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Case Report / Olgu Sunumu

A giant aneurysm of the left atrial appendage in childhood

Çocukluk çağında dev sol atriyal apendaj anevrizması

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Congenital aneurysm of the left atrial appendage is a very rare cardiac anomaly which can be accompanied with supraventricular rhythm disorders and life-threatening systemic thromboembolism. In this article, we present a six-year-old girl with a bulky mass of soft tissue density on the cardiac wall in chest X-ray which was adjacent to the left atrial appendage in two-dimensional transthoracic echocardiography. The patient was diagnosed with the left atrial appendage aneurysm. Surgery was performed to eliminate potential complications. The patient was uneventful in the postoperative follow-up.

Key words: Aneurysm; childhood; left atrial appendage; surgery.

Doğuştan sol atriyal apendaj anevrizması oldukça nadir, supraventriküler ritim bozuklukları ve yaşamı tehdit eden sistemik tromboembolinin eşlik edebildiği bir kalp anomalisidir. Bu yazıda, göğüs grafisinde kalp duvarında yumuşak doku yoğunluğu ve iki boyutlu transtorasik ekokardiyografide bu yoğunluğun sol atriyal apendaja komşu olduğu izlenen altı yaşında bir kız olgu sunuldu. Hastaya sol atriyal apendaj anevrizması tanısı kondu. Muhtemel komplikasyonları ekarte etmek için cerrahi yapıldı. Hasta ameliyat sonrası takiplerde sorunsuz idi.

Anahtar sözcükler: Anevrizma; çocukluk çağı; sol atriyal apendaj; cerrahi.

An isolated congenital intrapericardial aneurysmal dilatation of the left atrial appendage (LAA) is a very rare condition that is generally diagnosed in older patients and is usually accompanied by supraventricular rhythm disorders and life-threatening systemic thromboembolism.[1] Noninvasive imaging modalities, such as transthoracic and transesophageal echocardiography, computed tomography (CT), and magnetic resonance imaging (MRI), are reliable tools for diagnosing this condition,[2] but surgery is recommended after confirming the diagnosis in order to eliminate potential complications.

CASE REPORT

A six-year-old girl was referred to our hospital after being diagnosed with cardiomegaly. The child's development and vital signs were normal. However, a neurological examination revealed 3/5 motor deficit in the left upper and lower extremities because of an acute cerebrovascular event two years earlier. It was not clear why the patient had not been evaluated for that event, but she underwent her first cardiac evaluation at our facility. We found a large opacity at the left perihilar region on chest X-ray, and an initial transthoracic echocardiographic (TTE) evaluation revealed a preliminary diagnosis of a pericardial cyst. After detailed TTE and transesophageal echocardiography (TEE), an aneurysmal cavity measuring 74x50 mm was detected that was associated with the left atrium through a 16 mm orifice (Figure 1). In addition, color Doppler ultrasonography demonstrated that this chamber was also associated with the left atrium. Furthermore, no intracavitary mass suggestive of a thrombus or tumor was present within this aneurysmatic chamber, and there was no cardiac pathology associated with the cavity.

Cardiac MRI confirmed the diagnosis of an LAA aneurysm which needed to be surgically resected. Following a median sternotomy, the pericardium

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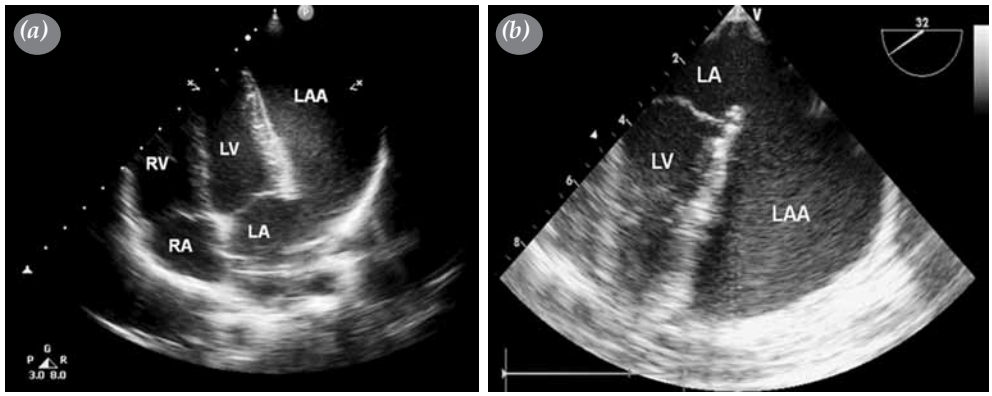

Figure 1. A view of the four-chamber transthoracic echocardiography on the left and the four-chamber transesophageal echocardiography on the right. LAA: Left atrial appendage; LA: Left atrium; LV: Left ventricle; RA: Right atrium; RV: Right ventricle.

