

PERSISTENT LEFT SIDED SVC WITH A BRIDGING / INTER-COMMUNICATING VEIN & STENOSIS OF THE BRIDGING VEIN ON CT

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Abstract:

Persistence of left superior vena cava (PLSVC) is a rare anomaly. The incidence of a persistent left SVC in general population is 0.3-0.5% whereas in congenital heart disease it is between 5-10%. In general, left superior vena cava seldom shows any intercommunication. There are very few cases of left SVC with an intercommunicating vein and even only few reports of stenosis in the bridging vein. We present an incidentally diagnosed case of left SVC with the presence of a bridging vein with stenosis in the intercommunicating vein proximal to left sided SVC in a case of 2 year old girl.

Key-words: Persistent left superior vena cava (PLSVC), Intercommunicating vein, Stenosis, Bilateral superior vena cava

INTRODUCTION

The coexistence of persistent left superior vena cava (pLSVC) and the normal right superior vena cava (RSVC) is called “Double Superior Vena Cava (DSVC)”. Duplication of superior vena cava (SVC) is a rare anomaly with higher incidence in CHD. The incidence of double SVC in the general population is 0.3-0.5%, whereas in patients with congenital heart disease it varies between 5-10%.¹ In general, cases of double superior vena cava mostly show no communication. There are very few cases of persistent left SVC with an intercommunicating innominate vein with stenosis in the bridging vein. Moreover, double SVC is surgically important in the presence of congenital heart disease.² In this article, we present a 2 years old girl who was incidentally diagnosed with double SVC, right draining directly into the right atrium & the left into the right atrium through the coronary sinus. There was an intercommunicating vein between

the right and the left sided SVC with stenosis of the intercommunicating vessel in the portion close to the left sided SVC.

CASE REPORT

A 2 year old girl presented in the pediatric OPD with fever since 15 days and rash over the body. Patient was a known case of rickets with history of delayed milestones. Family history was positive for a deaf and dumb sibling.

Her routine investigations revealed severe anemia with cardiomegaly on CXR. A 2D echo was done for further evaluation. It revealed situs solitus with supra-mitral membrane, moderate mitral stenosis and bilateral SVC with an abnormal communicating channel. A CT aortogram was done to confirm the findings. The CT study revealed a double SVC with the right SVC draining into the right atrium & the persistent left SVC draining into the right

atrium through the coronary sinus. The CT study could clearly delineate the course & calibre of the PLSVC & its relation to adjoining significant anatomical structures. It also revealed an intercommunicating vessel which was stenosed in the portion close to the left SVC. The left atrium was enlarged.

DISCUSSION

Embryologically, the right SVC originates from the junction of the right pre-cardinal vein and the right common cardinal vein. An oblique anastomosis develops, connecting with the pre-cardinal veins to become the left brachiocephalic vein. Typically, the left pre-cardinal vein and the left common cardinal vein undergo atrophy.^{3,4} In patients with a double SVC, this atrophy does not occur and the left SVC remains patent, usually draining into the coronary sinus and then into the right atrium. In approximately 10% of patients the left duplicated SVC attaches to the roof of the left atrium and may cause hemodynamic compromise.

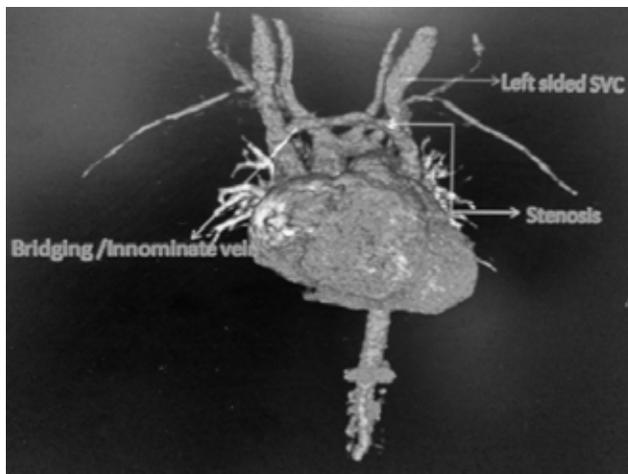
Persistent left SVC is the most common congenital thoracic venous anomaly with a prevalence of 0.3–0.5% in general population.¹ The thoracic embryonic venous system is composed of two large veins (the superior cardinal veins) which return blood from cranial aspect of embryo, and the inferior cardinal vein, which returns blood from the caudal aspect. Both pairs of veins join to form right and left common cardinal veins before entering the embryological heart. The left common cardinal vein persists to form coronary sinus and oblique vein of left atrium. During the 8th week of gestation, an anastomosis forms between right and left superior cardinal veins resulting in the innominate (or brachiocephalic) vein. The cephalic portion of superior cardinal veins form the internal jugular veins. The caudal portion of right superior vein forms the normal right-sided superior vena cava, while the portion of the left superior cardinal vein caudal to the innominate vein normally regresses to become “ligament of Marshall”.

If this normal regression of the left superior cardinal vein fails to occur, a persistent left-sided vascular structure that empties into the coronary sinus results and is called as PLSVC. The innominate vein may or may not degenerate in these cases leading to variations in anatomy. The most common subtype of PLSVC results in the presence of both left and right SVCs. A bridging vein may or may not be present. A PLSVC is associated with absence of a bridging

vein in 65% cases.² Even more rarely, the caudal right superior cardinal vein regresses leading to an absent right SVC with PLSVC.

