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Radiological Case Report / Caso Clínico

Left-Dominant Arrhythmogenic Cardiomyopathy: MRI Diagnosis in a High-Risk Phenotype

Miocardiopatia Arritmogénica Esquerda: Diagnóstico por RM em Fenótipo de Alto Risco

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Abstract

We report a 73-year-old male presenting with right-sided heart failure and atrial flutter with rapid ventricular response. Initial evaluation revealed elevated cardiac biomarkers and high-risk pulmonary embolism, complicated by in-hospital cardiac arrest requiring fibrinolysis.
Following transient clinical improvement, the patient was readmitted with syncope and ventricular arrhythmias, raising suspicion of an underlying arrhythmogenic condition. Cardiac magnetic resonance imaging demonstrated findings consistent with left-dominant arrhythmogenic cardiomyopathy.
Arrhythmogenic cardiomyopathy is a genetically determined disease with a wide phenotypic spectrum, including left ventricular variants that may present with non-specific features and delay diagnosis.[1,2]
This case highlights the key role of cardiac magnetic resonance in tissue characterization and in the detection of atypical forms, with important implications for risk stratification and management.

Keywords

Arrhythmogenic cardiomyopathy; Cardiac magnetic resonance; Arrhythmias.

Resumo

Descreve-se o caso de um doente do sexo masculino, 73 anos, admitido por insuficiência cardíaca de predomínio direito e flutter auricular com resposta ventricular rápida. A avaliação inicial revelou elevação de biomarcadores cardíacos e tromboembolismo pulmonar de alto risco, complicado por paragem cardiorrespiratória intrahospitalar, tendo sido submetido a fibrinólise.
Após melhoria clínica transitória, o doente foi readmitido por síncope e arritmias ventriculares, levantando suspeita de etiologia arrítmica subjacente. A ressonância magnética cardíaca demonstrou achados compatíveis com miocardiopatia arritmogénica com predomínio ventricular esquerdo.
A miocardiopatia arritmogénica é uma doença genética com amplo espectro fenotípico, incluindo variantes de predomínio esquerdo que podem apresentar-se de forma inespecífica e atrasar o diagnóstico.[1,2]
Este caso destaca o papel fundamental da ressonância magnética cardíaca na caracterização tecidular e identificação de formas atípicas, com implicações na estratificação de risco e abordagem terapêutica.

Palavras-chave

Miocardiopatia arritmogénica; Ressonância magnética cardíaca; Arritmias.

Clinical Case

A 73-year-old male patient, with no known cardiovascular risk factors, was admitted for right-sided heart failure (HF) of three weeks' duration. On admission, he presented with atrial flutter with a rapid ventricular response (~150 bpm), elevated NT-proBNP, D-dimer, and troponins. Chest radiography confirmed signs of decompensated HF (Figure 1).
CT pulmonary angiography confirmed acute pulmonary embolism (PE) (Figure 2). Echocardiography revealed severe biventricular dysfunction (LVEF ~27%). During hospitalization, the patient experienced cardiorespiratory arrest (asystole), with return of spontaneous circulation after emergency thrombolysis. Days after discharge, he was readmitted for syncope and episodes of non-sustained ventricular tachycardia. Given the suspicion of an underlying arrhythmic etiology, cardiac magnetic resonance imaging (CMR) was performed (Figure 3).

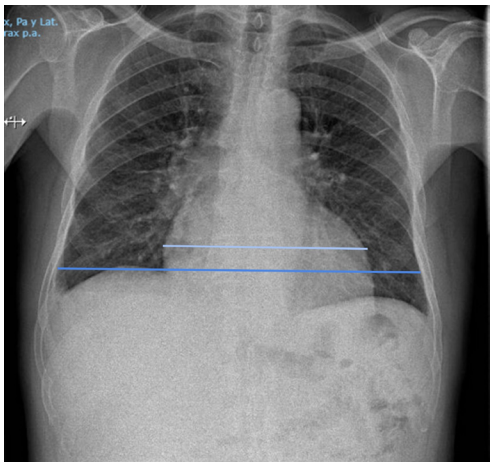


Figure 1 – Chest X-ray (AP): Marked cardiomegaly, signs of pulmonary venocapillary congestion and small bilateral pleural effusion, consistent with decompensated heart failure.

