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Original article

Arrhythmogenic Cardiomyopathy. Patterns of Ventricular Involvement Using Cardiac Magnetic Resonance

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ABSTRACT

Introduction and objectives: Biventricular arrhythmogenic cardiomyopathy and left dominant arrhythmogenic cardiomyopathy forms had recently been included in the spectrum of arrhythmogenic cardiomyopathy. The aim of the study was to describe, using cardiovascular magnetic resonance, the patterns of ventricular involvement as well as late gadolinium enhancement in these conditions.
Methods: Medical databases and records from the cardiology units of 3 hospitals were reviewed to obtain data from patients with arrhythmogenic cardiomyopathy.
Results: Twenty-six consecutive patients were included (40 [16] years, 16 males). Right ventricle involvement was present in 19 patients (73%). Among them, 13 patients (50%) had volumes over the upper limit of normality, 11 (42%) patients had late gadolinium enhancement in right ventricle and 6 patients (23%) had just mild involvement with wall motion abnormalities or microaneurysms. Left ventricle involvement was present in 24 patients (92%), all of them with late gadolinium enhancement. In 15 patients (57%) left ventricular systolic dysfunction was observed, and dilatation in 3 patients (11%). Late gadolinium enhancement was more frequent in the inferior, lateral, and inferolateral walls (65%, 57%, and 61% of patients, respectively) while septum was seldom affected (26% of cases). The pattern of late gadolinium enhancement was mainly subepicardial (46% of patients) or transmural (19%), and was intramyocardial in only 12% of the cases.
Conclusions: In this sample, left ventricle involvement is very common. The most frequent finding was late gadolinium enhancement, while the least frequent was dilatation. The pattern of late gadolinium enhancement was more frequently subepicardial and located in the inferior and inferolateral walls.

Resonancia magnética cardiaca en miocardiopatía arritmogénica. Tipos de afección y patrones de realce tardío de gadolinio

Palabras clave:
Miocardiopatía arritmogénica
Cardiorresonancia magnética
Realce tardío de gadolinio

RESUMEN

Introducción y objetivos: La miocardiopatía arritmogénica biventricular y la miocardiopatía arritmogénica izquierda han sido incluidas recientemente en el espectro de la miocardiopatía arritmogénica. El objetivo del estudio es describir con cardiorresonancia magnética el tipo de afección observada y describir los patrones de realce tardío de gadolinio.
Métodos: Se revisaron las bases de datos y la historia clínica informatizada de tres hospitales, para obtener datos de enfermos consecutivos con miocardiopatía arritmogénica.
Resultados: Se incluyó a 26 pacientes consecutivos, con una media de edad de 40 ± 16 años, de los que 16 eran varones (67%). Se observó afección de ventrículo derecho en 19 pacientes (73%), con volúmenes aumentados en 13 pacientes (50%), 11 pacientes (42%) con realce tardío de gadolinio en ventrículo derecho y 6 (23%) presentaban únicamente alteraciones de la contractilidad segmentaria. Se observó afección de ventrículo izquierdo en 24 pacientes (92%), todos con realce tardío de gadolinio; 15 pacientes (57%) presentaron disfunción sistólica ventricular izquierda. En 3 pacientes (11%) se observó dilatación de ventrículo izquierdo y ninguno de ellos fue diagnosticado de miocardiopatía arritmogénica izquierda. La localización del realce tardío de gadolinio fue predominantemente inferior (65%), inferolateral (61%) y lateral (57%), mientras que la localización septal fue menos frecuente (26%). El patrón de realce tardío de gadolinio fue fundamentalmente epicárdico (46%) y transmural (19%), raramente intramiocárdico (12%).
Conclusiones: En esta muestra, la afección del ventrículo izquierdo es muy frecuente; el hallazgo observado en el mayor número de pacientes fue el realce tardío de gadolinio y el menos frecuente, la

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dilatación. El patrón de realce tardío de gadolinio es subepicárdico y afecta a territorios inferior, inferolateral y lateral.

