CASE REPORT

Coexistence of Wolff–Parkinson–White and Brugada Syndrome: Mere Curiosity?

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The association between Brugada syndrome (BS) and ventricular preexcitation is a rare condition, with sporadic cases already reported. We report the case of a 29-year-old man, with palpitation unrelated to physical or emotional stress. The electrocardiogram of the first visit revealed a ventricular preexcitation pattern and an end-conduction delay, with negative T wave in V₁ and intraventricular conduction disturbance in V₂ (atypical for BS). The typical aspect of BS occurred after introduction of propafenone for the prevention of atrioventricular tachycardia. We discuss the recognition of this rare association, the proarrhythmic effects of some drugs, treatment options, and prognosis.

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arrhythmias; Brugada syndrome; Wolff–Parkinson–White syndrome

INTRODUCTION

You see only what you look for.

You recognize only what you know.

—Dr. Merril C. Sosman

Brugada syndrome (BS) is characterized by a specific electrocardiographic pattern in the precordial leads in the absence of ischemia, electrolyte disturbances, or structural heart disease. It increases the susceptibility to ventricular arrhythmias and sudden cardiac death (SCD).¹ Supraventricular arrhythmias are also common because the arrhythmogenic substrate should not be restricted to the ventricles and atrial fibrillation is the most frequent arrhythmia.^{2,3} Although many cases have been described, BS is a rare entity, with a global prevalence of 1–5 cases/10,000.⁴ Some patients come from affected families, but there are also sporadic cases.

The Wolff–Parkinson–White syndrome (WPW) has a higher prevalence, 10–30 cases/10,000. Based on these data, we conclude that the probability of association of these two entities (BS and WPW) is extremely low, around 1–15/10,000,000, different from the estimate of De Roy et al.⁵

However, in terms of the world population, this could involve a large number of patients at risk for arrhythmias and SCD. Early recognition of this concomitance could prevent any misconduct that might increase the risk in these patients.

CASE REPORT

A 29-year-old man was admitted to our hospital because of palpitation unrelated to physical or emotional stress. There was no personal or family history of syncope or early sudden death. The electrocardiogram (ECG) of the first visit revealed an intermittent ventricular preexcitation, with a short PR, delta wave, wide QRS with an end-conduction delay, intraventricular conduction disturbance in V₂, and negative T wave in V₁ (Fig. 1). Due to this electrocardiographic aspect, the higher intercostal space leads V₁ to V₃ suggested by Sangwatanaroj et al.⁶ were recorded, but they were inconclusive.

Laboratory tests and echocardiography were normal. Propafenone was started for the preventive treatment of atrioventricular tachycardia, with remission of the symptoms. At the 2 years follow-up visit, a Brugada type 1 pattern was observed (Fig. 2).

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Figure 1. Baseline 12-lead ECG of the patient at admission. Note the preexcitation with a suggestive “Brugada” pattern.

Figure 2. Twelve-lead ECG after oral propafenone therapy, without preexcitation, but showing type 1 Brugada pattern.

This resulted in the suspension of the drug. However, even after propafenone discontinuation, there was no change in the ECG, configuring a spontaneous type 1 Brugada pattern, although the patient was still complaining of palpitations. Ac-

cessory pathway radiofrequency catheter ablation (RFCA) and the electrophysiological study (EPS) were then indicated.

A successful left posterior accessory pathway RFCA was performed. After that, stimulation

Figure 3. Twelve-lead ECG after 2 years propafenone discontinuation.

with 430-ms cycle to S3 (200 ms) in the right ventricular outflow tract resulted in induced ventricular fibrillation (VF), which was promptly cardioverted. It was then indicated an implantable cardioverter defibrillator (ICD), implanted without any complications.

Two years after the RFCA, the patient remained asymptomatic, without drug therapy and the Brugada ECG pattern was still the same (Fig. 3). The patient had no ICD therapies until this period.

DISCUSSION

The association between BS and ventricular pre-excitation was first described in 2001 by Eckardt et al.⁷ Subsequently, six other cases of this association have been reported.^{5,8–12} The uniqueness of this case was that the typical ST segment elevation occurred only after administration of propafenone for pharmacological treatment of WPW syndrome. Propafenone, a sodium channel-blocking drug played a role of a provocative test, and could also potentially harm the patient. It is also known that in individuals with suspected BS certain drugs (i.e., propafenone, procainamide, lithium, and clomipramine) should be administered with caution and under more frequent surveillance. It

is debatable whether these drugs could induce such changes in these patients without a proper “genetic” substrate. In our case report, even after 2 years of RFCA and 4 years without propafenone, the type 1 BS ECG pattern was still maintained, ruling out the possibility of cardiac memory either by propafenone or the presence of anomalous pathway. Recently, Chevallier et al.¹³ described the electrocardiographic criteria, based on angular measures alpha and beta, to distinguish incomplete right bundle branch block of types 2 and 3 Brugada pattern. Of note, the application of these criteria, retrospectively in our case, could predict the positivity of pharmacological response.

Another question that arises is whether these patients with both syndromes would be more susceptible to arrhythmias and SCD, and could be candidates to receive a prophylactic ICD. Of the seven cases reported, four patients received an ICD,¹² two patients refused the implant, and one patient received no ICD because he was asymptomatic and ventricular tachycardia (VT) or VF was not induced at EPS. In individuals without BS, a sustained polymorphic VT or VF is exceptionally induced. Conversely, it is estimated that inducible VT/VF in patients with BS can occur in up to 80% of the cases. Of note, one-third of these individuals present with a first polymorphic

ventricular tachycardia or ventricular fibrillation within 2 years of follow-up.¹⁴ Although there are controversies regarding the ICD implantation and EPS in these patients,^{15,16} it is known that the prognosis could not be so benign in asymptomatic patients with spontaneous type 1 BS ECG pattern. The only available treatment is the ICD, because antiarrhythmic drugs, like amiodarone or beta-blockers, do not protect against SCD. Quinidine and cilostazol could be an option to prevent SCD, but these drugs still deserve more conclusive studies for this specific indication. In our particular case, we made the option for ICD implantation due to the spontaneous type 1 BS pattern, VF inducibility at EPS, and also by the fact that the patient lives far from a major urban center (a rural property).

CONCLUSION

Although rare, the coexistence of BS and WPW is a possibility that should be investigated in suspected cases. Propafenone, a common drug for preventing arrhythmia recurrences in patients with WPW could potentiate and unmask latent or unapparent BS. In these exceptional cases, restrictive recommendations should be made on the use of some drugs that may be even harmful to these patients. After RFCA of anomalous pathway, although controversial, EPS stratification and ICD should be considered as an option for these patients.

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