Case Report

Endocardial Late Potentials During Sinus Rhythm Define the Re-Entry Circuit of Ventricular Tachycardia in Arrhythmogenic Right Ventricular Cardiomyopathy

Michalis Efremidis[1], Efstathia Prappa[1], Charalampos Charalampous[1], Nikos Protonotarios[2], Konstantinos P. Letsas[1], Antonios Sideris[1]

[1]*Second Department of Cardiology, Evangelismos General Hospital, Athens,* [2]*Yannis Protonotarios Medical Centre, Naxos, Greece*

Key words:
Ventricular tachycardia, endocardial potentials mapping, ablation.

Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D) is an inherited cardiomyopathy characterized by progressive fibro-fatty replacement of the right ventricular myocardium. We report a case where mapping of endocardial potentials during sinus rhythm identified the re-entry circuit of a recurrent ventricular tachycardia in a patient with ARVC. The tachycardia was subsequently ablated successfully.

Manuscript received:
January 9, 2012;
Accepted:
February 1, 2012.

Address:
Michalis Efremidis

42 M. Alexandrou St.
171 22 Athens, Greece
e-mail: micbcar@otenet.gr

Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D) is a rare form of inherited cardiomyopathy that is an important cause of ventricular arrhythmias, especially in children and young adults. We report a case where mapping of endocardial potentials during sinus rhythm identified the re-entry circuit of a recurrent ventricular tachycardia (VT) in an adult patient with ARVC.

Case presentation

A 68-year-old female patient known to be a homozygous carrier for plakoglobin mutation (Naxos disease), a recessive form of ARVC/D, was referred to our hospital for management of recurrent, drug refractory VT. There was a positive family history for sudden cardiac death; her grandson, also suffering from ARVC/D, had died suddenly a few years before, despite having an implantable cardioverter defibrillator (ICD). Our patient had been diagnosed at that time through a family screening procedure. She was completely asymptomatic until recently, when she presented recurrent episodes of syncope due to sustained VT.

On physical examination, the cutaneous phenotype of the disease was apparent, with peculiar woolly hair and palmoplantar keratoderma. Heart sounds were normal, without any murmurs or gallops. The baseline ECG showed sinus rhythm, epsilon wave in lead V_1, negative T waves in leads V_1-V_3, and frequent premature ventricular complexes (PVCs) with a left bundle branch block (LBBB) morphology and inferior axis. The ECG during VT had the same morphology (cycle length 300 ms). Ambulatory ECG monitoring showed frequent monomorphic PVCs (>1000 per day) and short episodes of bigeminy. The signal average ECG was abnormal.

Transthoracic echocardiography revealed a severely dilated right ventricle (RV) and akinetic/dyskinetic areas in the RV outflow tract, apex, and postero-dia-

phragmatic wall. Left ventricular function was preserved. Cardiac MRI confirmed the RV echocardiographic findings and additionally revealed left ventricular involvement (subepicardial late gadolinium enhancement in the inferolateral wall). Coronary angiography revealed no significant coronary artery stenosis.

Our patient was considered to be at high risk for sudden cardiac death, and consequently a candidate for ICD implantation. Given the recurrent nature of the VT episodes, a radiofrequency (RF) catheter ablation procedure was also scheduled. The procedure was guided by a three-dimensional electroanatomical mapping system (CARTO 3, Biosense Webster, Inc., Diamond Bar, CA, USA). Since the VT was not well-tolerated, RV inflow and outflow voltage mapping was performed during sinus rhythm using a 4 mm tip deflectable catheter (NAVISTAR, Biosense Webster). A wide area of myocardium with low voltage (<0.5 mV) was depicted, especially in the free wall of the RV inflow tract (Figure 1). Notably, isolated late potentials were recorded in this area during sinus rhythm (Figure 2). The earliest activation site of the PVCs (preceding the onset of the surface QRS by 30 ms) was identified in the same area and pace mapping at this location demonstrated a "perfect match" with the morphology of the PVCs and the spontaneous VT (Figure 3). RF energy applications were delivered in this region (target temperature of 60°C, 30 W, 60 s) (Figure 1). Programmed ventricular stimulation failed to induce any PVCs or VT. Ambulatory ECG recordings performed 48 h following RF ablation failed to demonstrate any PVCs. The patient subsequently received an ICD because she had risk factors for sudden cardiac death (not well tolerated VT presenting with syncope, severe RV involvement, LV involvement on MRI). After a six-month follow-up period, the patient remains asymptomatic.

