Epsilon-Like Electrocardiographic Pattern in a Patient with Brugada Syndrome

Ozcan Ozeke, M.D.,* Umut Y. Cavus, M.D.,† Ilyas Atar, M.D.,‡
Bulent Ozin, M.D.,‡ and Erdogan Ilkay, M.D.*

From the *Department of Cardiology and †the Emergency Department, MESA Hospital, Ankara, Turkey; and ‡Department of Cardiology, Baskent University Faculty of Medicine, Ankara, Turkey

Both Brugada syndrome (BrS) and arrhythmogenic right ventricle dysplasia/cardiomyopathy (ARVD/C) can cause repolarization abnormalities in right precordial leads and predispose to sudden cardiac death (SCD) due to ventricular arrhythmias. Although there is controversy over whether BrS is distinct from ARVD/C, it is believed that both are different clinical entities with respect to both the clinical presentation and the genetic predisposition. The coexistence of these two relatively rare clinical entities is also reported, but, some hypothesized that it is more possible that disease of the right ventricular muscle might accentuate the Brugada electrocardiographic pattern. In clinic practice, there may be cases where the dividing line is not so clear. We report a 33-year-old male presenting with recurrent syncope, who has a peculiar pattern of coved-type ST-segment elevation (ST-SE) with epsilon-like wave in right precordial leads.

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CASE REPORT

A 33-year-old male, who had a history of one syncope episode after alcohol uptake, presented with a syncope event immediately after a large meal. Physical examination was unremarkable. Hematological and biochemical tests were within normal limits. Cardiac echocardiogram (ECG) was normal. His first ECG taken in the emergency department showed characteristic Brugada pattern with coved-type ST-segment elevation (ST-SE) in leads V₁–V₂ (Fig. 1A); and the patient was hospitalized. Later in the day, the ECG showed a discrete wave resembling epsilon wave just beyond the QRS complex (Fig. 2B); the second ECG taken to exclude baseline artifact revealed also similar tracing (Fig. 2B). We planned cardiac magnetic resonance imaging (MRI), which showed no cardiac abnormality. Subsequently, drug challenge test with propafenone (2 mg/kg intravenous infusion over 10 minutes) was performed the next day and revealed the conversion of type 3 to type 2 ST-SE (Figs. 1B–D). Electrophysiological study showed a

normal AH (110 ms) and HV (45 ms) intervals. Ventricular fibrillation was easily induced by programmed stimulation in right ventricular outflow tract. Given the recurrent syncope and these findings, this patient fulfilled the diagnosis of Brugada syndrome (BrS) and an internal cardiac defibrillator was implanted.

DISCUSSION

The Brugada syndrome is characterized by ST-SE in the right precordial ECG leads and a high incidence of sudden cardiac death (SCD) in patients with seemingly normal structural hearts. The syndrome manifests primarily during adulthood, with a mean age of SCD of approximately 40 years. ECG abnormalities constituting the hallmark of this entity have been recently proposed by a Consensus Report.¹ ECG pattern may be dynamic over time and may include transient normalization. Among the three types of ECGs, only type 1 Brugada ECG pattern in more than one right precordial lead in baseline ECG or during drug challenge with sodium

Address for reprints: Ozcan Ozeke, M.D., Department of Cardiology, MESA Hospital, Yasam Caddesi No: 5, Sogutozu, 06510 Ankara, Turkey. Fax: +90 292 99 10; E-mail: ozcanozeke@gmail.com

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Figure 1. Twelve-lead electrocardiogram tracings show characteristic Type 1 “coved-type” ST-segment elevation in V_1 – V_2 (A) and conversion of Type 3 ECG (B) to Type 2 ECG after 5 minutes (C) and 10 minutes (D) following intravenous administration of 150 mg propafenone.

channel blockers is diagnostic for the syndrome when this is presented in a patient with one of the following features: documented ventricular fibrillation (VF), polymorphic ventricular tachycardia (VT), a family history of SCD at <45 years of age, type 1 Brugada ECGs in family members, inducibility of VT with programmed electrical stimulation, syncope, or nocturnal agonal respiration. According to consensus report, drug challenge should not be performed in patients with baseline type I Brugada ECG in any documented ECG tracing, and should only be used in patients with symptoms related to the disease where Brugada ECG pattern is suspected (type 2 or 3 Brugada ECG). However, we performed the drug challenge to clarify the diagnosis since we could not be sure which symptoms related to the BrS or other causes such as vasovagal syncope precipitated by alcohol uptake. Indications for termination of drug challenge are development of a diagnostic type 1 Brugada ECG,

a ≥ 2 -mm increase in ST-SE in patients with a type 2 Brugada ECG, the development of ventricular premature beats or other arrhythmias, or widening of the QRS $\geq 30\%$ above baseline.¹

Arrhythmogenic right ventricle dysplasia/cardiomyopathy (ARVD/C) an inherited heart muscle disease that predominantly affects the right ventricle, is characterized pathologically by myocardial atrophy with fibrofatty replacement and clinically by ventricular electric instability with VT or VF that may lead to SCD, primarily in young people and athletes. Clinical diagnosis of this rare syndrome is often difficult because of the nonspecific nature of disease features and the broad spectrum of phenotypic manifestations.²

