Brugada Syndrome diagnosed in the Emergency Department: A Case Report and interesting ECG features

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Abstract

Brugada syndrome is believed to be the cause of up to 50% of sudden cardiac death (SCD) cases due to ventricular dysrhythmias in young healthy individuals with no structural heart disease. This syndrome was first reported in 1992 and is rarely seen in the Emergency Department (ED). Given the life-threatening nature of Brugada syndrome, we hope to increase awareness in Emergency Medicine practitioners. We report the case of a 26 year-old Bangladeshi male who was referred to the ED with a history of intermittent palpitations, syncope and electrocardiogram findings of RSR’ and ST elevation in V1–V2 characteristic of Brugada syndrome. It is crucial that emergency physicians search for this diagnosis, as an implantable cardioverter-defibrillator is the only recognized life-saving intervention, and the risk of SCD is high if the diagnosis is missed.

Introduction

Brugada syndrome is a rare, though clinically significant, condition that can have grave consequences if unrecognized. The syndrome is characterized by a history of syncope or cardiac arrest due to ventricular tachydysrhythmia and the electrocardiographic pattern that was first described by Brugada[1]. The finding of Brugada-like electrocardiographic variants in asymptomatic individuals without a personal or family history consistent with cardiac dysrhythmia (including syncope or sudden cardiac death) is termed the Brugada sign[2]. Whereas Brugada syndrome is manifested by a propensity for ventricular dysrhythmias due to sodium channel abnormalities, the sign simply refers to the findings on the electrocardiogram (ECG) without the accompanying symptoms or contributory family history[2].

The key to treatment is expedient recognition of the Brugada sign and immediate cardiology evaluation and intervention, when appropriate. Up to 27% of individuals with diagnosed Brugada syndrome will develop ventricular tachycardia (VT) or sudden cardiac death (SCD) during a 2–3 year follow-up period[3]. This case discussion focuses on the clinical symptoms of Brugada syndrome, its physiology, and the appropriate treatment.

Fig 1. This first ECG, post VF conversion, performed in the Emergency Department, demonstrates a RBBB pattern RSR’ with ST elevation in lead V1–V2, with negative T wave inversion, and AF (Type 1 Brugada)
Case Report
A 26 year-old-man presented to the Emergency Department (ED), with history of syncope, and had an episode of VF which was DC converted, referred for cardiac evaluation to rule out “acute MI [myocardial infarction]”. The review of systems revealed a 2weeks history of episodic palpitations associated with blurry vision and a feeling as if he was going to “pass out, and had 3 episodes of syncope during sleep, as noted by his roommates. The patient reported episodes lasting from 2 to 10 min that seemed to “come and go” without any specific triggers. He specifically denied chest pain, fever, chills, weight loss, cough, headache, abdominal pain, urinary symptoms, rash or joint pain. There was no prior history of syncope. ECG recorded after DC conversion is shown in Fig 1.

Fig 2. ECG showing type 2 Brugada syndrome another day.

Fig 3. Type 1 Brugada syndrome
His past medical history was negative for previous surgeries, medications, and drug allergies. In addition, there was no family history of sudden infant death syndrome, SCD, or syncope. The patient did have a 10-pack-year smoking history.

In the ED, the patient was mildly anxious. His blood pressure was 124/56 mm Hg, his respirations were 18 breaths/min, and his temperature was 36.5°C (97.8°F). His cardiac examination was normal, revealing a regular heart rate of 67 beats/min with normal S1 and S2, no murmur, gallop, or friction rub. The rest of his physical examination was unremarkable. The ED ECG revealed an RSR' pattern in V1–V2 with ST elevation in those leads (Figure 1B). Interestingly, these two ECGs recorded on the same day had the same pattern essentially.

In the ED, intravenous access was started and laboratory studies were sent. The patient was placed on a cardiac monitor, was given 325 mg of aspirin and administered 2 L oxygen via nasal cannula. The complete blood count, electrolytes, thyroid-stimulating hormone, free thyroxine level, and troponin were normal. After DC shock patient had a mild elevation of troponin levels 0.17; urine toxicology screen was negative, and a portable chest X-ray study was normal. A cardiologist was consulted and the patient was admitted to the coronary care unit for further monitoring and testing. The patient had no evidence of dysrhythmias while monitored in the CCU except brief atrial fibrillation. While on telemetry, the patient experienced episodes of paroxysmal atrial fibrillation. A transthoracic echocardiogram was performed, which demonstrated a structurally normal heart and no other abnormalities. Multiple ECGs on telemetry revealed an RSR' pattern in V1–V2 with ST elevations.

**Discussion**

Brugada syndrome, first described in 1992, consists of: 1) an electrocardiographic pattern resembling a RBBB pattern with static or dynamic ST segment elevation in leads V1–V3; 2) a structurally normal heart; 3) a propensity for ventricular tachydysrhythmias precipitating syncope when non-sustained, and sudden cardiac death when sustained; and 4) a familial occurrence in some cases. Brugada syndrome has been described worldwide, and has been estimated to account for 40–60% of all cases of idiopathic ventricular fibrillation (VF) in some countries in Southeast Asia, our patient was from Bangladesh.