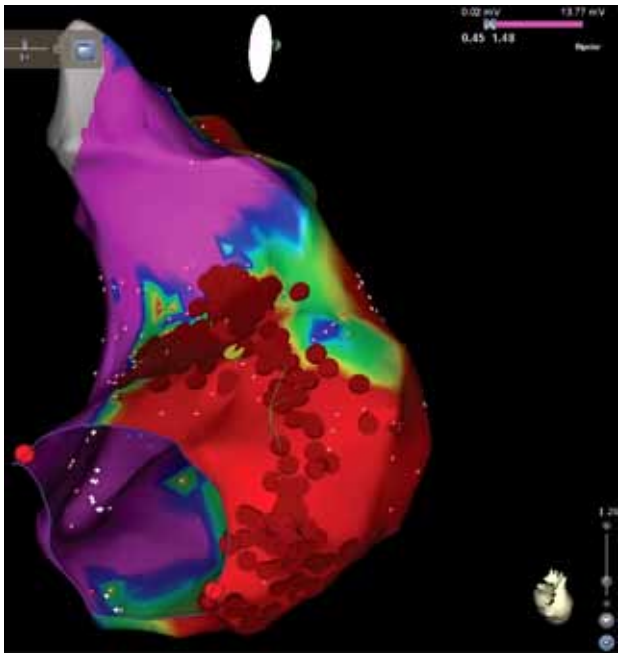

Figure 1. Electroanatomical voltage map (right lateral view) showing a wide area of myocardium with low voltage (<0.5 mV) at the free wall of the inflow tract of the right ventricle.

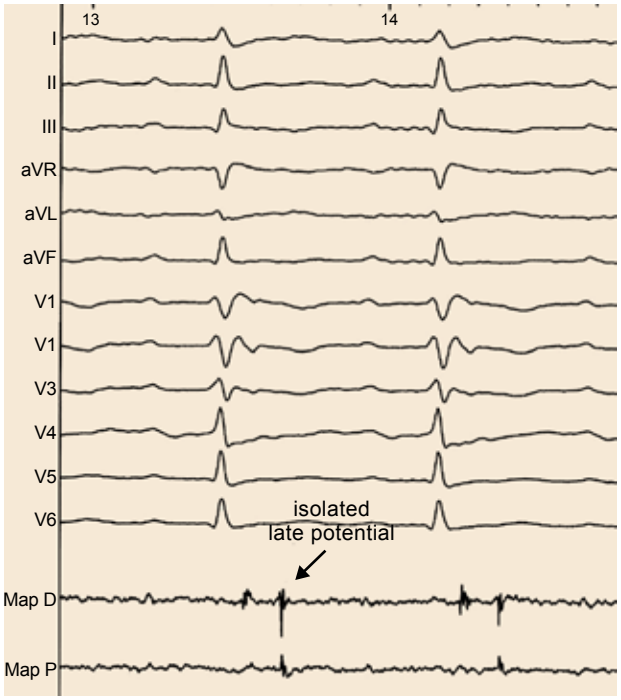

Figure 2. Intracardiac tracing during right ventricular mapping (map) showing isolated late potentials during sinus rhythm.

Discussion

ARVC/D is an inherited cardiomyopathy characterised by progressive fibro-fatty replacement of the RV myocardium.[1,2] Scar tissue within the myocardium provides the substrate for re-entrant VT.[1] Prevention of sudden cardiac death and VT recurrence is achieved with antiarrhythmic drug therapy, ICD and RF catheter ablation.[1-5] Several single-centre studies have reported conflicting results concerning the acute and long-term efficacy of catheter ablation in patients with ARVD/C.[2,3]

The ablation procedure in our case was guided by a three-dimensional electroanatomical mapping system. RV voltage mapping has been demonstrat-