Abbreviations

AC: arrhythmogenic cardiomyopathy
BVAC: biventricular arrhythmogenic cardiomyopathy
LDAC: left-dominant arrhythmogenic cardiomyopathy
LGE: late gadolinium enhancement
RDAC: right-dominant arrhythmogenic cardiomyopathy

INTRODUCTION

In arrhythmogenic cardiomyopathy (AC), myocardial tissue is replaced with fatty and fibrous tissue, initially in the epicardium. This process usually affects the regions posterior and inferior to the right ventricle (RV) entry tract, adjacent to the tricuspid valve. The infiltrate is an electrically unstable substrate that produces phenomena ranging from isolated ventricular extrasystoles to sustained ventricular tachycardia or ventricular fibrillation.[1]

Within the spectrum of AC, biventricular arrhythmogenic cardiomyopathy (BVAC) and left-dominant arrhythmogenic cardiomyopathy (LDAC) phenotypes have been reported.[2]

The introduction of cardiac magnetic resonance imaging (cMRI) in clinical practice has significantly improved assessment of the RV. This imaging technique is the most accurate and reproducible for quantifying volumes and biventricular function. Likewise, detection of late gadolinium enhancement (LGE) after administration of gadolinium-DPTA (gadopentate dimeglumine) acid represents progress in the characterization of myocardial tissue in a number of diseases.[3]

Recent data indicate the importance of LGE for identifying fibroadipose lesions. Tandri et al.[4] first reported the clinical usefulness of these sequences in AC. These investigators found LGE in the myocardium of the RV in 67% of patients with this condition, and this was associated with inducible monomorphic ventricular tachycardia in electrophsyiologic studies.

Subsequently, Sen-Chowdhry et al.[5] demonstrated that finding LGE in the left ventricle (LV) allows a more sensitive and specific diagnosis than LGE in the RV. Recently, an update of the Task Force diagnostic criteria has been published[6] and these now include genetic diagnosis. With the new criteria, more patients with LDAC will be diagnosed. Little has been published with regard to the clinical usefulness of LGE in the LV for diagnosis of AC and particularly in left phenotypes, and therefore it was not included in the 2010 diagnostic criteria.

The objectives of the present study were therefore to describe the type of disease observed with cMRI in patients with AC, describe the prevalence and patterns of LGE, and establish a scoring system to enable a structured diagnosis of the different phenotypes.

METHODS

This was a retrospective study of the databases of the cMRI units (Filemaker Pro, version 8.1, United States) of 3 hospitals (*Hospital Clínico Universitario, Hospital Universitario La Fe, Hospital General Universitario de Valencia*) between 2006 and 2010, and subsequently the computerized medical records of the patients selected from these databases.

Entries compatible with AC were selected from the databases of the cMRI units. The computerized medical records were then examined to extract clinical data (form of presentation, presence of arrhythmias, electrocardiographic [ECG] findings, and Holter-ECG monitoring).

Of the 5685 entries reviewed, 32 had suspected AC. In 6 of these, there was no conclusive clinical diagnosis and they were excluded (3 with severe dilatation and RV dysfunction, 2 with segmental contraction abnormalities and mild RV dysfunction, and 1 with RV segmental contraction abnormalities only).

Scoring System Biventricular Involvement

A scoring system for biventricular involvement was drawn up to define the distribution of morphological characteristics and to establish a structured diagnosis of the different phenotypes. To calculate the score, it was first necessary to diagnose single-ventricular involvement (LDAC or right-dominant arrhythmogenic cardiomyopathy [RDAC]) or biventricular involvement from examination of cMRI images. Subsequently, the following was assessed for both ventricles: segmental contraction abnormalities, ventricular dilatation, systolic dysfunction, and LGE.

If these characteristics were observed in the LV, they counted as -1 whereas if they pertained to the RV they counted as –1. The score was obtained by summing all individual components (Table 1). Thus, the presence of balanced characteristics in both ventricles yielded an overall score of 0, that is, purely biventricular involvement, whereas negative values or positive values corresponded to prominently left or right involvement, respectively. For example, LDAC diagnosed only by LGE in the LV would have a score of –1, as would BVAC with segmental contraction abnormality in the RV, LGE in the LV, and systolic dysfunction in the LV. To differentiate between them, they would be denominated BVAC with a score of –1 or an LDAC with a score of –1. The score comprised the diagnosis of single-ventricular or biventricular involvement based on inspection of the cMRI images (LDAC, RDAC, or BVAC) along with the score obtained. This score is not an indicator of severity but rather a morphological indicator of ventricular predominance that enables a structural definition of purely biventricular involvement (score 0), biventricular involvement with left or right predominance (any score other than 0), and single-ventricular forms with limited involvement (score 1, 2, –1, or –2) or multiple characteristics of involvement (score –3, –4, 3, or 4).

Table 1
Morphologic Score of Ventricular Predominance (Sum of Morphological Characteristics)

	LV	RV
Segmental contraction abnormalities	–1	1
Ventricular dysfunction	–1	1
Dilatation	–1	1
Late gadolinium enhancement	–1	1

LV, left ventricle; RV, right ventricle.