Int J Cardiovasc Sci. 2024; 37:e20230081

CASE REPORT

Inflammatory Cardiomyopathy as a Cause of Recurrent Ventricular Tachycardia Requiring Heart Transplant

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Introduction

The most common setting for ventricular tachycardia (VT) is ischemic heart disease, in which myocardial scar tissue is the substrate for arrhythmia.[1,2] However, in some myocardial diseases (i.e., myocarditis, arrhythmogenic and dilated cardiomyopathies [DCM]), electrical abnormalities can precede apparent structural changes rendering diagnosis and treatment difficult.

The present study describes a case of inflammatory cardiomyopathy that appeared as a recurrent VT and required a heart transplant as the only therapeutic alternative after 20 years of the disease.

Case presentation

In 1999, a 47-year-old woman with well-controlled hypertension experienced the first episode of palpitations provoked by physical emotion and exertion. There was no family history of premature cardiovascular disease or sudden cardiac death. Serial electrocardiograms (ECG) revealed monomorphic premature ventricular contractions and VT with right bundle branch block (BBB) morphology (Figure1). Transthoracic echocardiography (TTE) showed 9 mm atrial septal defect (ASD) with no other morpho-functional changes. Laboratory tests (electrolyte, endocrine, inflammatory) were normal. Episodes of ventricular arrhythmia became more frequent and symptomatic. The patient underwent ASD repair. Subsequently, TTE showed an absence of cardiac remodeling. Coronary angiography revealed no anomaly. The patient continued to experience recurrent symptomatic VT resistant to antiarrhythmic drugs and endocardial catheter ablation (CA). An invasive electrophysiology study demonstrated inducible VT of 3 different morphologies (Figure 2). In 2009, the patient underwent redo CA, but with no effect. A repeat TTE demonstrated dilation of the LV with reduced ejection fraction (36%, diffuse hypokinesis). To assess myocardial perfusion and stratify the risk of cardiovascular events, myocardial perfusion scintigraphy was performed. This action revealed regions of hypoperfusion. The patient underwent implantable cardioverter-defibrillator (ICD) implantation. After, several appropriate ICD shocks were applied. In 2012, TTE showed signs of left ventricular noncompaction (LVNC) but there was no dilation of the heart, and ejection fraction was mildly reduced (Figure 3). Multislice spiral computed tomography coronary angiography (MSCT-CA) revealed increased LV trabeculation and several adipose foci in the myocardium of the right ventricle. Laboratory investigations showed elevated antimyocardial antibodies. Viral and additional immune tests were negative. The patient received evidence-based heart failure treatment and antiarrhythmic drugs.

In 2018, the patient developed electrical storm that required ICD reimplantation due to battery depletion, redo CA, and renal sympathetic denervation. TTE showed signs of left chamber dilation and reduced ejection fraction (33%, diffuse hypokinesis) of the LV. The patient's condition progressively worsened, and three days later, the transplant was performed. The transplant biopsy revealed a mild (grade 1A) rejection. At the time of this report the patient had no signs of arrhythmia and heart failure. Follow-up biopsy and TTE showed no transplant rejection or dysfunction.

Keywords

Ventricular Tachycardia; Heart Failure; Myocarditis; Heart Transplantation.

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DOI: https://doi.org/10.36660/ijcs.20230081

Manuscript received May 24, 2023; revised manuscript October 12, 2023; accepted November 8, 2023.

