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CASE REPORT

MYOCARDIAL ARRHYTHMOGENIC CARDIOMYOPATHY – DIAGNOSTIC AND THERAPEUTIC CHALLENGES

Tamaz KHELADZE[1], Mariam LOMIDZE[1], Giorgi BAZERASHVILI[1], Vache SHIOLASHVILI[1], Tengiz VERULAVA[2]✉

[1] Chapidze Emergency Cardiology Center, Tbilisi, Georgia
[2] School of Medicine, Caucasus University, Tbilisi, Georgia

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Abstract

Introduction. Arrhythmogenic cardiomyopathy (ACM) is a progressive myocardial disease of genetic or acquired origin, characterized by ventricular arrhythmias and increased risk of sudden cardiac death. Once considered a right ventricular disorder, ACM also includes left-dominant and biventricular phenotypes. This case report aims to describe the clinical presentation and diagnostic challenges of an atypical biventricular form of ACM developing after a prolonged asymptomatic period.
Case presentation. We report a 44-year-old woman with a remote history of myocarditis who presented with recurrent syncope and sustained ventricular tachycardia degenerating into ventricular fibrillation. Electrocardiography demonstrated epsilon waves and widespread T-wave inversion. Cardiac magnetic resonance imaging revealed severe biventricular systolic dysfunction, marked right ventricular dilatation, and non-ischemic subepicardial myocardial fibrosis. Based on clinical, electrocardiographic, and imaging findings, a diagnosis of biventricular ACM was established. Given the high risk of sudden cardiac death, a dual-chamber implantable cardioverter-defibrillator was implanted for secondary prevention.
Conclusions. This case highlights the potential for delayed and life-threatening arrhythmic manifestations

Résumé

Cardiomyopathie arythmogénique du myocarde – défis diagnostiques et thérapeutiques

Introduction. La cardiomyopathie arythmogène (CMA) est une maladie myocardique progressive d'origine génétique ou acquise, caractérisée par des arythmies ventriculaires et un risque accru de mort subite cardiaque. Autrefois considérée comme une pathologie du ventricule droit, la CMA comprend également des phénotypes à prédominance gauche et biventriculaires. Ce cas clinique décrit la présentation et les difficultés diagnostiques d'une forme biventriculaire atypique de CMA, apparue après une longue période asymptomatique.
Présentation du cas. Nous rapportons le cas d'une femme de 44 ans, ayant des antécédents de myocardite, qui a eté consultéé pour des syncopes récurrentes et une tachycardie ventriculaire soutenue dégénérant en fibrillation ventriculaire. L'électrocardiogramme a mis en évidence des ondes epsilon et une inversion généralisée de l'onde T. L'imagerie par résonance magnétique cardiaque a révélé une dysfonction systolique biventriculaire sévère, une dilatation ventriculaire droite marquée et une fibrose myocardique sous-épicardique non ischémique. Sur la base des données cliniques, électrocardiographiques et d'imagerie, un diagnostic

✉ Address for correspondence: Tengiz VERULAVA
School of Medicine, Caucasus University, Tbilisi, Georgia
Address: Paata Saakadze street 1, 0102 Tbilisi, Georgia
Email: tverulava@cu.edu.ge; Phone +9955577284849

of ACM in patients with prior myocarditis. It underscores the pivotal role of cardiac magnetic resonance imaging, long-term clinical surveillance, and timely risk stratification with implantable cardioverter-defibrillator therapy to prevent sudden cardiac death.

Keywords: arrhythmogenic cardiomyopathy, ventricular arrhythmias, cardiac magnetic resonance imaging.

List of abbreviations:
ACM – arrhythmogenic cardiomyopathy
CMR – cardiac magnetic resonance
ECG – electrocardiogram
ICD – implantable cardioverter-defibrillator
LGE – late gadolinium enhancement
LV – left ventricle
LVEF – left ventricular ejection fraction
RV – right ventricle
RVEF – right ventricular ejection fraction
RVEDV – right ventricle end-diastolic volume
RVEDV/BSA – right ventricle end-diastolic volume/body surface area
ESC – European Society of Cardiology

de cardiomyopathie arythmogène biventriculaire (CAB) a été établi. Compte tenu du risque élevé de mort subite cardiaque, un défibrillateur automatique implantable double chambre a été mis en place à titre préventif secondaire.
Conclusions. Ce cas souligne le risque de manifestations arythmiques tardives et potentiellement mortelles de la CAB chez les patients ayant des antécédents de myocardite. Il met en évidence le rôle crucial de l'imagerie par résonance magnétique cardiaque, d'une surveillance clinique à long terme et d'une stratification précoce du risque, associée à la mise en place d'un défibrillateur automatique implantable, pour prévenir la mort subite cardiaque.

