Persistent Left Superior Vena Cava

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Persistent left superior vena cava (LPSVC) is a rare and important congenital venous anomaly and it is caused by a defect in the closure of the left anterior cardinal vein during cardiac development. It can lead to coronary sinus (CS) dilatation when it drains to right atrium. In another perspective, this dilatation is very important to understand the way of drainage of LPSVC. Additionally, knowledge of LPSVC's physiopathology is also important before some interventional and surgery procedures.
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The superior vena cava (SVC) is the great venous trunk that receive deoxygenated blood from the upper half of the body, above the diaphragm to the right atrium of the heart. It is a large diameter (24 mm), yet short vein. It is located in the anterior right superior mediastinum.

Duplication of superior vena cava (SVC) is a rare anomaly. The incidence of double SVC in general population is 0.3% whereas in patients with congenital heart disease it varies between 10-11%. Double SVC cases have clinical importance if especially the one on the left side drains into the left atrium. Moreover, double SVC is surgically important in the presence of congenital heart disease.[1]

The presence of double SVC is important from a clinical and surgical perspective as it may cause physiological and hemodynamic alterations. It is important to recognize this abnormality when cardiac catheterization, echocardiography, angiography, and open-heart surgery is being undertaken.[2]

The precise and elegant rendering of the human anatomy achieved by Computed Tomography (CT) has, for over a decade,

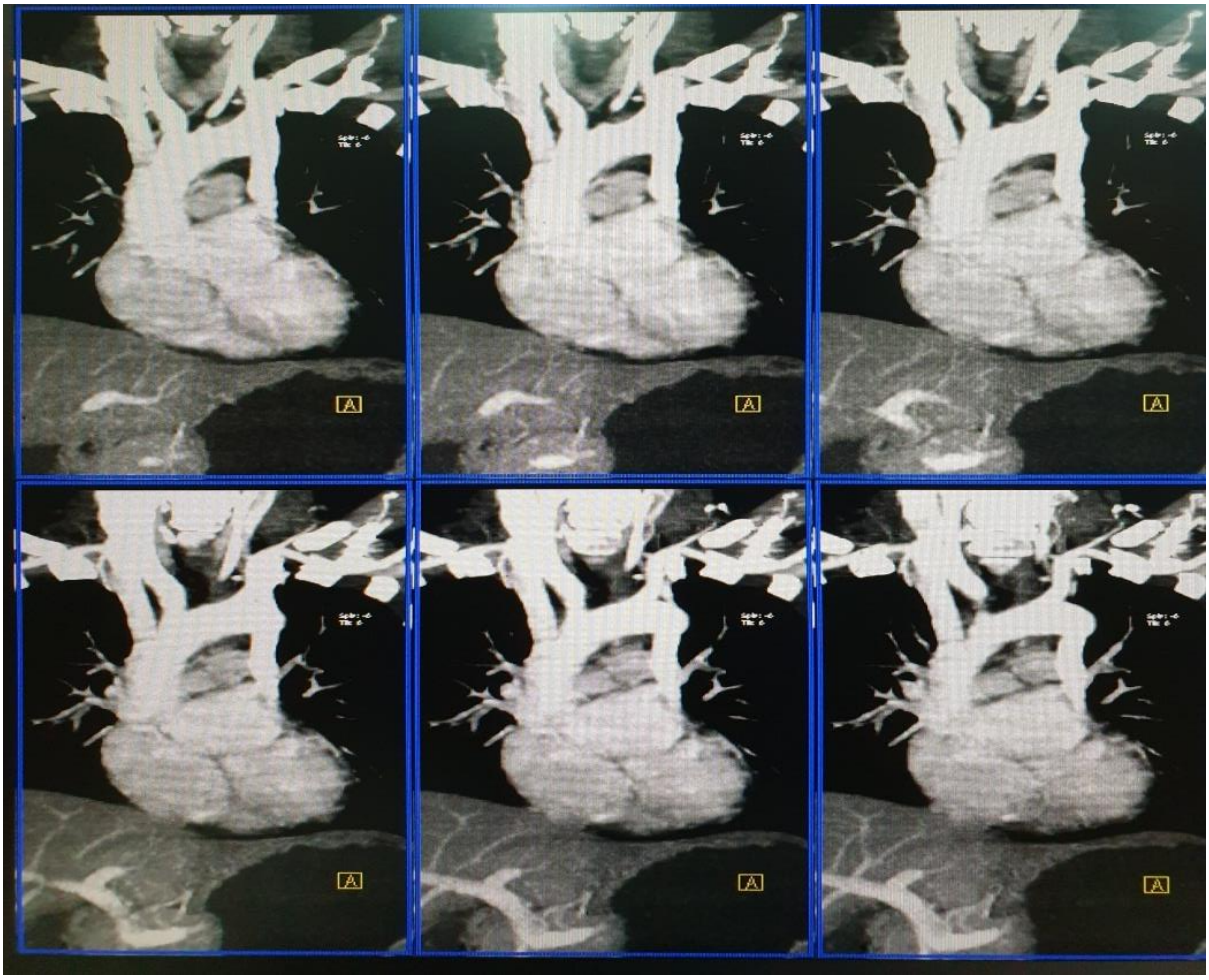

Figure 1. Left persistent superior vena cava arising directly from the confluence of internal jugular vein and subclavian vein.

