Letters to the Editor

Concealed Channelopathy in a Patient With Recurrent Ventricular Fibrillation Treated With Catheter Ablation

Fibrilación ventricular recorrente tratada mediante ablación con catéter en paciente con canalopatía oculta

To the Editor,

Idiopathic ventricular fibrillation (VF) is responsible for approximately 8% of the cases of sudden cardiac death, and occult channel defects have been reported to be among the possible causes. On rare occasions, it is triggered by an electrical storm, in which case catheter ablation is the only alternative when drug therapy fails.

We present the case of a 53-year-old woman with nothing remarkable in her medical history, who had been asymptomatic until she was admitted to the hospital with a syncopal episode. Monitoring demonstrated the presence of polymorphic ventricular tachycardia with spontaneous cessation and frequent monomorphic premature ventricular contractions (PVC), with left bundle branch block and left superior axis morphology and a short coupling interval that triggered repeated episodes of VF, requiring defibrillations on 20 occasions over a period of 72 h.

The baseline electrocardiogram was normal. Metabolic disorders, toxic substance intake and structural heart disease were ruled out by means of echocardiography, coronary angiography, cardiac magnetic resonance imaging, and the determination of adrenaline and flecaïnide concentrations.

A first electrophysiology study (EPS) was carried out as an emergency measure because the frequent recurrences of VF persisted despite amiodarone infusion, requiring defibrillations. The last episode occurred during transfer to the electrophysiology laboratory where, after introduction of catheters into the His bundle and the apex of the right ventricle (RV), neither PVC nor other arrhythmias developed.

Four hours after completion of the EPS, the VF-inducing PVC reappeared, requiring further defibrillations that were not prevented by the infusion of isoprenaline, beta blockers, or procainamide; for this reason, the EPS was repeated the next day. From mid septum to apical septum of the RV, Purkinje potentials were recorded in relation to the distal insertion of the right branch, potentials that preceded the initiation of the QRS of the self-limiting VF-inducing PVC by 45 ms on one occasion (Fig. 1). Applications of radiofrequency suppressed the PVC and the episodes of VF. Seven days later, a single-chamber automatic defibrillator was implanted and, after 10 months of drug-free follow-up, the device has recorded no ventricular arrhythmias.

Figure 1. A: electrocardiographic tracing at hospital admission with an episode of ventricular fibrillation initiated by premature ventricular contraction. B: electroanatomic mapping of right ventricle, using the CARTO® system, in which no low voltage zones were observed. The sites to which radiofrequency energy was applied are indicated in red. C: right oblique fluoroscopic view showing catheters in right atrium and right ventricular apex and ablation in apical septum. D: Purkinje potential at the site of the origin of the premature ventricular contractions. ABL, ablation; RA, right atrium; RV, right ventricle; P, Purkinje potential.

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genetic study detected a novel frame-shift mutation in the sodium channel alpha subunit gene SCN5A (D1816fs), which results in early transcriptional arrest and produces a truncated or shortened protein.

The major mechanism of sudden cardiac death is VF and the most common cause is ischemic heart disease. Ventricular fibrillation is considered to be idiopathic when there is no evidence of structural heart disease, cardiotoxicity, electrolyte disorders, or a predisposing hereditary condition.\(^1\)

Although PVC is a benign arrhythmia, there are cases in which PVC originating in the Purkinje system or in the RV outflow tract initiates VF episodes in individuals without heart disease, with structural heart disease, or with channel defects. The absence of heart disease and the early development of PVC fit well with the entity described by Leenhardt et al.\(^1\) Haissaguerre et al reported the efficacy of radiofrequency ablation in PVC in the prevention of new episodes of VF over the short and long terms.\(^5\)

This case is singular for 2 reasons. On one hand, it highlights the efficacy of ablation in this desperate situation and the difficulty derived from the mechanical suppression of the focus via catheter. On the other hand, it describes a novel mutation in the SCN5A gene as a cause of idiopathic VF with a normal electrocardiogram. Thus, it involves an occult channel defect of which VF was the first sign. The incomplete penetration of channel defects explains the fact that individuals with VF who have an apparently normal heart and in whom there is no electrocardiographic evidence of this condition exhibit electrical instability in relation to a genetically determined occult arrhythmogenic disease.\(^1\)

