter monitoring.^{7,14}

Management of CVC malposition must be based on several considerations: the indication of insertion, the location of insertion, and the condition of the patient.¹³ If there are difficulties as in this case, the risks and benefits of the inserted CVC must be considered.⁶ The patient in this case

Figure 3 Schematic illustration of persistent left superior vena cava. Redrawn with data from these studies.^{7,11,19}

was critically ill, so immediate removal of the catheter was considered to worsen the condition. Removal of the catheter in conditions that are not optimal can increase the risk of complications, morbidity and mortality.¹⁸

Conclusions

PLSVC is a very rare congenital vena cava disorder, mostly with no symptoms, and is generally found incidentally during invasive procedures such as hemodialysis catheter insertion. The location of the tip of the catheter that is attached via PLSVC can be confirmed when it drains into the right atrium, does not provide complications and then can be used for

short-term hemodialysis with tighter monitoring. By understanding this condition, a nephrologist is expected to be more aware, can make the diagnosis, determining appropriate management, and preventing complications, thereby improving patient safety.

Statement of Ethics

This study was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. The information used did not identify any individual, and the subject’s identity was kept confidential. A written informed consent was obtained from the patient’s family for publication of this manuscript. Institutional approval was not required to publish the case details.

Disclosure

The authors report no conflicts of interest in this work.

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Table 1 Classification of Superior Vena Cava

Types	Description
I	Normal (right) superior vena cava present, PLSVC absent
II	Right superior vena cava absent, PLSVC present
IIIa	Both superior vena cava present with left brachiocephalic vein in between
IIIb	Both superior vena cava present without left brachiocephalic vein in between

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