A 52-year-old woman with a history of systemic lupus erythematosus presented to our institution with nonhealing ulcers on her left lower extremity and toes. She had no cardiac symptoms, with a normal ECG and chest radiograph (Figures 1 and 2). As part of her evaluation she underwent a transesophageal echocardiogram to look for endocarditis, which demonstrated a right-to-left shunt with Valsalva that appeared to originate from the vicinity of the lateral aspect of the left atrium (Movie I in the online-only Data Supplement). There was also a vascular structure seen adjacent to the aorta that typically would represent a remnant of the cardinal vein, such as a persistent left superior vena cava. There were no findings of endocarditis.

A magnetic resonance angiogram was performed to further evaluate the origin of the right-to-left shunt and demonstrated a levoatriocardinal vein that drained the left upper lobe pulmonary veins. This levoatriocardinal vein connected the left innominate vein with the left atrium via the left superior pulmonary vein (Figure 3). The flow in the levoatriocardinal vein was demonstrated to be primarily directed cephalad, creating a left-to-right shunt, as would be expected. However, cine phase contrast imaging demonstrated a reversal of flow in the levoatriocardinal vein after Valsalva (Figure 4). This was the source of the right-to-left shunt detected on transesophageal echo. Transthoracic echo confirmed the bidirectional flow in the levoatriocardinal vein (Figures 5 and 6, Movies II and III in the online-only Data Supplement). There were no findings of volume overload in the right heart because the left pulmonary veins were still connected to the left atrium, with only partial drainage of these veins into the innominate vein.

Further clinical history revealed that the patient was having clusters of transient ischemic attacks, most recently after coughing spells during hospitalization for pneumonia while she had an intravenous line in her left arm. This raised the possibility of paradoxical embolism due to reversal of the shunt by severe coughing.

This anomaly was determined to be treated best using percutaneous embolization of the levoatriocardinal vein. The
planning angiogram nicely demonstrated the patient’s variant anatomy (Movie IV in the online-only Data Supplement). The levoatriocardinal vein was closed with a 12-mm Amplatzer vascular plug (AVP II) (Figure 7 and Movie V in the online-only Data Supplement). This successfully occluded the levoatriocardinal vein and eliminated the right-to-left and left-to-right shunts. Follow-up transthoracic echo demonstrated complete occlusion of the levoatriocardinal vein with no residual right-to-left shunt.

As described by McIntosh in 1926, the levoatriocardinal vein is an abnormal persistent connection between the pulmonary venous system and the cardinal (systemic) venous system through the splanchnic plexus of the embryonic foregut. It is most commonly associated with left obstructive lesions such as mitral atresia, hypoplastic left heart, and cor triatriatum. Approximately 50 cases have been reported in the literature, only 3 of which had otherwise normal cardiac anatomy.

Patients with levoatriocardinal veins and no other cardiovascular abnormality presented much later in life (aged 5–52 years) than those with left obstructive lesions (aged <4 months). Two of these patients, aged 15 and 24 years, were being evaluated for clinically suspected atrial septal defects. The third patient, aged 5 years, was being evaluated for an asymptomatic murmur. Our patient had no cardiac symptoms at the time of her transesophageal echocardiogram.

Levoatriocardinal vein is a rare anomaly that can be seen in patients with otherwise normal cardiac anatomy, although it is more common in patients with other left obstructive abnormalities. It should be considered in the differential diagnosis of abnormal vascular structures adjacent to the aorta along with a left superior vena cava and a vertical vein with partial anomalous pulmonary venous return.

Disclosures

None.

References

Figure 4. Magnetic resonance images from a cine phase contrast examination showing reversal of flow in the levoatriocardinal vein during Valsalva. Arrows indicate the levoatriocardinal vein. At rest the vein is black indicating flow toward the patient’s head. With valsalva the flow reverses and is displayed as white, which indicates flow direction toward the patient’s feet.

Figure 5. Color Doppler images of the levoatriocardinal vein flow at rest (top) and reversal of the levoatriocardinal vein flow during Valsalva (bottom).

Figure 6. Spectral Doppler demonstrates the changes in the levoatriocardinal vein flow at rest (left), with Valsalva (middle), and with release of Valsalva (right).
Figure 7. After deployment of the Amplatz vascular plug into the levoatriocardinal vein.