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Congenital left ventricular aneurysm: a case report

Doğuştan sol ventriküler anevrizma: Olgu sunumu

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Congenital left ventricular aneurysm is a rare anomalous of the heart. Association with other congenital heart defects is common. Imaging modalities have an important role in the diagnosis of the disease, due to its asymptomatic nature. In this article, we report a one-month-old girl with congenital left ventricular aneurysm, a rare condition existing together with congenital heart disease, which was diagnosed by two-dimensional echocardiography and angiography.

Key words: Aneurysm; angiography; congenital; echocardiography; left ventricle.

Doğuştan sol ventriküler anevrizma, nadir görülen bir kalp anomalisidir. Diğer doğuştan kalp defektleri ile birlikteliği sıktır. Asemptomatik olması nedeni ile hastalığın tanısında görüntüleme yöntemleri önemli yer tutmaktadır. Bu yazıda, nadir görülen ve doğuştan kalp hastalığı ile birlikte olan, iki boyutlu ekokardiyografi ve anjiyografi ile tanısı konulan, doğuştan sol ventriküler anevrizması olan bir aylık kız olgu sunuldu.

Anahtar sözcükler: Anevrizma; anjiyografi; doğuştan; ekokardiyografi; sol ventrikül.

Congenital left ventricular aneurysms (LVAs) are a rare anomaly of the heart usually seen in conjunction with other congenital heart diseases. The terms "congenital aneurysm" and "congenital diverticulum" of the heart have been used interchangeably, but some authors define them differently and make a distinction. Patients with congenital LVAs are usually asymptomatic, and their differential diagnosis includes the left ventricular (LV) diverticulum. The former has a large LV neck that connects to a fibrous wall along with a paradoxical motion. In contrast, most cases involving the latter have a narrow neck with a muscular wall and a synchronous contraction with the LV.[1,2] This makes it difficult to clinically diagnose the disease. Left ventricular aneurysms are most often detected incidentally during imaging procedures performed for other indications. Although congenital LVAs are usually asymptomatic, on rare occasions, they can cause arrhythmia, chest pain, cardiac rupture, and sudden death.[2] The etiology of congenital LVAs is not totally clear, but they are thought to result from either congenital epicardial cysts or embryological defects of the LV muscle. In addition, in adults, they are associated with past myocardial infarction (MI).[3] Furthermore, aneurysms may also occur due to myocarditis, dilated cardiomyopathy, and chest trauma. Two-dimensional (2D) or three-dimensional (3D) echocardiography (ECHO), angiography, and magnetic resonance imaging (MRI) can be used to diagnose aneurysms. Herein, we present a case with a congenital LVA that was brought to our attention on 2D ECHO and definitively diagnosed via angiography along with the results of other imaging methods.

CASE REPORT

A one-month-old girl baby was referred to our clinic because she was suspected of having congenital heart disease due to a cardiac murmur. A physical examination revealed no abnormal pathological findings, except for a 3/6 systolic ejection murmur at the pulmonary focus. Cardiomegaly was detected on the chest X-ray. A classic, apical four-chamber image on 2D ECHO demonstrated focal hypertrophy of the interventricular septum (IVS) and showed

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a suspicious defect in the IVS region close to the atrioventricular valves (Figure 1). A color Doppler examination revealed no turbulence flow, which normally accompanies a ventricular septal defect. Based on the ECHO performed at modified angles, we believed that the defect might be the mouth of the aneurysm which had been hidden and concealed by the focal hypertrophy of the IVS. Biventricular hypertrophy, more prominent in the right ventricle, was also observed on the 2D ECHO. Additionally, the hypertrophy had caused a slight obstruction with a 23 mmHg gradient in the path of the LV outlet, and another obstruction had caused a 60 mmHg gradient in the supravalvular region in the path of the right ventricular (RV) outlet. However, the systolic function of the LV was normal. In addition, a 6 mm secundum-type defect was detected in the interatrial septum. Contrast enhanced angiography (Figures 2) of the LV demonstrated a 32x12 mm aneurysmatic configuration arising from the IVS and extending to the left side of the RV and posterior aspect of the sternum. We also observed that the hypertrophy of the IVS had caused a filling defect and a 23 mmHg gradient in the subaortic region. In addition, the aortic valve was bicuspid, and the right coronary artery (RCA) was dominant. After consulting with the cardiology and surgical departments, it was decided that intervention was necessary, and the patient was referred to a surgical center.