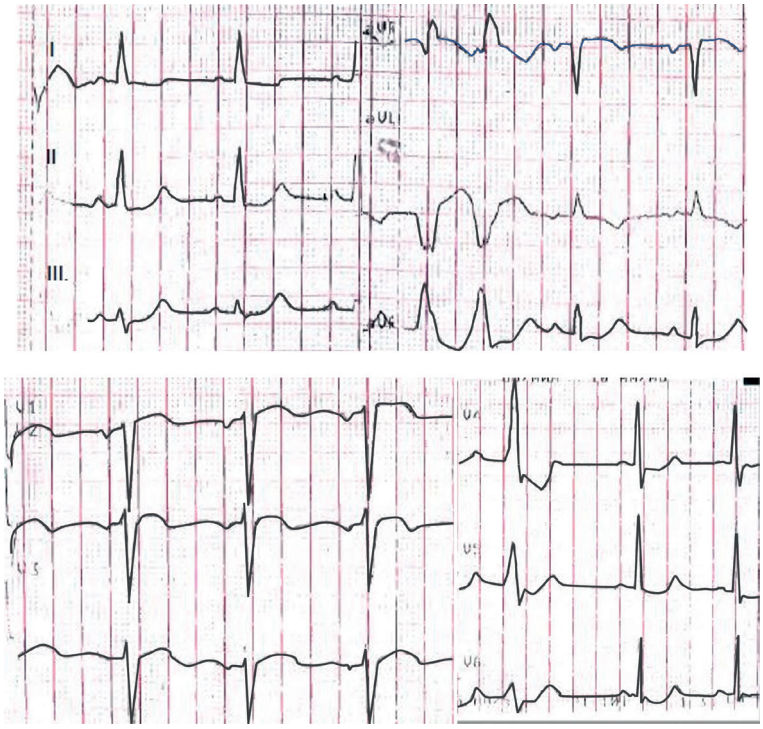

Figure 1 – ECGs.
ECG (25 mm/s, 10 mm/mV) at baseline: sinus rhythm at 75 bpm, normal axis, poor progression of R from V1 to V3, and frequent premature ventricular contractions.

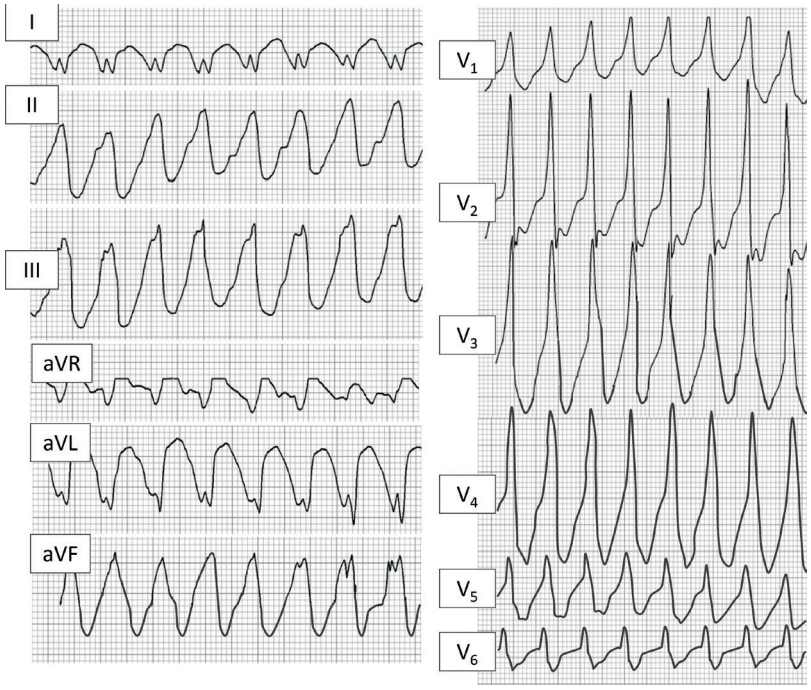

Figure 2 – ECG during VT episode (25 mm/s, 10 mm/mV): monomorphic wide complex tachycardia (166 bpm) with right BBB pattern.

