Acute Myocarditis Presenting with Ventricular Arrhythmias: The Role of CMR in the Differential Diagnosis of ARVD

Olivier Huttin¹, Zied Frikha¹, Béatrice Bremilla-Perrot¹, Jean-Marc Sellal¹, Damien Mandry², Etienne Aliot¹, Yves Juilliere¹, Nicolas Sadoul¹ and Christine Selton-Suty¹

Abstract

We herein present the findings of the case of a 23-year-old man who was hospitalized for ventricular tachycardia (VT) with no previous history of cardiac disease or any family history of sudden death. Based on the clinical features as well as the echographic and MRI results, the patient was diagnosed with both acute viral myocarditis and arhythmogenic right ventricular dysplasia (ARVD). The patient underwent implantation of an automatic cardioverter defibrillator. There was no recurrent VT during the 24 month follow-up. This case demonstrates the link between ARVD and myocarditis, and highlights the importance of conducting an RV assessment through a cardiac magnetic resonance (CMR) study in the context of arrhythmia and myocarditis.

Key words: ventricular tachycardia, ARVD, myocarditis


Introduction

Myocarditis has emerged as a frequent etiological cause of sudden arrhythmic death over the past several decades. The diagnosis of myocarditis is a challenging process in everyday clinical practice. The development of new imaging techniques, cardiac magnetic resonance (CMR) in particular, has enabled the increased recognition of myocarditis as a cause of ventricular arrhythmias. This technique is an important tool in the improvement of the differential diagnosis, and it can provide valuable knowledge as to the pathological substrates that may give rise to the arrhythmic syndromes.

Case Report

A 23-year-old man presented with palpitations that had started 6 hours prior to admission and dizziness that arose immediately after playing soccer. He was an active smoker with no previous history of cardiac disease or any family history of sudden death. There was neither context of fever, nor chest pain nor any previous syncopal episodes. A physical examination revealed a heart rate of 190 bpm and a low blood pressure of 72/53 mm Hg with no symptoms of right heart failure. The 12-lead electrocardiogram (ECG) that was performed at his admission showed a ventricular tachycardia (VT) with an inferior axis and left bundle branch block morphology (Fig. 1). The VT could not be stopped by either intravenous lidocaine or amiodarone, and it was interrupted by rapid pacing. His baseline ECG, which was performed following the resumption of his sinus rhythm, showed Q waves in the inferior leads and negative T waves in the anterior leads (Fig. 2). The blood tests revealed an elevated troponin level (34 ng/mL, normal <5 ng/mL) and an elevated creatine phosphokinase (CPK) level (1,240 IU/L, normal <120 IU/L) at 36 hours after his admission. His electrolyte levels as well as the inflammation markers were normal (C-reactive protein (CRP) <5 mg/L). Echocardiography showed a mild left ventricular systolic dysfunction (LVEF 45%) as well as left ventricular (LV) wall motion abnor-
met the criteria for both acute viral myocarditis and ARVD. Therefore, he was treated with a low dose of sotalol as well as ACE inhibitors and aspirin due to the spontaneous contrast in his LV cavity. At a follow-up appointment 2 months later, a programmed ventricular stimulation was performed and induced the same ventricular tachycardia despite the administered antiarrhythmic therapy. A new CMR demonstrated the recovery of the LV systolic function (LVEF=0.56) and a partial regression of the edema, but with the persistence of the major ARVD criterion (Fig. 6). The patient agreed to receive an automatic implantable cardioverter defibrillator (AICD). He was discharged on sotalol following the implantation of the AICD. There was no recurrent VT during the 24-month follow-up.

