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Hemiazygous continuation of inferior vena cava draining into the coronary sinus *via* persistent left superior vena cava: a rare anomaly

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Abstract

We present a case of left sided inferior vena cava with hemiazygous continuation draining into the coronary sinus via the left persistent superior vena cava. This was incidentally found in an individual referred to our centre for evaluation of palpitations. These caval anomalies are rare, and are often associated with no clinical manifestations. However, it is necessary to recognize them during routine workup to avoid diagnostic and procedural pitfalls.

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Introduction

Left persistent superior vena cava (LPSVC) is an uncommon incidental finding in 0.3% of general population. Amongst subjects with congenital heart disease, it is relatively more common and is found in 4.3% patients. Similarly, left persistent inferior vena cava (LPIVC) has been documented in 0.2-0.5% of the general population [1]. LPIVC running posterior to aorta with azygous or hemiazygous continuation is extremely rare, with only few cases described. This is found in 0.3% of general population and 0.6-2% in subjects with congenital heart disease [2]. The normal development of caval system during embryogenesis involves several anastomoses. Some vessels may fail to develop or regress during this complex process, and lead to numerous caval anomalies, including LPSVC and LPIVC [2].

We report a case of a LPIVC (transposed) communicating with the hemiazygous vein which was draining into the dilated coronary sinus *via* persistent LPSVC.

Case Report

A 26-year-old man was referred to our center for echocardiography in view of occasional palpitations and atypical chest pain. General physical examination and cardiovascular examination were normal. An electrocardiogram showed sinus tachycardia with right bundle branch block. A chest X-ray was done which showed mild cardiomegaly with prominent bronchovascular markings. Echocardiography demonstrated mild dilation of left and right atrium (RA), with mild tricuspid regurgitation. A pulse-wave Doppler of the tricuspid regurgitation jet showed peak pulmonary peak systolic pressure of 50 mmHg. A dilated coronary sinus was seen behind the left atrium (Figure 1, Video 1). Additionally, hepatic veins were separately draining into the RA. An atrial septal defect could not be excluded thus patient was taken for cardiac catheterization.

After advancing the catheter from right femoral vein, we found that the catheter tracked posteriorly and to the left of abdominal aorta. Following this, contrast injection was done from the right femoral vein to delineate the correct anatomy. Interestingly we found that, instead of continuing as azygous vein, inferior vena cava passed posteriorly and to the left of abdominal aorta, and was not draining into the RA (LPIVC). This LPIVC continued as hemiazygous vein, leading to LPSVC which was seen draining into the dilated coronary sinus (Figure 2, Video 2). With another contrast injection from right cephalic vein, a normal caliber of right superior vena cava (SVC) was seen draining directly into the RA, with absent azygous vein and no evidence of

communication with the LPSVC (Figure 3, Video 3). The right heart catheterization was completed, and atrial septal defect was ruled out. We also performed a coronary angiography which did not show any evidence of luminal narrowing. The patient was discharged in good condition with a low dose beta blocker (bisoprolol 1.25 mg) to control the heart rate.

Discussion

Due to numerous transformations during the development of superior and inferior vena cava, variations may be found in their adult forms. The development of SVC occurs from two symmetric

