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CASE REPORT

Vascular access dysfunction associated with type 3 persistent left superior vena cava in an adult patient. Case report

Disfunción de acceso vascular secundaria a vena cava superior izquierda persistente tipo 3 en un paciente adulto. Reporte de un caso

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Abstract

Introduction: Persistent left superior vena cava (PLSVC) is a congenital venous anomaly with an estimated prevalence of 0.5% in the adult population. This condition is usually asymptomatic, does not require treatment, and is usually diagnosed incidentally. However, knowledge of its occurrence is essential for the proper planning of some procedures and surgeries.
Case report: A 59-year-old man with stage 5 chronic kidney disease was referred to the Hospital Universitario Nacional de Colombia due to central venous hemodialysis catheter dysfunction (right jugular vein). A new central venous catheter was inserted in the left jugular vein, but it failed shortly after, so imaging studies were performed (chest X-ray and CT angiography), detecting type 3 PLSVC draining into the left atrium. Considering this finding and the fact that the CT angiography showed no signs of obstruction in the right jugular vein, the catheter was removed from the left jugular vein and a new one was placed in the right jugular vein, which allowed continuing hemodialysis without complications and discharging the patient 24 days after his admission to hospital.
Conclusions: Although PLSVC is a rare anomaly, especially type 3, its diagnosis is usually incidental, as in the case of the patient described here. Knowledge of vascular anomalies in patients with chronic kidney disease is fundamental, given that it allows selecting the proper vascular access and reducing the risk of complications such as arrhythmia, thrombosis, or even perforation of the heart. This case report is highly relevant since blood flow to the heart and lungs may be altered in patients with PLSVC, and this may require hemodynamic adjustments during hemodialysis and in the follow-up of this population.

Resumen

Introducción. La vena cava superior izquierda persistente (VCSIP) es una anomalía venosa congénita con una prevalencia estimada de 0.5% en población adulta. Por lo general, esta condición es asintomática, no requiere tratamiento y su diagnóstico suele realizarse de manera incidental. Sin embargo, saber de su presencia es fundamental para la adecuada planeación de algunos procedimientos y cirugías.
Presentación del caso. Hombre de 59 años con enfermedad renal crónica etapa 5 remitido al Hospital Universitario Nacional de Colombia por disfunción del catéter venoso central de hemodiálisis (vena yugular derecha). Se insertó un nuevo catéter venoso central en la vena yugular izquierda, pero presentó disfunción temprana, por lo que se realizaron estudios imagenológicos (radiografía de tórax y angioTAC) en los que se detectó una VCSIP tipo 3 con drenaje en la aurícula izquierda. Teniendo en cuenta este hallazgo, y que en la angioTAC no se observaron signos de obstrucción en la vena yugular derecha, se retiró el catéter de la vena yugular izquierda y se colocó uno nuevo en la vena yugular derecha, lo que permitió continuar la hemodiálisis sin complicaciones y dar el alta tras 24 días de hospitalización.
Conclusiones. Si bien la PSLVC es una anomalía inusual, en especial la de tipo 3, su diagnóstico suele ser incidental, como ocurrió en el paciente aquí presentado. Conocer las anomalías vasculares en pacientes con enfermedad renal crónica es fundamental dado que permite seleccionar el acceso vascular adecuado y disminuir el riesgo de complicaciones como arritmias, trombosis, o incluso perforación vascular. La presentación de este caso es de gran relevancia ya que en los pacientes con PSLVC es posible que el flujo sanguíneo hacia el corazón y los pulmones se altere, lo que podría requerir ajustes hemodinámicos durante la hemodiálisis y en el seguimiento a esta población.

Introduction

Persistent left superior vena cava (PLSVC) is the most common congenital venous anomaly in adults, occurring in 0.3% to 0.5% of the general population. However, its prevalence can be as high as 4-8% in patients with congenital heart disease[1-7] such as heterotaxy syndrome, atrial septal defect, endocardial cushion defects, tetralogy of Fallot, transposition of the great arteries, Brugada syndrome, among others.[4,5,8]

This anomaly is caused by a failure of regression of the left anterior cardinal vein at 12 weeks of gestation, which, together with the right anterior cardinal vein, constitutes the venous drainage of the cephalic portion[1,9] (normal regression, Figure 1). It has been proposed that this happens because of abnormalities in cardiac progenitor cells from the secondary heart field, which is derived from the splanchnic mesoderm and is involved in the formation of multiple internal organs such as blood vessels and the visceral serosa of the heart.[1]

