

Case Report

Persistent Left Superior Vena Cava in a Patient who is Hemodialysis Dependent: A Rare Anomaly of Venous Drainage that Interventional Nephrologists Should be Cognizant of

Awad Magbri*, Eussera El-Magbri, Mariam El-Magbri, Christi Teets, Cory Casey, Amanda Sinks, Aimee Blankenship, Deanna Szymanski, Shaukat Rashid and Balhinder Brar

Toledo Vascular Access Center, Toledo, OH, USA

*Address for Correspondence: Awad Magbri, Toledo Vascular Access Center, Toledo, OH, USA, E-mail: elmagbri@hotmail.com

Received: 19 September 2018; Accepted: 18 October 2018; Published: 20 October 2018

Citation of this article: Magbri, A., EL-Magbri, E, EL-Magbri, M, Teets, C., Casey, C., Sinks, A., et al.(2018) Persistent Left Superior Vena Cava in a Patient who is Hemodialysis Dependent: A Rare Anomaly of Venous Drainage that Interventional Nephrologists Should be Cognizant of. Arch Nephrol, 1(1): 001-003.

Copyright: © 2018 Magbri, A, et al. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

ABSTRACT

This is a case of 54 year-old African American man who is dialysis dependent secondary to multiple myeloma involving the kidneys. He has left internal jugular central venous catheter. He came to Toledo vascular access center for exchange of his catheter due to mal-function. During the exchange of his catheter we noted that the patient has persistent left superior vena cava with porta-cath in the right superior vena cava. His coronary sinus was hugely dilated on the angiogram. His persistent left superior vena cava drains in the right side of the heart via the coronary sinus. Interventionalists including interventional nephrologists should be aware of this anomaly as they deal with central catheter insertion and exchange.

Keywords: Persistent left superior vena cava, Central venous catheter, Arrhythmia, Cardiac anomaly, Coronary sinus

Case History

The patient is 54 year-old African American man with history of anemia of chronic disease and iron deficiency, diagnosed recently with multiple myeloma which affected the kidney and he is now on hemodialysis (HD) via a left internal jugular tunneled dialysis catheter. The patient has a history of hypertension and hyperlipidemia. He came to Toledo Vascular Access Center for exchange of his mal-functioning catheter. The left internal jugular catheter had been in for the last 2 weeks. This catheter was placed in outside hospital and the hospital record is not available. The patient was prepped and draped for the procedure. The pre-procedure fluoroscopy revealed persistent left superior vena cava where the catheter is lying. Before placing the new catheter a contrast angiogram reveals that the coronary sinus was

dilated and the persistent left superior vena cava draining into the right side of the heart, (Figures 1-5). It is not known if the clinician who placed the central venous catheter in the hospital is aware of the anomaly or he had used the fluoroscopy to check the position of the catheter for malposition or any complication results from the procedure.

Nonetheless, the position of the previous catheter is very suggestive for persistent left sided superior vena cava. The patient has a porta-cath in the right superior vena cava demonstrating the existence of the right SVC.

Discussion of the Case

Persistent left superior vena cava (PLSVC) is a rare but the most

common thoracic vein anomaly. Its frequency is 0.3 to 0.5% of the general population, and it is associated with congenital heart disease in up to 12%. There are 2 types of PLSVC;

- A) The most common type, the PLSVC connected to the right atrium via coronary sinus (90% of this anomaly)