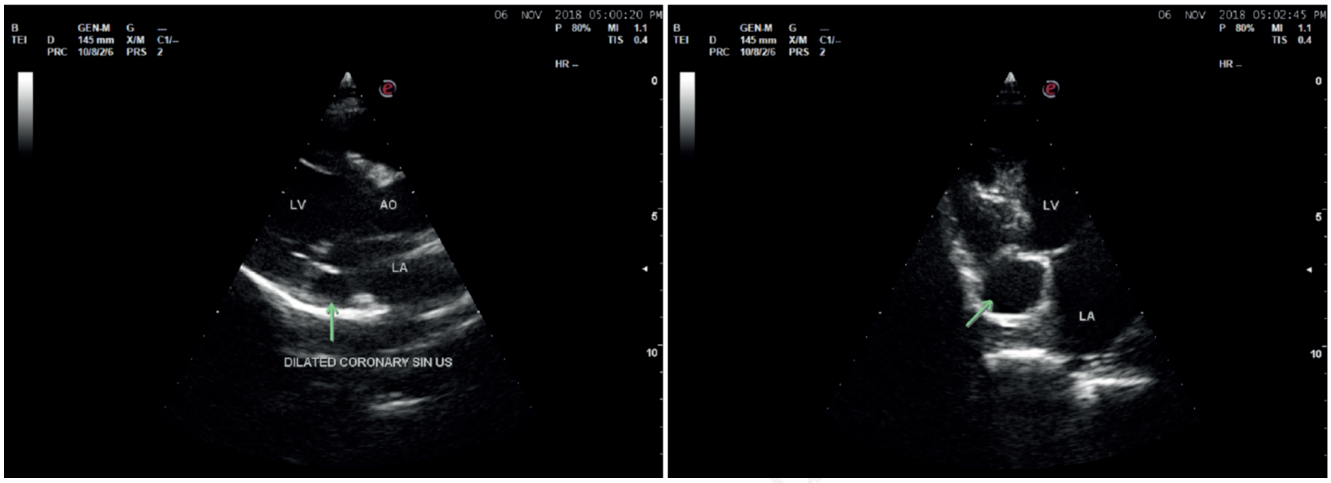

Figure 1, Video 1. Echocardiogram showing dilated coronary sinus (green arrow) in parasternal long axis view (left) and modified three chamber view (right). LA, left atrium; LV, left ventricle; AO, aorta.