**Discussion**

This case illustrates that the link between ARVD and myocarditis remains to be elucidated and that the differential diagnosis can be challenging. Our patient fulfilled 2 of the major criteria that were set forth by the task force for ARVD (1): imaging criteria (RV involvement noted from both CMR and echocardiography) and repolarization abnormalities (inverted T waves in right precordial leads in the absence of complete right bundle-branch block). In addition, he met one minor ARVD diagnosis criteria: arrhythmia (sustained ventricular tachycardia). However, he also fulfilled the criteria for a diagnosis of acute myocarditis based on his clinical symptoms (palpitations), troponin release and cardiac MRI results (an increased myocardial T2 signal on an inversion recovery sequence and a delayed contrast enhancement following a gadolinium infusion). At this point, the controversy regarding the link between the two conditions has not yet been settled.

Several studies have demonstrated that myocarditis may affect the right ventricle, (RV) causing structural abnormalities, including microaneurysms, as well as the arrhythmic manifestations that are typical of ARVD (2, 3). According to Hofmann et al. (2) and Pieroni et al. (3), myocardial inflammatory infiltrates may lead to the functional and structural changes of the right ventricular myocardium that resemble those produced by fibrofatty replacement, and provide the
necessary substrate for producing ventricular arrhythmias. Previous studies have also suggested that myocardial inflammation may play a key role in the progression of genetically determined arrhythmic syndromes and in their subsequently adverse clinical course.

Myocardial inflammation may be seen in up to 75% of ARVD hearts at autopsy, and this inflammation likely plays a role in triggering ventricular arrhythmias (4). Viruses have been detected in the myocardium of some ARVD patients, thus leading to the claim of an infective etiology of the disease (5). Other authors have suggested that the viruses are innocent bystanders or that spontaneous cell degeneration may serve as a milieu favoring the viral settlement in the myocardium (6). It should be noted that there is a prevalence of biventricular involvement in ARVD (7, 8). Recently, Sen-Chowdhry et al. published a report detailing the clinical courses of patients with predominant mutations in the desmoplakin gene and marked LV involvement with the relative sparing of the RV (9).

Recent reports indicate that the clinical presentations of ARVD and myocarditis are similar (3, 10). Pieroni et al. suggests that acute episodes of inflammation or myocarditis may be a step in the evolution of ARVD, and that myocarditis may even mimic ARVD up to the point of even fully satisfying the original 1994 International Task Force criteria (3).

Both myocarditis and ARVD can be studied by CMR, and a very specific set of criteria for myocarditis with a dominant role for CMR was recently described (11). According to this consensus, a diagnosis of myocarditis can be made when at least two of the following features are present: one area of non-ischemic late gadolinium enhancement, global or regional signal increase in T2-weighted images and global early gadolinium enhancement. CMR study can also provide additional information about the morphological and functional evaluation of the RV.
Concerning ARVD, CMR has emerged as an important imaging modality in the diagnosis and evaluation of this disease. In this situation, CMR allows a clear visualization of the RV in view of its three dimensional, multi-planar capabilities with excellent spatial resolution and improved contrast between the blood pool. According to the revised Task Force (1), there is no mention of fatty infiltration, wall thinning, delayed enhancement or LV involvement. The major criteria for ARVD as outlined by the Task Force are the regional RV akinesia or dyskinesia or the dyssynchronous RV contraction that is commonly associated with severe global/segmental RV dilation or global systolic dysfunction. The minor criteria include the mild global/segmental dilation of the RV, regional contraction abnormalities and global diastolic dysfunctions.

This case report highlights the importance of an RV assessment (global function and segmental analysis) through a CMR study in the context of arrhythmia and myocarditis. These data indicate a need for the inclusion of sequences for the detection of myocardial inflammation with a morphological and functional evaluation of the RV when CMR is performed as a part of the non-invasive diagnostic work-up of patients who present with ventricular arrhythmias. In these cases, it is important to report all of the detected abnormalities, whether they suggest either myocarditis or ARVD. Finally, the enhancement patterns seen on CMR may also serve as a map for the exact location of ablation or an endomyocardial biopsy, if necessary; thereby enhancing the diagnostic accuracy of an endomyocardial biopsy.

The authors state that they have no Conflict of Interest (COI).

References


© 2013 The Japanese Society of Internal Medicine
http://www.naika.or.jp/imonline/index.html

1918