

Arrhythmogenic Right Ventricular Cardiomyopathy: An Unusual Possible Cause of Arrhythmia in a 78 year old Man with a 40 year History of Palpitations

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For Editorial see page 375

A long history of undocumented regular palpitations occurring in the elderly with ostensibly normal heart and a normal resting electrocardiogram is usually related to paroxysmal supraventricular tachycardia. We report the case of a 78 year old man with a 40 year history of palpitations that were most probably due to ventricular tachycardia secondary to arrhythmogenic right ventricular cardiomyopathy/dysplasia.

PATIENT DESCRIPTION

An apparently healthy 78 year old man presented with a 40 year history of rapid, regular, non-documented palpitations. These episodes, lasting up to 15 minutes, were provoked by stress and were relatively well tolerated apart from some dizziness at their onset. The patient recalled that one of these episodes was terminated by carotid sinus massage administered by a physician before any electrocardiographic documentation could be done. While on atenolol therapy the patient continued to experience palpitations. His medical history was unremarkable apart from hypercholesterolemia. During these four decades of palpitations, repeated non-invasive cardiologic workups were nor-

mal. These included resting ECG, exercise test, echocardiogram and 24 hour Holter monitoring. His routine medications were aspirin, atenolol and atorvastatin.

Several possible diagnoses to explain the palpitations were discussed with the patient, focusing on the high probability of PSVT. Trans-telephonic ECG monitoring or diagnostic electrophysiologic study was recommended but the patient preferred to wait. About 1 month later, following a stressful event, the patient exhibited rapid palpitations resembling those he had experienced previously; however, this time the episode was prolonged (2 hours), prompting hospitalization. The ECG in the emergency room [Figure A] showed a rapid (220/min), regular, wide QRS tachycardia with left bundle branch block and left QRS axis. The patient was hypotensive (90 mmHg systolic blood pressure). A diagnosis of VT was made. A synchronized direct current shock was administered and sinus rhythm was restored. The ECG performed immediately after cardioversion showed transient atrioventricular conduction disturbances and ST-T depression in inferolateral leads. An echocardiogram showed normal left and right ventricular size and function with no other abnormalities. In addition, a coronary angiography showed normal coronary arteries and normal left ventricular angiography.

A diagnostic EPS was performed with the patient off drugs. No antegrade dual atrioventricular nodal physiology could be demonstrated. There was no retrograde

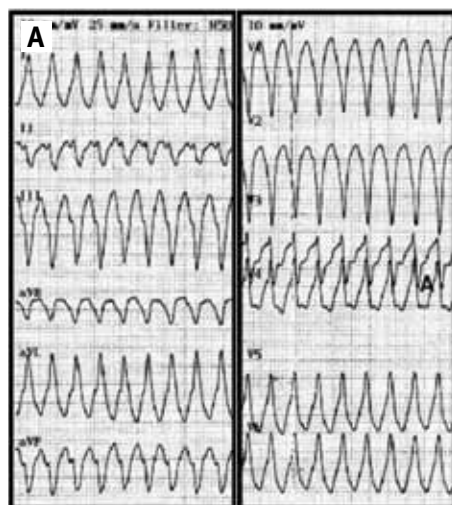
PSVT = paroxysmal supraventricular tachycardia
 VT = ventricular tachycardia
 EPS = electrophysiologic study

conduction. Supraventricular tachycardia could not be induced with rapid atrial pacing. However, sustained monomorphic VT (237/min) was easily induced with triple extra-stimulation (600/280/250/210) delivered from the RV apex. The induced VT grossly resembled the spontaneous arrhythmia with only a slight difference in ECG lead II. VT was easily terminated with rapid RV pacing. At this point, we reached two conclusions: the first, that the spontaneous episodes of palpitations that occurred in our patient during a 40 year period were likely due to rapid VT; the second, that in the absence of both coronary artery disease and LV abnormalities, and based on the LBBB morphology of the VT, a right ventricular cardiomyopathy should be sought. Otherwise, the cause of VT would be classified as “idiopathic.”

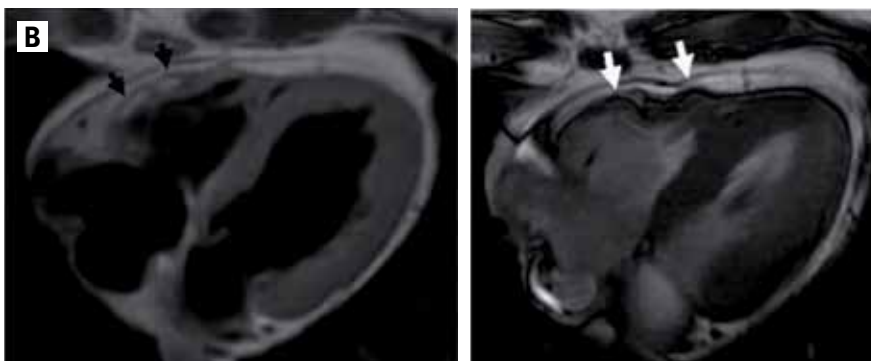
A repeat echocardiogram was performed and visualized by several echocardiographic experts with special focus on the RV. Again no abnormalities were found. In contrast, magnetic resonance imaging findings were consistent with the diagnosis of localized RV dysplasia [1,2] without left ventricular involvement [Figure B]. We therefore offered the patient three therapeutic options: a) implantation of an automatic cardioverter defibrillator, b) EPS-guided anti-arrhythmic therapy with sotalol, and c) radiofrequency ablation. The patient chose the second option, which we recommended.

