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An unusual Brugada syndrome case

Nadir görülen bir Brugada sendromu vakası

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Introduction

Brugada syndrome (BS) is characterized by right bundle branch block (RBBB) pattern with ST-segment elevation in right precordial leads and a propensity for sudden cardiac death due to ventricular arrhythmias. We present the case of a 20-year-old man with an episode of syncope followed by ventricular fibrillation (VF)-induced cardiac arrest.

Case Report

A 20-year-old man who had developed syncope recovered completely during his transport to the nearest hospital. Electrocardiogram (ECG) findings in the emergency department were as follows: 1) J waves in many leads without ST elevation, with a simultaneous Mobitz type II second-degree atrioventricular (AV) block (Fig.1), 2) atrial fibrillation (AF), and 3) monomorphic ventricular tachycardia (VT). During his examination, the patient suddenly lost consciousness and his ECG revealed VF. Direct current countershocks were delivered immediately. Cardiopulmonary resuscitation was performed and the patient was intubated. He was transferred to our hospital for further examination and therapy. Shortly after his arrival at our hospital, he was extubated.

Our evaluation of the patient's medical history revealed that he had been the victim of a traffic accident one week prior. He had been rarely taking analgesic medication (Tenoxicam, 20 mg orally) for widespread aches produced by the accident. He had no personal history of arrhythmia, syncope or seizure, and no family history of sudden death. He was not taking any other medications beyond the prescribed analgesic. His electrolyte levels, chest x-ray, echocardiography, and coronary angiogram were normal. His cerebral computerized tomography was also normal. His ECG revealed a RBBB and coved-type marked ST elevation together with negative T waves in the V1-V3 leads (Fig. 2). The patient was diagnosed with type 1 BS.

The patient was followed for recurrence of arrhythmias for 5 days. No antiarrhythmic drugs were prescribed. He was then transferred to another hospital for electrophysiological study (EPS). According to the EPS report, the related times and intervals were within the normal ranges. Ventricular tachycardia was not induced by extra-stimulus. Due to typical Brugada type ECG and cardiac arrest attack, implantable cardioverter defibrillator was implanted.

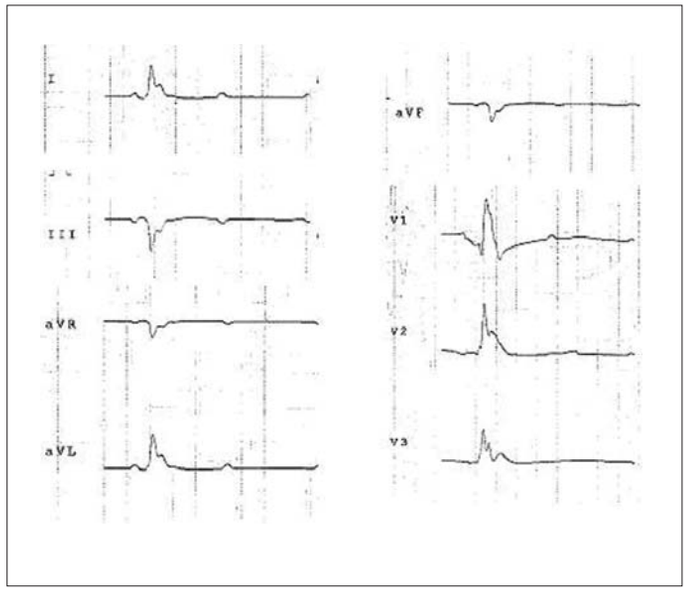

Figure 1. J waves in many leads and Mobitz type II second-degree atrioventricular block

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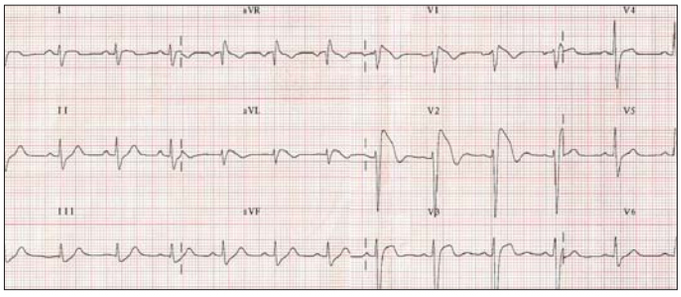

Figure 2. Brugada type I electrocardiogram

Discussion

The presently described BS patient presented some unusual features. Firstly, an arrhythmia episode occurred one week after a traffic accident. Secondly, his ECG exhibited J waves in many leads that were simultaneous with a Mobitz type II second-degree AV block.

Brugada syndrome is an inherited cardiac disorder with variable ECG features. Its clinical diagnosis is based on ECG pattern, syncope, and sudden death from VF in patients without any structural heart abnormalities. The ECG features of BS are dynamic and often concealed. Three types of ECG abnormalities have been recognized. Type 1 BS is characterized by a coved-type ST-segment elevation of ≥2 mm followed by a negative T-wave in leads V (1) to V(3). Type 2 BS is characterized by a high take-off, ≥2 mm, ST-segment elevation that gives rise to a gradually descending ST-segment elevation in a saddleback configuration, followed by a positive or biphasic T-wave. Type 3 BS involves a right precordial ST-segment elevation of ≤1 mm with a saddleback and/or coved configuration.

The J wave is produced by a transmural voltage gradient during initial ventricular repolarization, which results from the presence of a prominent action potential notch mediated by the transient outward potassium current (I_{to}) in the epicardium (1). Clinical conditions that are associated with J waves include early repolarization syndrome, BS and idiopathic VF. A previously described male Japanese patient with BS showed a prominent J wave in multiple leads (2). Some cases of BS patients exhibiting J waves and ST-segment elevation in inferior leads have recently been reported (3, 4).

Several mutations in the SCN5A gene on chromosome 3 have been implicated in genetically-derived BS. Such mutations account for 10%-30% of BS in genotyped families (5). Mutations in SCN5A associated with the BS phenotype usually result in a loss of channel function by reduction in Na currents. Interestingly, SCN5A mutations have been reported to generate several types of disease entities distinct from BS, such as congenital type 3 long-QT syndrome, idiopathic VF, congenital sick sinus syndrome, atrial fibrillation, and even overlap syndrome. The existence of these commonly derived disease entities is consistent with the view that BS and its associated bradyarrhythmic complications may be caused by a single SCN5A mutation (6).

It has been reported that ST elevation and ventricular arrhythmogenicity are augmented by the parasympathetic tone in BS (7). Our patient may have developed an increased vagal tone due to his ongoing body aches. Several factors (i.e., vagal tone, body temperature, meals, anti-arrhythmic and other drugs, habits, hormonal status) are known to influence ECG pattern in BS (7). Our patient was not taking any arrhythmogenic drugs.

In the present case, AF and monomorphic VT were observed. Typically, BS presents with either VF or polymorphic VT. Few cases of BS with monomorphic VT have been reported (8,9). Approximately 20% of BS patients develop supraventricular arrhythmias associated with AF (10).

Conclusion

We present the case of a 20-year-old man with type I BS who suffered cardiac arrest. J waves accompanied by an AV block may serve as an important diagnostic sign in detection of high-risk individuals. Clinical and genetic studies are warranted to fully elucidate this new concept.

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