In our case, CT revealed a vein which was seen to the left of the main pulmonary artery & coursing anterior to the proximal left main pulmonary artery to eventually open into the right atrium through the coronary sinus. This vascular structure was in continuity with the left IJV. It had a maximum diameter of about 7 mm at the level of thoracic inlet. It paralleled the course of the right sided normal SVC till the level of the hilar sections & was nearly of the same size as right sided normal SVC. Diagnosis of PLSVC was confirmed.

The study further revealed an intercommunicating vein which was stenosed in its portion close to PLSVC. This communicating vessel was extending from the inner aspect of the left sided SVC to the proximal portion of the right sided SVC and coursing anterior to the branches of the aortic arch & posterior to the thymic tissue. It measured 4.4 mm in its maximum diameter & was stenosed in the portion close to the left sided SVC where it measured about 2.4 mm in diameter.



Variations have also been reported in the insertion of PLSVC. In 80–90% of individuals, the persistent LSVC drains into the right atrium via the coronary sinus as in our case and is of no hemodynamic consequence. In the remaining cases, it may drain into the left atrium resulting in a right to left sided shunt. Drainage of the right SVC to the LA is an exceedingly rare congenital malformation of systemic veins,⁵ In a previously published case report, Van Praagh and colleagues were able to identify 18 cases of LA drainage of the right SVC, and an additional seven cases of bi-atrial drainage of right SVC in viscer-atrial

situs solitus.⁶

PLSVC is rarely symptomatic in patients without congenital heart disease, however during cardiac catheterization, angiography, echocardiography, and open heart surgery, it is important to be aware of anomalies of the great thoracic veins, including a duplicated SVC. Duplicated SVC can be suspected on chest radiograph if a catheter or line is in the left para mediastinal area and can be diagnosed on CT scan by tracking the anomalous vessel from the left jugular and subclavian veins to the coronary sinus.

In the absence of congenital heart disease, duplicated SVC is benign and no treatment is necessary. A double superior vena cava may make it difficult for internal jugular or subclavian venous catheterization, radiofrequency ablation, pacemaker insertion or coronary artery bypass graft. If a right to left shunt is present then cyanosis, sepsis and cerebral abscess may possibly occur. Thus, the prognosis is excellent although usually asymptomatic, the congenital anomalies of the vena cava must be correctly identified by radiologists. This can both avoid misinterpretation of unexpected imaging findings and allow a correct planning of surgical and interventional treatments when an anomalous venous map is known in advance.

Other than direct observation of the vessel, there are some radiological clues that can point to the existence of PLSVC, including (i) an enlarged and densely opaque coronary sinus on CT when intravenous contrast is injected from the left arm, (ii) the presence of focal mediastinal widening superior to the left side of the aortic knob on chest radiography and (iii) the aberrant course of the intravenous catheter approaching the right atrium from the left arm.

When a double superior vena cava is diagnosed and if other congenital anomalies are discovered then comprehension of the risks related to catheterization and selection of a safe route should be considered. A double SVC may account for aberrant appearances while positioning a central venous catheter placed from the left side of the body. It is safer to have a definite venous access with the help of ultrasound guidance. The injection of contrast will help delineate the venous anatomy when a variation in the venous anatomy is suspected.³

Almost 40% of patients with PLSVC can have a variety of associated cardiac anomalies,^{4,7} such as atrial septal defect, bicuspid aortic valve, coarctation of aorta, coronary sinus

ostial atresia, and cor triatriatum. The presence of associated anomalies is more common with concomitant absence of right SVC the notation of which warrants appropriate investigation to rule out other anomalies. The PLSVC has been associated with anatomical and architectural abnormalities of the sinus node and conduction tissues. Both sinus and AV node can have persistent fetal dispersion in the central fibrous body in subjects with PLSVC,⁸ but in this case a supra-mitral membrane & mitral stenosis was seen.

REFERENCES

1. Wood P: Diseases of heart and circulation. 1956, Philadelphia: JB Lippincott, 2Google Scholar
2. Webb W, Gamsu G, Speckman J, Kaiser J, Federle M, Lipton M: Computed tomographic demonstration of mediastinal venous anomalies. *AJR Am J Roentgenol.* 1982, 139 (1): 157-161. View ArticlePubMedGoogle Scholar
3. Voci P, Luzi G, Agati L: Diagnosis of persistent left superior vena cava by multiplane transesophageal echocardiography. *Cardiologia.* 1995, 40 (4): 273-275. PubMedGoogle Scholar
4. Sarodia B, Stoller J: Persistent left superior vena cava: case report and literature review. *Respir Care.* 2000, 45 (4): 411-416. PubMedGoogle Scholar
5. Winter F: Persistent left superior vena cava; survey of world literature and report of thirty additional cases. *Angiology.* 1954, 5 (2): 90-132. 10.1177/000331975400500207. View Article Pub Med Google Scholar
6. James T, Marshall T: XVIII. Persistent fetal dispersion of the atrioventricular node and His bundle within the central fibrous body. *Circulation.* 1976, 53 (6): 1026-1034. View ArticlePubMedGoogle Scholar
7. Gorenflo M., Sebening C., Ulmer H.E. Anomalous connection of the right superior caval vein to the morphologically left atrium. *Cardiol Young.* 2006;16(2):184–186. [PubMed]

8. Van Praagh S., Geva T., Lock J.E., Nido P.J., Vance M.S., Van Praagh R. Biatrial or left atrial drainage of the right superior vena cava: anatomic, morphogenetic, and surgical considerations—report of three new cases and literature review. *Pediatr Cardiol.* 2003;24(4):350–363. [PubMed]