A 13-year-old boy was referred to cardiology for evaluation of intermittent exertional breathlessness. Previous respiratory investigations had revealed normal lung function and no evidence of asthma on exercise spirometry. There was no past medical history of note. Cardiorespiratory examination was unremarkable except for a soft ejection systolic murmur at the lower left sternal border. ECG showed sinus rhythm and normal P-wave morphology and QRS axis, with a T-wave inversion in leads III and aVF. Chest radiograph demonstrated normal cardiothoracic ratio, but the heart was noted to be globular in shape (Figure 1).

Transthoracic echocardiography revealed the presence of a large, thin-walled aneurysmal structure in continuity with the free wall of the right atrium, discrete from the caval veins (Figure 2). There was mild tricuspid regurgitation, with a Doppler velocity of 2.1 m/s. Both ventricles were of normal size and function. To further characterize this cardiac abnormality, the patient was referred for cardiac magnetic resonance (CMR) imaging. Cine imaging using a balanced steady-state free precession sequence showed a noncontractile, smooth-walled, right atrial appendage aneurysm (RAAA) extending anterior to the body of the right atrium (Figure 3A and 3B and Movie I of the online-only Data Supplement). The aneurysm had a broad-based pyramidal shape and was partially separated from the main atrial chamber by a membrane (total indexed right atrial volume 118 mL/m² at end-ventricular systole). There was mild distortion of the right ventricular (RV) inlet, but no significant compression. No intracavity thrombus was identified. The pulmonary arteries were unobstructed and of normal caliber. There was a small pericardial effusion.

The patient subsequently underwent exercise tolerance testing, managing 12 minutes and 15 seconds on the standard Bruce protocol and achieving a workload of 13.8 metabolic equivalents. The test was stopped because of dyspnea, with no ST-segment changes detected. A 1-week cardiac rhythm monitor revealed no evidence of arrhythmia. On review in the outpatient clinic, the patient had symptomatically improved. He was therefore commenced on aspirin for thromboembolic prophylaxis and managed conservatively with annual imaging surveillance.

Figure 1. Chest radiograph (posteroanterior film) showed that the cardiac silhouette was of normal size, but globular in shape.

Figure 2. Transthoracic echocardiography (apical 4-chamber view) demonstrated an aneurysmal cavity (asterisk) contiguous with the main body of the RA. RA indicates right atrium; RV, right ventricle.
During the next 2 years the patient was asymptomatic, with no significant change in the size of the RAAA on transthoracic echocardiography. However, repeat CMR imaging performed 3 years after the initial scan revealed a marked increase in the size of the aneurysm that was disproportionate to somatic growth (total indexed right atrial volume increased to 151 mL/m²) (Figure 3C and 3D). In addition, there was evidence of clear compression of the RV inlet by the RAAA (Movie II of the online-only Data Supplement), with flattening of the tricuspid annulus during ventricular diastole. Although the patient remained well, in view of the increase in relative size of the RAAA and associated RV inlet diastolic displacement, the patient was referred for surgical resection.

After median sternotomy and pericardectomy, the aneurysm was visible, arising from the caudal part of the right atrial free wall in close proximity to the inferior vena cava and right coronary artery (Figure 4). In view of the closely related vascular structures, the RAAA was partially resected with plication of the residual aneurysmal tissue without complication. Pathological examination of the resected specimen. Macroscopic examination revealed a paper-thin, semitransparent membrane with chord-like structures on its endocardial surface.

Figure 3. Serial T1-weighted bSSFP CMR images in mid-diastole from 2005 (A and B), 2008 (C and D), and 2010 (E and F). Four-chamber (A) and transverse axial (B) views initially showed a thin-walled RAAA (asterisk) without major compression of the RV inlet (arrowhead). No intracavity thrombus was identified. Repeat images acquired 3 years later (C and D) revealed a marked increase in the size of the RAAA, which was now compressing the RV inlet (white arrow). After partial resection, there was reduction in the size of the RAAA and relief of RV inlet compression (E and F).

Figure 4. Intraoperative view of the intrapericardial RAAA (asterisk). RAAA indicates right atrial appendage aneurysm.

Figure 5. Histopathologic appearance of the resected specimen. Macroscopic examination revealed a paper-thin, semitransparent membrane with chord-like structures on its endocardial surface.

Figure 6. Transmural microscopic section revealed that the wall of the RAAA was predominantly composed of bands (arrow) of collagen and smooth muscle with scattered residual atrial myocytes (arrowhead). Residual papillary muscle (asterisk) is also visible. RAAA indicates right atrial appendage aneurysm.
imen revealed a round, paper-thin strip of fibrous tissue measuring 40×35 mm and <1 mm in thickness (Figure 5). Transmural histological section confirmed that the wall of the aneurysm consisted primarily of layers of collagen and smooth muscle cells, with interspersed islands of residual atrial myocytes (Figure 6). At the outpatient review 2 months after surgery, the patient had made a good recovery. Repeat CMR showed that the RAAA was significantly reduced in size (total indexed right atrial volume decreased to 83 mL/m²), with no residual RV inlet compression (Figure 3E and 3F and Movie III of the online-only Data Supplement).

Right atrial appendage aneurysm is a rare condition that is most often congenital in etiology. Potential complications include arrhythmia, thrombosis, and rupture. Clinical experience of this malformation is limited, with the result that the natural history and optimal management strategy are unclear. In this case, serial CMR demonstrated significant relative enlargement of the RAAA and associated compression of the RV inlet during diastole. Our report therefore highlights the importance of longitudinal follow-up of RAAA and the value of CMR imaging in performing this task.

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