Levoatriocardinal vein with normal intracardiac anatomy and pulmonary venous return

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ABSTRACT

Levoatriocardinal vein (LACV) is characterized by an abnormal connection between pulmonary and systemic venous return. This extremely rare cardiac malformation is usually associated with left-sided obstructive lesions including mitral atresia, hypoplastic left-heart syndrome, and abnormal pulmonary venous connection. Patients may have low systemic cardiac output and pulmonary venous obstruction symptoms. In this manuscript, we report a case with LACV and normal pulmonary venous return with absence of any intracardiac pathology. LACV was demonstrated with echocardiography, angiography, and computed tomography. Surgical correction was made successfully.

Keywords: Abnormal pulmonary venous connection, cardiac magnetic resonance, levoatriocardinal vein

INTRODUCTION

Levoatriocardinal vein (LACV) is a rare cardiac malformation that represents a connection between pulmonary venous and cardinal systems. Embryologically it is a result of abnormal persistency of splanic plexus, which is a connection between pulmonary venous plexus and cardinal system. It was first described by McIntosh in 1926[1] and encountered mostly every time in patients with left-sided obstructive lesions such as cor triatriatum, mitral stenosis or atresia, hypoplastic left-heart syndrome, aortic atresia, and coarctation.[2]

Clinically, the symptoms of pulmonary venous obstruction and low cardiac output[2] could be diagnosed within early infants. In this report, we describe a patient with late clinical presentation of LACV without pulmonary venous obstruction and additional cardiac malformation.

CASE REPORT

A 24-year-old patient suffered from fatigue and palpitations during his military service. He was diagnosed with atrial septal defect and referred to our center for correction.

In his physical exam, 2/6 heart murmur and fixed splitting of second heart sound was heard at the upper-left sternal border. Telecardiography revealed cardiomegaly and ECG was unremarkable except right-bundle branch block.

Echocardiographic examination showed dilation of the right atrium, right ventricle, and main pulmonary artery. Apical four chambers and high parasternal views demonstrated an anomalous vein with an ascending course that was initially interpreted as an abnormal drainage of the left-sided pulmonary veins. Drainage of the right pulmonary veins to left atrium was normal and interatrial septum was intact. He was diagnosed to have an abnormal partial pulmonary venous connection with these features.

In catheterization, all pulmonary veins were draining to the left-atrium. LACV was taking off from the left atrium, and was draining to the right-sided superior vena cava via the left brachiocephalic vein [Figure 1]. Qp/Qs was 2.8/1. Pulmonary artery pressure was normal and no gradient was detected between pulmonary veins and the left atrium. Angiograms showed no left sided obstruction at any level. Magnetic resonance imaging showed a distinct LACV taking off from the left atrium and coursing into the right atrium. MRI also demonstrated normal return of pulmonary veins to the left atrium [Figure 2a and b].

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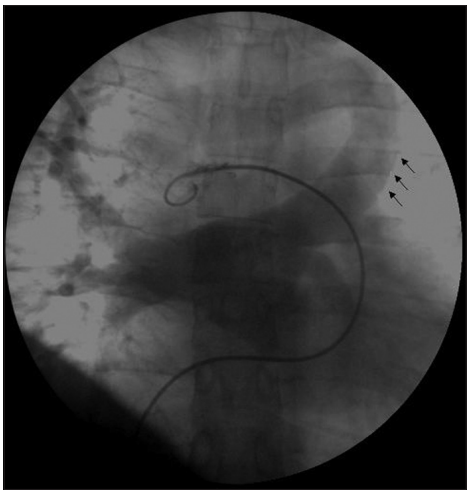

Figure 1: Pulmonary artery injection shows an abnormal drainage of left atrium by LACV in late venous return phase (Arrow: LACV)

