CASE REPORT

Giant Congenital Aneurysm of the Left Atrial Appendage

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ABSTRACT

Left atrial appendage aneurysm, either congenital or acquired, is a very rare anomaly. It is caused by congenital dysplasia of the atrial muscles, or it may be the result of other systemic or heart diseases.
This anomaly is mostly asymptomatic and usually diagnosed incidentally, but occasionally it may present with atrial tachyarrhythmias and/or thromboembolic events.
This report describes the case of a patient with giant congenital aneurysm of the left atrial appendage.

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Key words > Aneurysm - Atrium - Atrial appendage

Abbreviations >

LA	Left atrium	**TTE**	Transthoracic echocardiography
LAAA	Left atrial appendage aneurysm	**MRI**	Magnetic resonance imaging
ECG	Electrocardiography	**CT**	Computerized tomography
TEE	Transesophageal echocardiography		

INTRODUCTION

Left atrial appendage aneurysm (LAAA) is a rare condition. To our knowledge, only 78 cases are reported so far in literature. (1)

Most of these cases are of congenital etiology, attributed to dysplasia of the left atrial (LA) pectinate muscles. They are rarely acquired, and occur due to a condition secondary to inflammatory or degenerative processes (2, 3) and/or to mitral valve disease. (4)

Many of the cases have been diagnosed from incidental findings through chest X-rays. (2, 5) However, patients may present with palpitations due to supraventricular arrhythmia, thromboembolism or other unusual symptoms such as dyspnea and angina pectoris. (5)

This report describes the case of a patient with giant congenital aneurysm of the left atrial appendage.

CLINICAL REPORT

A 32-year-old male patient was admitted to the emergency service with an episode of sudden, rapid palpitations while playing soccer, with no other associated symptoms. Physical examination showed a rapid irregular pulse, and the ECG confirmed atrial fibrillation with high ventricular response. The patient recovered his sinus rhythm spontaneously. A subsequent ECG showed signs of left atrial overload.

Lab tests were normal. Chest X-ray (Figure 1) revealed a prominent left heart border. Transthoracic echocardiography (TTE) showed a severely enlarged left atrial (LA) appendage.

The patient had a history of self-limiting, rapid, irregular episodes of palpitations of several years evolution associated with sports practice, for which he had never consulted before. By prior chest X-rays, he knew he had cardiomegaly due to "athlete's heart". He underwent a transesophageal echocardiography (TEE) revealing significant LA appendage enlargement (4.16 cm x 8.95 cm) which confirmed the LAAA diagnosis. There was no evidence of atrial thrombi, but a thick membrane at the appendage entrance was detected, with no significant pressure gradient registered by pulsed Doppler (Figure 2).

A 64-row multidetector computed angiotomography (CT) and a magnetic resonance imaging (MRI) were performed to confirm the diagnosis and rule out associated congenital diseases and coronary artery disease. The patient received oral anticoagulants and amiodarone and was referred to cardiovascular surgery for surgical treatment.

Resection of the LAAA was performed under extracorporeal circulation (Figure 3). The patient progressed

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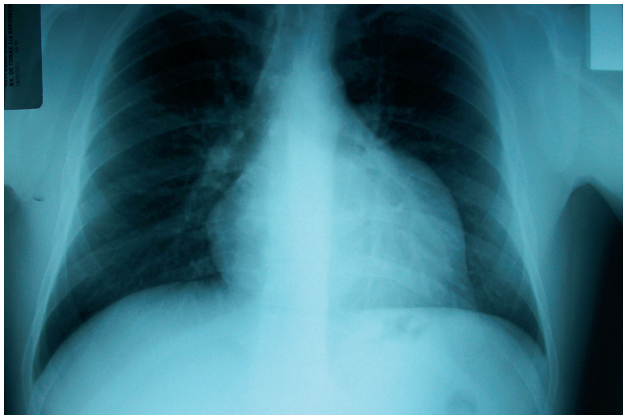

Fig. 1. Chest X-ray showing prominent left ventricular border.

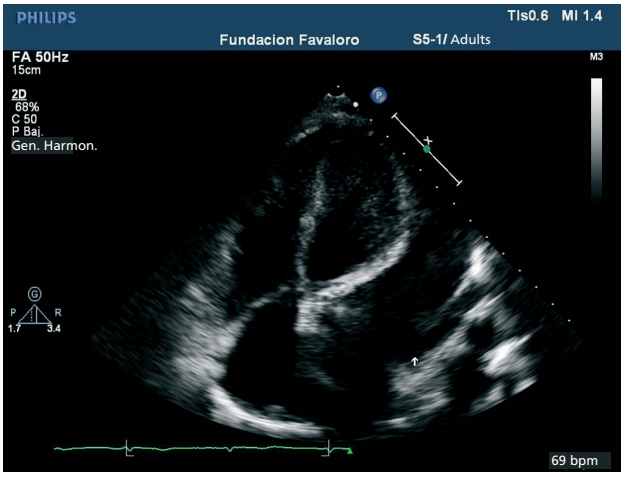

Fig. 2. Four-chamber view transthoracic echocardiography showing giant aneurysm of the left atrial appendage, close to the left ventricle.

uneventfully, and three months later he remained asymptomatic and in sinus rhythm without any medication. The control TTE was normal.

DISCUSSION

We report the case of a 32-year-old man with a history of intermittent, fast, irregular episodes of palpitations of many years evolution associated with sports practice, and an event of atrial fibrillation documented in an emergency service. Subsequent studies revealed a giant LAAA, with no evidence of any other associated systemic or cardiac disease.