DISCUSSION

Congenital LVAs or diverticulae are rare. In a study by Skapinger,[4] nonselective catheterization of an adult population revealed an aneurysm prevalence of 0.26% while an autopsy series revealed a prevalence of 0.40%. It may be difficult to diagnose congenital LVAs since they are asymptomatic, but treatment via anti-arrhythmic medical therapy, surgical resection, or radiofrequency ablation should begin as soon as possible after diagnosis because of the risk for arrhythmia, ventricular fibrillation, and sudden death. An implantable cardioverter-defibrillator can also be inserted in suitable cases when needed. Spontaneous rupture of the LVA may cause sudden death, with the risk being higher for aneurysms that are rich in fibrous tissue rather than muscular tissue. However, since the risk for thromboembolus is high for giant aneurysms that are rich in muscular tissue, anti-aggregant and/or anticoagulant therapy should be initiated when there is no surgical intervention.[5] In our case, electrocardiography demonstrated no arrhythmia, and there was no sign of thromboembolus. Furthermore, no symptoms associated with congenital LVA were present, and no pathological abnormalities were found other than the systolic ejection murmur related to the ventricular hypertrophy.

Although congenital LVAs are usually seen in the apex and free wall of the LV,[6] they may also occur on the septal side,[7] as in our case. Imaging methods gain importance in the diagnosis of aneurysms. In addition to ECHO and angiography, cardiac tomography and MRIs can be used. However, ECHO is often preferred because it is noninvasive.[4,8]

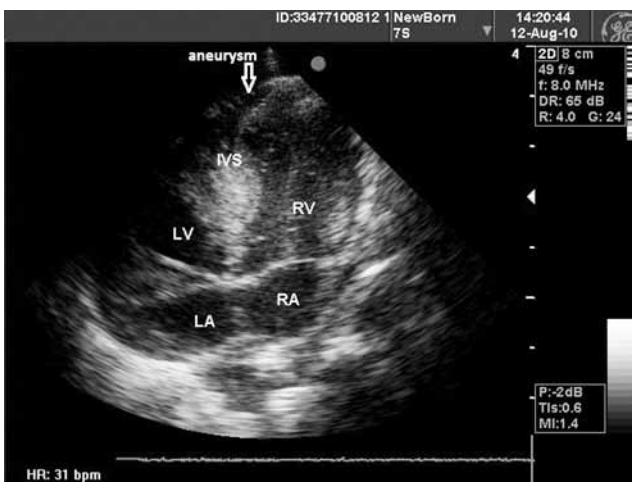

Figure 1. Apical four-chamber echocardiography image of left ventricular aneurysm. IVS: Interventricular septum; LV: Left ventricular; RV: Right ventricular; LA: Left atrium; RA: Right atrium.

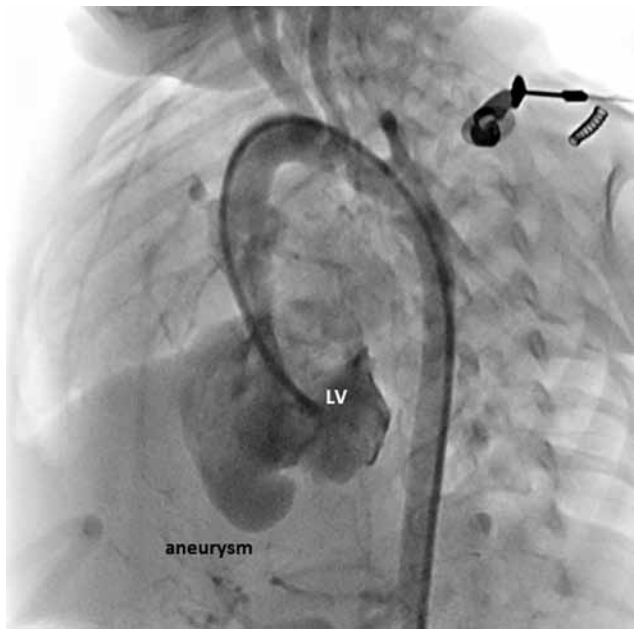

Figure 2. Angio image of left ventricular aneurysm. LV: Left ventricular.

In this case, ECHO showed the mouth and a part of the aneurysm. Subsequent angiography was then performed which demonstrated that the aneurysm was larger than we had assumed and revealed its localization, size, and area around the congenital LVA more clearly. If we had not examined the ECHO results so carefully, we might have suggested further clinical monitoring of the patient and attributed the existing murmur to pulmonary or aortic obstruction. Thus, the patient might have died before the surgical intervention.

In conclusion, we believe that aneurysms can be detected on ECHO when it is performed precisely and carefully. In addition, selective LV angiography can be used in some cases to clearly demonstrate the outlet, size, and location of the aneurysm without the need for cardiac tomography or an MRI.

Declaration of conflicting interests

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