Mots-clés: cardiomyopathie arythmogène, arythmies ventriculaires, imagerie par résonance magnétique cardiaque

Introduction

Arrhythmogenic cardiomyopathy (ACM) is a myocardial disease characterized by non-ischemic fibrotic or fibro-fatty replacement of the myocardium, creating a substrate for ventricular arrhythmias and sudden cardiac death. The foundational diagnostic criteria for this condition were first proposed in 1994 and have since evolved with advances in imaging and genetics[1]. ACM is most associated with pathogenic variants in genes encoding desmosomal proteins, leading to myocyte detachment, progressive myocardial remodeling, and electrical instability[2]. However, increasing evidence suggests that ACM may also occur in non-genetic forms, including post-inflammatory or idiopathic variants, expanding the traditional understanding of the disease.

Clinically, these structural and electrical abnormalities predispose affected individuals to lifelong ventricular arrhythmias, often presenting with syncope or sudden cardiac arrest[3]. The risk of sudden cardiac death is particularly high in young and physically active individuals, frequently preceding overt structural heart disease.

The estimated population prevalence of ACM ranges from 1:1,000 to 1:5,000[4]. Phenotypically, approximately 50–60% of cases predominantly involve the right ventricle (RV), while 20–30% demonstrate left ventricular (LV) or biventricular involvement, reflecting the growing recognition of ACM as a disease spectrum rather than a purely right-sided cardiomyopathy[5].

The aim of this case report is to describe the clinical presentation and diagnostic challenges of an atypical biventricular form of ACM in a middle-aged woman with a remote history of myocarditis. This case highlights the progression from an initially asymptomatic phase to life-threatening ventricular arrhythmias and underscores the importance of long-term follow-up, advanced cardiac imaging, particularly cardiac magnetic resonance (CMR), and structured risk stratification in patients with prior myocarditis or unexplained arrhythmic events.

Case presentation

A 44-year-old woman with a history of two pregnancies and two deliveries, employed as a cook, was admitted following recurrent episodes of syncope and palpitations. Five years prior to the current presentation, she had been hospitalized with acute myocarditis, during which transthoracic echocardiography revealed a non-mobile thrombus in the left ventricle. She was treated with warfarin, and subsequent follow-up demonstrated complete resolution of the thrombus and improvement in echocardiographic findings. The patient did not receive regular medical follow-up and was not on chronic cardiovascular therapy in the ensuing years (Table 1).

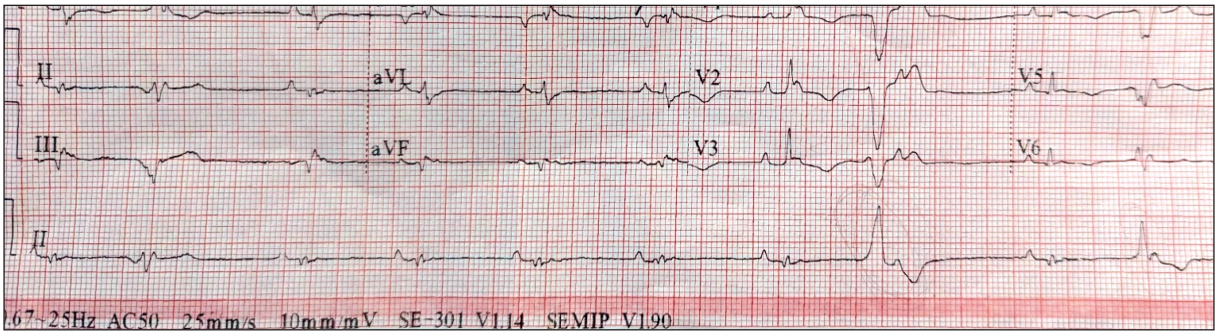


Figure 1. Electrocardiogram showing epsilon waves in leads V1, V2, V3, as well as (-T) – V1, V2, V3, V4, V5, V6. Ventricular extrasystole. Right bundle branch block. Low QRS voltage (<0.5 mV) in standard leads.