was opened and the aneurysm was found at the left myocardial border. It was compressing the left ventricle, which was consequently deformed (Figure 2). Under cardiopulmonary bypass (CPB) and left atrial decompression with venting by the right upper pulmonary vein, cardiac arrest was established with antegrade cold blood cardioplegia. The aneurysmal sac was then entered from the outside. Its wall was fragile, but there was no thrombus within the sac or the left atrium. Next, the aneurysmal appendage tissue was resected, and the neck of the aneurysmal sac was closed with a continuous prolene suture. Then the remaining appendage tissue was closed over this. Afterwards, the patient was taken off of CPB in sinus rhythm, and the operation was completed uneventfully. She was discharged from the hospital on postoperative day six. The pathology report of the resected LAA revealed an extremely thin-walled, dilated LAA aneurysm, and a microscopic view showed predominant endomyocardial fibrosis without any signs of inflammation. At the edge of the aneurysm, the myocardium was replaced by scar tissue with old, obliterated vessels and necrotic myocardial cells. At the postoperative one- and six-month follow-ups, the patient was in sinus rhythm and her echocardiographic findings were normal.

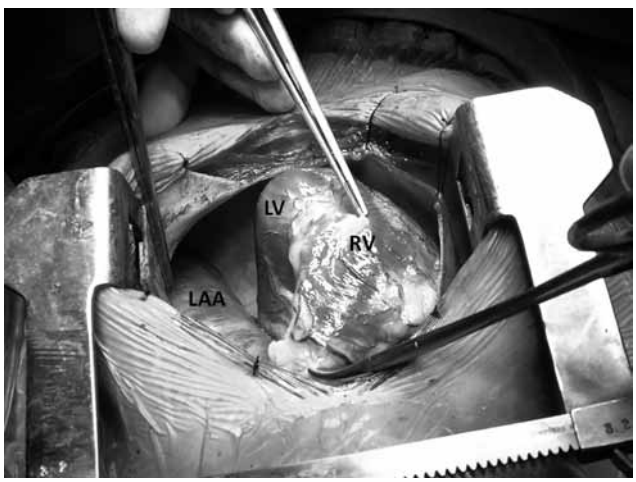

Figure 2. Surgical view after opening the pericardium. LAA: Left atrial appendage; LV: Left ventricle; RV: Right ventricle.

DISCUSSION

A LAA aneurysm is very rare and may be either congenital or acquired; however, congenital aneurysms are extremely uncommon. When these occur, the auricle is not usually enlarged, and the aneurysms are not associated with other pathologies. The most frequent location is the LAA, although they have also been found in the wall of the left atrium and on very rare occasions, in the right atrium.

Generally, congenital aneurysms occur intrapericardially, but there have been a few cases in which partial defects were seen in the pericardium. The origin of these aneurysms is unknown, but some authors have attributed them to dysplasia of the musculi pectinati.[1] Histological studies of the LAA wall found that they are mostly composed of a collagenous atrial substance and cardiac atrial muscle, but endocardial endothelial tissue may also be observed.[2]

In the majority of cases, the main symptom leading to diagnosis of congenital LAA is recurrent or continuous supraventricular arrhythmia; however systemic embolisms are also common. Cardiac insufficiency and chest pain are seen much less often. A routine chest roentgenogram can be used to determine whether the left border of the heart is enlarged, and an exact diagnosis is relatively easy to make via contrast echocardiography, MRI, and/or CT angiography.[2]

Because patients are usually symptomatic at the time of diagnosis, prompt surgery via an aneurysmectomy is indicated. Even for those without symptoms, surgery is necessary to eliminate potential complications. Various approaches to the aneurysmectomy have been successfully used and have been previously described in the literature. For example, access to the aneurysm has been achieved by a median sternotomy, a left thoracotomy, and a minithoracotomy[3,4] while the resection has been carried out both with and without extracorporeal circulation.[5] Our reason for choosing a median sternotomy was to use the most viable approach to reach the aneurysm in case CPB was needed. It was not easy to resect the aneurysm without CPB because of the adhesions between the LAA aneurysm and the LAA at the posterior side. In addition, cardiac arrest was performed to reach the most posterior part of the neck of the aneurysm because it was distended.

In conclusion, a child with opacity at the left perihilar region on chest X-ray may have a congenital aneurysm of the LAA. Even though cases involving an isolated LAA aneurysm are unusual in childhood, surgical resection is preferable to ensure a favorable outcome because it can prevent systemic embolization, rupture of the aneurysmal sac, or a mass affect in the pericardium from occurring.

Declaration of conflicting interests

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