

Journal of INTERNATIONAL MEDICAL RESEARCH

Case report of double superior vena cava and double odd vein with hypoplastic left brachiocephalic vein Journal of International Medical Research 2022, Vol. 50(9) 1–6 © The Author(s) 2022 Article reuse guidelines: sagepub.com/journals-permissions DOI: 10.1177/03000605221119655 journals.sagepub.com/home/imr



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Abstract

Case Reports

Persistent left superior vena cava (PLSVC) is a common venous variation that is usually accompanied by an absence of the left brachiocephalic vein, and displays a higher incidence in patients with congenital heart disease. Here, the case of a 57-year-old male patient who was found to have PLSVC on chest computed tomography (CT) during screening for gastric cancer metastasis at the Affiliated Hospital of Qinghai University, is described. Further coronal CT and three-dimensional reconstruction of the chest revealed the patient's double superior vena cava (DSVC), double odd veins, and left brachiocephalic vein dysplasia. The patient did not have congenital heart disease and the case was associated with dysplasia of the left brachiocephalic vein, indicating an unusual and rare venous abnormality. At the time of writing, the patient was receiving antitumour therapy.

Keywords

Double superior vena cava, hypoplastic left brachiocephalic vein, persistent left superior vena cava, congenital heart disease, venous abnormality, chest CT

Date received: 9 February 2022; accepted: 27 July 2022

Introduction

Anomalies of the superior vena cava (SVC) are usually congenital, and are sometimes combined with congenital heart disease. However, most patients have no obvious symptoms. The SVC drains blood from the head and upper extremities into the right atrium. Normally, the right side

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drains through the right brachiocephalic vein, while the left side drains through the left brachiocephalic vein. In some cases, a double superior vena cava (DSVC) may be encountered, and these vessels are referred to as left and right SVC.¹ Here, the case of a patient with persistent left SVC (PLSVC) is described.

Case report

All patient details were deidentified for this case report, thus, the patient's informed consent was not considered necessary, and ethics approval was not required for report publication. Informed consent for treatment was obtained from the patient and the reporting of this study conforms to CARE guidelines.²

A 57-year-old male patient was admitted to the Oncology Department of the Affiliated Hospital of Qinghai University, Xining, China in September 2021, to undergo gastroscopy and pathological confirmation of gastric cancer due to abdominal discomfort. The patient was screened for gastric cancer metastasis, and during plain and enhanced computed tomography (CT) of the chest (Revolution CT scanner; GE Healthcare, Chicago, IL, USA), the left internal jugular vein and subclavian vein were found to be converged behind the left sternoclavicular joint to form the left SVC, and did not converge downward to the right with the right brachiocephalic vein. The patient's right SVC had a normal course with an underdeveloped left brachiocephalic vein (Figure 1). Coronary artery reconstruction was performed with GE AW Workstation software, version 4.7 (GE Healthcare) to determine the direction of the left SVC, and revealed that the merged left SVC extended downward, passing through the left side of the aortic arch, the root of the left pulmonary artery, and the inside of the left pulmonary vein (Figure 2). Three-dimensional (3D) reconstruction showed that the left SVC travelled rightward



Figure 1. Computed tomography mediastinal window of the chest from a 57-year-old male patient, showing the left cephalic brachial vein with hypoplasia (red arrow), and the left superior vena cava (white arrow).



Figure 2. Computed tomography coronal reconstruction of the thorax from a 57-year-old male patient, showing the hypoplastic bridging left cephalic brachial vein (red arrow) and the left and right superior vena cava (white arrow).

along the coronary sulcus behind the left auricle and merged into the enlarged coronary sinus, which opened into the right atrium (Figure 3). During the 3D reconstruction, double odd veins were also observed. The patient's left odd vein started from the left lumbar ascending vein, ascended along the left side of the spine, and merged into the left SVC posteriorly at the level of the 4th thoracic vertebral body, via the left pulmonary root posteriorly



Figure 3. Three-dimensional reconstruction of the heart and blood vessels based on chest computed tomography from a 57-year-old male patient, with white arrows marking the course of the left superior vena cava.



Figure 4. Computed tomography mediastinal window of the chest from a 57-year-old male patient, showing the left odd vein (white arrow).

above the left SVC, before it entered the coronary sinus (Figures 4 and 5). The patient required no treatment for vascular abnormalities, and at the time of writing, was receiving antineoplastic treatment.

