Journal of Clinical and Experimental Research in Cardiology
Volume 6| Issue 1
ISSN: 2394-6504

Case Report Open Access

Giant Right Atrial Aneurysm and Right Atrial Appendage Aneurysm with Atrial Septal Defect and Atrial Fibrillation

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Citation: Enshi Wang, Xuesong Fan, Yunhu Song, Hongbin Wu (2022) Giant Right Atrial Aneurysm and Right Atrial Appendage Aneurysm with Atrial Septal Defect and Atrial Fibrillation. J Clin Exp Res Cardiol 6(1): 102

Abstract

The study was approved by the The Medical Ethics Committee of the Cardiovascular Institute and our Hospital. All methods were performed in accordance with relevant guidelines and regulations. The written informed consent was obtained from the patients.

Keywords: Atrial, Septal Defect, Atrial Fibrillation, Cardiology

Introduction

The right atrial aneurysm (RAA) and right atrial appendage aneurysm (RAAA) were rare cardiac anomalies. The RAA was firstly surgically corrected by Bailey in 1955 [1]. Binder and his colleges reported 17 patients in 60 RAAs were surgically repaired [2]. 48% of the RAA and RAAA were asymptomatic.[2] However, atrial arrhythmia, right atrial thrombosis, tricuspid insufficiency, and heart failure developed in some RAA or RAAA patients. In this case, one patient diagnosed as RAA, RAAA, atrial septal defect (ASD), and atrial fibrillation (AF) at the same time undergone surgery in our hospital.

Case

The patient was a 38-year-old female, who was admitted into our hospital for palpitation and chest tightness after activity since one year ago. The cardiothoracic ratio was 0.75 in chest X-ray (Figure 1A). The electrocardiography (ECG) suggested atrial fibrillation and incomplete right bundle branch block (Figure 1B). The patient's exercise was limited. Even one simple action such as getting up from the bed could cause the AF rate increasing from 70-80 bpm to 170-180 bpm and the patient felt severe palpitation. The echocardiography showed right atrial and appendage dilation (90.2mm×85.7mm and 59.2×49.5mm in dimensions respectively) (Figure 1C, 1D), two central type ASDs of 2mm and 10mm, and tricuspid annulus compression by the enlarged RAA (the dimension of tricuspid annulus was 28 mm during systolic period and 19mm during diastolic period). The following computed tomography (CT) and magnetic resonance imaging (MRI) revealed the similar results (RAA 129.6mm×82.3mm) (RAA 153×87×116mm).

