Levoatriocardinal vein in association with tetralogy of Fallot: a rare pulmonary to systemic connection

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DESCRIPTION

A 1-year-old-male child presented with cyanosis and failure to thrive. A transthoracic echocardiogram established the diagnosis of tetralogy of Fallot, and the patient was referred for a CT angiography (CTA) for further evaluation. CTA confirmed the diagnosis of tetralogy of Fallot. An anomalous venous channel (*) was seen connecting the left superior pulmonary vein (LSPV) and the left brachiocephalic vein (figure 1). The LSPV and the left inferior pulmonary vein were seen draining normally into the left atrium (LA). No left heart obstructive lesions were noted.

A levoatriocardinal vein (LACV) is a rare form of pulmonary to systemic connection. Embryologically, it represents a persistent communication between the left cardinal vein and the pulmonary vein (PV), in the presence of a normal communication between PVs and LA. Most frequently, it is observed in association with left-sided obstructive lesions such as hypoplastic left heart syndrome, mitral stenosis/atresia, cor triatriatum, aortic stenosis/atresia and aortic coarctation.¹ In such cases, it serves as an alternate drainage pathway for the pulmonary venous blood, thus decompressing the LA. The presenting symptoms usually depend on the associated congenital heart disease. An isolated

LACV may present with an asymptomatic murmur or be detected in a patient with a left-to-right shunt with a clinical suspicion of atrial septal defect. In the presence of a bidirectional shunt, the patients may present with symptoms due to paradoxical embolism and would require surgical/endovascular closure of the anomalous channel.

Very rarely, an LACV may be seen in the absence of left-sided obstructive lesions, such as in this case, where it is seen association with tetralogy of Fallot.

Learning points

- ▶ Levoatriocardinal vein (LACV) represents a persistent embryologic communication between the left cardinal vein and the pulmonary vein, in the presence of a normal communication between pulmonary veins and left atrium.
- ▶ It is frequently observed in association with left-sided obstructive lesions, such as hypoplastic left heart syndrome, mitral stenosis/atresia, cor triatriatum, aortic stenosis/atresia and aortic coarctation, serving as an alternate drainage pathway for the pulmonary venous blood and decompressing the left atrium.
- ▶ While an isolated LACV may present with an asymptomatic murmur, in the presence of a bidirectional shunt, the patients may present with symptoms due to paradoxical embolism.

Figure 1 Oblique coronal maximum intensity projection image (A) and volume-rendered image (B) depict the levoatriocardinal vein (*) connecting the left superior pulmonary vein (LSPV) and the left brachiocephalic vein (LBCV). The LSPV and the left inferior pulmonary vein (LIPV) are seen to drain normally into the left atrium (LA).

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