Persistent Left Superior Vena Cava in Hematological Malignancy Requiring Central Venous Catheter Insertion for Intensive Chemotherapy

Osamu Imataki a Hiroyuki Kubo a Yukiko Hamasaki a Maki Oku a Jun-ichiro Kida a Makiko Uemura a Harumi Matsuka b

aDivision of Hematology and Stem Cell Transplantation, Faculty of Medicine, Kagawa University, and bNursing Division, Kagawa University Hospital, Kagawa, Japan

Key Words
Persistent left superior vena cava · Coronary sinus atresia · Central venous catheter · Chemotherapy · Hematological malignancy

Abstract
Persistent left superior vena cava is a congenital vascular anomaly, which is possibly arrhythmogenic and thrombogenic, rarely complicated with coronary sinus atresia. We treated a 42-year-old male with Hodgkin’s lymphoma requiring central venous catheter placement for intensive chemotherapy. Persistent left superior vena cava was revealed after the insertion of the central venous catheter by the radiological finding of the catheter tip cannulated into the vena cava cavity. The relationship between coronary sinus atresia and persistent left superior vena cava induced by central venous catheterization remains unclear; however, the hematologist should pay attention to the malpositioning of the central venous catheter.

Background
Persistent left superior vena cava (PLSVC) is a congenital but benign vascular anomaly [1] with a prevalence of 0.3–0.5% in the general population [2, 3]. PLSVC is one of the most...
common cardiovascular anomalies but is generally asymptomatic, and it is thus often identified accidentally during cardiovascular ultrasound/CT scan screening or coronary interventional angiography [4]. Anatomically, PLSVC causes the aberrant backflow vein to drain into the retrograde positioned coronary sinus [5].

Some researchers have called attention to the risk of atresia of the coronary sinus among the anomalies associated with PLSVC [4, 6], but there are no recommendations regarding anticoagulation therapy for patients with PLSVC. Some anecdotal case reports describe PLSVC-associated arrhythmia such as atrial fibrillation [7, 8]. Accordingly, PLSVC is understood as possibly arrhythmogenic. Nevertheless, coronary sinus atresia (CSA) due to PLSVC is a rare complication.

In patients with hematological malignancies, the insertion of a central venous catheter (CVC) is required for the patient to undergo intensive chemotherapy. Hematologists must make a decision whether to conduct a CVC insertion for patients with a given complication. What if the patient has a cardiac abnormality, e.g., PLSVC? There are a few reports of CSA in PLSVC induced by central venous catheterization [5], pacemaker wire placement, or cardiac surgery [9]. The placement of a CVC might evoke arrhythmia due to an additive enhancement of the patient’s arrhythmogenic condition. Considering such a situation, it may not be advisable to insert a CVC in a PLSVC patient with a hematological malignancy.

**Case Presentation**

We treated a 42-year-old male with stage IV Hodgkin’s lymphoma with bone marrow involvement. The initial chemotherapy (ABVd, doxorubicin 25 mg/m², bleomycin 9 mg/m², vinblastine 6 mg/m², and dacarbazine 250 mg/m², day 1 and 15) was effective, resulting in complete remission after 6 cycles of ABVd. However, after 15 months he relapsed. Although the ABVd chemotherapy had been administered via a peripheral vein, salvage chemotherapy was initiated via an implantable venous access port catheter because his peripheral blood accesses were ultimately unusable. He received an indwelling port catheter from the left subclavian vein, since the right subclavian approach was not possible. The port catheter happened to be cannulated PLSVC (fig. 1a), which was found at the time for the first time in his life.

The patient successfully completed a total of 3 courses of salvage chemotherapy constructed with AraC, carboplatin, etoposide, and methylprednisolone (ACES). Although obstruction of the coronary venous drainage caused by PLSVC has been suggested, it is not always thrombogenic [10]. The formation of a thrombus may depend on the PLSVC flow. A small PLSVC flow can easily connect to hemostasis of the venous circulation return. In our patient, the PLSVC cavity had enough space for a port catheter to be cannulated and positioned (fig. 1b). Ideally the back flow and diameter of the PLSVC to the coronary sinus should be evaluated by cardiovascular ultrasound or cardiography [11].

**Conclusions**

Although PLSVC is a rare congenital anomaly, some risks including thrombosis and occlusion should be annotated when the patient receives a CVC insertion. The relationship between CSA and PLSVC remains unclear; however, the hematologist should pay attention to the malpositioning of the CVC tip.
Acknowledgements

We thank KN International, Inc., which provided medical writing services.

Statement of Ethics

Informed consent to participate in the study was obtained from the participant. Written informed consent was obtained from the patient for publication of this case series and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Disclosure Statement

The authors declare that they have no competing interests.

References

1. Shum JS, Kim SM, Choe YH: Multidetector CT and MRI of ostial atresia of the coronary sinus, associated collateral venous pathways and cardiac anomalies. Clin Radiol 2012;67:47–52.
2. Tak T, Crouch E, Drake GB: Persistent left superior vena cava: incidence, significance and clinical correlates. Int J Cardiol 2002;82:91–93.
3. Biffi M, Boriani G, Frabetti L, Bronzetti G, Branzi A: Left superior vena cava persistence in patients undergoing pacemaker or cardioverter-defibrillator implantation: a 10-year experience. Chest 2001;120:139–144.
4. Santoscoy R, Walters HL 3rd, Ross RD, Lyons JM, Hakimi M: Coronary sinus ostial atresia with persistent left superior vena cava. Ann Thorac Surg 1996;61:879–882.
5. Saranteas T, Mandila C, Poularas J, Papanikolaou J, Patriankos A, Karakitsos D, Karabnis A: Transeosophageal echocardiography and vascular ultrasound in the diagnosis of catheter-related persistent left superior vena cava thrombosis. Eur J Echocardiogr 2009;10:452–455.
6. Qanadli SD, Rolf T, Glaser F, Delay D, Beigelman-Aubry C, Prêtre R: Coronary sinus atresia with persistent left superior vena cava: unusual clinical presentation and endovascular management. Cardiovasc Intervent Radiol 2014;37:825–828.
7. Hsu LF, Jais P, Keane D, Wharton JM, Deisenhofer I, Hocini M, Shah DC, Sanders P, Scavée C, Weerasooriya R, Clémenty J, Haïssaguerre M: Atrial fibrillation originating from persistent left superior vena cava. Circulation 2004;109:828–832.
8. Luik A, Deisenhofer I, Estner H, Ndrepepa G, Pflaumer A, Zrenner B, Schmitt C: Atresia of the coronary sinus in patients with supraventricular tachycardia. Pacing Clin Electrophysiol 2006;29:171–174.
9. Lim PC, Baskaran L, Ho KL, Teo WS, Ching CK: Coronary sinus ostial atresia and persistent left-sided superior vena cava: clinical significance and strategies for cardiac resynchronization therapy. Int J Angiol 2013;22:199–202.
10. Siddiqui AM, Cao LB, Movahed A: Side matters: an intriguing case of persistent left superior vena-cava. World J Clin Cases 2013;1:159–161.
11. Goyal SK, Punnam SR, Verma G, Ruberg FL: Persistent left superior vena cava: a case report and review of literature. Cardiovasc Ultrasound 2008;6:50.
Fig. 1. a The patient’s chest X-ray indicated the positioning of the catheter tip in the left mediastinum (arrowhead), suggesting that the catheter was aberrantly placed (i.e., not into the normal insertion site, the right-sided superior vena cava). b CT scan depicting an indwelling venous catheter port placed into the PLSVC (arrowhead); coronal (top) and transverse view (bottom).