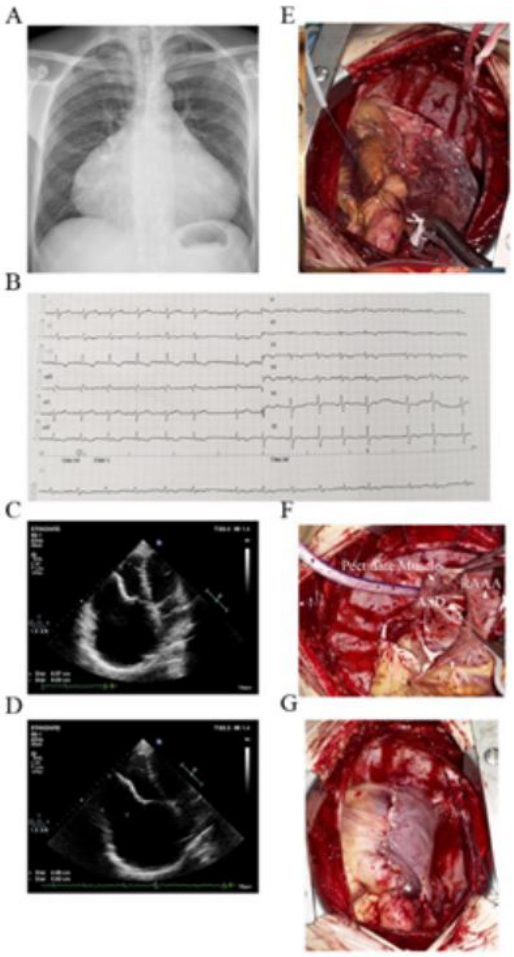

Figure 1: Figure 1A, chest X-rays before operation; Figure 1B, the ECG before operation; Figure 1C, dimensions of RAA in echocardiography before operation; Figure 1D, dimensions of RAAA in echocardiography before operation; Figure 1E, the overall look of RAA and RAAA after pericardiotomy; Figure 1F, the inside appearance after RAA and RAAA opening; Figure 1G, the overall look of heart after RAA and RAAA surgically corrected

The right femoral artery and vein were used to establish cardiopulmonary bypass (CPB) by open puncture approach. The dilated RAA and RAAA almost occupied the anterior and the diaphragmatic surface of the heart (Figure 1E). The right atrioventricular groove (RAVG) was pushed to the left side (Figure 1F). After full flow of CPB, the RAA and RAAA was completely collapsed. 2 ASDs located in the central atrial septum (2mm and 10mm respectively) (Figure 1F). The RAA wall was resected along the left side (5 cm away from the RAVG) and the right side (the boundary of the pectinate muscle and the membranous aneurysm wall). The front wall of the RAAA was resected too. One 5/0 prolene continuous suture closed the ASDs. Two 6/0 prolene continuous sutures closed the right atrial and appendage incision (Figure 1G). The resected RAA wall is larger (12.5cm×8.0cm) and thinner (0.1-0.2mm thin) than the RAAA wall (5.8cm×2.6cm×0.5cm) (Figure 2A). In pathological examination, most of the atrial wall was thinned, the local myocardial cells were atrophied and disappeared, myocardial fibrosis, and local fibrous tissue hyperplasia and thickening of endocardium were observed (Figure 2B, 2C). The sinus rhythm recovered after operation and persisted until discharge from hospital (Figure 2D).

The X-ray showed diminished cardiothoracic ratio of 0.50 (Figure 2E). Echocardiography suggested distinct dimension reduction of the right atrium (52.4mm×42.0mm) (Figure 2F) and right atrial appendage (25.6mm×17.1mm) (Figure 2G).

