Atypical Clinical Presentation of Arrhythmogenic Biventricular Cardiomyopathy

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Introduction

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an inherited disease affecting the heart muscle. It is clinically characterized by life-threatening ventricular arrhythmias. Although it primarily affects the right ventricle, the left ventricle might also be involved[1,2]. However, the clinical manifestations and prognosis of biventricular arrhythmogenic cardiomyopathy not yet been established. We present a case of ARVC with left ventricle (LV) involvement, with no typical symptoms and no relevant medical history.

Case Report

A 57-year-old hypertensive woman presented with a 6-month history of atypical chest pain. In addition, ongoing maintenance therapy with azathioprine for Crohn's disease and a nonfunctional adrenal adenoma were observed. Family history was unremarkable, and physical examination results were normal.

Electrocardiography revealed a normal sinus rhythm, left axis deviation, and T-wave inversion in precordial leads from V2 to V6. Further, an epsilon wave in leads V1–V3 (Figure 1) led to the suspicion of ARVC. Monitoring using a 24-h Holter electrocardiogram documented only occasional ventricular extrasystoles with left bundle branch block morphology and no significant cardiac arrhythmia. Further evaluation included cardiac magnetic resonance (CMR) imaging, which revealed right ventricular (RV) enlargement with an indexed end-diastolic volume of 110 ml/m^2 and mild dysfunction (ejection fraction = 44%). No signs of fatty tissue infiltration were observed, but regional RV dyskinesia in the free wall and outflow tract was identified (Figure 2A). LV was mildly enlarged (90 ml/m^2), and systolic function was at the lower limit of normal. Flow analysis revealed no shunts in the aorta and pulmonary artery. Subepicardial delayed enhancement was observed the lateral basal and middle segments of LV as well as a small focal area of the RV free wall (Figure 2B). These findings suggested a diagnosis of ARVC with LV involvement.

In this case involving a patient with atypical symptoms, no clinical arrhythmia, and a negative family history, three major criteria led to the definite diagnosis of ARVC: inverted T waves in the precordial leads in the absence of complete right bundle branch block; epsilon wave in the right precordial leads; and regional RV dyskinesia with indexed end-diastolic volume ≥ 100 ml/m^2.

LV involvement in ARVC has been recognized in several studies and can occur in >75% patients with disease progression[1]. CMR is an ideal technique to aid in the diagnosis of this condition[3]. Particularly, in asymptomatic patients, the differential diagnosis of ARVC should be considered. Future investigations should clarify the clinical relevance of these findings and the prognosis of patients with biventricular arrhythmogenic cardiomyopathy.

Author contributions

Conception and design of the research: Rangel I; Acquisition of data: Rangel I, Madureira AJ; Analysis and interpretation of the data: Vasconcelos M, Campelo M, Frutuoso C, Madureira AJ; Writing of the manuscript: Rangel I, Vasconcelos M; Critical revision of the manuscript for intellectual content: Vasconcelos M, Campelo M, Frutuoso C, Madureira AJ, Maciel MJ.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any post-graduation program.

Keywords

Arrhythmogenic Right Ventricular Dysplasia / complications; Arrhythmogenic Right Ventricular Dysplasia / diagnosis; Heredity, Death, Sudden, Cardiac.