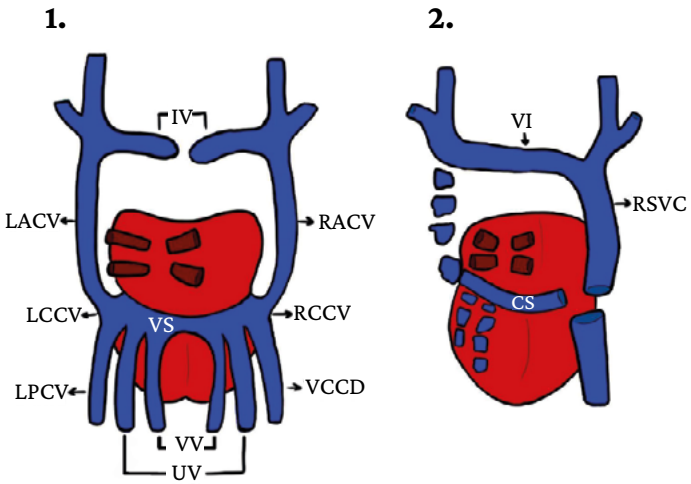

Figure 1. Heart development. A) Embryonic heart, where the anterior and posterior cardinal veins drain through common right and left cardinal veins. B) Normally developed heart with regression of the left superior vena cava, uterine veins, and vitelline veins.
IV: innominate veins; VV: vitelline veins; UV: umbilical veins; LACV: left anterior cardinal vein; RACV: right anterior cardinal vein; LCCV: left common cardinal vein; RCCV: right common cardinal vein; LPCV: left posterior cardinal vein; RSVC: right superior vena cava; VS: venous sinus; CS: coronary sinus.
Source: Elaborated based on Laasri *et al.*[10]

There are four morphological types of PLSVC: type 1: left superior vena cava (SVC) with an absent or atretic right SVC, type 2A: double SVC with anastomosis between right and left SVC, type 2B: double SVC without anastomosis, and type 3: left SVC draining directly into the left atrium (LA) without anastomosis between the SVC11 (Figure 2).

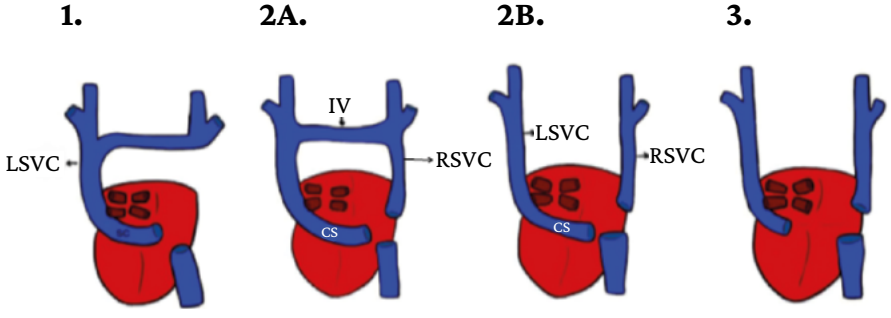

Figure 2. Anatomical types of persistent left superior vena cava.
LSVC: left superior vena cava; IV: innominate veins; RSVC: right superior vena cava; CS: coronary sinus.
Source: Elaboration based on Demșa *et al.*[11]

Most patients are asymptomatic since PLSVC drains into the right atrium (RA) through the coronary sinus with no clinical implications other than the anatomical variant.[2,8] However, patients with type 3 PLSVC may have a right-to-left cardiac shunt with oxygen desaturation[5,12,13] and, therefore, may present with dyspnea on exertion, limitation in physical activity,[12,13] and impaired quality of life.[6] It is important to keep in mind that type 3 PLSVC is the least common, occurring in about 10% of individuals with this condition.[5]

However, some patients may require correction, which can be performed by ligation, intra-atrial baffle bypass, reimplantation in the RA, and interposition of the graft in the RA.[8,14] Despite this, knowing about the presence of this anatomical variant in patients is essential to plan and perform some interventional procedures or cardiothoracic surgeries.[2,5]

The following is the case of an adult patient with chronic kidney disease on hemodialysis with a temporary central catheter in the left jugular vein, in whom type 3 PLSVC was detected incidentally.