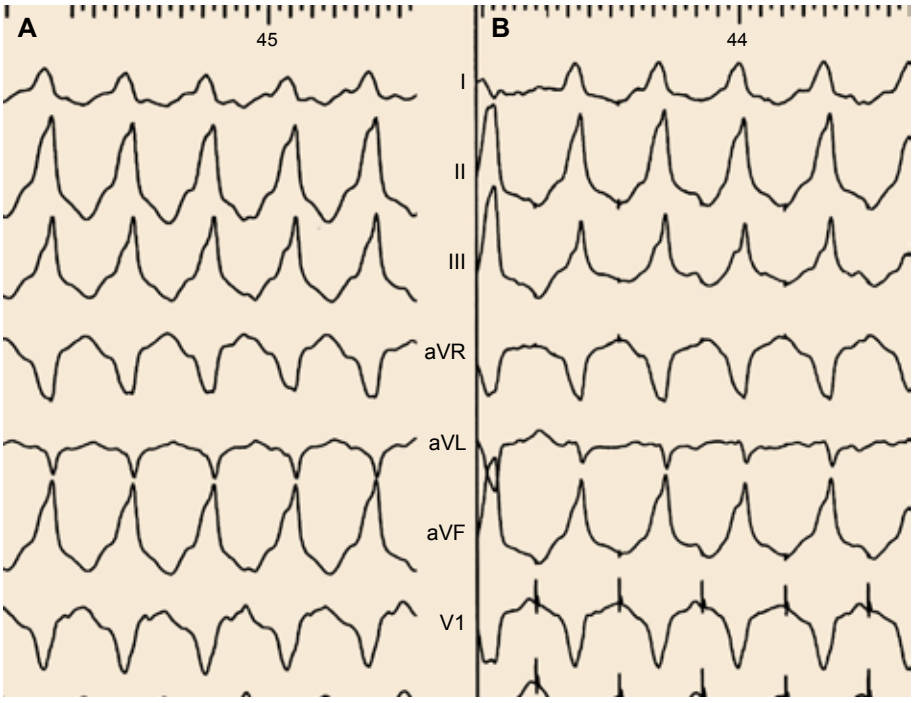

Figure 3. Pace-mapping (B) demonstrating a "perfect match" with the morphology of the ventricular tachycardia (A).

ed to accurately identify the presence, location, and extent of the pathological substrate of ARVC/D by detection of low-voltage regions that reflect RV fibrofatty myocardial atrophy.[2,3] At the site of earliest endocardial activity (free anterior wall of the RV inflow tract), an area of isolated late potentials was found. Reduced conduction velocity and nonuniformly anisotropic conduction produce areas of delayed/fractionated electrical activity that can persist after the inscription of the QRS complex during sinus rhythm.[4,5] In our case, pace mapping in this region reproduced exactly the morphology of the QRS of the PVCs and the spontaneous VT. This entire area was ablated using linear lesions connecting viable with fibrotic tissue (tricuspid valve area). Ablation of this localised source of delayed isolated electrograms abolished all PVCs and resulted in non-inducibility of the VT after ablation. However, data from previous studies showed that these late potentials can be recorded from relatively large areas, which may not correlate with the site of origin of VT. Additionally, some sites yielding late potentials during sinus rhythm are bystander sites during VT.[4,5] This case presentation demonstrates how ablation of VT in a patient with ARVC/D may be facilitated by accurate delineation of the arrhythmogenic substrate.

References

1. Protonotarios N, Tsatsopoulou A, Anastasakis A, et al. Genotype-phenotype assessment in autosomal recessive arrhythmogenic right ventricular cardiomyopathy (Naxos disease) caused by a deletion in plakoglobin. J Am Coll Cardiol. 2001; 38: 1477-1484.
2. Avramides D, Protonotarios N, Asimaki A, Matsakas E. Arrhythmogenic right ventricular cardiomyopathy/dysplasia. Hellenic J Cardiol. 2011; 52: 452-461.
3. Gatzoulis KA, Archontakis S, Dilaveris P, et al. Ventricular arrhythmias: from the electrophysiology laboratory to clinical practice. Part I: malignant ventricular arrhythmias. Hellenic J Cardiol. 2011; 52: 525-535.
4. Marcus FI, McKenna WJ, Sherrill D, et al. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia: proposed modification of the Task Force Criteria. Eur Heart J. 2010; 31: 806-814.
5. Marchlinski FE, Zado E, Dixit S, et al. Electroanatomic substrate and outcome of catheter ablative therapy for ventricular tachycardia in setting of right ventricular cardiomyopathy. Circulation. 2004; 110: 2293-2298.
6. Satomi K, Kurita T, Suyama K, et al. Catheter ablation of stable and unstable ventricular tachycardias in patients with arrhythmogenic right ventricular dysplasia. J Cardiovasc Electrophysiol. 2006; 17: 469-476.
7. Kühne M, Abrams G, Sarrazin JF, et al. Isolated potentials and pace-mapping as guides for ablation of ventricular tachycardia in various types of nonischemic cardiomyopathy. J Cardiovasc Electrophysiol. 2010; 21: 1017-1023.
8. Hsia HH, Lin D, Sauer WH, Callans DJ, Marchlinski FE. Relationship of late potentials to the ventricular tachycardia circuit defined by entrainment. J Interv Card Electrophysiol. 2009; 26: 21-29.