Arrhythmogenic Right Ventricular Dysplasia with Right Atrial Thrombus

Displasia Arritmogênica do Ventrículo Direito com Trombo em Átrio Direito

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Abstract

Arrhythmogenic right ventricular dysplasia is an autosomal dominant genetic disease characterized by the progressive replacement of the myocardium with fibrous fat tissue. It is clinically characterized by arrhythmia, heart failure, syncope and, in some cases, sudden death. We report the case of a patient with such pathology, at an advanced stage, with atrial flutter and thrombus formation in the right atrial appendage, whose therapeutic option was anticoagulation and subsequent electrical cardioversion.

Introduction

Arrhythmogenic right ventricular dysplasia (ARVD) is a genetic cardiomyopathy triggered by mutations in the genes that code for desmosomes.[1] These mutations lead to apoptosis of cardiac muscle cells, causing them to be later replaced by progressive fatty infiltration.[2] This predisposes patients to an arrhythmogenic condition, causing sudden death, and ventricular and supraventricular arrhythmia.[3,4] Supraventricular arrhythmias are present in 25% of patients with ARVD who have arrhythmias.[2] However, the incidence of thromboembolic complications is very low.[4]

Case report

A 54-year-old male patient who had an episode of arrhythmic syncope at the age of 25 during a soccer game was investigated and diagnosed with arrhythmogenic right ventricular dysplasia. Implantable cardioverter defibrillator (ICD) was implanted. The patient presented dilatation of right chambers, severe right ventricular thinning and dysfunction, maintaining functional class 2–3 despite optimized clinical therapy. In November 2017, symptoms got significantly worse, when the patient had atrial flutter. During that period, heart rate control was attempted through adjustment of medication and anticoagulation. On the same occasion, transesophageal echocardiography (TEE) found the presence of a 2.5 x 3 cm thrombus in the right atrial appendage (Figure 1). Anticoagulation with 150 mg dabigratan twice a day for 30 days was administered and TEE was repeated (Figure 2).

After 30 days of anticoagulation, there was thrombus resolution with spontaneous contrast in the right atrium only. Electrical flutter cardioversion was performed and the patient presented improvement of symptoms and concomitant quality of life. Patient remains on optimized medication and anticoagulation and is evaluated for queuing for heart transplantation.

Discussion

We report the case of a patient with ARVD and atrial flutter with right atrial thrombus.

Incidence/prevalence of ARVD in the general population ranges from 1: 2,000 to 1: 5,000 people, being more common in men, youth people and athletes.[2,3,5]

In decreasing order of frequency, supraventricular arrhythmias in these patients include: atrial fibrillation, tachycardia and atrial flutter.[2] It is speculated that most of these patients have a low CHADS2 or CHA2DS2-VASc score, representing low risk of thromboembolism in patients with AF.[6] Patients with ARVD are typically younger, therefore, the prevalence of hypertension and diabetes mellitus is lower than in the general population of AF.[3]

One of the complications presented by patients with such pathology is thrombi formation as, with the replacement of myocytes by fibrous fat tissue, an abnormality in cardiac motility is installed, predisposing to their formation. This complication is believed to have an incidence of 0.5 to 100 patients, and is even more common in patients with more extensive cardiac involvement.[4,7] Most of these thrombi are located at the apex of the right ventricle in a proportion of 7:10.

In the literature, there have been only a few reports of right atrial thrombi in patients with ARVD.[8] In this case, atrial thrombus was detected in the right atrium, not in the left atrium. The incidence of left atrial thrombi in patients with AF/FL has been widely investigated. However, so far, little attention has been paid to right atrial thrombi in these patients.[7]

Several reports indicate that right atrial appendage thrombi is detected in 0.7% to 2.4% of patients with AF/FL. However, it is less frequent than in the left atrial appendage.[9]

In patients with ARVD complicated by supraventricular

Keywords

Arrhythmogenic Right Ventricular Dysplasia; Thrombosis; Atrial Flutter; Heart Transplantation.

