

An incidentally diagnosed asymptomatic congenital left atrial appendage aneurysm

Tanısı tesadüfen konulan asemptomatik doğuştan sol atriyal apendiks anevrizması

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Left atrial appendage aneurysm is a rare cardiac anomaly. It can be congenital in origin or acquired secondary to inflammatory or degenerative processes. As it may cause potentially lethal arrhythmias or thrombi, surgical treatment is needed immediately after the diagnosis. In this article, we report a case of an asymptomatic three-year-old boy with a congenital left atrial appendage aneurysm. The patient was successfully treated with surgical resection of the aneurysm.

Keywords: Child; congenital; left atrial aneurysm.

Left atrial appendage aneurysm is a rare cardiac anomaly. It can be congenital in origin or can develop secondary to inflammatory or degenerative processes. Most patients with congenital left atrial aneurysm are asymptomatic until the second decade. Symptomatic patients usually present with arrhythmia or systemic embolization.^[1] Herein, we report a case of congenital left atrial aneurysm which was an incidentally diagnosed.

CASE REPORT

A three-year-old boy was referred to the pediatric cardiology unit for the detection of a murmur during routine pediatric examination. Physical examination revealed non-specific findings except grade 1/6 soft, systolic murmur heard at the left lower sternal border. The resting electrocardiography (ECG) was normal, while chest radiograph showed cardiomegaly with a prominent bulge on the left upper cardiac contour.

Sol atriyal apendiks anevrizması, nadir rastlanan bir kardiyak anomalidir. Doğuştan oluşabileceği gibi, enflamatuvar veya dejeneratif süreçlere sekonder edinsel olarak da gelişebilir. Potansiyel ölümcül aritmilere veya trombüs oluşumuna neden olabileceği için, tanı konulur konulmaz cerrahi tedavi gereklidir. Bu yazıda üç yaşında asemptomatik bir erkek çocukta saptanan doğuştan sol atriyal apendiks anevrizma olgusu sunuldu. Hasta anevrizmanın cerrahi eksizyonu ile başarılı bir şekilde tedavi edildi.

Anahtar sözcükler: Çocuk; doğuştan; sol atriyal anevrizma.

Echocardiographic examination revealed a large aneurysm of the left atrial appendage communicating with the left atrium (Figure 1). Associated congenital heart defects were not present. Magnetic resonance imaging (MRI) of the heart revealed an atrial aneurysm of 4.2x3x3.5 cm in size (Figure 2).

Surgical removal of the aneurysm was performed through a median sternotomy, on cardiopulmonary bypass (CPB) with moderate hypothermia and crystalloid cardioplegia (Figure 3). The aneurysm was located at the posteroinferior section of the heart, occupying a large space within the pericardial sac, displacing the left ventricle anteriorly. The operation was performed on CPB with moderate hypothermia and antegrade crystalloid cardioplegia with aortic cross clamping at 32 °C. The aneurysmal sac was resected and inspection was performed to check for gross pathology or thrombus formation. The remnant of left atrial appendage was closed with a double running



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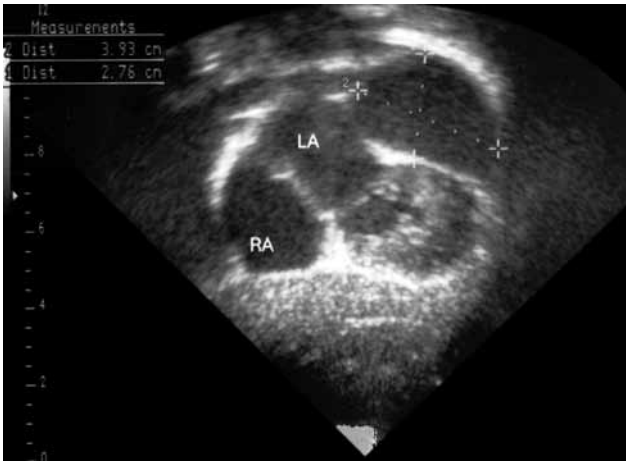


Figure 1. Echocardiographic view of the left atrium and the left atrial appendage aneurysm from subcostal window.

layer of 5-0 prolene. Total CPB time was 33 minutes, whereas cross-clamp time was 13 minutes. Heart beat of the patient recovered spontaneously after the aortic cross clamp was released. Weaning from CPB was smooth. The postoperative course was uneventful and the patient was discharged on the sixth postoperative day. At one year during the follow-up, the patient still remained asymptomatic.

Pathologic examination demonstrated full layers of endocardial, myocardium and the epicardium with focal subendocardial fibrous hyalinization, which were



Figure 2. Magnetic resonance imaging of the left atrial aneurysm.

consistent with the diagnosis of congenital atrial aneurysm.

DISCUSSION

Congenital left atrial appendage aneurysm is a rare congenital anomaly first described in 1953 by Fry.^[2] Acquired aneurysms are more often encountered than the congenital ones. Acquired aneurysms may be associated with diseases which cause mitral stenosis such as rheumatic heart disease or congenital mitral stenosis. Acquired atrial aneurysm associated with tuberculosis or syphilitic myocarditis has been previously reported.^[2]

Congenital left atrial appendage aneurysm is rarely symptomatic in childhood. In addition, it is rarely symptomatic in the infancy and even prenatally.^[3,4] Our patient was asymptomatic and the aneurysm was incidentally detected by transthoracic echocardiography (TTE) which was in consistent with the literature findings. During childhood, left atrial aneurysm may cause heart failure, respiratory distress or cardiac tamponade. It mostly presents after the second decade with palpitations caused by atrial fibrillation or supraventricular tachycardia, symptoms of mitral regurgitation due to the annular dilation and findings of systemic thromboembolism. One third of the cases were reported to already have a thrombus formation within the aneurysmal sac during surgery.^[5] Stasis within the aneurysmal sac accounts for the thrombus formation and systemic embolization.^[1]

The following diagnostic criteria for congenital left atrial aneurysm were proposed: (i) the origin from an otherwise normal left atrium, (ii) a well-defined communication with the left atrium,



Figure 3. Intraoperative view of the aneurysm.

(iii) intrapericardial location and (iv) displacement or distortion of the left ventricle by the aneurysm.^[6] Our patient fulfilled all of the four criteria for the diagnosis of a left atrial aneurysm. Another diagnostic criterion is the linear measurement of the aneurysm exceeding 3 cm.^[7]

Several imaging studies such as TTE or transesophageal echocardiography (TEE), computed tomography, angiography and MRI can be used for diagnosis.^[8] With the introduction of technological improvements in imaging studies in recent years, conventional angiography is rarely performed as a diagnostic tool.

Complications associated with aneurysm including arrhythmias and thromboembolism, in particular, can be devastating. Therefore, prompt surgical excision upon diagnosis is recommended, even in asymptomatic patients. A thorough characterization of the aneurysmal sac should be performed before surgery either with MRI or echocardiography. Aneurysms which have a thrombus formation or a large connection with the left atrium require CPB. On the other hand, aneurysms with a small connection can be surgically excised without CPB.

Approaching the left atrium via left thoracotomy without CPB has been previously reported in the literature.^[9,10] However, we used median sternotomy technique, since we believe that median sternotomy with cardiac arrest and CPB is a safer method in small children, particularly.

In conclusion, given the fact that atrial aneurysms have potential for further devastating complications; such as mitral regurgitation due to annular dilatation, supraventricular tachycardia/atrial fibrillation or systemic thromboembolism, surgical excision should be performed upon diagnosis before the complications develop.

Declaration of conflicting interests

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