The majority of LAAA are congenital, although there are reports of aneurysms acquired in relation to mitral valve disease or systemic pathologies. (2, 3)

The origin of congenital aneurysms would be attributed to dysplasia of the LA pectinate muscles. (1)

Left atrial appendage aneurysm is rarely diagnosed during childhood and generally becomes manifest during the second or third decades of life, as was the case of our patient. (1)

Most cases are silent and found incidentally, but others are symptomatic, including palpitations related with supraventricular arrhythmias, dyspnea, angina pectoris (probably due to compression of the left coronary artery trunk or branches), and stroke or sudden death related to thromboembolism. Our patient suffered from palpitations and had at least one documented episode of atrial fibrillation.

There are extremely rare associations of LAAA with other congenital diseases, such as septal defect, persistent left superior vena cava, anomalies of the renal arteries or anomalous pulmonary venous return. (6)

The majority of LAAA are found incidentally in a chest X-ray as an anomalous mass on the left border of the heart silhouette. Many of the diagnostic techniques, such as TTE, TEE, CT and MRI, have proved to be useful for the diagnosis of LAAA in terms of differentiating it from other pathologies. The diagnostic method of choice will depend on the patient's characteristics, local availability, and operators' expertise of the different methods. (6)

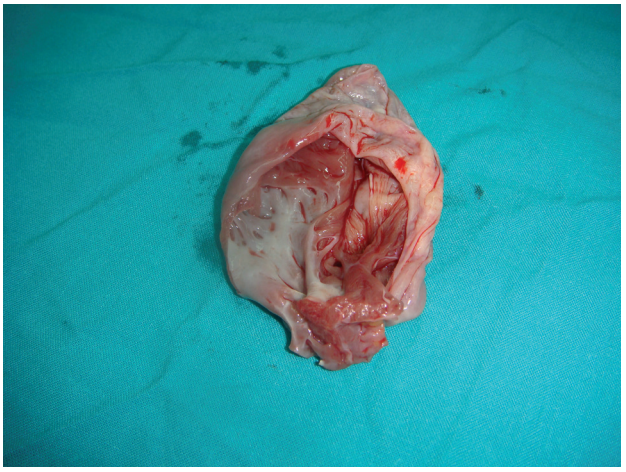

Fig. 3. Surgical piece of extirpated left atrial appendage aneurysm.

Diagnostic criteria for LAAA include: 1) absence of any other concomitant cardiac pathology; 2) its origin from a normal LA; 3) a direct continuity of flow between the LA and the atrial appendage; 4) distortion of the left ventricular free wall by the aneurysmal body; and 5) absence of pericardial defects. Furthermore, some authors incorporate its complete inclusion within the pericardium. Its size has also been proposed as another diagnostic criterion for this pathology, taking into consideration a longitudinal diameter of 3 cm, but this is not generally accepted. (7)

The LA appendage is usually the most affected by the aneurysm; however, other localizations within the LA have been described. Moreover, localization in the right atrium is even less common. (3)

Left atrial appendage aneurysm requires differential diagnosis from other pathologies such as pericardial effusion, pericardial cyst, coronary artery aneurysm, left ventricular pseudoaneurysm and coronary sinus dilation. All these entities have a similar

appearance, since they are echolucent and can have the same localization, but only the LAAA communicates directly with the LA chamber. (6) In the few cases of acquired LAAA reported, diagnosis is performed when the LAAA coexists with conditions that increase pressure and atrial dimensions, such as heart failure or mitral valve stenosis, (2) or due to weak LA appendage wall secondary to myocarditis. (3)

Surgical resection of the aneurysm is the treatment of choice, even in totally asymptomatic patients. (6) Resection of the LAAA is a safe and effective treatment that eliminates arrhythmogenic foci and potential sources of systemic embolism. (6)

There is little evidence in the literature about isolated medical treatment. Some authors have chosen medical treatment for some patients with giant LAAA with no evidence of arrhythmias or thrombi, (8) but there are no long-term follow-up reports for these cases, except for a case that resulted in stroke 2 years after diagnosis. (9) Based on case reports, we could argue that isolated resection of the LAAA results in successful sinus rhythm and absence of arrhythmias, provided the substrate of the irritable focus has been fully resected. Absence of arrhythmias after resection is reported in several publications, with up to 8 years follow-up. (9)

We believe percutaneous closure techniques can become an optional treatment for congenital LAAA in the future, as was described in a case of a LAAA pseudoaneurysm. (10)

CONCLUSIONS

Left atrial appendage aneurysm is a rare entity, usually congenital, but acquired in some cases. The majority of congenital LAAA are diagnosed incidentally during the second or third decades of life. Surgical resection is the recommended treatment, eliminating the risk of arrhythmias and thromboembolism.

RESUMEN

El aneurisma de la orejuela de la aurícula izquierda, ya sea congénito o adquirido, es una anomalía extremadamente rara. Es causado por la displasia congénita de los músculos auriculares o puede ser consecuencia de otras enfermedades cardíacas o sistémicas.
Esta anomalía cardíaca generalmente es asintomática y su diagnóstico suele hacerse de manera incidental, pero en ocasiones puede manifestarse por taquiarritmias auriculares y/o por eventos tromboembólicos.
En esta presentación se describe el caso de un paciente con un aneurisma congénito gigante de la orejuela de la aurícula izquierda.

Palabras clave > Aneurisma - Apéndice atrial - Aurícula

Conflicts of interest:
None declared.

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