Case presentation

A 59-year-old man was referred to the Hospital Universitario Nacional de Colombia (quaternary level of care in Bogotá) in January 2023 due to temporary central venous hemodialysis catheter dysfunction of obstructive origin placed in the right jugular vein. The patient had type 2 diabetes *mellitus* (treatment with insulin glargine and glulisine, basal-bolus injection regimen), primary arterial hypertension (treatment with calcium channel blockers), and stage 5 chronic kidney disease (renal replacement therapy since November 2022 after being admitted to the hospital due to gram-positive bacteremia with an isolate of methicillin-susceptible *Staphylococcus aureus*). During his hospital stay in November 2022, monoclonal gammopathy of unknown significance was detected, and it was being followed up on an outpatient basis by the hematology service.

The following findings were reported during his physical examination on admission: blood pressure of 137/77mmHg, heart rate of 102bpm, respiratory rate of 16rpm, temperature of 36.5°C, beginning of nail clubbing in both upper limbs, pain in the dorsal region on palpation, and limitation of spinal mobility (extension and flexion). The patient also reported subjective weight loss in the last 2 months (approximately 7kg), night sweats, occasional chills, occasional respiratory functional impairment with physical activity, and low back pain in the last 3 months.

On the second day of admission, he was evaluated by specialists from the nephrology service, who, after confirming the dysfunction of the central venous catheter lodged in the right jugular vein, inserted a new central venous dialysis catheter in the left jugular vein guided by ultrasound; however, early dysfunction of the catheter was reported on the same day of insertion after low flow was observed on the dialysis machine. In view of this situation, the nephrology service immediately requested a chest X-ray that showed an anomalous vascular pathway in the catheter (Figure 3). Therefore, that same day a computed tomography angiography (CTA) of the chest and neck was performed (Figure 4), showing a left jugular vein catheter (subclavian insertion) and type 3 PLSVC (drainage of the LSVC to the LA and of the RSVC to the RA). Additionally, opacification of the right internal jugular vein was observed due to a thrombus (cephalocaudal extension: 27mm) that generated occlusion of less than 50% of the lumen in the lower third of the vessel, so it was decided to remove the dysfunctional right catheter.

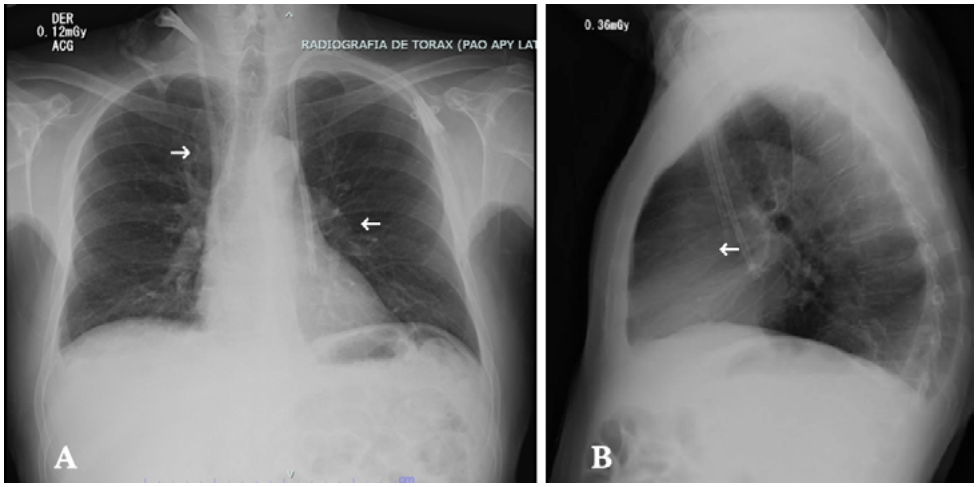

Figure 3. Chest X-rays confirming bilateral catheterization of the internal jugular vein and revealing that the catheter in the left vein (subclavian insertion) shows signs of a non-vascular pathway. A) anteroposterior view; B) lateral view.