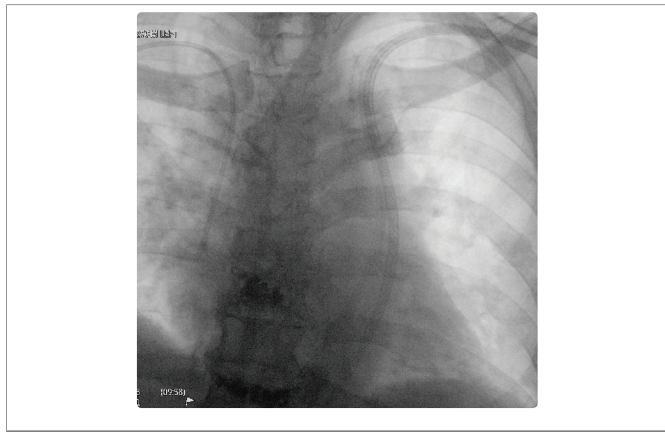


Figure 1: The previous catheter in the left internal jugular and ending in the persistent left superior vena cava. Porta-cath is also seen in the right internal jugular vein with its end in the right superior vena cava.

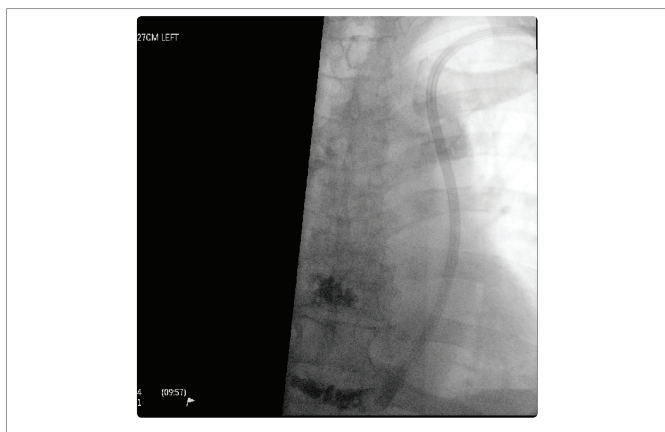


Figure 2: The new catheter in the left internal jugular vein with its tip near the coronary sinus of the persistent left superior vena cava.

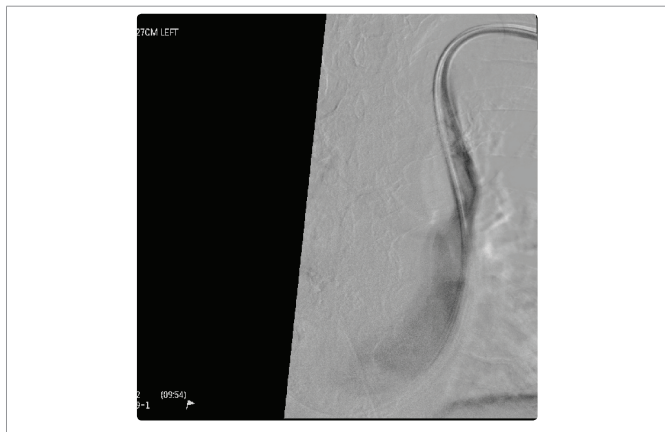


Figure 3: An angiogram through the previous catheter showing the persistent left superior vena cava.

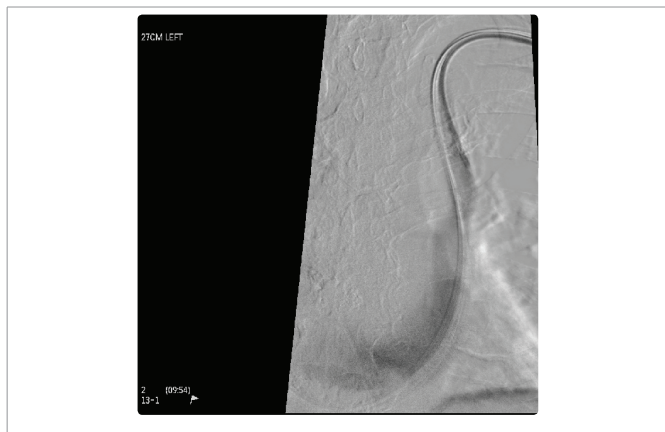


Figure 4: The angiogram showed dilated right coronary sinus.