During repeat EPS on sotalol (80 mg 3 times/day), only non-sustained VT (200/min, lasting ≤ 3 sec) could be induced

LBBB = left bundle branch block
 RV = right ventricular



[A] Twelve-lead ECG recorded at hospital admission showing a wide QRS complex tachycardia at 220/min and a pattern of left bundle branch block with left QRS axis



[B] Magnetic resonance imaging. Upper panel: axial T1 weighted black-blood image showing high signal intensity consistent with fatty replacement of the underlying basal free wall of the myocardium (arrows). Lower panel: axial balanced steady-state free precession (bright-blood) image showing focal aneurysms in the right ventricular free wall

with an aggressive protocol of ventricular stimulation including up to three extra-stimuli using two cycle lengths, two RV sites, and repetition ($n=5$) of the shortest coupling intervals, allowing ventricular capture. However, sustained well-tolerated VT (200/min) could still be induced at the eighth trial of triple extra-stimulation from the RV outflow tract. This VT could easily be terminated with rapid RV pacing.

Taking into account the relatively protective effect of sotalol on VT inducibility and the fact that the patient had a relatively benign 40 year course without anti-arrhythmic medication, we prescribed sotalol and decided against an ICD. During a follow-up of 4½ years the patient has remained asymptomatic on sotalol. Only a few ventricular premature complexes have been documented on serial Holter ECGs performed during that period.

COMMENT

In our patient the association of spontaneous and induced VT of LBBB-left QRS axis morphology and the MRI findings indicated a borderline probability that he suffered from arrhythmogenic right ven-

tricular cardiomyopathy/dysplasia according to the modified Task Force Criteria [2]. Another possible but less defined etiology is healed focal myocarditis. Nonetheless, in the absence of histologic confirmation or genotyping, a definite diagnosis cannot be made. ARVC/D is an inherited cardiomyopathy characterized by progressive fibro-fatty replacement of the RV myocardium [1] with involvement of the left ventricle usually late in the course of the disease. ARVC/D is seen predominantly in males, and 30–50% of cases have a familial distribution. Up to 80% of individuals with ARVC/D present with syncope or sudden cardiac death. The remainder frequently present with palpitations in association with ventricular premature complexes or VT. Symptoms are usually exercise-related.

The patient described here was 78 years old when the diagnosis of ARVC/D was suspected, i.e., much older than the age at which it is usually reported (between the second and fifth decades of life) [1]. A few cases of elderly people with this cardiomyopathy have been reported in the literature. The two oldest patients reported were both 82 years old: one was a man

ARVC/D = arrhythmogenic right ventricular cardiomyopathy/dysplasia

with a previous history of syncope that began 5 years earlier [3] and the second was a woman with sustained VT due to an initially diagnosed ARVC/D [4]. In fact, many of the reported diagnoses in patients older than 70 were made on autopsy [3].

The most interesting finding in our patient is that he had an ostensibly normal heart with a normal resting ECG and presented with a 40 year history of regular stress-induced palpitations, including at least one episode apparently terminated by carotid massage. This scenario is usually related to any of the major causes of PSVT, such as atrioventricular nodal reentry, reentry involving a concealed accessory bypass tract or, less commonly, atrial tachycardia. The fact that sustained VT but not SVT could be induced at EPS strongly suggests that the recurrent episodes of palpitations were caused by VT. Therefore, ARVC/D should be added to the list of possible causes of undocumented regular palpitations in ostensibly healthy patients. The apparent termination of one episode by carotid massage could be explained simply by a non-sustained VT event.

Another interesting observation in our patient was the lack of ECG abnormalities frequently observed in ARVC/D patients

ICD = implanted cardioverter defibrillator

(epsilon wave, negative T-waves in right precordial leads, right intraventricular conduction delay) [1], as well as the normal RV anatomy assessed by repeated echocardiograms. This contrasted with the MRI findings supporting the diagnosis of ARVC/D. The focalized character of the RV involvement may explain the lack of the typical ECG and echocardiographic abnormalities and the relatively excellent long-term clinical course. In addition, it supports the idea that the progression of disease over the years in some forms of ARVC/D may remain relatively limited. Finally, although implantation of a cardioverter defibrillator is increasingly recommended in the management of ARVC/D

patients, our case suggests that EPS-guided therapy with sotalol may also be a reasonable option [5]. Our patient was fortunate that his palpitations were diagnosed 40 years after they began. During that period, he enjoyed an active professional life with no any anti-arrhythmic medication, ICD or ablation. It is highly unlikely that he would have enjoyed such a peaceful clinical course otherwise.

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