Table 1. Clinical timeline of the patient

Time point	*Clinical events and findings*
~5 years before admission	Hospitalization for acute myocarditis. Transthoracic echocardiography revealed a non-mobile LV thrombus. Warfarin therapy initiated.
Following months	Resolution of LV thrombus on follow-up echocardiography. Improvement in cardiac findings. Anticoagulation discontinued.
Intervening years	No regular cardiology follow-up. No chronic cardiovascular medication. Patient clinically asymptomatic.
Day of admission	Onset of palpitations, recurrent syncope, dyspnea, tachypnea, and retrosternal discomfort. Emergency medical services activated.
Pre-hospital phase	Documented sustained ventricular tachycardia and ventricular fibrillation during ambulance transport. Multiple electrical defibrillations performed with restoration of sinus rhythm.
Hospital admission	ECG showed epsilon waves (V1–V3), T-wave inversion (V1–V6), right bundle branch block, ventricular extrasystoles, and low QRS voltage.
Initial investigations	Echocardiography: RV dilatation, biventricular systolic dysfunction, restrictive LV diastolic dysfunction, mild mitral and moderate tricuspid regurgitation.
Acute differential workup	Coronary angiography: no significant coronary artery disease. CT pulmonary angiography: no pulmonary embolism.
Advanced imaging	CMR: LVEF 33%, RVEF 21%, severe RV dilatation (RVEDV/BSA 138 mL/m^2), subepicardial LGE in LV lateral wall → diagnosis of biventricular arrhythmogenic cardiomyopathy.
In-hospital management	Classified as high risk for sudden cardiac death. Dual-chamber ICD implanted according to ESC Class I recommendations.
1-month follow-up	Ventricular tachycardia episode successfully terminated by ICD antitachycardia pacing.
Subsequent management	Optimization of guideline-directed heart failure therapy. Consideration of electrophysiological study, catheter ablation, genetic testing, and thrombophilia screening.
5-month follow-up	Echocardiography showed recovery of LV systolic function (EF 58%) and clinical improvement.

Five years after the initial myocarditis diagnosis, the patient experienced several episodes of rapid heartbeat and syncope, accompanied by retrosternal discomfort, progressive dyspnea, and tachypnea, prompting emergency medical services activation. During transport, continuous cardiac monitoring revealed recurrent episodes of sustained ventricular tachycardia degenerating into ventricular fibrillation, requiring multiple electrical defibrillations, after which sinus rhythm was restored.

On admission, electrocardiography demonstrated epsilon waves in leads V1–V3, T-wave inversion in leads V1–V6, ventricular extrasystoles, right bundle branch block, and low QRS voltage (<0.5 mV) in the limb leads (Figure 1).

Transthoracic echocardiography revealed RV dilatation, biventricular systolic dysfunction, and restrictive LV diastolic dysfunction, without evidence of regional wall motion abnormalities. The LV was not dilated. Mild mitral regurgitation and moderate tricuspid regurgitation were noted. The aortic root and pulmonary artery dimensions were within normal limits, and pulmonary artery systolic pressure was at the upper normal limit.

Given the presence of recurrent malignant ventricular arrhythmias and chest discomfort, an acute coronary syndrome was excluded despite mildly elevated high-sensitivity troponin T levels (0.104 ng/mL). Emergency coronary angiography demonstrated no hemodynamically significant coronary artery

disease. Due to dyspnea, tachypnea, desaturation, and elevated D-dimer level (1.4 μg/mL), pulmonary embolism was excluded based on a negative computed tomography pulmonary angiography.

CMR imaging revealed severely impaired LV systolic function (LV ejection fraction (LVEF) 33%, LV end-diastolic volume 140 mL) and marked right ventricular dysfunction (RV ejection fraction (RVEF) 21%, with significant RV dilatation (RV end-diastolic volume (RVEDV) 246 mL; RVEDV/body surface area (BSA) 138 mL/m^2)). Late gadolinium enhancement imaging demonstrated patchy subepicardial enhancement in the lateral wall of the LV, consistent with non-ischemic myocardial fibrosis. These findings supported the diagnosis of biventricular ACM.

In view of the severely reduced ventricular function and documented ventricular fibrillation, the patient was classified as being at high risk for sudden cardiac death. According to European Society of Cardiology (ESC) class I recommendations, a dual-chamber implantable cardioverter-defibrillator (ICD) was implanted following informed consent.

At one-month follow-up, an episode of ventricular tachycardia was successfully detected and terminated by antitachycardia pacing via the ICD. Due to limited efficacy of antiarrhythmic pharmacotherapy, invasive electrophysiological study and catheter ablation were considered. Genetic testing and thrombophilia screening were recommended.