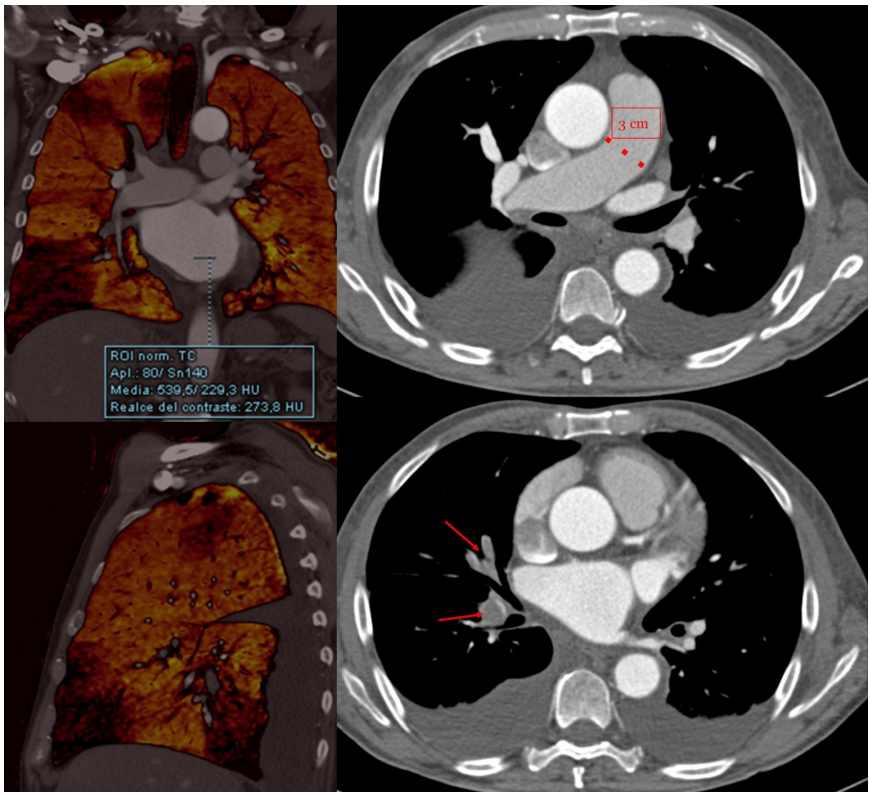


Figure 2 – CT Pulmonary Angiography (PE Protocol) with Iodine Map (Dual Energy): CT angiography and iodine mapping demonstrating filling defects in segmental branches of the right pulmonary artery (red arrows), consistent with acute pulmonary embolism. Dilation of the main pulmonary artery (3 cm) and areas of wedge-shaped hypoperfusion in the corresponding lung parenchyma are observed.