provided a novel perspective to the medical diagnostic armamentarium. CT has had a significant impact on almost every avenue in the daily practice of medicine and has gained broad acceptance by our clinical colleagues. A wide variety of congenital vascular anomalies of the upper mediastinum exist. Since most of these are clinically silent, they are often unsuspected and discovered incidentally on radiographic studies done for other reasons. Familiarity with the CT appearance of such anomalies may aid in the interpretation of otherwise potentially confusing CT images.[3]

A thorough knowledge of thoracic and cervical vascular anatomy is extremely important for vascular surgeons, surgical oncologists, interventional radiologists and other physicians involved in management of oncology patients who sometimes need to perform procedures to obtain deep venous access. Persistent left superior vena cava (PLSVC) is the most common congenital thoracic venous anomaly and knowing about it is critical to the success of implanting invasive devices, in terms of minimizing the risks of potential complications.[4]

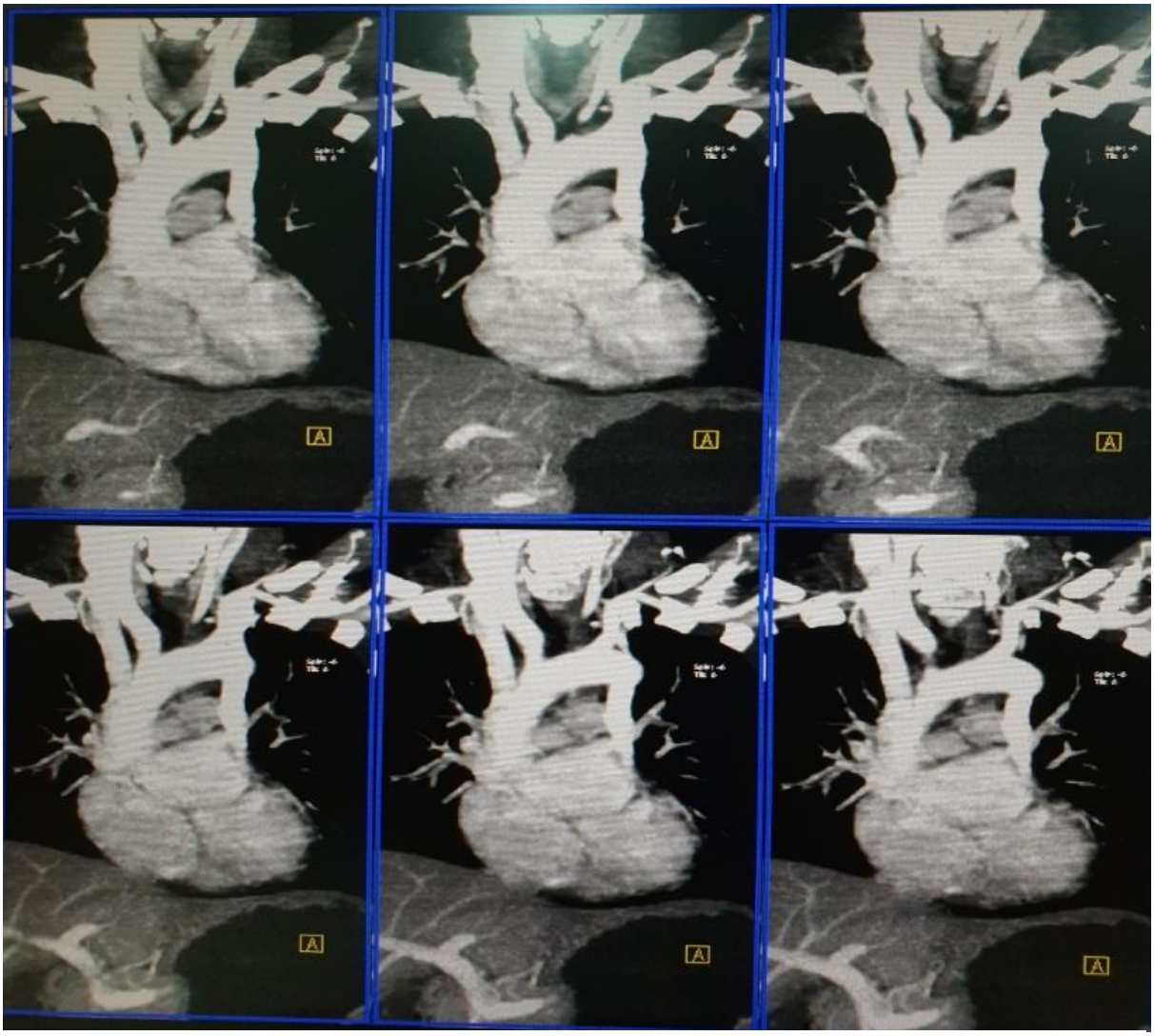

Figure 2. Normal side superior vena cava arising from the right confluence of internal jugular vein and subclavian vein

Case report

A 48-year-old lady visited an orthopaedic surgeon with right supraclavicular region pain. She was advised chest x-ray which was normal. Then she was referred to a physician. She had duodenitits on endoscopy and was under medication for the same. On follow up she had no relief of symptoms and the pain in supraclavicular region persisted. On examination she had anemia and her blood report showed decreased hemoglobin level. She was then advised Contrast Enhanced Computed Tomography (CECT) chest and abdomen.

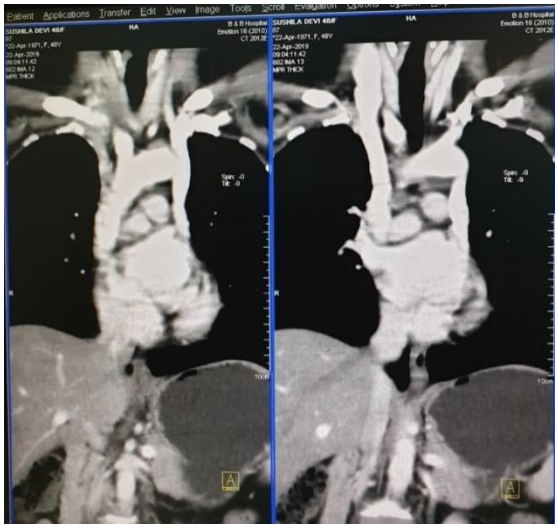

Figure 3. Left persistent SVC enters into the right atrium along with right SVC.

Discussion

LPSVC is a rare organogenesis abnormality. The ratio of LPSVC is 0.3-0.5% in the general population. It is due to the persistence of the terminal part of the left anterior cardinal vein, which normally involutes in the sixth month of uterine life. The major venous drainage system of the embryo is constituted by the cardinal veins. The oblique anastomotic branch joins to the anterior cardinal veins and two of them forms left brachiocephalic vein. Normally the