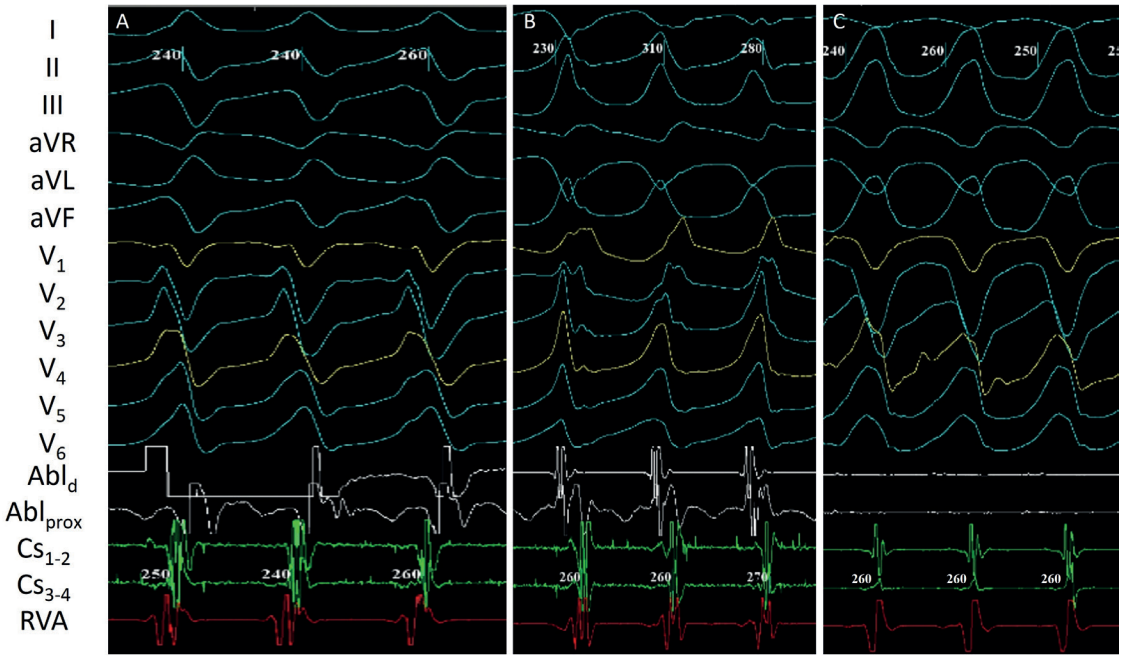

Figure 3 – Electrophysiological study: VT with three different morphologies (150 mm/s).
A) VT with superior axis and left BBB morphology; B) VT with inferior axis and right BBB morphology; C) VT with inferior axis and left BBB morphology. Ablp/Abld – proximal and distal ablation catheter, Cs1-2/3-4 – distal coronary sinus. RVA: right ventricular apex.

Anatomopathological macroscopic study of the explanted heart revealed a dilation of both ventricles. Serial slices of the coronary arteries showed initial atherosclerotic lesions. Histological study demonstrated diffuse oedema and extensive interstitial inflammatory infiltrates with the predominance of lymphocytes; cardiomyocyte dystrophy, and diffuse interstitial sclerosis in both ventricles, with no cardiomyocyte necrosis. No signs of LVNC were observed, and there was no fibrofatty replacement of myocytes characteristic of arrhythmogenic cardiomyopathy.

Discussion

In this case the disease appeared with sustained monomorphic VT. It is important to note that the main clinical symptom remained throughout a 20-year history of current disease arrhythmia.

Potentially reversible causes of VT were excluded during first-line diagnostic testing. ASD is not thought to be an etiological factor for arrhythmia, since VT progressed after surgical correction.

Idiopathic VT was ruled out, given the subsequent remodeling of the left ventricle and increase in complexity of ventricular arrhythmia.

Based on further diagnostic findings (adipose foci in the myocardium of the right ventricle by MSCT-CA,[3] signs of LVNC by echocardiography,[4,5] decreased cardiac perfusion by cardiac perfusion imaging,[1,2,4] and elevated levels of antimyocardial antibodies), it was later hypothesized that VT could be of mixed origin due to the coexistence of arrhythmogenic cardiomyopathy (right or biventricular), LVNC, and chronic myocarditis. However, only myocarditis complicated by DCM was confirmed by the histological study.

The substantial variability in LV morpho-functional measures (size, signs of LVNC, systolic function) upon repetition of the TTE is likely to be attributed to disease-related factors, such as alternating episodes of progression, attenuation of myocardial inflammation, and intermittent arrhythmia burden.

