

Right Atrial Aneurysm with Supraventricular Tachycardia and Frequent Atrial Premature: A Case Report

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Case report

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Abstract

Background: Right atrial aneurysm (RAA) is a rare form of congenital heart disease with a wide range of clinical presentation varying from asymptomatic patients to those with refractory atrial arrhythmias. Diagnosis is often confused with other causes of right atrial dilation such as Ebstein disease or idiopathic right atrial enlargement. **Case presentation:** We present the case of right atrial aneurysm with supraventricular tachycardia and frequent atrial premature in our institution and discuss the diagnostic challenges, the research progress of right atrial aneurysm and follow-up situation.

Conclusion: RAA should be considered in the differential diagnosis of RA enlargement. Accurate diagnosis can be achieved by echocardiograph, and our report also highlights the importance of longitudinal follow-up of RAA over 5 years and the long-term regular conservative treatment for patients with supraventricular tachycardia and frequent atrial premature could be useful.

Introduction

Right atrial aneurysm (RAA) is a rare form of congenital heart disease defined as an isolated enlargement of the right atrium in the absence of known additional cardiac lesions which causes right atrial dilatation. With development of ultrasound technology and advancing ultrasonic machine, the accurate diagnosis of RAA has been made more frequently with echocardiography. However, related case reports in the current literature describe a highly variable presentation, and the management of RAA remains controversial and varies between different medical centers and areas.

Case Presentation

A 27-year-old male patient was admitted to our institution with a history of paroxysmal palpitation for 11 years, and progressively exacerbated one day. Physical examination revealed normal blood pressure (115/75 mmHg), no abnormal cardiac murmur and jugular venous distention. ECG showed supraventricular tachycardia and frequent atrial premature. In 2015, the patient underwent intracardiac radiofrequency ablation, right bypass with atrioventricular reciprocating tachycardia (AVRT), a typical atrial flutter were identified using electrophysiological testing, and catheter radiofrequency ablation was successfully for treating the right bypass. In the follow-up examination after radiofrequency ablation, he was diagnosed as Ebstein's anomaly in community hospital. In 2018, in order to get more accurate diagnosis, he came to our department. Transthoracic echocardiography revealed the right atrium was enlarged and the anterior leaflet of the tricuspid valve was displaced downward into the right ventricle (Fig. 1). The distance between the attachment of tricuspid diaphragm and anterior mitral valve was 30 mm. However, the morphology of tricuspid valve appeared normal, and the size and function of both ventricles were normal. There was mild tricuspid valve regurgitation without stenosis. The peak flow velocity of tricuspid regurgitation was 2.6 m/sec. In the apical four-chamber view, as an outpouching of

the RA was proximal to the normally appearing TV leaflets. In the non-standard subxiphoid four-chamber view and oblique parasternal four-chamber view, there was an obvious echo drop-out in RA lateral wall and extrinsically impinging onto the RV free wall. To further clarify this cardiac abnormality, the patient was conducted a contrast-enhanced ultrasound examination of cardiac, which demonstrated a saccular aneurysm of the lateral free wall of the right atrial measuring 53mm*27 mm with intense spontaneous echo contrast (Fig. 2), but thrombus was not found in any part of the heart. Similar findings were visualized on the transesophageal echocardiogram (Fig. 3). We compared the echocardiographic images of the patient in 2015, which showed that the right atrium was enlarged, with normal tricuspid anatomical attachment and no local exocoele of right atrial wall. In most cases, it's recommended to reduce the size of the RA surgically in order to prevent thrombus formation in the RA and prevent embolism. However, the patient did not accept surgery. From 2018 to now, he was asymptomatic with clinically follow-up and a long-term regular anticoagulation therapy.

Discussion

- RAA is an extremely rare congenital cardiac condition that can present anytime between fetal life and old age^{1,2}. Most right atrial aneurysm manifested no obvious symptoms³. However, atrial fibrillation, supraventricular arrhythmias(in this case) and intracardiac thrombus may occur^{2,4-5}. Our report details a 6-years disease progression with a single RAA who was symptomatic with supraventricular tachycardia and no history of embolic event.