Under guideline-directed medical therapy for heart failure, the patient showed marked clinical improvement. Follow-up echocardiography at five months demonstrated recovery of LV systolic function, with an LVEF of 58%.

Discussion

Arrhythmogenic cardiomyopathy (ACM) remains a diagnostic challenge, particularly in its early stages, as the disease may remain clinically silent for years before manifesting with malignant ventricular arrhythmias or sudden cardiac arrest[6]. Early diagnosis is further complicated by phenotypic variability and overlap with other forms of non-ischemic cardiomyopathy. Characteristic electrocardiographic features—including T-wave inversion in the right precordial leads, epsilon waves, and ventricular arrhythmias—provide important diagnostic clues but may be absent or subtle in early disease. In this context, CMR imaging plays a pivotal role by enabling the detection of ventricular dilation, regional wall motion abnormalities, and non-ischemic myocardial fibrosis, which are central to contemporary ACM diagnosis.

The present case illustrates a delayed and complex clinical expression of ACM in a 44-year-old woman with a remote history of myocarditis and left ventricular thrombus. After a prolonged asymptomatic period without medical follow-up, the patient developed life-threatening ventricular tachyarrhythmias and biventricular systolic dysfunction, underscoring the progressive nature of the disease and the potential for late clinical deterioration. The occurrence of ventricular fibrillation as the initial manifestation highlights the importance of maintaining long-term vigilance in patients with prior myocardial inflammatory injury and unexplained arrhythmic symptoms.

Although ACM has historically been associated with genetic desmosomal mutations and predominantly RV involvement, increasing evidence supports the concept of ACM as a spectrum disorder, with left-dominant and biventricular phenotypes now well recognized. In this patient, CMR demonstrated severe dysfunction of both ventricles (LVEF 33%, RVEF 21%), marked RV dilation, and subepicardial late gadolinium enhancement of the LV lateral wall—findings consistent with advanced biventricular ACM as defined by contemporary ESC and Padua criteria. Genetic testing was not performed in this case, which represents a limitation of the diagnostic evaluation.

Recent studies have demonstrated a substantial genetic overlap between myocarditis and inherited cardiomyopathies[7]. Notably, pathogenic or likely pathogenic variants in cardiomyopathy-associated genes have been identified in approximately one in thirteen patients with acute myocarditis, suggesting that myocardial inflammation may act as a phenotypic trigger or disease modifier in genetically susceptible individuals[8]. This observation supports the growing recognition of genetic testing as a valuable tool in selected patients with myocarditis, ventricular arrhythmias, or unexplained cardiomyopathy, although its routine use requires further validation.

Conclusions

This case illustrates a late and severe clinical presentation of biventricular arrhythmogenic cardiomyopathy, manifesting with life-threatening ventricular arrhythmias after a prolonged asymptomatic period in a patient with a remote history of myocarditis. While a direct causal relationship cannot be definitively established, prior myocardial inflammation may have acted as a trigger or modifier of disease expression, contributing to progressive structural and electrical remodeling.

The case highlights the critical role of CMR imaging in identifying non-ischemic myocardial fibrosis and biventricular involvement, thereby facilitating accurate diagnosis and risk stratification. Although

genetic testing was not performed, its potential value in similar clinical scenarios is increasingly recognized.

Overall, this report emphasizes the importance of long-term surveillance in patients with prior myocarditis or unexplained arrhythmic symptoms, as well as the need for timely diagnostic evaluation and prophylactic interventions, particularly ICD implantation, to prevent sudden cardiac death and preserve quality of life.

Author contributions

Conceptualization, TK, ML, GB, TV; Methodology, TK, ML; Software, TK, TV; Validation, TK, GB; Formal Analysis, TK, ML, GB, TV; Investigation, TK, ML, GB, TV; Resources, TK; Data Curation, TK, ML; Writing – Original Draft Preparation, TK, ML, GB, TV; Writing – Review & Editing, TK, ML, GB, TV; Visualization, TK, TV; Supervision, TK; Project Administration, TK, ML, GB, TV. All the authors have read and agreed with the final version of the article.

Compliance with Ethics Requirements:

"The authors declare no conflict of interest regarding this article"

"The authors declare that all the procedures and experiments of this study respect the ethical standards in the Helsinki Declaration of 1975, as revised in 2008(5), as well as the national law. Informed consent was obtained from the patient included in the study"

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