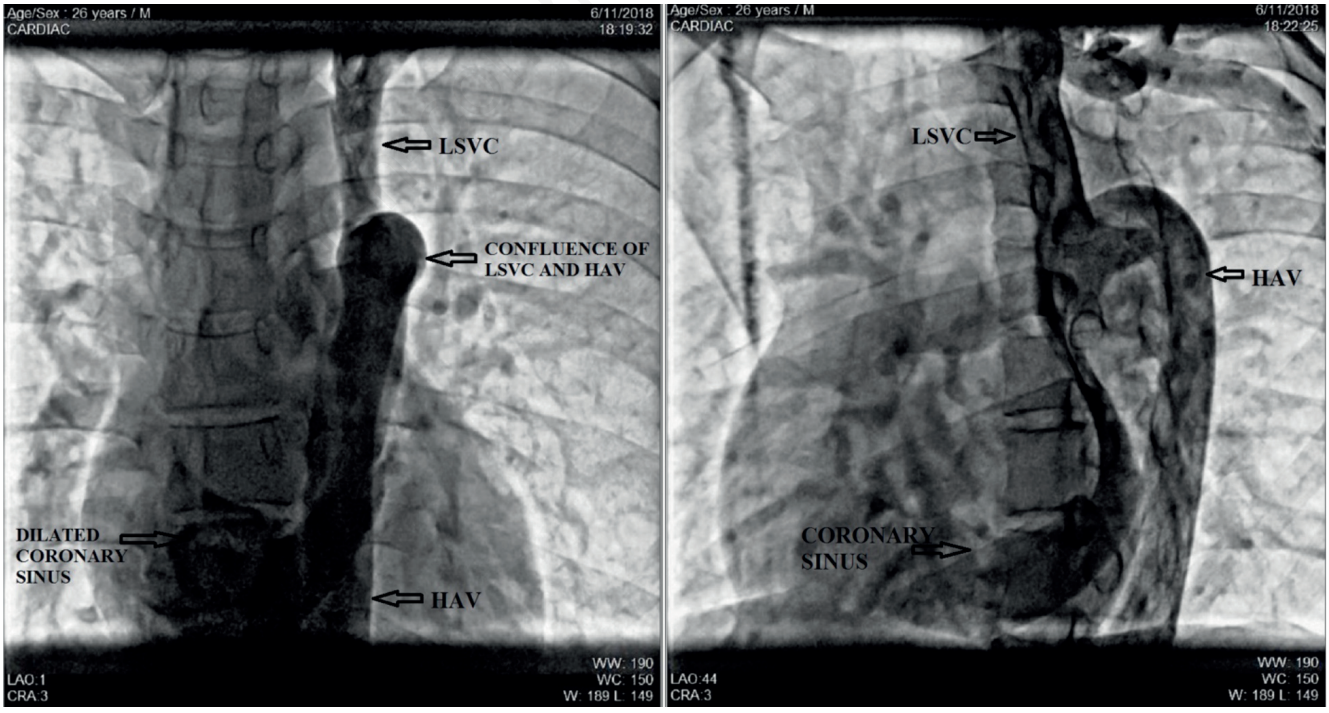

Figure 2, Video 2. Antero-posterior (left) and left anterior oblique (right) fluoroscopic projection demonstrating left persistent inferior vena cava (LPIVC) continuing as hemiazygous vein (HAV), leading to left persistent superior vena cava (LSVC), draining into dilated coronary sinus.

right and left anterior cardinal veins, which drain into a common cardinal vein and finally to the embryological heart. These two anterior cardinal veins are connected by an oblique vein anastomosis. Due to right-to-left shunting of blood in the embryological life, the left-sided common cardinal vein regresses and becomes the ligament of Marshall, the oblique vein anastomosis becomes the left brachiocephalic (innominate) vein, and the right anterior and right common cardinal veins form the right SVC [3]. In our case, the failure of regression on left common cardinal vein resulted in LPSVC.

The LPIVC is most frequently formed due to the persistence of left supracardinal vein, which lies posterior to the embryological aorta. The left postcardinal or subcardinal veins may also lead to LPIVC, but these are situated anterior to the aorta [4]. In our case, the LPIVC was most likely due to persistence of left supracardinal vein, since it was posterior to the aorta in its abdominal course.

LPIVC may drain into RA through one of the following pathways [2]:

a. It crosses over to the right side passing anterior to the aorta after receiving the left renal vein, and eventually drains into RA.
b. It continues as azygous vein, which drains into right SVC, and finally into RA.
c. It continues as hemiazygous vein which may finally drain to RA *via* azygous vein → right SVC ending in RA, accessory hemiazygous vein → superior intercostal vein → azygous vein → right SVC ending in RA or *via* LPSVC → coronary sinus ending in RA.

In our case the least common course of LPIVC (i.e., posterior to aorta), and least common drainage pathway (via hemiazygous vein → LPSVC → coronary sinus) was observed.

The association of venous anomalies has been linked to heterotaxy syndromes in some cases. Heterotaxy is the nomenclature used to define discordant position of midline structures like thoracoabdominal organs, and vessels in cases of complex congenital heart disease. This term has however been also applied to isolated anomalies like LPSVC and interrupted aortic arch. Such anomalies have been shown to have coexisting splenic anomalies. The association of LPIVC and heterotaxy syndromes has not been described [5].

There are only few similar cases reported in medical literature. Ojha et al reported an isolated left inferior vena cava with hemiazygos continuation in a 1-month-old child being worked up for congenital heart disease. He had situs ambiguous with left isomerism, a midline liver and right polysplenia [6]. Kabakus *et al*. reported a left sided IVC with hemiazygous continuation to left SVC and absent right SVC, in a 30-year-old female [7]. Kim *et al*. have also reported a similar case of hemiazygous continuation of LPIVC draining into RA *via* LPSVC [8]. Lastly, another similar case report of interrupted IVC with hemiazygous continuation with a LPSVC in association with left single coronary artery has been described [9].

LPIVC is asymptomatic in most cases, and is discovered incidentally on routine imaging, right heart catheterization or surgery. It is important to recognize this normal anatomical variant, and should be differentiated on imaging from lymphadenopathy, abdominal masses and dilated gonadal veins. Surgical procedures like Glenn Shunt, Mustard's repair etc. need to be modified accordingly to accommodate for LPIVC and dilated coronary sinus. Additionally, LPIVC and LPSVC may be accidentally injured during operative procedures and may result in serious hemorrhage. In these cases of LPIVC, the insertion of caval filters poses a challenge, due to reduced diameter of the transverse IVC. The placement of temporary and permanent pacemakers may be difficult in patients with LPSVC or LPIVC. Hence, it is recommended that such percutaneous interventions should always be done under fluoroscopic guidance. These venous anomalies may lead to inadequate venous drainage from lower limbs, thus increasing the risk of venous stasis and deep venous thrombosis [10-12].