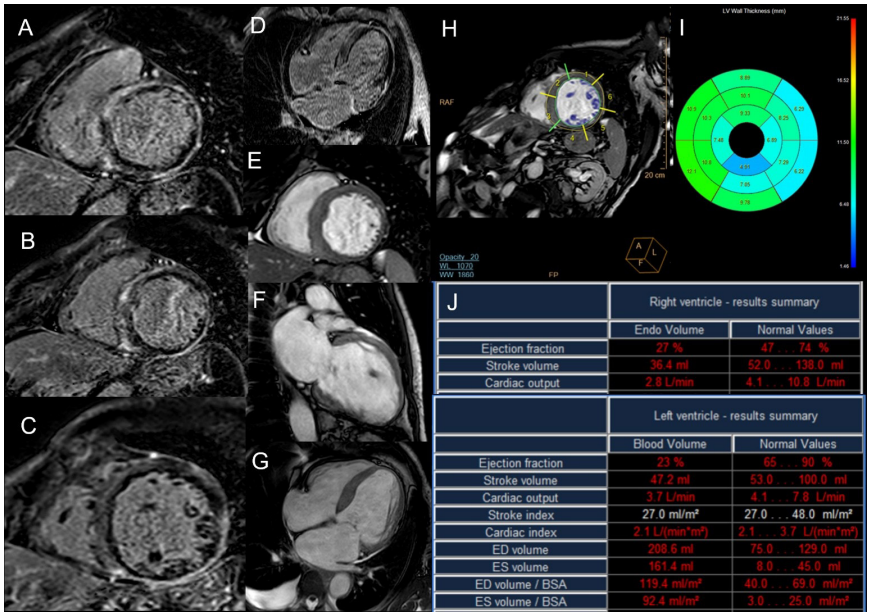


Figura 3 – Cardiac Magnetic Resonance (CMR) Imaging: Functional and tissue characterization study. A-C, short-axis and four chambers late gadolinium enhancement (LGE) reveal extensive non-ischemic (midcardial and subepicardial) ring-like enhancement in the basal and mid-segments. D-G, cine-steady-state free precession (SSFP) images demonstrate marked biventricular dilation with spherical remodeling of the left ventricle (LV). H-J, parametric wall thickness map and volumetric analysis confirm severe biventricular systolic dysfunction (LVEF 23% and RVEF 27%), associated with marked biatrial dilatation and right pleural effusion. The fibrosis pattern and biventricular involvement favor this entity over other differential diagnoses, such as chronic myocarditis or sarcoidosis, and are compatible with the phenotype of left-dominant arrhythmogenic cardiomyopathy.

Discussion

Phenotypic Spectrum and Genetics

Arrhythmogenic cardiomyopathy (ACM) is a complex clinical entity of predominantly genetic etiology, characterized by the progressive replacement of the myocardium by fibroadipose tissue.[2-5] This infiltration creates an arrhythmogenic substrate that predisposes to potentially lethal ventricular arrhythmias and heart failure.[2-5] Although the classic phenotype involves the right ventricle (ARVC), recent advances in imaging and genetics have allowed the identification of left-dominant variants (ALVC) and biventricular forms.[3,4]

Most cases are caused by pathogenic variants in desmosomal genes (such as PKP2, DSP, DSG2).[3,4] However, the ALVC variant, as suggested in this case, is frequently associated with pathogenic variants in desmoplakin gene (DSP) or in non-desmosomal genes, such as filamine C (FLNC).[3,4] These variants have been associated a particularly aggressive phenotype, with a high propensity for fatal ventricular arrhythmias, often preceding evidence of severe systolic dysfunction or marked ventricular dilation.[3,4]

The Evolution of Diagnostic Criteria: From Task Force to Padua

Historically, diagnosis was based on the 2010 Task Force Criteria, which had limited sensitivity for left-sided variants.[2-5] The introduction of the Padua criteria in 2020 transformed the approach, establishing specific criteria for ALVC.[1,3,4]

Under the Padua criteria, the diagnosis of ALVC is based on morphofuncional criteria such as left ventricular systolic

dysfunction (with or without dilation) or regional walls motion abnormalities, structural criteria such as late gadolinium enhancement (LGE) in the left ventricle (major structural criterion), and electrocardiographic abnormalities such as inverted T waves in V4-V6 or ventricular arrhythmias with right bundle branch block morphology.[1] In the present case, the presence of LGE, an independent marker of structural involvement, even in phases where ventricular dilation may not be prominent, and biventricular systolic dysfunction fulfill the Padua criteria for ALVC, supporting a diagnosis that would be omitted by older protocols.

