Congenital left atrial appendage aneurysm: Atypical presentation

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ABSTRACT

Congenital left atrial appendage aneurysm is a rare condition caused by dysplasia of the atrial muscles. We report a case of a 14-year-old boy, with a 5-month history of cough and in sinus rhythm. Transthoracic echocardiography and computerized tomographic angiography confirmed the aneurysm of the left atrial appendage which was resected through median sternotomy on cardiopulmonary bypass. This case is presented not only for its rarity but also for its atypical clinical presentation.

Keywords: Aneurysmectomy, congenital heart disease, left atrial appendage aneurysm

INTRODUCTION

Left atrial appendage aneurysm (LAAA) is a rare condition characterized by localized or diffuse dilatation of the left atrial appendage. Congenital LAAA must be separated from other forms of dilatation of the left appendage secondary to rheumatic or degenerative process involving mitral valve. While most patients reported in the literature present with signs of atrial tachyarrhythmias and/or systemic thromboembolism, we present a case of a 14-year-old boy presenting with a 5-month history of cough, in sinus rhythm, and found to have a large LAAA.

CASE REPORT

A 14-year-old boy presented to our institution with a 5-month history of dry cough. On presentation, he had an unremarkable lung and cardiac examination. Chest roentgenogram showed a prominent upper left heart border with a normal cardiothoracic ratio [Figure 1a]. Electrocardiogram confirmed a sinus rhythm associated with left atrial enlargement. He underwent a transthoracic echocardiography that revealed a large cyst adjacent to the left ventricle; the cavity size was 8 cm × 6 cm with a surface area of 24 cm². It was free from any echodensity suggesting the presence of thrombus. There was a connection between this cavity and the left atrium [Figure 1b]. Computed tomography (CT) angiography demonstrated the LAAA with no thrombus in it. CT coronary acquisition ruled out any compression of the left coronary arteries' tree from the mass [Figure 1c]. The patient was referred for surgical removal of the aneurysm: aneurysmectomy.

The aneurysm was approached through median sternotomy on cardiopulmonary bypass (CPB). On inspection, a deformation of the pulmonary infundibulum and the lateral wall of the left ventricle were noted [Figure 2a]. After cardioplegic arrest, the aneurysm was opened, no thrombus was noted inside it [Figure 2b], and then resected and sutured at its base. The postoperative course was uneventful. The anatomicopathological report showed that the aneurysmal wall has three layers: endocardium, myocardium, and pericardium with endomyocardial fibrosis. The cough has completely resolved in the immediate postoperative course and at follow-up.

DISCUSSION

Congenital LAAA is characterized by a diffuse enlargement of the appendage caused by dysplasia of musculi pectinati

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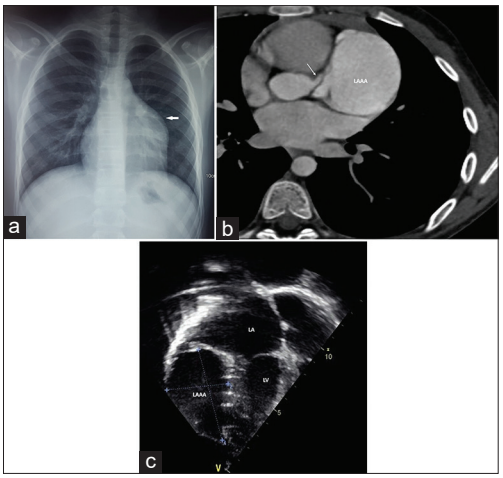

Figure 1: (a) Chest X-ray demonstrating prominent upper left heart border. (b) Transthoracic echocardiogram showing an apical four chamber. The left atrial appendage aneurysm is communicating with the left atrium. (c) Coronary computed tomography angiography demonstrating a distinct separation between the mass (arrow) and the left main coronary artery (arrow)

and related atrial muscle bands. The left appendage preserves its normal three layers' configuration without any microscopic anomalies in the nucleus. Congenital forms must be separated from acquired forms where the atrial enlargement is secondary to mitral stenosis or regurgitation. Congenital LAAA is a rare condition with approximately 50–80 cases reported in the published English literature; this prevalence is varying depending on the studied era.[1,2]

Clinical presentation ranges from asymptomatic patients to complications such as systemic thromboembolic events. The most common clinical presentations were palpitations reported in 43%, followed by dyspnea in 22%, arrhythmia in 15%, stroke in 11%, and nonspecific chest pain in 7%.[1] Chest pain is due to tension on the pericardium or to a temporary compression by the aneurysm on the coronary arteries. Finally, reduced left ventricular compliance by aneurysm compression is leading to raising ventricular filling pressures and diastolic dysfunction with dyspnea and heart failure.

Our patient, despite a deformity seen in the lateral wall of the left ventricle [Figure 2a], had normal left ventricular compliance and filling pressures. Thus, the symptom of cough present in this case is not related to diastolic dysfunction and heart failure. Since our patient had a giant LAAA, it may result in a compression on the left mainstem bronchi and/or on the left recurrent laryngeal nerve leading to episodes of cough. This symptom resolved completely after surgery supporting the role of the aneurysm mass effect on these structures. CT coronary angiography [Figure 1c] showed a distinct separation between the aneurysm and the coronary arteries explaining that chest pain and signs of ischemia in the electrocardiogram were absent in our patient.

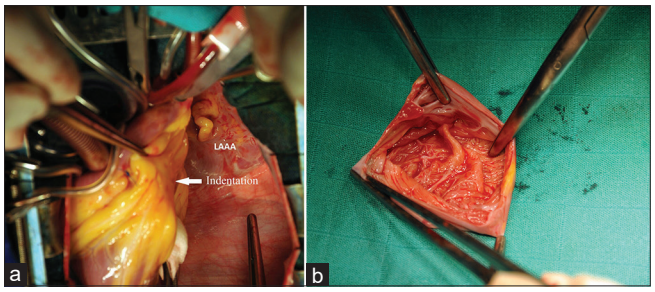

Figure 2: (a) Intraoperative pictures demonstrating a large aneurysm of the left appendage (LAAA) with the indentation deformity in the pulmonary infundibulum and the left ventricle (arrow). (b) The resected aneurysm without thrombus inside it

Like other rare conditions, no guidelines exist concerning the management of these aneurysms. However, most of the authors proposed surgical resection as the standard treatment modality regardless of the presence or absence of symptoms.[3] Median sternotomy with CPB is preferred when the aneurysm is large with thrombus, and minimally invasive techniques should be considered for small LAAA in centers familiar with these procedures.[4] Although there are no long-term follow-up studies, freedom from atrial fibrillation has been reported for up to 8 years after surgery.[5]

This case report demonstrates that even large LAAA may present in sinus rhythm and without signs of heart failure. The symptom of cough present is probably related to compression on adjacent airway or nervous structures. Further studies are needed to understand all aspects of the disease and then to establish guidelines for the management of this rare entity.

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Conflicts of interest

There are no conflicts of interest.

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