Luz Muñoz,\(^a\) Luis Tercedor,\(^b, c\) Miguel Álvarez,\(^b\) and Ricardo Sebastián Galdeano\(^d\)

\(^a\)Servicio de Cardiología, Hospital Carlos Haya, Málaga, Spain
\(^b\)Unidad de Arritmis, Servicio de Cardiología, Hospital Universitario Virgen de las Nieves, Granada, Spain

*Corresponding author:
E-mail address: luis.tercedor.sspa@juntadeandalucia.es (L. Tercedor).

Available online 17 April 2011

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doi:10.1016/j.rec.2010.11.023

Does the Metabolic Syndrome Need More Descriptive Studies or More Evidence of Its Implication in Secondary Prevention?

¿El síndrome metabólico en España necesita más estudios descriptivos o más evidencia de su implicación en prevención secundaria?

To the Editor,

We have read with interest the article “Connection Between the Early Phases of Kidney Disease and the Metabolic Syndrome” recently published in Revista Española de Cardiología. The authors describe the association of the metabolic syndrome (MS) and early kidney disease (EKD) with carotid intima-media thickening (CIMT).\(^1\) Interestingly, although they assessed the relationship between a group of risk factors and the presence of subclinical and early vascular lesions, they did not discuss its actual implications regarding prevention or the treatment of these patients. In fact, it remains to be clarified whether these lesions should be considered to be the 2 diagnostic criteria of the MS; this has already been proposed for C-reactive protein, which, besides presenting higher levels in subjects with the MS, improves risk prediction of cardiovascular disease.\(^2\)

In addition, EKD and CIMT are 2 disorders that can be stabilized but are only resolved with difficulty, especially when the values are close to normal: the mean glomerular filtration rate was 87 mL/min/1.73 m\(^2\) and CIMT ranged between 0.6 mm and 0.7 mm. Moreover, clinical trials that have investigated CIMT regression or changes in EKD have not found a correlation with improved cardiovascular prognosis.\(^3, 4\) Suggesting that they behave as markers of vascular injury rather than as risk factors. In contrast, studies based on the MESYAS (Metabolic Syndrome in Active Subjects) registry have shown that the lipid components of the MS, as measured by the ratio of triglyceride to high-density lipoprotein (HDL), are very specific markers for the presence of other components of the MS\(^5\) and, more importantly, one of the major risk factors for myocardial infarction.\(^6\) This association has also been described both for the presence of the MS and for the additive effect of its components.\(^7\)

Finally, the authors considered high blood pressure and diabetes as exclusion criteria in the study.\(^1\) when it has been reported that these are 2 of the main factors associated with the presence of the MS in the Spanish workforce.\(^8\) Furthermore, they state that they are unaware of the existence of previous data that associate the MS with EKD. Nevertheless, in 2004 and 2005, respectively, data from the NHANES III (National Adult Health Examination Survey) registry\(^9\) and the MESYAS registry\(^10\) were published regarding this association and coincided regarding the lack of an association between HDL and kidney damage.

We believe that the authors describe an association with few implications for cardiovascular risk stratification, while the actual implication of the MS in patients with established cardiovascular disease remains to be elucidated. During the last decade, the MS has given rise to great interest due to its relevance in the prevention of diabetes mellitus and cardiovascular disease; however, its relevance in patients with established cardiovascular disease remains undefined and unaccepted.

Alberto Cordero,* José Moreno-Arribas, Vicente Bertomeu-González, and Vicente Bertomeu-Martínez

Departamento de Cardiología, Hospital Universitario de San Juan, San Juan de Alicante, Alicante, España

*Corresponding author:
E-mail address: acorderofort@gmail.com (A. Cordero).

Available online 11 August 2011