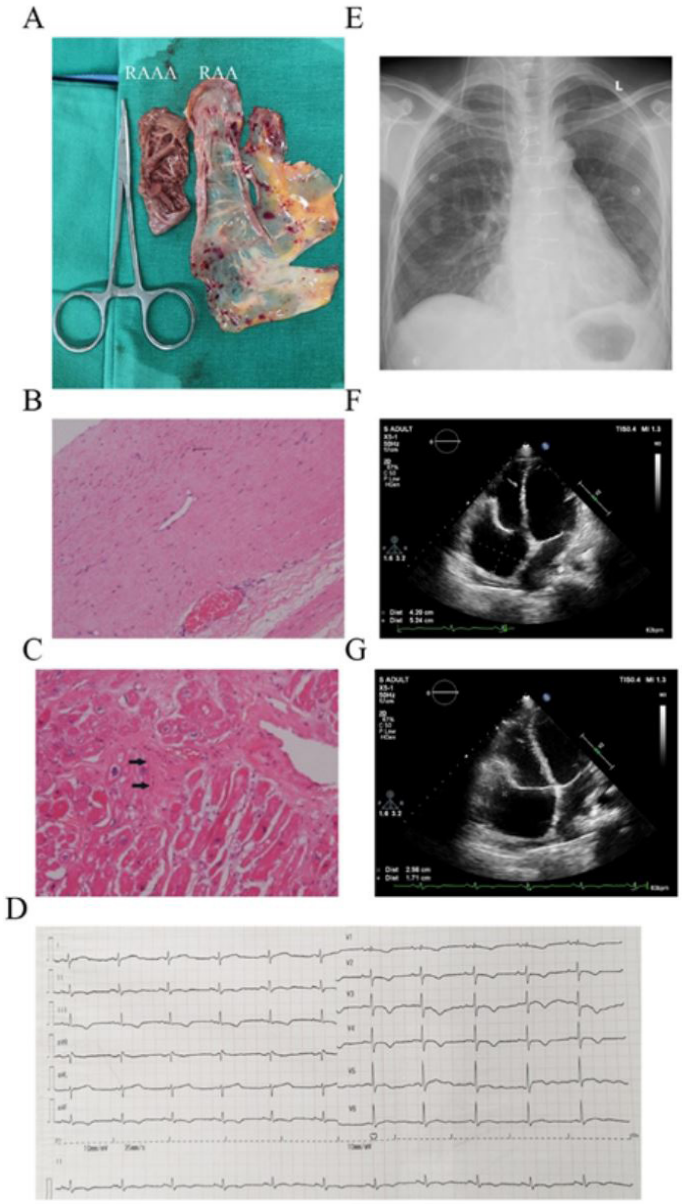

Figure 2: Figure 2A, the specimens resected from RAA and RAAA; Figure 2B, the pathological examination of RAA in HE staining (10×); Figure 2C, the pathological examination of RAAA in HE staining (10×); Figure 2D, the ECG after operation; Figure 2E, chest X-rays after operation; Figure 2F, dimensions of RAA in echocardiography after operation; Figure 2G, dimensions of RAAA in echocardiography after operation

Discussion

RAA and RAAA are all rare cardiac malformations, while patients with RAA and RAAA concomitantly were rarer still. In this case, the female patient suffered from both of them. This supplied a special vision for us to compare the RAA and RAAA in the same patient. In this case, the RAA was larger in dimension and thinner in aneurysm wall than the RAAA. From surgical finding, the RAA wall was mainly compose of fibrosis while the RAA was constituted by atrial muscular macroscopically. Then in the microscopical level, most of the myocardial cells in the RAA wall were atrophied and replaced by the fibrous tissue hyperplasia. The same histological finding was also seen in the RAAA wall, but the change of RAAA was focal. The similar histological change in the RAAA was also found in other report [3]. This comparison revealed that the RAA has severer pathological change than the RAAA.

The ASD is not large enough to cause the dilation of right atrium and right atrial appendage. The reason of RAA and RAAA was mostly the dysplasia of the muscular wall of the RA. In this case, we found similar pathological findings in RAA and RAAA that showed dysplasia of atrial muscular and myocardial fibrosis, which could interpret the enlargement of the right atrium and right atrial appendage.

The RAA and RAAA could be asymptomatic in the early time, but in the third and fourth decades of life, the main manifestations of this disease appear, which result from thromboembolism, right heart failure, or refractory atrial arrhythmia, particularly atrial flutter/fibrillation. The symptoms of this patient was mainly caused by AF. The sinus rhythm restored from AF since spontaneous recovery of heart beat and remained until discharge. It suggested that the preoperative AF mainly caused by the RAA and RAAA. When the RAA and RAAA resected, the sinus rhythm restored. In another report, the original atrial tachyarrhythmia was successfully abolished after RAAA surgical resection in four patients [4]. This case and other reported cases before validated that most of the atrial or supraventricular arrhythmia could be abolished after surgical repair.

Conclusion

RAA or RAAA mainly displayed as the enlargement of the right atrium or right atrial appendage due to the dysplasia of atrial muscular and fibrosis of aneurysm wall. For RAA or RAAA patients with symptoms, especially with the atrial or supraventricular arrhythmia, surgical repair is safe and effective and should be the priority recommendation.

Funding

This work was supported by the National Natural Science Foundation of China (Grants no. 81400242) and Beijing excellent talents training support (2018000021469G243).

Conflict of interest

The authors report no relationships that could be construed as a conflict of interest.

Data availability statement: All data are available as required

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