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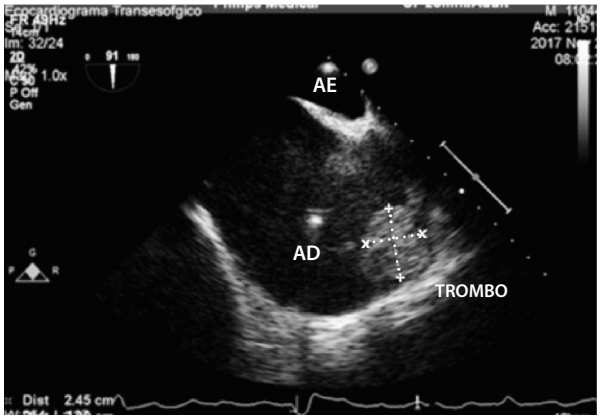

Figure 1 – *2.5 x 3 cm thrombus in right atrial appendage.*

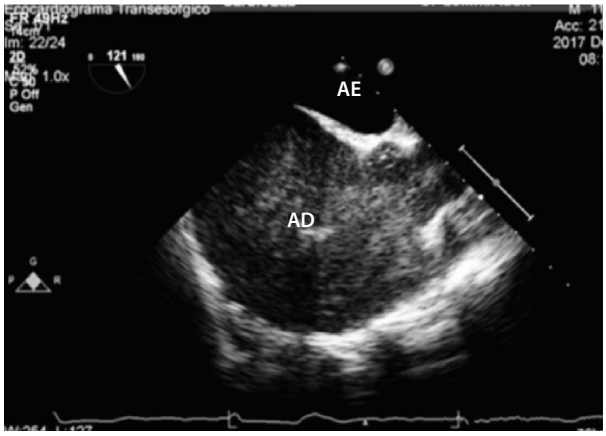

Figure 2 – *After 30 days of anticoagulation, the thrombus was resolved, despite the presence of very significative spontaneous contrast.*

tachyarrhythmia, the risk of thrombus formation may be higher in the right atrium than in the left atrium.[8]

Right atrial thrombus formation can lead to a fatal thromboembolic complication.[7] Anticoagulation and restoration of sinus rhythm should be considered to prevent this complication and prevent worsening of ventricular function.[7]

Authors contribution

Data acquisition: Bolonhez AC, Mangili OC, Santos CB. Data analysis and interpretation:

Santos CB. Manuscript writing: Santos CB. Critical revision of the manuscript for important in-tellectual content: Bolonhez AC.

References

1. Corrado D, Link MS, Calkins H. Arrhythmogenic Right Ventricular Cardiomyopathy. N Engl J Med. 2017;376(1):61-72.
2. Feitosa Filho FH, Lino DOC, Néri AKM, Queiroz REB, Sales M. Cardiomiopatia arritmogênica do ventrículo direito. Rev. Brasileira de Clinica medica. 2011;9(1):67-73.
3. National library of Medicine: Arrhythmogenic right ventricular dysplasia, atualizado em novembro de 2018. Acesso em: 01 de Agos 2019. Disponível em: <https://www.dynamed.com/topics/dmp~AN~T116117>
4. Kemmner S, Lesevic H, Reents T, Schunkert H, C. Right ventricular thrombus formation in a patient with arrhythmogenic right ventricular dysplasia following radiofrequency ablation. Clin Case Rep. 2016;4(6):554–7.

5. Mckenna WJ, Calkins H, Downey BC. Arrhythmogenic right ventricular cardiomyopathy: Anatomy, histology, and clinical manifestations, 2018. Acesso em: 25 de Mar 2018. Disponível em: https://www.uptodate.com/contents/arrhythmogenic-right-ventricular-cardiomyopathy-anatomy-histology-and-clinical-manifestations

6. Lip GY, Nieuwlaat R, Pisters R, Lane DA, Crijns HJ. Refining clinical risk stratification for predicting stroke and thromboembolism in atrial fibrillation using a novel risk fator- based approach. Chest. 2010 Feb;137(2):263-72.

7. Wlodarska EK, Wozniak O, Konka M, Rydlewska-Sadowska W, Biederman A, Hoffman P. Thromboembolic complications in patients with arrhythmogenic right ventricular dysplasia/cardiomyopathy. Europace. 2006 Aug;8(8):596-600.

8. Kazmierczak J, Kornacewicz-Jach Z, Wojtarowicz A. Atrial Epicardial Pacing with long stimulus to P wave interval in a patient with Arrhythmogenic right ventricular dysplasia complicates by right atrial Thrombosis. PACE. 1998;22.

9. Mckenna WJ. Arrhythmogenic right ventricular cardiomyopathy: Treatment and prognosis, 2018. Acesso em: 25 de Mar 2018. Disponível em https://www.uptodate.com/contents/arrhythmogenic-right-ventricular-cardiomyopathy-treatment-and-prognosis#H17