right anterior cardinal vein and the right common cardinal vein merge to form the SVC while the left anterior cardinal vein and the left common cardinal vein must undergo atrophy. Patency of embryonic veins lead to multiple variations for example double SVC, double inferior vena cava (IVC) or other venous connections which don't normally exist. The PLSVC represents failure of obliteration of the left anterior cardinal vein in early embryological development.[5]

Radiological features

Contrast enhanced CT chest show an anomalous vessel coursing inferiorly to the left of the aortic arch and anterior to the left hilum. It is in direct continuation with the confluence of the left subclavian vein and the left internal jugular vein (**Figure 1**). Right SVC is also in direct continuation with the confluence of right subclavian and right internal jugular vein **(Figure 2)**. There is no communication seen between right and left SVC. Inferiorly both SVC drain in to the right atrium separately **(Figure 3)**. Findings are consistent with double SVC due to the presence of a persistent left SVC (anatomical variant) along with a normal right SVC.

Conclusion

We present a case of PLSVC which is a rare congenital variation. Our patient had no any cardiac symptoms and was an incidental finding. If not associated with congenital heart disease, it is benign condition without any clinical manifestation. In few other studies the incidence rate of double SVC in normal population was 0.12% and congenital heart disease, they found the incidence of bilateral SVC as 11%. Identification of the left SVC is pertinent during cardiopulmonary bypass surgery. The informed and alert cardiac surgeon must correctly assess the presence of a left SVC and take the necessary steps to occlude or drain it into the venous reservoir. The discovery of a left SVC during cardiac catheterization and angiocardiography may induce the investigator to suspect a possible association with other congenital anomalies and especially a left-to-right shunt at the atrial level. It is recommended that in this clinical situation a contrast enhanced computer tomography scan be undertaken before any invasive diagnostic procedure. This investigation readily demonstrates the presence of a left-sided SVC and alerts the attendant clinician to this vascular abnormality.

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