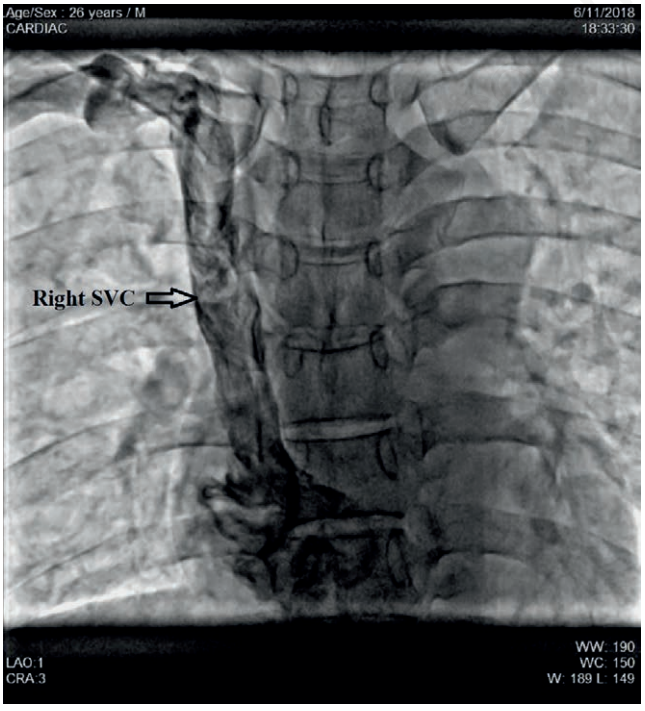

Figure 3, Video 3. Antero-posterior fluoroscopic projection demonstrating right superior vena cava (SVC) draining directly into right atrium.

Conclusions

The congenital anomalies of caval system, as described in our case, are not uncommon as they have no cardinal symptoms and usually do not require any specific treatment. However, clinicians need to be mindful of such anomalies with an adequate index of suspicion to prevent undesirable complications.

References

1. Dahnert W. In: 2nd ed, Radiology review manual, Baltimore: Williams and Wilkins; 1996, p. 433.
2. Bass JE, Redwine MD , Kramer LA, et al . Spectrum of congenital anomalies of the inferior vena cava: cross-sectional imaging findings. Radiographics 2000;20:639-52.
3. Sonavane S, Milner D, Singh S, et al. Comprehensive imaging review of the superior vena cava. Radiographics 2015;35:1873-92.
4. Knudtzon J, Svane S. Left sided inferior vena cava. Acta Chir Scand 1986;152:547–9.

5. Alharthi M, Mookadam F, Collins J, et al. Images in cardiovascular medicine. Extracardiac venous heterotaxy syndrome: complete noninvasive diagnosis by multimodality imaging. Circulation 2008;117:e498-503.
6. Ojha V, Pandey NN, Jagia P. Hemiazygos continuation of isolated left-sided inferior vena cava into persistent left superior vena cava: rare association of left isomerism. BMJ Case Rep 2019;12:e230350.
7. Kabakus I, Kocher M, Agha A, Burt JR. Left-sided inferior vena cava with hemiazygos continuation to left superior vena cava. Cureus 2019;11:e6503.
8. Kim HJ, Ahn IO, Park ED. Hemiazygos continuation of a left inferior vena cava draining into the right atrium via persistent left superior vena cava: demonstration by helical computed tomography. Cardiovasc Intervent Radiol 1995;18:65-7.
9. Kim YJ, Kwon HS, Ahn SE et al. Interrupted inferior vena cava with hemiazygos continuation in an adult with a persistent left superior vena cava and left single coronary artery: A case report. J Korean Soc Radiol 2016;74:394-8.
10. Freedom RM. Angiography of congenital heart disease, London: Collier Macmillan; 1984, pp. 46-61.
11. Kraimps JL, Dib H, Raynier P, et al. Left-sided inferior vena cava and thrombosis: case report. Eur J Surg 1993;159:441-3.
12. Brener BJ, Darling RC, Frederic PL, et al. Major venous anomalies complicating abdominal aortic surgery. Arch Surg 1974;108:159-65.