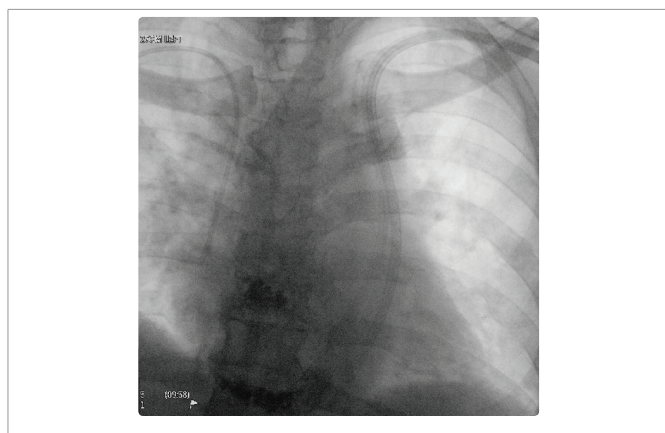


Figure 5: The new catheter in the persistent left superior vena cava with the porta-cath in the right superior vena cava.

- B) In the other 10% of cases the PLSVC connects to the left atrium, this type is associated with intra-cardiac shunts like atrial septal defect (ASD), ventricular septal defect (VSD), or heterotaxy syndromes. In latter cases of anomaly the right to left shunt is usually of small magnitude.

The abnormality is due to failure of regression of the left anterior and common cardinal veins and the left sinus horn in embryo life. The course of the PLSVC is that in 90% of cases it drains into the right atrium where it starts at the junction of the left subclavian vein and the left internal jugular vein [1-3]. It passes lateral to the aortic arch where it receives blood from the left superior intercostal vein. Anterior to the left hilum it is joined by the hemizygous system and crosses adjacent to the posterior wall of the left atrium. Here it receives the great cardiac vein to form the coronary sinus (CS).

In 10-20% of cases the PLSVC drains into the left atrium. Here the PLSVC adopts the same course as previously mentioned but passes between the left atrial appendages (anteriorly) and the left superior pulmonary vein posteriorly [4].

The PLSVC is associated with absent or small left brachiocephalic vein in 65% of cases as in the case under discussion. This anomaly produces bilateral superior venae cava which is present in the majority of individuals with this anomaly. In small percentage of patients, the right superior vena cava is absent (10-18%).

When the PLSVC joins the coronary sinus, it can cause dilatation of the coronary sinus and partially occludes the mitral valve which can impair the left ventricular inflow. This can cause sluggish of blood flow into the left ventricle with a risk of thrombus formation in the left atrium [5,6].

Most patients with PLSVC are asymptomatic, however, cardiac arrhythmia due to sinus node and atrioventricular node dysfunctions can occur. This arrhythmia is caused by enlargement and dilatation of the right atrium or the coronary sinus [7,8]. The PLSVC drains about 20% of the whole venous return and therefore significantly enhanced venous return to the CS forces an increase in its dimensions [9].

Most PLSVC are accidentally found during central venous cannulation, pacemaker insertion, intra-cardiac defibrillator (ICD) implantation, cardiopulmonary bypass, or autopsy [10,11]. Irritation of the coronary sinus during insertion of cardiac devices can cause severe hypotension, cardiac arrhythmia, myocardial ischemia, and cardiac arrest. PLSVC is commonly the only anomaly, however, it can be associated with ASD, ventricular septal defect (VSD), pulmonary stenosis, Tetralogy of Fallot, and anomalous pulmonary drainage [4].

The failure of the left posterior cardinal vein to degenerate in the eighth week of the embryo life results in a persistent left superior vena cava state. This anomaly can be associated with a left azygous vein on some occasions [12,13]. The implications of PLSVC could be important for clinicians who are involved in placement of central venous-access devices [14]. Interventional nephrologists because of the nature of their jobs of performing interventional procedures like insertion or exchanges of tunneled dialysis catheters on patients with end-stage renal disease should be more cognizant of the PLSVC.

