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Transcatheter Ablation of Idiopathic Ventricular Fibrillation in a Patient with ICD and Arrhythmic Storm

We report the case of a 38-year-old male patient without coronary risk factors who was admitted in February 2012 at Hospital El Cruce after cardiac arrest secondary to ventricular fibrillation (VF). Anamnesis ruled out a family history of sudden death (SD) and other conditions; physical examination and ECG were normal, showing only signs of early repolarization in the inferior and lateral wall (slur). Cardiac magnetic resonance imaging (MRI) and coronary angiography (CAG) ruled out structural heart disease, and after 45-day recovery, a single-chamber cardioverter defibrillator was implanted (ICD). During the second year of follow-up, the patient underwent two shock therapies that corresponded to effective therapies of ventricular fibrillation episodes, according to the telemetry device interrogation.

In February 2015, the patient was readmitted at Hospital el Cruce due to arrhythmic storm. After shock therapies, the ECG showed signs of increased early ventricular repolarization (notch) in inferior and anterolateral leads, and ventricular premature beats (VPB) with short coupling interval on the ascending limb of the T wave (Figure 1A). Device telemetry data showed VPB with very short coupling interval causing VF episodes that reversed with shock therapy (Figure 1B). A subsequent Holter monitoring study revealed a high-density of VPB with R-on-T phenomenon.

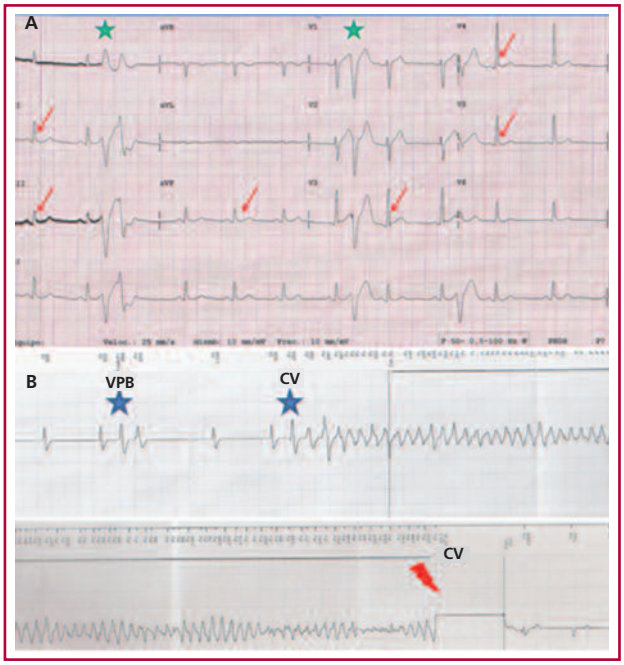

Fig. 1. A. 12-lead electrocardiogram showing ventricular premature beats (asterisks) with left bundle branch block, late R-wave transition in precordial leads, and R-on-T phenomenon. Early repolarization (slur and notch) are observed in the inferolateral wall (arrows). **B.** Implantable cardioverter defibrillator telemetry showing the episode of ventricular fibrillation gated by ventricular premature beats (VPB) that reverts with cardioversion (CV).

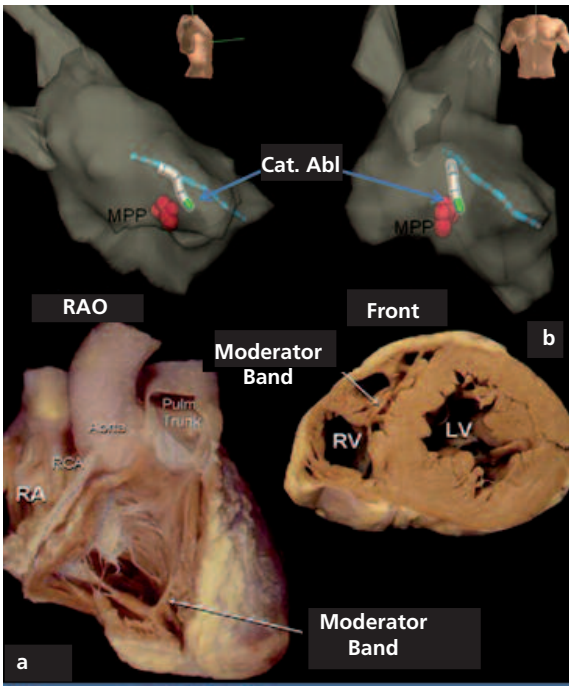

Fig. 2. a. Three-dimensional electroanatomical reconstruction of the right ventricle in right anterior oblique (RAO) and front views, showing the site of radiofrequency application (red spheres). **b.** Anatomical preparation showing the location of the moderator band in the right ventricle (RV).

