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Asymptomatic Right Atrial Appendage Aneurysm Associated with Tricuspid Regurgitation in a 7-year-old Boy

A 7-year-old boy, without any symptoms, was referred to our center due to a cardiac murmur. Transthoracic echocardiography demonstrated an enlarged right atrium (143 × 91 mm) and moderate tricuspid regurgitation (TR) (Figure 1A-1C). Imaging confirmed the diagnosis as a right atrial appendage aneurysm (RAAA) (Figure 1D and E). Electrocardiogram and Holter monitor test showed sinus rhythm (Figure 1F). The patient underwent surgical intervention through a median sternotomy. Intraoperatively, an extremely enlarged right atrial appendage and a thin atrial wall were revealed (Figure 2A and 2B). The annulus of the tricuspid valve was dilated. The saline test revealed obvious centric regurgitation without insufficiency or prolapse of the leaflets (Video 1). The subvalvular apparatus, with intact cords and papillary muscles, appeared to be typical. Annuloplasty was performed with interrupted sutures with pledges, and the latter saline test showed minimized TR (Video 2). The RAAA was resected (Figure 2C), and the right atrium was reconstructed (Figure 2D). Transthoracic echocardiography confirmed satisfying tricuspid valve performance as trivial TR and no stenosis. Postoperative recovery was uneventful, and the patient is currently under follow-up for 16 months.

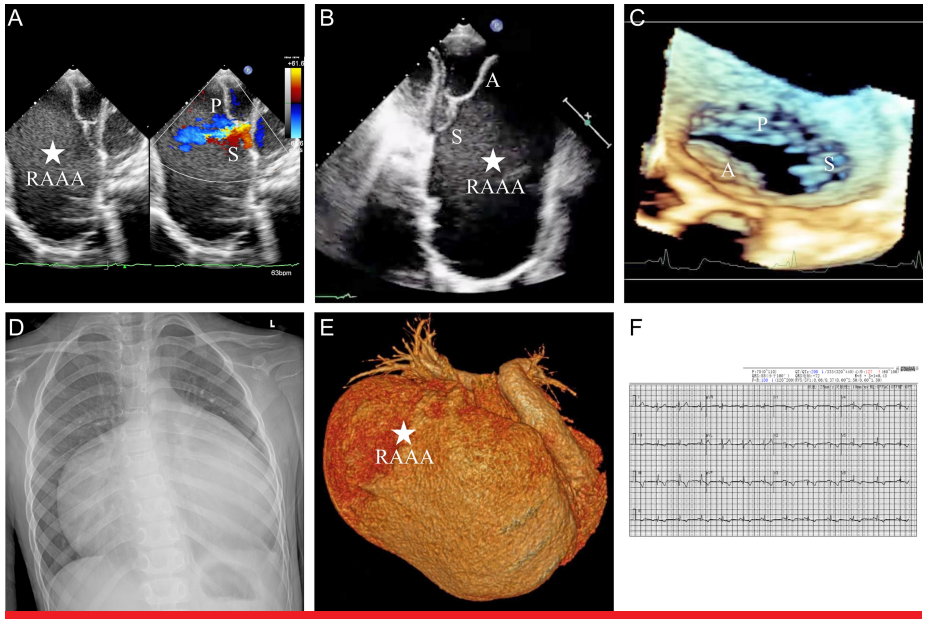

Figure 1. (A-C) Preoperative transthoracic echocardiography showing a significantly enlarged right atrium and a stream of tricuspid regurgitation between the septal and posterior leaflets. The three leaflets (A: anterior, P: posterior, S: septal) with normal shapes adhered to the annulus level, distinguishing this patient from Ebstein anomaly. (D, E) Preoperative chest x-ray and CT imaging revealing the size and spatial location of the RAAA. The cardiothoracic ratio was about 0.9. (F) Preoperative electrocardiogram as sinus rhythm. RAAA, right atrial appendage aneurysm.

