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The Fifth Chamber of the Heart: Congenital Aneurysm of the Atrial Appendage

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This case series reports on four patients who presented with giant aneurysms of the atrial appendages (AA).

Patient 1 was a 4.5-month-old male, who was referred due to an "enlarged heart shadow on chest radiography." Transthoracic echocardiography showed an aneurysm of the left atrial appendage measuring 4.5x4.5 cm, which communicated with the atrial cavity via a 1.2 cm neck. This aneurysmal cavity was described as a "fifth chamber" (Figure 1a) and is consistent with previous reports.[1]

Patient 2 was a 15-year-old, 45 kg female who presented with palpitations and syncope attacks. Echocardiography revealed a giant aneurysm measuring 16x10 cm, which communicated with the right atrium (Figure 1b). Computerized tomography (CT) angiography confirmed the diagnosis of a giant right atrial appendage aneurysm (RAAA) (Figure 1c).

Patient 3 was a 24-year-old male who was diagnosed with Ebstein's anomaly during his military service examination. Echocardiography revealed a giant aneurysm measuring 6.1x5.6 cm) of the right AA, which communicated with the atrial cavity. The diagnosis was confirmed by CT and magnetic resonance imaging (MRI) (Figure 1d).

Patient 4 was an 11-year-old female who applied for a second opinion and was found to have an aneurysmal enlargement of the right atrium appendage measuring 11.5x9.5 cm. The patient complained of intermittent palpitations, but no rhythm disorder was found during Holter monitoring (Figure 1e). None of the patients had a thrombus formation in the appendage aneurysms.

Atrial appendage aneurysm resection was performed on all patients via a median sternotomy under cardiopulmonary bypass. One patient (patient 2) underwent a right atrial maze procedure for atrial arrhythmias in addition to atrial aneurysm resection.

Atrial appendage aneurysms are uncommon conditions believed to result from dysplasia of the atrial appendage musculature, leading to localized or diffuse dilatation of the AA.[2] The right appendage aneurysm is more common than the left.[1] Although this condition can occur in people of all ages, it is rarely diagnosed in childhood[1-5] is usually found incidentally. The aneurysms can increase in size over time and may present as atrial arrhythmias or systemic thromboembolic events, which are potentially life-threatening complications.[2]

Echocardiography is the primary diagnostic method for AA aneurysms, and cardiac CT or MRI can provide more detailed evaluation. Surgical treatment is recommended even in asymptomatic patients to avoid atrial arrhythmias or thromboembolic events.[3,4] Antiarrhythmic therapy may be administered during concomitant atrial appendage resection to reduce the risk of thromboembolic complications, depending on the patient's clinical condition.

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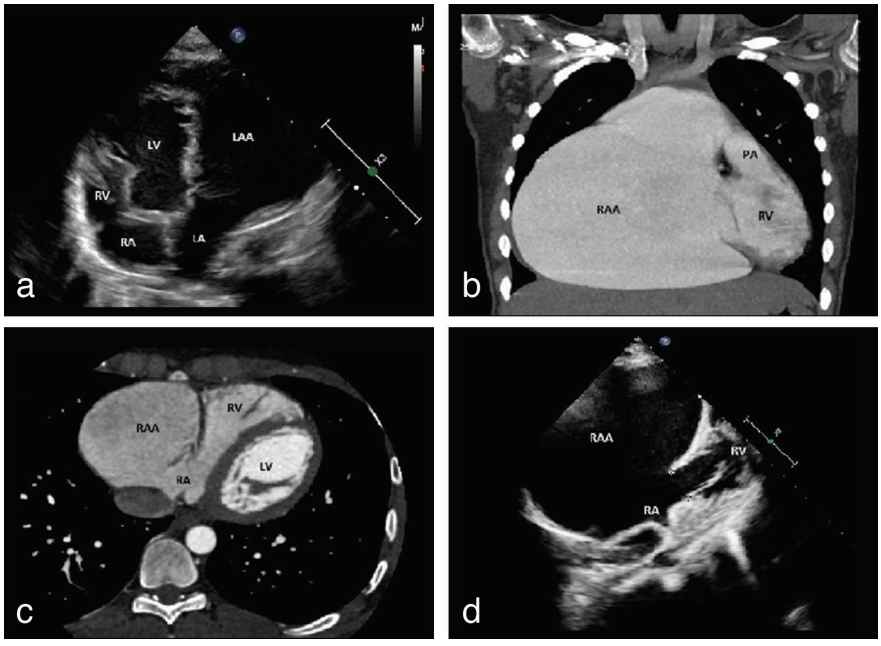

FIG. 1.
a: Patient 1: A transthoracic echocardiography showing a left atrial appendage aneurysm.
b: Patient 2: CT angiography coronal section showing a giant aneurysm associated with the right atrium.
c: Patient 3: Aneurysm formation associated with the right atrium is seen on the axial section of CT angiography.
d: Patient 4: A giant aneurysm associated with the right atrium is seen on the transthoracic echocardiography.

CT: computerized tomography; LA: left atrium; LAAA: left atrial appendage aneurysm; LV: left ventricle; PA: pulmonary artery; RA: right atrium; RAAA: right atrial appendage aneurysm; RV: right ventricle

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