Discussion

Persistent left superior vena cava is formed due to failure of the left anterior main vein

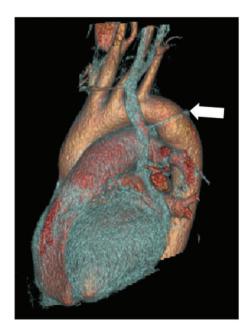


Figure 5. Three-dimensional reconstruction of the heart and blood vessels based on chest computed tomography from a 57-year-old male patient, showing the left odd vein (white arrow).

to close during embryonic development, the development of Cuvier's canal, or the poorly developed traffic branch between the left and right anterior main veins.³⁻⁶ The incidence of DSVC in the normal population is 0.3%, but is higher, at around 10-11%, in patients with congenital heart disease;⁷ therefore, the combination of asymptomatic congenital heart disease and other vascular anomalies should alert to the potential presence of DSVC.⁸ In most cases DSVC is asymptomatic, however, in patients with DSVC combined with a cardiac defect, such as an atrial septal defect, the right-to-left shunt has haemodynamic implications.9 Because DSVC can present with severe cardiac anomalies, further cardiology investigations (e.g., transthoracic ultrasonography, magnetic resonance imaging, or cardiac CT using contrast) are required.¹⁰ In the present case, enhanced

CT of the chest did not show any evidence of cardiac defects.

In 1965, Nandy et al.¹¹ classified PLSVC into various types according to its relationship with the right SVC and left brachiocephalic vein, and its confluence site, as follows: (1) DSVC without left brachiocephalic vein; (2) DSVC with left brachiocephalic vein; (3) absence of right SVC and persistence of left SVC; and (4) DSVC with bilateral odd veins. Of note, the present case does not match the above classification, as although DSVC with bilateral azygos veins meets the definition classified as type 4 PLSVC, the left brachiocephalic vein is not described as present. The left brachiocephalic vein has been shown as missing in more than half of cases with PLSVC.¹² In most patients with left brachiocephalic vein absence. PLSVC is formed because absence of the left brachiocephalic vein leads to persistence of the left anterior main vein. In the current case, PLSVC and brachiocephalic veins were present, but there was no congenital heart disease. We considered whether the brachiocephalic vein had developed due to the absence of congenital heart disease in the fetus, but the brachiocephalic vein exhibited dysplasia and stenosis due to the presence of PLSVC. Thus, the DSVC and bilateral azygos veins with left brachiocephalic vein dysplasia observed in the current patient are a rare anomaly. In the present case, no discomfort was observed in the patient, which may be related to the lack of congenital heart disease.

In the absence of a comorbid heart valve defect, DSVC is generally asymptomatic and is found incidentally during examination of the patient at the time of presentation for different reasons. However, DSVC may be detected as a component of complex cardiology due to PLSVC, resulting in enlargement of the venous sinuses, and this enlargement may lead to compression of the AV node and the bundle of His, possibly inducing arrhythmias. This enlargement may also lead to compression of the left atrium and reduced cardiac output.^{13,14}

During cardiac surgery in patients with DSVC, recognition of the PLSVC, and use of appropriate cannulation techniques to eliminate the large amount of venous blood entering the heart through the coronary sinus in the body circulation, are important. In addition, it is important to determine whether anastomotic or collateral vessels exist between the left and right SVC, or, in some cases, whether the right SVC is absent, in which case ligation of the PLSVC may result in venous filling of the head and arm (superior vena cava syndrome).¹⁵ In patients with DSVC, very rare cases of PLSVC with coronary sinus agenesis have been reported, usually associated with atrial defect. This abnormality often leads to cyanosis and reduced stress, with retrograde arrest being an absolute contraindication.^{16,17}

Amongst patients with DSVC, serious complications may occur when left subclavian vein cannulation is required, such as in oncology or dialysis patients, or during pacemaker implantation. Complications may include left subclavian vein thrombosis, arrhythmia, coronary sinus perforation, pericardial tamponade, cardiogenic shock, or even death.^{11,18–22} Therefore, amongst this group of patients, more detailed treatment plans are required.

Conclusion

The incidence of PLSVC is usually high in patients with congenital heart disease. Therefore, fetuses with an abnormal SVC or left brachiocephalic vein that is identified during prenatal examination should be screened for congenital heart disease to facilitate early intervention and reduce complications. Echocardiographic examination remains necessary during screening of adult patients with PLSVC. Decisions on whether or not to provide treatment may be reached according to the results of echocardiography and the presence or absence of clinical symptoms. In addition, amongst patients with PLSVC, clinicians should fully understand the vascular anatomy when performing an invasive operation, with the aim of reducing the occurrence of complications.

Data accessibility

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Acknowledgements

We thank our colleagues in the relevant clinical departments for providing us with test and imaging guidance.

Author contributions

HL and HW wrote the original manuscript; XH and GL helped design the project; GL supervised the manuscript; all authors reviewed the final version.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Funding

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

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