The patient underwent transcatheter ablation guided by three-dimensional electroanatomical mapping (Ensite system). Once the procedure was initiated under general anesthesia, total absence of VPB was found despite several methods of basal stimulation and high-dose isoproterenol infusion, and even after discontinuation of anesthetic drugs, representing a serious limitation to treatment. During continuous infusion of high esmolol doses (500 μg/kg in 1 minute, followed by 100 μg/kg/min), return of VPB similar to those causing VF was achieved and localized on the moderator band in the right ventricle (Figures 2 a & b).

In this anatomic site, the endocavitary electrogram showed greater precocity, fascicular initial fast deflection, and perfect pace-mapping. The use of radiofrequency on that area accelerated the occurrence of ventricular rhythm similar to the morphology at VF onset, but was relieved after a few seconds due to permanent disappearance of VPB (Figure 3).

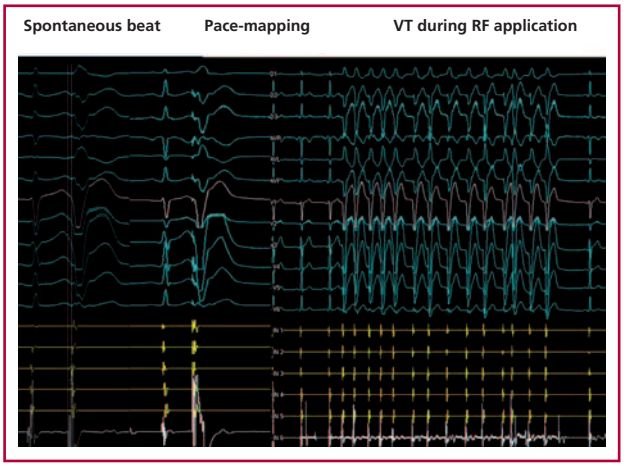

Fig. 3. Simultaneous surface and endocavitary electrocardiographic recordings showing, from left to right, clinical ventricular premature beats, pace-mapping at the site of application and time of radiofrequency application.

Since then, the patient did not repeat events, and absence of ventricular arrhythmia was confirmed both in stress test and Holter monitoring.

Sudden death accounts for 50% of cardiovascular deaths and 25% in adults, out of which 6-14% are individuals with no structural heart disease, many of them as debut. Most of those deaths are associated with known electrocardiographic patterns, such as long or short QT interval, Brugada syndrome, etc. However, in some cases the ECG signs are unclear, being identified as idiopathic ventricular fibrillation. (2)

For decades, early repolarization characterized by baseline J-point elevation on the 12-lead ECG has been considered as benign. However, its prevalence particularly in inferior and/or lateral leads has been associated with ventricular fibrillation vulnerability.

To avoid confusion with the early-repolarization pattern commonly found in young adults and trained athletes, whose J point and ST segment are elevated in V2-V4 precordial leads, the "inferolateral J-point elevation syndrome" associated with ventricular fibrillation is defined as J-point elevation manifested as a slow transition from the QRS segment to the ST segment (slur) or as a positive deflection inscribed at the end of the R wave (notch), with ST-segment elevation with upper concavity >1 mm in inferior leads (II, III, aVF) or lateral leads (DI, aVL, V5, V6), or in both. (2-5) This pattern is usually associated with sinus bradycardia and increased vagal tone, U wave, relatively short QT interval, vertical QRS axis, and attenuation or disappearance with exercise.

Evidence has associated it with idiopathic VF, increasing 4 times the risk of cardiac death in young men. When it occurs in inferior and anterior leads associated with greater magnitude of J-point elevation (>2 mm) and horizontal/descending ST segment, it identifies an ECG profile with 10-fold higher risk of arrhythmic death, especially in young men. However, due to its high prevalence in the general population, detecting those criteria in a routine ECG in asymptomatic individuals with no family history of SD is not enough to account for a preventive therapy or a special follow-up.

Recently, the results of some few series of patients undergoing transcatheter ablation were published, whose origin was strongly associated with the moderator band of the right ventricle. (6)

Evidence suggests that radiofrequency ablation, although technically difficult, can be initially successful but nearly 50% of the patients require a second procedure. Initially successful ablation does not rule out cardioverter defibrillator implantation due to the high incidence of late relapses reported. (7)

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