The etiology of RAA is still unclear, it has been speculated that an intrinsic structural protein defect, dysplastic pectinate muscle or abnormal collagen were prone to dilate the right atrial⁶.

Histological changes of resected aneurysmal tissue had revealed a variety of changes adjacent to entirely normal findings, including fibrosis, focal lymphocytic infiltration and focal myxoid changes¹.

Abnormal enlargement of the RA was often confused with other more common conditions such as Ebstein's anomaly⁷, diverticulum and idiopathic right atrial enlargement. As in our case, it is difficult to distinguish Ebstein's anomaly from idiopathic right atrial enlargement, when the whole right atrium was enlarged. But when the right atrium side wall locally expanded and pushed the corresponding anterior tricuspid valve ring, resulting in the false downward movement of the anterior leaflet, it was easily misdiagnosed for Ebstein's anomaly, especially the insertion points of mitral and tricuspid valves in the apical four-chamber view as the distance between these two points greater than 15 cm. However, differential diagnosis can be made by carefully observing the structure of the right atrial wall and the relationship between the out pouching of right atrial and tricuspid valve.

The natural course of RAA remains unclear, but reported complications included thrombus formation in the RA cavity, predisposing to thromboembolic complications such as stroke and pulmonary embolism. Arrhythmias and atrial rupture are the other potential complications.

Most authorities recommended surgical resection in the patients with marked RA enlargement complicated by atrial fibrillation, evident thrombus formation or signs of pulmonary embolization. At present, there is no unified recommended treatment standard in the world. In general, conservative treatment is recommended for asymptomatic patients⁸. Patients with congenital heart defects, or large and symptomatic tumors were recommended surgery. But some patients with small and symptomatic tumors, radiofrequency ablation can be tried, and the effect is also acceptable. However, the current histological findings showed right atrial wall of right atrial aneurysm was thin and degenerative, so the risk of perforation is higher than normal ventricle or atrium during ablation. According to these recommendations, he had received radiofrequency ablation to maintain his sinus rhythm five-year ago, now the best therapy of choice was to reduce the size of RA surgically, but he was received a long-term regular anticoagulation therapy, because she did not accept surgery.

Conclusion

RAA should be considered in the differential diagnosis of RA enlargement. Accurate diagnosis can be achieved by echocardiograph, and our report also highlights the importance of longitudinal follow-up of RAA over 5 years and the long-term regular conservative treatment for patients with supraventricular tachycardia and frequent atrial premature could be useful.

Abbreviations

Right atrial aneurysm (RAA)

atrioventricular reciprocating tachycardia (AVRT)

electrocardiogram (ECG)

right atrium (RA)

right ventricle (RV)

Declarations

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Figures

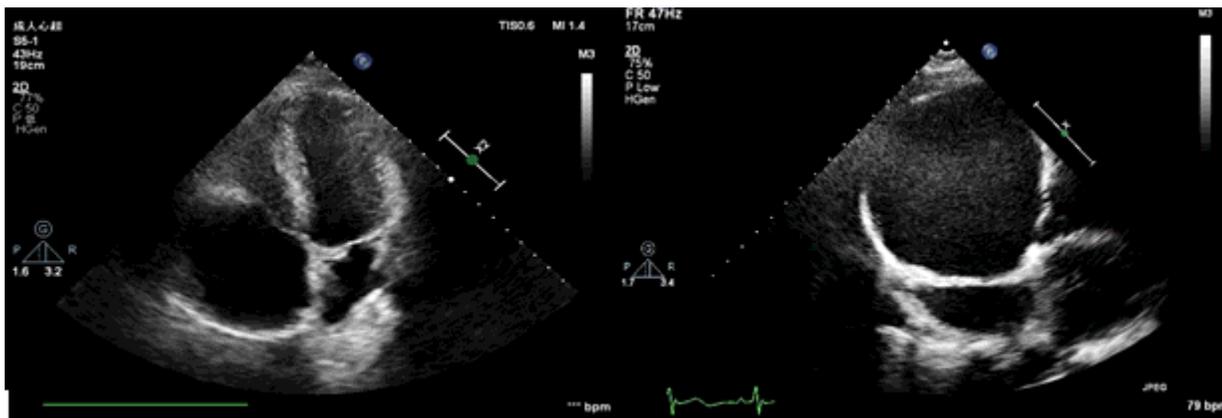


Figure 1

Apical four-chamber view and subxiphoid four-chamber view: anterior leaflet of the tricuspid valve is displaced downward into the right ventricle, the right atrium was enlarged and the attachment of tricuspid anatomical was normal.