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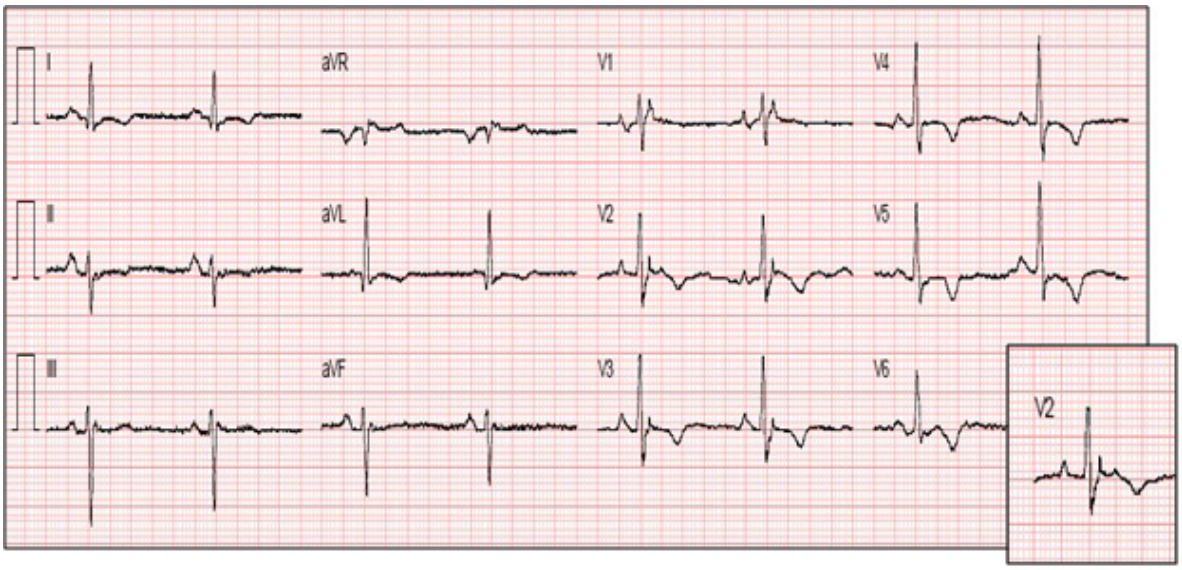

Figure 1 - *Resting 12-lead ECG showing T-wave inversion from V2 to V6 (major diagnostic criteria) and epsilon waves in V1–V3 (major diagnostic criteria).*

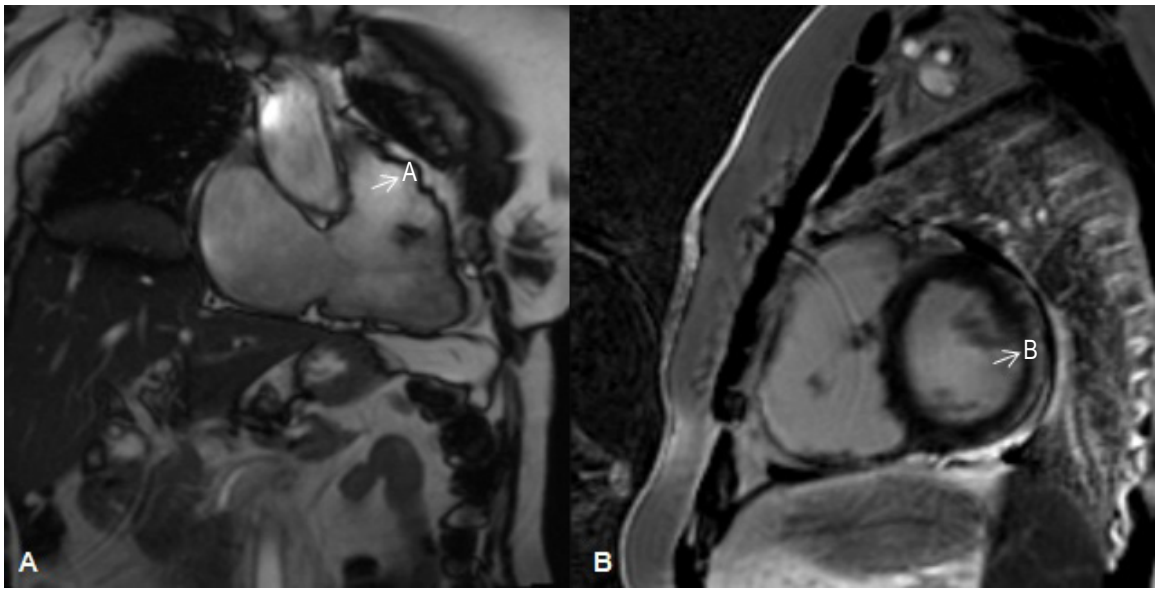

Figure 2 - A) *Steady-state free precession cine magnetic resonance imaging (RV long axis view) indicated regional RV dyskinesia (arrows) at end-systole.* **B)** *CMR imaging acquired 10 min after gadolinium injection (0.1 mM). Note the subepicardial late enhancement in the LV lateral wall (arrows) proximal to a small area of late enhancement in the RV.*

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