Left Hepatic Vein and Persistent Left Superior Vena Cava Drainage into the Coronary Sinus with Subaortic Valve Stenosis

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ABSTRACT

Persistent left superior vena cava (PLSVC) and left hepatic vein drainage into the coronary sinus is a very rare combination of thoracic venous anomalies. A literature search revealed only two previously reported cases. PLSVC is associated with congenital heart disease and impulse conduction abnormalities. Subaortic valve stenosis is among these abnormalities. We present an extremely rare case of a 19-year-old woman who presented with PLSVC and left hepatic vein drainage into the coronary sinus associated with subaortic valve stenosis. The vascular anomalies were noted on a computed tomography angiogram of the chest performed for a history of shortness of breath. The patient had been previously diagnosed with and underwent treatment for subaortic valve stenosis at ages 2 and 8. PLSVC increases the incidence of congenital heart disease.

KEYWORDS: Persistent left superior vena cava, left hepatic vein, subaortic valve stenosis, enlarged coronary sinus, left ventricular outflow tract obstruction

A persistent left superior vena cava (PLSVC) is a vascular anomaly rarely seen in the normal population. It is reported to be present in between 0.3 and 0.5% of the population. It is seen more commonly in patients with congenital heart disease and is reported to be present in around 3 to 5% of these cases.¹ A left hepatic vein draining into the coronary sinus is an even rarer vascular anomaly. This combination of anomalies is extremely rare and has been previously described only twice.^{2,3} PLSVC can impair the normal development of the left ventricle (LV). Subaortic valve stenosis is associated with PLSVC, which can cause both inflow and outflow tract obstructions in the LV.⁴ We present a case involving PLSVC and a left hepatic vein draining into the coronary sinus with secondary subaortic valve stenosis.

CASE REPORT

A 19-year-old woman presented to the emergency department with a chief complaint of shortness of breath. She had a medical history that included subaortic valve stenosis for which she underwent surgical repair at ages 2 and 8. The patient underwent computed tomography angiography (CTA) of the chest. The study revealed an isolated persistent left superior vena cava (SVC) (Figs. 1A, B, and 2A) and an anomalous left hepatic vein both of which drained through an enlarged coronary sinus (Figs. 1B, C, and 2B). Contrast was seen refluxing inferiorly through the left hepatic vein and into its contributing branches in the left lobe of the liver (Figs. 1D and 2A, B). The right lobe of the liver as well as the right and middle hepatic veins contained minimal contrast (Fig. 1D). In this case the contrast was seen

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Figure 1 (A) Contrast is seen in the persistent left superior vena cava (PLSVC) (white arrow). Note the complete absence of the right superior vena cava (SVC). (B) Thick slab maximum intensity projection CT image shows contrast in the PLSVC (white arrow), left hepatic vein (broken black arrow), and right atrium (solid black arrow.) The contrast also refluxes into the contributing branches of the left hepatic vein in the left lobe of the liver. The inferior vena cava is seen separate from the left hepatic vein. Note the complete absence of the right SVC. (C) Contrast is seen within the enlarged coronary sinus (black arrow) draining into the right atrium. Note that there is no communication between the coronary sinus and the left heart. The enlarged coronary sinus runs just inferior to the region of the mitral valve. This is the basis for the proposed limitation of atrioventricular flow in the left heart during development leading to congenital heart defects. (D) Contrast is seen refluxing through the left hepatic vein (broken black arrow) and into its contributing branches in the left lobe of the liver. Note the difference in contrast phase between the right and left lobes of the liver.

flowing through the coronary sinus and into the right atrium (Figs. 1B, C, and 2B). There was no evidence of connection of the coronary sinus with the left atrium.

DISCUSSION

In the majority of patients with PLSVC and left hepatic vein drainage into the coronary sinus, both a right and left SVC are present. Very rarely there is an isolated PLSVC with complete absence of the right SVC. In these patients, the PLSVC drains blood from the head and upper extremities bilaterally. The blood returns in most cases to the right atrium through an enlarged coronary sinus.⁵ These patients show no evidence of hemodynamic compromise and in the absence of con-

genital heart disease will go undetected. In 8% of patients, the PLSVC will drain into the left atrium creating a left to right shunt.³ These patients may show signs of hemodynamic compromise.