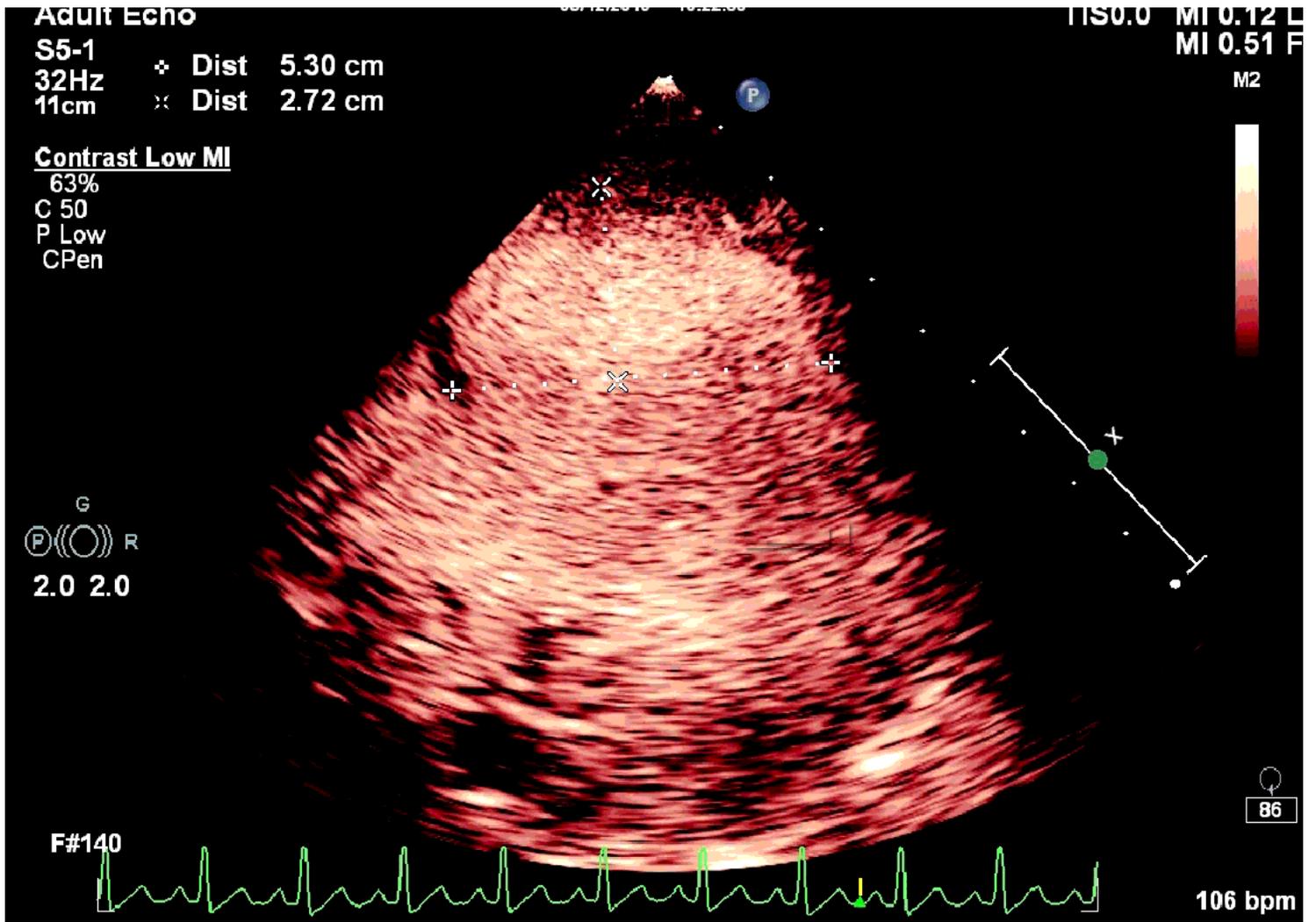


Figure 2

Contrast-enhanced ultrasound, Nonstandard subxiphoid four-chamber view demonstrated an aneurismal.