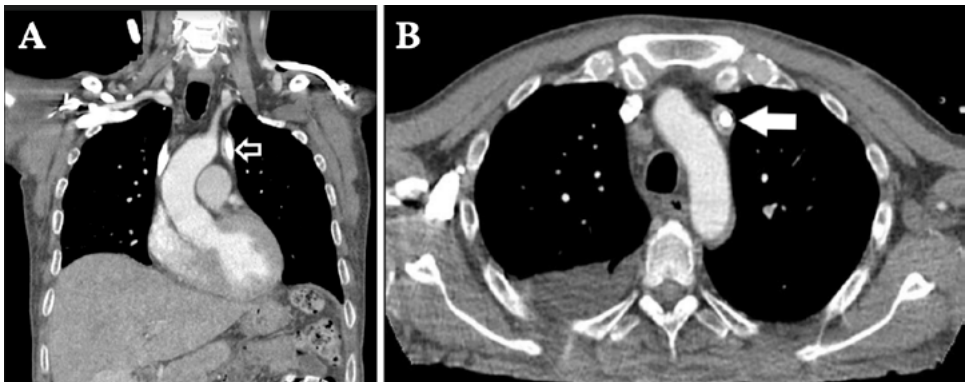

Figure 4. Computed tomography angiography. A) coronal plane; B) cross-sectional plane.

In view of these findings, seven days after being admitted to hospital, an electrocardiogram (Figure 5) and a transthoracic echocardiogram were requested, finding left ventricle ejection fraction of 57%, as well as grade II diastolic dysfunction with increased filling pressures and increased LA volume (volume index of 36mL/m²).

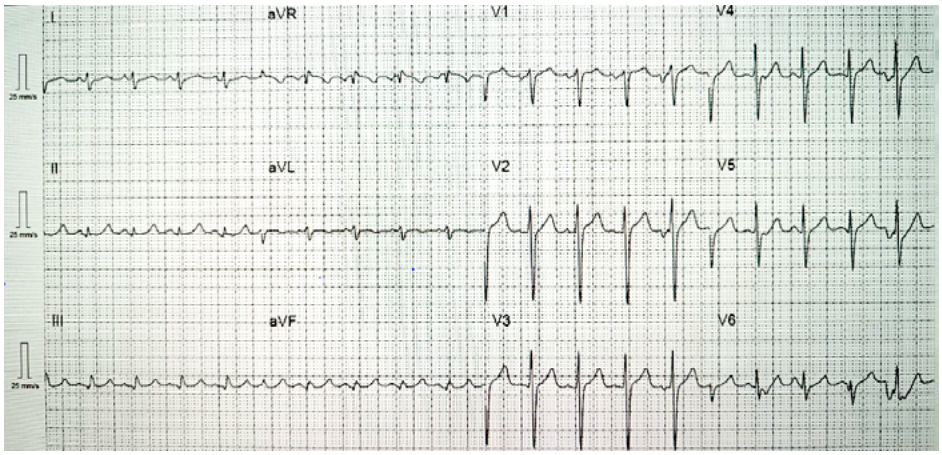

Figure 5. 12-lead electrocardiogram showing heart rate of 110bpm, sinus rhythm, right axis, normal narrow QRS, left posterior fascicular block, and preserved PR and QT intervals; no signs of atrial or ventricular hypertrophy, pathological Q waves or alterations in the ST segment were observed.

Nine days after being admitted to hospital, the vascular surgery service staff removed the catheter from the left jugular vein and, since the CTA showed no signs of obstruction in the right jugular vein or other cardiovascular malformations, they proceeded to insert a new dialysis catheter in this vein, which allowed performing hemodialysis without complications.

On day 12 of hospitalization, a bone marrow biopsy was performed due to suspicion of monoclonal gammopathy of unknown significance, ruling out malignancy. Due to the persistence of low back pain, 2 days later, magnetic resonance imaging (MRI) of the spine was performed, in which findings suggestive of spondylodiscitis in T10-T11 were observed.

On day 16 of hospitalization, specialists from the infectious disease department evaluated the patient considering a possible relationship between spondylodiscitis and the episode of bacteremia that occurred in November 2022. Taking into account the microbiological isolates identified in that hospital stay, in-hospital treatment with cefazolin 2g IV every 24 hours plus 1g additionally after the dialysis sessions was indicated. Outpatient treatment consisted of cephalexin 500mg every 12 hours and trimethoprim-sulfamethoxazole 160/800mg every 12 hours for 6 weeks, adjusted to the patient's renal function.