ARVD/C and BrS are distinct clinical entities with respect to both the clinical presentation and the genetic predisposition.³ In BrS, imaging techniques such as echocardiography, angiography, MRI, and radionuclide scintigraphy show no

Figure 2. ECG tracings taken different times (A and B) show epsilon-like pattern (arrows) in V₂ (B).

evidence of overt structural heart disease, whereas ARVD/C patients characteristically display right ventricular morphological and functional changes (such as global dilatation, bulgings/aneurysms, and wall motion abnormalities). Ventricular arrhythmias in ARVD/C are most commonly "monomorphic" VT (left bundle branch block type), often precipitated by catecholamines or exercise, accounting for SCD of young competitive athletes. In contrast, ST-SE and arrhythmias in Brugada patients are enhanced by vagotonic agents or β -adrenergic blockers, and "polymorphic" VT most commonly occurs during rest or sleep. Unlike BrS, the ECG abnormalities in ARVD/C are not dynamic, displaying a constant T-wave inversion, epsilon waves, and in the progressive stage, reduction of the R amplitude, which are largely unaffected by sodium channel blocker administration.^{1,4} However, clinically there is some crossover between these conditions, and some investigators feel that the BrS is really a form of ARVD/C and an association between these two syndromes can be supposed according to several reports.⁵⁻¹¹ Ep-

silon wave is said to be pathognomonic of ARVD/C; but, coexistence of the epsilon and Brugada waves on the same tracing has been reported in patients with ARVD/C.⁸ Corrado et al. presented data in support of the hypothesis that a subpopulation of patients with ARVD/C, referred to as "concealed forms," present with the typical clinical and electrocardiographic features of the BrS, including the presence of type 1 ST-segment elevation and polymorphic VT. The concealed forms of ARVD/C might be a cause of so-called "idiopathic" ventricular arrhythmias in subjects with apparently "normal hearts." These findings show an overlap in clinical manifestation and mechanisms of ventricular arrhythmias between patients with ARVD/C and BrS.⁸ It is not impossible, given the fact that in early stage of ARVD/C, fatty deposits can be so minimal or segmental lesion that they may not be detectable, therefore, it is necessary to consider the possibility of ARVD/C in patients with the BrS. In this case, since we could not detect any structural heart disease with echocardiography or cardiac MRI, we used the

"epsilon like" term, and the patient was diagnosed as BrS.

REFERENCES

1. Antzelevitch C, Brugada P, Borggrefe M, et al. Brugada syndrome: Report of the second consensus conference: Endorsed by the Heart Rhythm Society and the European Heart Rhythm Association. *Circulation* 2005;111:659-667.
2. Corrado D, Fontaine G, Marcus FI, et al. Arrhythmogenic right ventricular dysplasia/cardiomyopathy: Need for an international registry. Study Group on arrhythmogenic right ventricular dysplasia/cardiomyopathy of the working groups on myocardial and pericardial disease and arrhythmias of the European Society of Cardiology and of the Scientific Council on Cardiomyopathies of the World Heart Federation. *Circulation* 2000;101:E101-E106.
3. Pérez Riera AR, Antzelevitch C, Schapacknik E, et al. Is there an overlap between Brugada syndrome and arrhythmogenic right ventricular cardiomyopathy/dysplasia? *J Electrocardiol* 2005;38:260-263.
4. Peters S, Trümmel M, Denecke S, et al. Results of ajmaline testing in patients with arrhythmogenic right ventricular dysplasia-cardiomyopathy. *Int J Cardiol* 2004;95:207-210.
5. Bjerregaard P, Molgaard H. A patient with biventricular dysplasia. *J Electrocardiography* 2002;35:289-290.
6. Schmidt T, Gerckens U, Ortmeyer D, et al. [Brugada syndrome or ARVD (arrhythmogenic right ventricular dysplasia) or both? Significance and value of right precordial ECG changes]. *Z Kardiol* 2002;91:416-422.
7. Erkapic D, Neumann T, Schmitt J, et al. Electrical storm in a patient with arrhythmogenic right ventricular cardiomyopathy and SCN5A mutation. *Europace*. 2008;10:884-887.
8. Corrado D, Basso C, Buja G, et al. Right bundle branch block, right precordial ST-segment elevation, and sudden death in young people. *Circulation* 2001;103:710-717.
9. Marras E, Basso C, Sciarra L, Delise P. Unexplained syncope, Brugada-like ECG and minimal structural right ventricular abnormalities: which is the right diagnosis? *J Cardiovasc Med (Hagerstown)*. 2009;10:273-275.
10. Das MK, Zipes DP. Fragmented QRS: a predictor of mortality and sudden cardiac death. *Heart Rhythm*. 2009;6(3 Suppl):S8-14.
11. Ahn J, Hurst JW. Worrisome thoughts about the diagnosis and treatment of patients with Brugada waves and the Brugada syndrome. *Circulation* 2004;109:1463-1467.