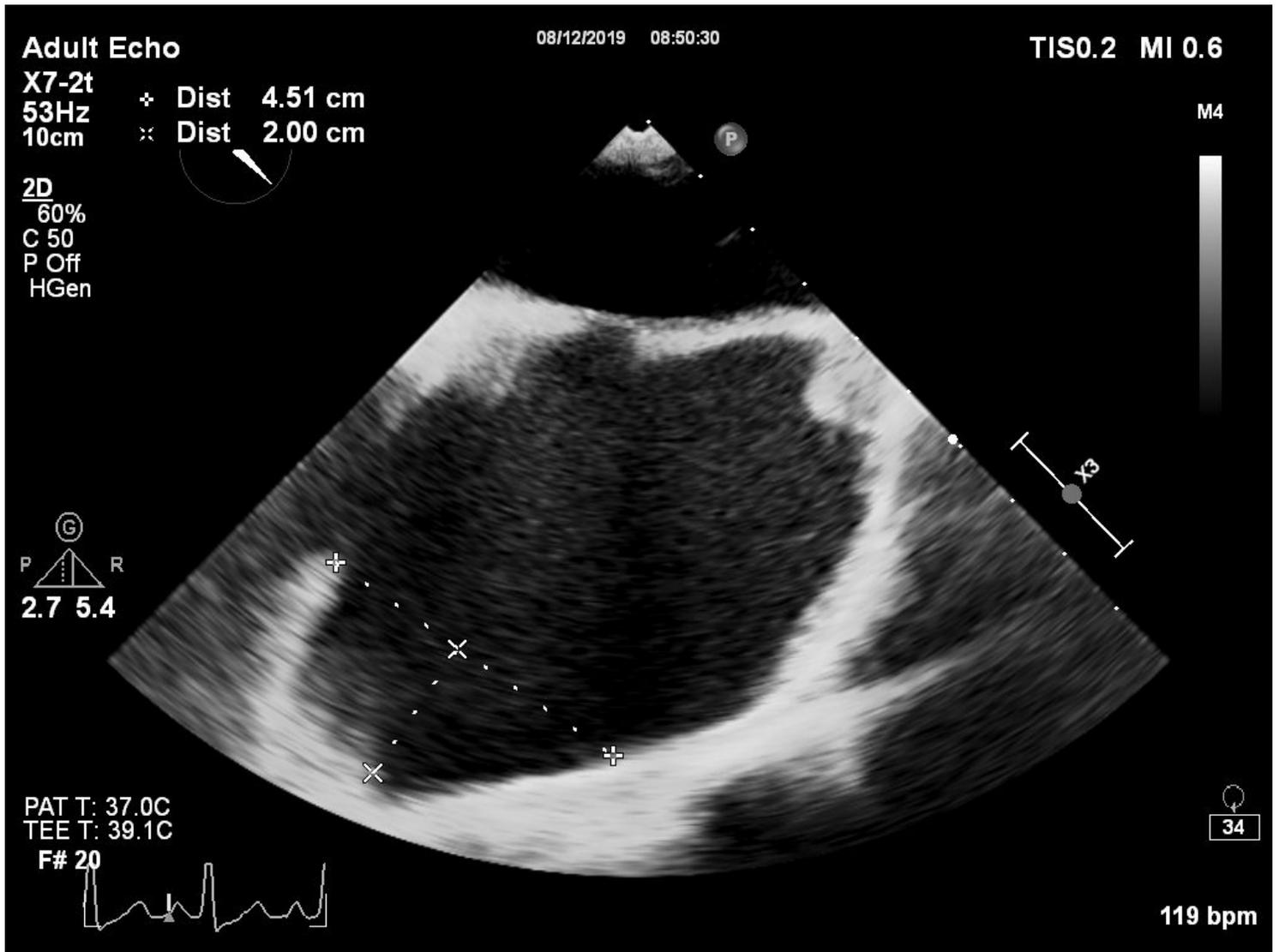


Figure 3

transesophageal echocardiogram demonstrated an aneurysmal cavity.

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