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CASE REPORT

Epicardial ablation for prevention of ventricular fibrillation in a patient with Brugada Syndrome☆

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PALAVRAS-CHAVE
Síndrome de Brugada;
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Mapeamento

Abstract We present the case of a 60-year-old woman with Brugada syndrome, permanent type 1 electrocardiographic pattern, who had previously received an implantable cardioverter-defibrillator. She suffered frequent syncopal episodes and multiple appropriate shocks (around five per month) due to polymorphic ventricular tachycardia/ventricular fibrillation, refractory to quinidine therapy. Combined epicardial and endocardial electroanatomical mapping was performed with a view to substrate ablation. An area of abnormal fractionated electrograms, lasting up to 370 ms and up to 216 ms after the end of the surface QRS, was identified in the epicardium in the lower anterior part of the right ventricular outflow tract. Extensive epicardial ablation of this area, which eliminated the fractionated electrograms, led to the disappearance of the Brugada electrocardiographic pattern six weeks after ablation. Despite discontinuation of quinidine, no further ventricular arrhythmias occurred during follow-up, which is still of short duration.

KEYWORDS
Brugada syndrome;
Catheter ablation;
Epicardial ablation;
Electrophysiology;
Mapping

Ablação epicárdica para prevenção da fibrilhação ventricular em doente com síndrome de Brugada

Resumo É apresentado o caso de uma doente de 60 anos com síndrome de Brugada, padrão tipo 1 permanente, portadora de cardioversor-desfibrilhador, com episódios frequentes de síncope por taquicardia ventricular polimórfica/fibrilhação ventricular (cerca de cinco por mês), refratários à terapêutica com quinidina e com múltiplos choques apropriados. Foi efetuado

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mapeamento eletroanatómico endocárdico e epicárdico do ventrículo direito, em ritmo sinusal, confirmando-se a presença de uma área epicárdica na região anterior da câmara de saída ventricular direita com eletrogramas anómalos, fracionados e de longa duração (até 370 ms), que se prolongavam até 216 ms após o término do QRS de superfície. A ablação epicárdica alargada dessa área, com abolição dos eletrogramas anómalos, conduziu ao desaparecimento do padrão de Brugada na reavaliação eletrocardiográfica efetuada às seis semanas. Apesar da suspensão da terapêutica com quinidina, não ocorreram novas disritmias ventriculares, durante o seguimento, ainda de curta duração.

Introduction

It is now over twenty years since Pedro and Josep Brugada first described the association between primary ventricular fibrillation (VF) and an electrocardiographic pattern of right bundle branch block with coved ST-segment elevation in the right precordial leads in individuals with no structural heart disease.[1] Since then, there has been major progress towards better understanding of the genetic and pathophysiological mechanisms of Brugada syndrome, particularly the hypothesis of a voltage gradient between the endocardium and the epicardium during depolarization as the mechanism behind the electrocardiographic abnormalities and associated ventricular arrhythmias.[2]

The fact that electrocardiographic abnormalities were observed in the right precordial leads, and thus originating in the right ventricular outflow tract (RVOT), soon raised the suspicion that this was the origin of arrhythmias in these patients. However, initial attempts at endocardial mapping of this region failed to identify the arrhythmic substrate. Various cases were reported in the literature in which endocardial ablation was attempted to prevent recurrence of VF in patients with Brugada syndrome and frequent ventricular extrasystoles originating in the RVOT.[3–6] Nevertheless, such an approach has important limitations, given that frequent ventricular extrasystoles are not common in these patients, making them difficult to map.

Nademanee et al.[7] recently described epicardial ablation of areas of delayed depolarization located in the anterior RVOT in Brugada syndrome. This study was a landmark in increasing knowledge of the pathophysiology of the syndrome, as well as providing evidence for an apparently effective therapeutic approach.

Case report

A 60-year-old woman had a history of frequent syncopal episodes for the past 10 years, unrelated to exertion or posture and usually preceded by dizziness and palpitations. Five years previously, she had suffered respiratory arrest during sleep, with generalized tonic-clonic movements. Four years ago, one of her sons suffered sudden death at the age of 39, which prompted investigation of the family. This revealed type 1 Brugada electrocardiographic pattern in the patient and another son, and both received an implantable cardioverter-defibrillator (ICD) in May 2009.

Direct sequencing of the SCN5A gene identified no pathogenic mutation. The polymorphisms c.87A>G (p.Ala29Ala) and c.3183A>G (p.Glu1061Glu) in homozygosity and c.5457T>C (p.Asp1819Asp) in heterozygosity, previously described as non-pathogenic variants, were identified.

Frequent recurrences of polymorphic ventricular tachycardia (VT) were subsequently documented coinciding with syncopal episodes, and three appropriate shocks for VF. The frequency of arrhythmic events increased progressively, despite quinidine therapy up to the maximum tolerated dose of 400 mg/day. In the six months prior to the ablation procedure, the patient had a mean of five episodes (4–8) of polymorphic VT/VF per month.

In July 2013, still under quinidine therapy, the patient underwent electrophysiological study. She presented sinus rhythm, with bifascicular block (complete right bundle branch block and left anterior hemiblock) and type 1 Brugada repolarization abnormalities in the right precordial leads and in the frontal plane leads DII, DIII and aVR (Figure 1). Occasional monomorphic ventricular extrasystoles were documented, with left bundle branch morphology, inferior axis, and QRS transition in V4, the frequency of which decreased spontaneously during the exam. During programmed ventricular stimulation, only self-limited runs of polymorphic VT (lasting up to 7 s) were induced, without VF being triggered.

Endocardial mapping of the right ventricle was then performed in sinus rhythm, using the CARTO 3 system (Biosense Webster, Diamond Bar, CA) with an irrigated catheter (Thermocool SF, Biosense Webster). No low-voltage areas were identified, nor zones with local electrograms prolonged beyond the end of the QRS complex (Figure 2). Epicardial access was obtained through subxiphoid puncture under fluoroscopic guidance, and a 9F introducer connected to a passive drainage system was inserted. High-density epicardial electroanatomical mapping of the right ventricle (304 points) and left ventricular (LV) anterior, lateral and posterior walls was performed. Voltage mapping showed normal epicardial voltages in both the left ventricle and RVOT (Figure 3). Mapping of the duration of the bipolar electrogram showed that this ended during the QRS complex in all LV regions mapped. By contrast, the bipolar electrogram terminated after the end of the QRS complex