E-PAGE ORIGINAL IMAGE

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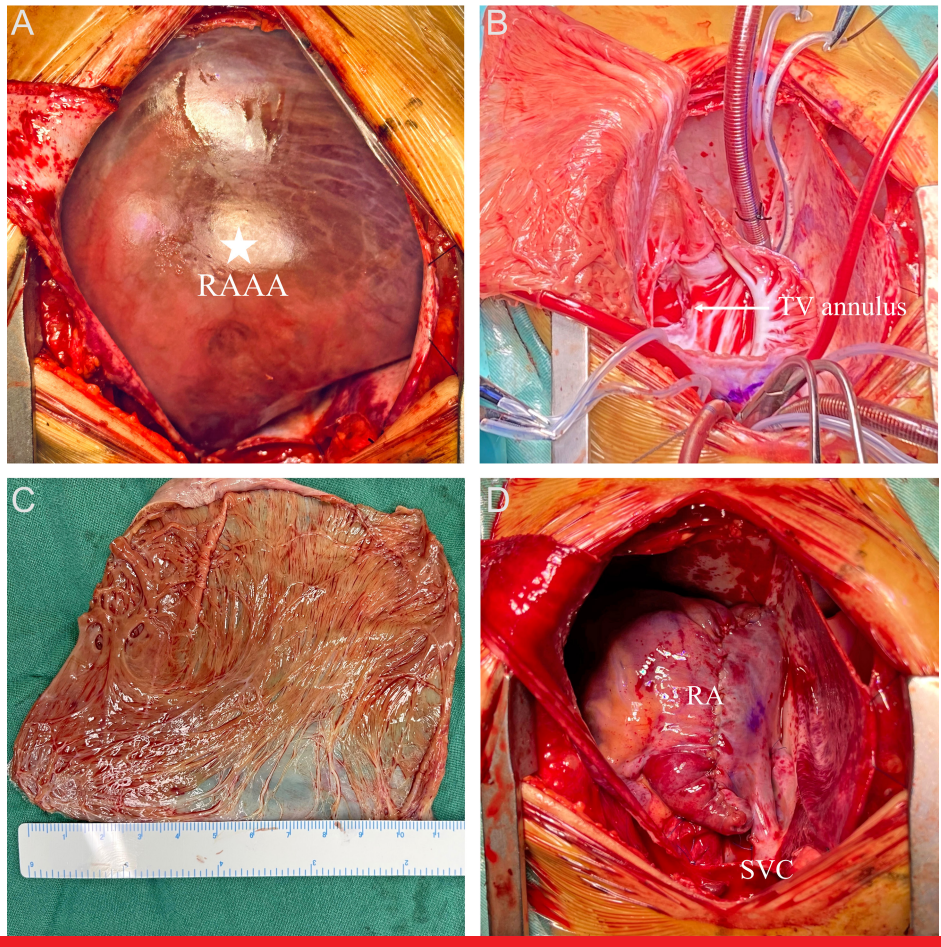

Figure 2. (A) Intraoperative view after a median sternotomy. The thoracic cavity was mostly occupied by the extremely enlarged right atrial appendage. (B) Intraoperative view after opening the right atrium. The thin atrial wall of the right atrium and a dilated tricuspid valve annulus were revealed. (C) The tissue of the right atrium aneurysm. The removed specimen was about 11 cm × 10 cm in size. (D) Intraoperative view after right atrium reconstruction. The adjacent structure, including the superior vena cava, could be observed after RAAA removement. RAAA, right atrial appendage aneurysm; TV annulus, tricuspid valve annulus; RA, right atrium; SVC: superior vena cava.

Right atrial appendage aneurysm, a rare cardiac malformation, is divided into 2 subtypes: (i) congenital RAAA due to dysplasia of the atrial muscles; (ii) acquired RAAA led by other structural cardiac diseases such as rheumatic heart disease. Tricuspid regurgitation has seldom been reported in RAAA cases, which might be either the cause or the consequence. Considering the patient's age, medical history, and intact structure of the tricuspid valve, the RAAA was more likely to be congenital, and TR was presumably related to distortion of the annulus. Therefore, annuloplasty was performed to minimize regurgitation.

Informed Consent: Consent was obtained from the patient's family.

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Video 1: Saline test after opening the right atrium showing obvious tricuspid regurgitation.

Video 2: Saline test after annuloplasty showing minimized tricuspid regurgitation.