Since there were no new complications, the patient was discharged after 24 days of hospitalization with the indication to start the previously described outpatient antibiotic treatment and continue with the hemodialysis sessions, for which he already had a renal unit assigned before admission to our institution. He was also recommended to attend follow-up appointments with the internal medicine, infectious disease, and hematology services, as well as an evaluation by specialists in physical rehabilitation and occupational therapy to initiate a physical therapy plan. However, no follow-up information was found in the database of the hospital's outpatient service.

Discussion

PLSVC usually drains into the right atrium through a dilated coronary sinus[8,15] and, although this condition in adults is usually asymptomatic,[8] it commonly causes arrhythmia because the dilatation elongates the atrioventricular node tissue, favoring the occurrence of reentrant tachycardia. Likewise, PLSVC has been associated with atrial and ventricular tachycardia and fibrillation.[16]

Since PLSVC is asymptomatic in most cases, its detection in adults is usually incidental[3,8,16] and occurs during invasive cardiothoracic procedures such as pacemaker placement;[5,16] hemodialysis catheter insertion in the internal jugular vein[2] and extracorporeal membrane oxygenation;[17] during pre-transplant testing[5] or imaging tests such as computed tomography (CT) or contrasted MRI in patients with a suspected heart mass;[17] or during the performance of tests prior to heart transplantation[5] or imaging tests such as computed tomography (CT) or contrasted MRI in patients with a suspected chest mass.[18]

PLSVC types 2A and 2B are the most common, accounting for 42-45% of cases.[15] It has been reported that less than 10% of cases are classified as type 1, with about half of these patients presenting with congenital heart defects such as atrial septal defect and tetralogy of Fallot.[16] Type 3 accounts for approximately 8% of cases, with around 40% of affected patients presenting with congenital heart defects such as atrial septal defects, interatrial or interventricular communication defects, bicuspid aortic valve, cor triatriatum, coarctation of the aorta, and coronary sinus ostial atresia;[7-9,19] this has resulted in the association of this type of PLSVC with the presence of hypoxemia and cyanosis, as well as

with the development of thrombosis and arrhythmia,[6,8,20] particularly atrial fibrillation,[18 and] consequently, it is generally diagnosed during infancy.[8]

This article presents the case of a patient with asymptomatic type 3 PLSVC that was detected incidentally after the insertion of a hemodialysis catheter in the jugular vein. It is worth noting that, despite the type of PLSVC, the patient never presented with cyanosis nor required oxygen therapy. In addition, given the high frequency of congenital heart disease in patients with type 3 PLSVC, an echocardiogram was performed, showing no signs of interatrial communication defects, but revealing a dilated LA and grade II diastolic dysfunction with increased filling pressures. It should be noted that in cases such as that of our patient (type 3 PLSVC and LA dilatation), strict clinical follow-up should be performed after diagnosis, as these conditions imply a greater predisposition to the development of arrhythmia and thrombosis.

Moreover, it has been described that performing imaging studies such as CT or MRI before invasive interventional procedures is useful because PLSVC may hinder the central venous catheterization process, and manipulating the guide wire near this defect can cause complications such as hemodynamic instability, arrhythmia, tamponade, and perforation of the heart, among others.[16]

Finally, once PLSVC is detected, the finding should be conveyed to all individuals involved in the patient's medical care, mainly intensivists, cardiologists, and anesthesiologists, as it is a critical factor in assessing the feasibility and planning of interventional procedures.[5]

Conclusions

In the present article, we report on the case of an adult with chronic kidney disease receiving hemodialysis through a central venous catheter inserted in the jugular vein, in whom type 3 PLSVC was detected incidentally. PLSVC is a rare congenital venous anomaly that, although generally asymptomatic, may hinder the performance of medical and surgical procedures. The incidental detection of this anomaly in an adult patient with chronic kidney disease on hemodialysis underscores the importance of early identification of anatomic variations, as this not only allows for a safer management of vascular accesses, but also reduces the risk of complications such as arrhythmia, thrombosis, and even perforation of the heart. Furthermore, this case highlights the need for rigorous clinical follow-up in patients with type 3 PLSVC given the potential risk of long-term cardiac complications.

Informed Consent

The patient signed an informed consent form accepting the use of his clinical data for the preparation and publication of this case report.

Conflicts of interest

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