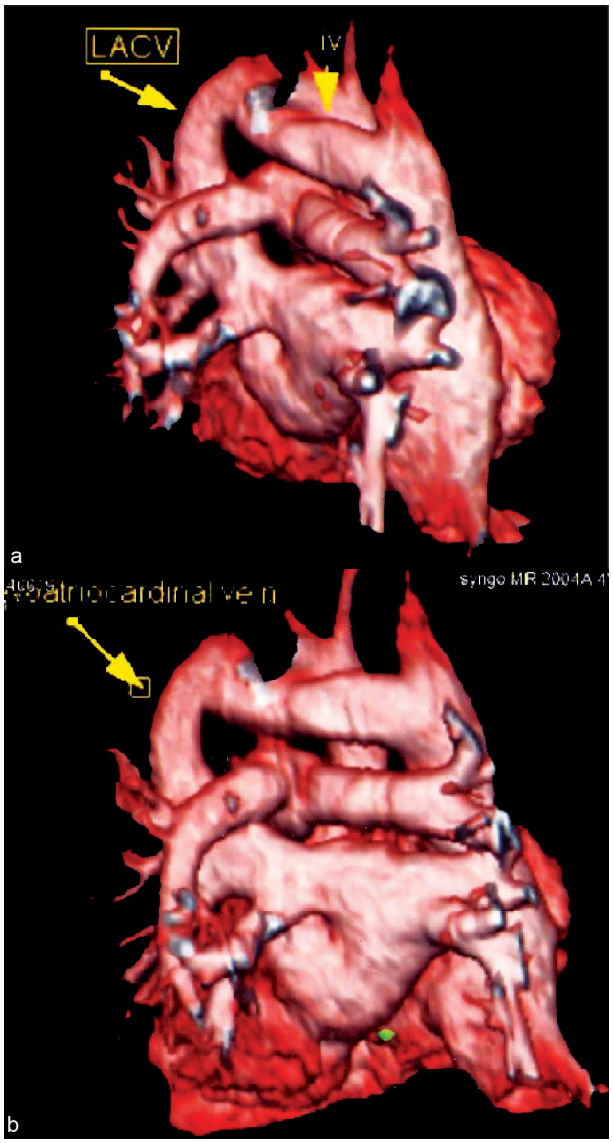

Figure 2a-b: Cardiac magnetic resonance imaging shows normal return of pulmonary venous system and connection between LACV and left atrium in posterior views

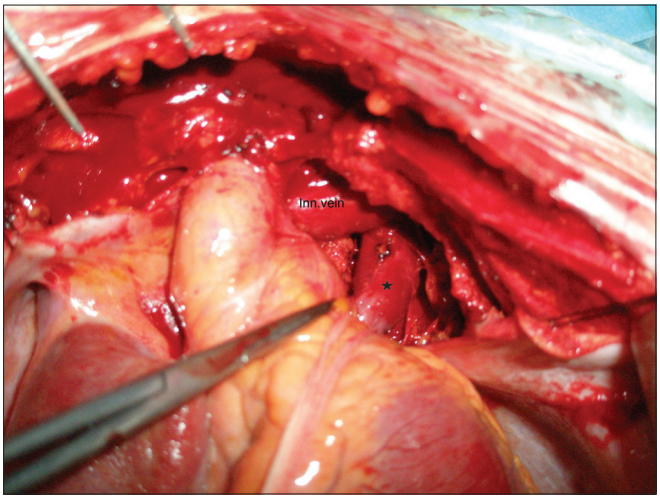

Figure 3: Connection between LACV and left innominate vein was observed at median sternotomy

Surgery was performed by a median sternotomy approach. LACV was surgically ligated [Figure 3]. The patient was discharged at the sixth postoperative day without any complications.

DISCUSSION

LACV is an extremely rare pathology that was reported only as case reports.[1-4] It is a type of interatrial communication that connects the pulmonary venous system directly to any part of the cardinal system such as brachiocephalic vein, superior vena cava, or another systemic vein. In most of cases, left-sided obstructive lesions were observed and LACV was the only route of communication between two atrias. In these cases, the only significant way to "unload" the left side of the heart was by LACV. It was thought that high left atrial pressure caused the patency of splanic plexus that is a natural connection between cardinal system and pulmonary veins in early intrauterine life.[2]

According to this theory, most of the cases with LACV have left-sided obstructive lesions.[5] Only one case has been reported, concerning LACV without any intracardiac pathology and normal pulmonary venous connection.[6] In practice this case is the second one concerning LACV without additional cardiac abnormality.

The levoatriocardinal vein should also be considered an entity distinct from a left superior vena cava. The latter forms from the left precardinal vein system. Some of the previously reported patients had both a left superior vena cava and a levoatriocardinal vein that drained to it. Two entities could be differentiated with showing the blood flow direction by echocardiography or other imaging techniques. Flow direction is from left atrium to innominate vein in LACV, whereas the left superior vena cava usually drains to the coronary sinus.[3]

Typical presentation can be found at the beginning of

a person's life and is characterized by the symptoms of low cardiac output. Our case was with no symptoms shown until adulthood. Then, the patient suffered from fatigue and exertional dyspnea, which were related to chronic right heart volume overload mimicking atrial septal defect. Diagnosis of LACV can be made easily with echocardiography especially with existence of left-sided obstructive lesions. The LACV is seen on parasternal and suprasternal views as an abnormal ascending vertical venous image. Contrast injection into the left atrium or LACV during diagnostic cardiac catheterization is a reliable method for diagnosis. Magnetic resonance imaging and computerized tomography are other diagnostic methods to reveal anatomical details of pathology.

Although transcatheter occlusion of LACV by some devices is possible without surgery, surgical ligation of LACV was made easily without cardiopulmonary bypass.

In conclusion, LACV is an extremely rare cardiac pathology existing almost always with left-sided obstructive lesions. The diagnosis of LACV may be missed, when no intracardiac abnormality exists. Therefore, we recommend a detailed examination for the patients with right-heart volume overload, in the case of the absence of an atrial septal defect and an abnormal pulmonary venous connection, even when no left-sided obstructive lesion is present.

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