Conclusion

This case showed bilateral superior venae cavae due to PLSVC. PLSVC is a rare anomaly that can have significant implications for the internationalists including interventional nephrologists who deal with insertion and exchange of central venous catheters for hemodialysis patients. The anomaly can be associated with right SVC and azygos veins and care should be exercised when placing catheters or cardiac devices on the left venous system of the body. The left brachiocephalic vein (BCV) is not seen in this patient denoting a small or vestigial BCV. The PLSVC in this case drains into the right coronary sinus, the latter is hugely dilated as depicted in the accompanying angiogram. Heart failure and arrhythmia can be associated with PLSVC.

References

1. Sarodia, BD., Stoller, JK. (2000) Persistent left superior vena cava: case report and literature review. *Respir Care*, 45(4): 411-416.

2. Gaynor, JW., Weinberg, PM., Spray, TL. (2000) Congenital heart surgery nomenclature and data project: systemic venous anomalies. *Ann Thorac Surg*, 69(3): 70-76.
3. Cormier, MG., Yedlicka, JW., Gray RJ., Rogelio, M. (1989) Congenital anomalies of the superior vena cava: a CT study. *Semin Roentgenol*, 24(2): 77-83.
4. MacDonald, ST., Emmanuel, Y., Myerson, S., Prendergast, B., Neubauer, S., Leeson, P. (2009) Absent right superior vena cava: multimodality imaging of upper body venous drainage via left-sided superior vena cava and azygos venous system. *Circ Cardiovasc Imaging*, 2(5): 34-36.
5. Santoscoy, R., Walters, HL 3rd., Ross, RD., Lyons, JM., Hakimi, M. (1996) Coronary sinus ostial atresia with persistent left superior vena cava. *Ann Thorac Surg*, 61(3): 879-882.
6. Muster, AJ., Naheed, ZJ., Backer, CL., Mavroudis, C. (1998) Is surgical ligation of an accessory left superior vena cava always safe? *Pediatr Cardiol*, 19(4): 352-354.
7. Pachon, M JC., Pachon, M EL., Pachon, M JC., Lobo, TJ., Pachon, MZ., Vargas, RN., et al. (2004) A new treatment for atrial fibrillation based on spectral analysis to guide the catheter RF-ablation. *Europace*, 6(6): 590-601.
8. Hsu, LF., Jais, P., Keane, D., Wharton, JM., Isabel, D., Méléze, H., et al. (2004) Atrial fibrillation originating from persistent left superior vena cava. *Circulation*, 109(7): 828-832.
9. Pivoski, SP., Khabiri, H. (2011) Persistent left superior vena cava: review of the literature, clinical implications, and relevance of alterations in thoracic central venous anatomy as pertaining in the general principles of central venous access device placement and venography in cancer patients. *World J Surg Oncol*, 9: 173.
10. Elison, B., Evans, D., Zanders, T., Jeanmonod, R. (2014) Persistent left superior vena cava draining into the pulmonary venous system discovered after central venous catheter placement. *Am J Emerg Med*, 32(8): 943.
11. Luckianow, G., Cole, D., Kaplan, L. (2009) Anatomic variant found during catheter insertion. *J Am Acad Phys Assist*, 22(9): 60-63.
12. Uemura, M., Takemura, A., Ehara, D., Yasumitsu, H., Ohnishi, Y., Suwa, F. (2009) Left superior vena cava with left azygos vein. *Okajimas Folia Anat Jpn*, 86(2): 55-60.
13. Abdulla, R., Blew, GA., Holterman, MJ. (2004) Cardiovascular embryology. *Pediatr Cardiol*, 25(3): 191-200.
14. Pivoski, SP. (2000) A prospective analysis of the cephalic vein cutdown approach for chronic indwelling central venous access in 100 consecutive cancer patients. *Ann Surg Oncol*, 7(7): 496-502.