In some cases, VT may be the only symptom of myocarditis, requiring many differentials to be considered.[1,2] There are no pathognomonic criteria

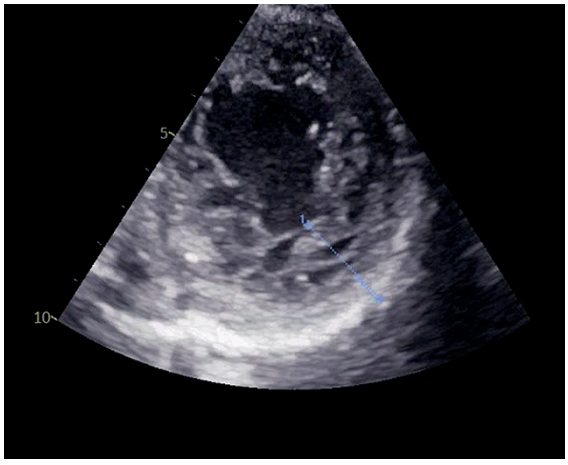

Figure 4 – Two-dimensional TTE (parasternal short-axis view): the ratio between the noncompacted and the compacted wall was more than 2 in the diastole..

associated with myocarditis; therefore, it should be suspected in all patients with ventricular arrhythmias, especially in young patients with no history of cardiac disease.[1,2] In this case, increased antimyocardial antibodies and reduced myocardial perfusion by scintigraphy suggested the diagnosis of myocarditis. However, the specificity of these findings is low.[1,2] Cardiac magnetic resonance imaging (MRI), considered the most reliable method for diagnosing myocarditis, was not performed, given that, at the time of the onset of the disease (2000), cardiac MRI was not widely available in the country of residence and was not included in local guidelines. After, cardiac MRI was impossible due to implantation (and reimplantation in 2018) of the non-MRI-conditional defibrillator. Endomyocardial biopsy (EMB), the gold standard for diagnosis, is rarely used in suspected myocarditis, as it is an invasive procedure, and there is a high probability of sampling error and variability in observer interpretation. Current position statements of the European Society of Cardiology[1] and the American Heart Association[4] vary widely regarding indications for biopsy. According to the Brazilian Society of Cardiology (guidelines on myocarditis; 2022) EMB was specifically indicated in the present case due to the development and progression of heart failure of undefined etiology associated with ventricular arrythmia.[6] However EMB was not performed.

Myocarditis can be an acute, subacute, or chronic disorder, and may present focal or diffuse involvement of the myocardium.[1,2] According to existing literature, in up to 30% of all patients with biopsy-proven myocarditis, progression to DCM can occur and is commonly associated with a poor prognosis.[4,7] In the present case, persisting diffuse myocardial inflammation, which is likely to be the main cause of DCMs, was detected by the pathohistological study of the explanted heart. Thus, the term 'inflammatory cardiomyopathy' can be used when referring to this case.[7] According to the medical history and further evaluation, there were no signs of other underlying causes (infiltrative, endocrine, or systemic diseases) of DCM. However, to date, it is impossible to exclude frequent premature ventricular contractions and VT, as well as genetic factors, as mechanisms contributed to heart remodeling.[1,2,4,8]

In summary, this case highlights that, in the absence of an ideal diagnostic tool, myocardial inflammation should be highly suspected as the potential cause of poorly controlled arrhythmia and cardiac dilation. Despite all high-tech treatments, in the end, the patient required life-saving heart transplant. Myocardial inflammation was confirmed by the study of the explanted heart.

Author Contributions

Conception and design of the research, acquisition of data and writing of the manuscript: Cabello Montoya FE, Stavtseva YV, Teterina M; analysis and interpretation of the data: Cabello Montoya FE, Stavtseva YV, Teterina M, Vorobiev A, Kobalaba Z; critical revision of the manuscript for intellectual content: Cabello Montoya FE, Teterina M, Vorobiev A.

Potential Conflict of Interest

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

Sources of Funding

There were no external funding sources for this study.

Study Association

This study is not associated with any thesis or dissertation work.

Ethics Approval and Consent to Participate

This article does not contain any studies with human participants or animals performed by any of the authors.

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