PLSVC is associated with congenital heart defects. These defects include atrial septal defects, ventricular septal defects, pulmonary stenosis/atresia, D-transportation of the great vessels, anomalous pulmonary drainage, mitral atresia, tetralogy of Fallot, coarctation of the aorta, absent right SVC, situs inversus, coronary sinus atresia/enlargement, subaortic stenosis, and obstruction of the left atrioventricular flow.^{3,4,6,7} Cardiac arrhythmias can occur due to persistence of left-sided fetal pacemaker tissue causing ectopic pacemaker activity.³ There can also be stretching of the

Figure 2 (A) Sagittal three-dimensional reconstructed image demonstrating the left SVC (white arrow) and left hepatic vein (broken black arrow) communicating with the coronary sinus (black arrow). (B) Thick slab sagittal maximum intensity projection image demonstrates contrast reflux into the left hepatic vein. The left hepatic vein communication with the coronary sinus is demonstrated on this image (black arrow).

atrioventricular node and bundle of His. The latter of these types of cardiac arrhythmias and the left atrioventricular flow obstruction is likely secondary to coronary sinus enlargement.⁷ A left hepatic vein draining into the coronary sinus may contribute to its further enlargement.

The embryology of the left hepatic vein anomaly may stem from persistence of the left vitelline connection with the left horn of the sinus venosus.^{2,3,8} The left horn of the sinus venosus eventually becomes the coronary sinus. The embryology of the PLSVC stems from the persistence of the left anterior cardinal vein. During normal development, the anterior cardinal vein joins the posterior cardinal vein. The left anterior cardinal vein drains into the left horn of the sinus venosus. Normally, flow is shunted through the right side and the left side resolves.^{5,7} In this patient, flow continued through the left side and the right side completely resolved. The sum results of these anomalies in this patient were an isolated PLSVC and anomalous left hepatic vein draining into an enlarged coronary sinus.

These vascular anomalies were seen in conjunction with secondary subaortic valve stenosis in this patient. The enlarged coronary sinus may cause flow

limitations in the developing LV. Flow within the developing left ventricular cavity helps direct the development of the LV. Impeded flow in the developing LV is responsible for LV and aortic hypoplasia and obstructive lesions in the LV, including secondary subaortic valve stenosis.⁴ Agnoletti et al as well as Macedo et al suggest that the flow limitation is due to compression of atrioventricular flow in the left heart due to the enlarged coronary sinus.^{6,9}

Imaging was essential in the diagnosis of this patient. PLSVC can be definitively diagnosed through echocardiography, conventional angiography, CTA, and magnetic resonance angiography (MRA). Conventional angiography used to be considered the gold standard for diagnosis of PLSVC. This procedure requires prolonged sedation and is invasive. Overlap of the great vessels in the thorax can cause some difficulty in identifying the individual structures.¹ Echocardiography can be employed easily. The use of air bubble contrast can help identify the 8% of patients who have a left to right shunt through an unroofed left coronary sinus. The drawbacks to echocardiography include a small visual window, operator dependency, and limited evaluation of extra-cardiac structures.¹

CTA and MRA are both noninvasive techniques that show the full extent of the vascular anomaly. These techniques depend on the ability of the patient to cooperate during the examination. These two modalities along with conventional angiography usually require the use of intravascular contrast agents that are sometimes contraindicated. There are some MRA techniques that do not require contrast but often yield lower quality images. MRA takes longer than CTA, and therefore requires longer sedation in very ill or uncooperative patients. MRA is also far more expensive than CTA. MRA, however, does not expose the patient to ionizing radiation. CTA and MRA images have become equal to, if not superior to those obtained through conventional angiography. In addition, they provide anatomic information that includes other major thoracic structures. In cases of vascular anomalies this information is important for planning surgical interventions.

CONCLUSION

In unique cases like the one discussed above, it is important to understand how to best utilize diagnostic imaging modalities to obtain the desired information. Which test is the best depends on the individual patient variables. These include the patient's age, allergies, ability to cooperate, and overall clinical condition. CTA with multiplanar reconstructions has replaced conventional angiography as the "gold standard" in diagnostic imaging and will provide the quickest and most complete evaluation of the great vessels unless contraindications are present.

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