The Central Role of Cardiac Magnetic Resonance (CMR)

CMR is the imaging modality of choice owing to its superior ability to characterize tissue. In the left-dominant pattern, the characteristic finding is non-ischemic (subepicardial or midmyocardial) LGE, highly suggestive, but not exclusive, of this entity, since overlapping patterns can occur in myocarditis and cardiac sarcoidosis.[5] Therefore, its interpretation should integrate the segmental distribution of enhancement, the clinical context, family history, and the other Padua criteria.

The "ring-like" (circumferential) pattern observed in this case is of particular relevance.[3,4] In addition to its diagnostic value, recent literature describes has been associated with pathogenic variants in the desmoplakin (DSP) or filamin C (FLNC) genes.[3,4] This ring of fibrosis creates the substrate for electrical reentry phenomena, explaining the episodes of ventricular tachycardia and syncope in the patient, and is associated with a greater propensity for malignant arrhythmias and sudden death, reinforcing the need for rigorous risk stratification and early intervention with an implantable cardioverter-defibrillator (ICD).[3,4]

In addition to LGE, parametric mapping techniques (T1 and T2 mapping) add diagnostic value:

- T1 Mapping and ECV: Increased native T1 time and Extracellular Volume (ECV) reflect diffuse interstitial fibrosis, allowing quantification of tissue replacement burden.[3,4]
- T2 Mapping: Essential for detecting myocardial edema. In ALVC, edema can signal a "hot phase," clinically mimicking acute myocarditis with chest pain and elevated troponins.[3,4]

The "hot phase" deserves particular attention due to its clinical impact. It corresponds to episodes of acute myocardial injury, characterized by chest pain, elevated troponins, and sometimes myocardial edema and LGE with subepicardial distribution, overlapping the clinical and imaging findings of acute myocarditis and representing one of the most common causes of initial diagnostic error).[3,4] This error can lead to delayed diagnosis and inappropriate treatment, particularly with corticosteroids or other immunosuppressants, without established indication in this entity, simultaneously delaying arrhythmic risk stratification and consideration of ICD.[3,4]

In patients with apparent myocarditis, the presence of disproportionate ventricular arrhythmias, recurrent episodes, a family history of sudden death or cardiomyopathy, T-wave inversion in the left precordial leads, or LV ring-like late enhancement should raise suspicion of ACM and prompt genetic testing and follow-up at a specialized center.[1,3,4] In the present case, the troponin elevation occurred in the setting of confouding factors (PE and decompensated heart failure), which illustrates the difficulty in interpreting this biomarker and reinforces the role of CMR in tissue characterization.

Differential Diagnosis and Associated Complications

The differential diagnosis of ALVC is broad, including chronic myocarditis, cardiac sarcoidosis, and idiopathic dilated cardiomyopathy. The specific distribution of LGE and family history are key to distinguishing the disease.[1-5]

A point of particular interest in this case was the initial presentation with (PE. Severe biventricular dysfunction (LVEF 23% and RVEF 27%) coupled with major atrial dilation promotes blood stasis and a prothrombotic state (Virchow Triad). This phenomenon demonstrates that ACM should not be considered merely as an arrhythmic disease, but as a structural pathology with the potential for serious thromboembolic complications, which can mask the primary diagnosis.

Conclusion and Teaching Points

- Diagnostic Sensitivity: ALVC is an underdiagnosed entity; the Padua criteria should be applied in patients with ventricular arrhythmias and non-ischemic pattern LGE.
- Prognostic Value of Imaging: The ring-like pattern of LGE is a high-risk marker for sudden cardiac death, regardless of ejection fraction.
- Therapeutic Decision: Cardiac magnetic resonance (CMR) was the decisive factor for risk stratification and the indication for ICD implantation, altering the patient's prognosis.

Divulgações Éticas / Ethical Disclosures

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

Conflicts of interest: The authors have no conflicts of interest to declare.

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Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

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