The ECG abnormalities are the hallmark of the Brugada syndrome. There are three types of repolarization variants recognized, based on the appearance of the ST segments in leads V1–V3. Type 1 has J-point or ST segment elevations > 2 mm at its peak, followed by a negative T wave, giving rise to a “coved” appearance. Type 2 has a J wave amplitude of > 2 mm, giving rise to a downsloping ST segment elevated ≥ 1 mm above baseline resulting in a “saddle back” ST configuration. Type 3 repolarization variants have the same saddleback appearance as Type 2 variation, but the J-point elevation gives rise to a ST segment that is elevated < 1 mm above the baseline. Coinciding with the correct clinical scenario, the diagnosis of Brugada syndrome can be made when a type 1 pattern is observed. When type 2 or type 3 ECG patterns are observed, a conversion to the diagnostic type 1 pattern must occur after sodium channel blocker administration to make a diagnosis of Brugada syndrome. A complete or incomplete RBBB pattern also has been discussed in association with the Brugada syndrome. The RBBB, however, is currently thought to be non-essential for diagnosis, and its presence could be attributed, at least in part, to early repolarization rather than to impulse delay or conduction block in the right bundle. More literature is emerging to further delineate ECG criteria to aid in determining risk of sudden cardiac death or ventricular fibrillation in patients with the Brugada sign. One retrospective Japanese study of 60 patients diagnosed with Brugada syndrome found that an S wave width ≥ 0.08 s in V1 and an ST elevation ≥ 0.18 mV in V2 correlated well with documented ventricular fibrillation (positive predictive values of 40.5% and 37.8%, respectively, with negative predictive values of 100%). These ECG criteria, in addition to the patient’s history, may be helpful in identifying patients at risk for tachydysrhythmias.

The prevalence of Brugada-like ECG (the Brugada sign) in the general asymptomatic population has been found to range from 0.05% to 0.7% in some studies.
patients are predominantly male (92%) and Asian (58%). At the time of diagnosis, they are usually in their mid-to-late thirties, with age ranging from 4 to 70 years. One study showed that 22% of patients had a family history of syncope, documented VF, or sudden death of suspected cardiac origin. A separate study showed that among patients demonstrating the Brugada sign after an aborted sudden death, 62% experienced SCD or ventricular fibrillation within a mean follow-up period of 54 months. Patients presenting with syncope had a 19% rate of SCD or documented VF. Those presenting solely with the abnormal ECG spontaneously had an 8% rate of these dysrhythmic events during a mean follow-up period of 27 months. Although patients in this last group were asymptomatic at the time of abnormal ECG recording, 72% of them had a family history of SCD. It is important to note that the ECG findings of the Brugada sign do not have to remain static. They can transform from one type to another over time or may normalize completely, making the diagnosis difficult. A normalized ECG, however, does not indicate a better prognosis in patients with the syndrome.

The disease is genetically determined with an autosomal dominant pattern. The alpha subunit of the sodium channel, SCN5A, on chromosome 3 has been linked to the disease. Several dozen SCN5A mutations and recently, another locus on chromosome 3, have been linked to Brugada's syndrome, indicating genetic heterogeneity. These mutations have been shown to result in either a total loss of the function of the sodium channel, or in acceleration of the recovery from the sodium channel activation. Twenty percent of patients with Brugada syndrome have documented SCN5A mutations. Penetrance of the disease is not 100%; some affected family members do not show the typical ECG or ventricular dysrhythmias spontaneously or under provocative testing.

Canine heart studies support the idea that heterogeneity of repolarization across the wall of the right ventricular (RV) outflow tract contributes to the ECG patterns and genesis of dysrhythmias in the Brugada syndrome. The presence of ST segment elevation in only the right precordial leads results from the higher density of transient outward currents in the RV epicardium. Usually, the action potential (AP) of epicardial cells but not endocardial cells display a pronounced phase 1, referred to as a "spike and dome morphology," which is the result of predominantly inward Ca currents (I_{Ca}) and some weak outward currents (I_{to}). When the I_{Ca} currents are overwhelmed by the I_{to} currents (due to sodium or calcium current blocking agents, ischemia, or metabolic inhibition), a reduction of up to 70% in the duration of the action potential is possible. This causes a loss of the dome of the AP at some areas at epicardial level (with ST segment elevation), whereas other areas such as the endocardium maintain the normal duration of the AP. The result is severe heterogeneity of duration of APs and, consequently, in refractory periods. Cells with short duration of AP can be re-excited by adjacent cells, with a normal duration of the AP triggering a re-entrant dysrhythmia. Brugada syndrome patients already have an ion channel malfunction that disrupts the natural equilibrium of the ventricular epicardium; however, certain other disruptions such as beta-adrenergic stimulation, febrile states, accelerated vagal activity, sodium channel blockade, and muscarinic stimulation will augment the ST segment elevations. This observation has led to the evaluation for Brugada syndrome in the electrophysiology laboratory using Class IA antidysrhythmic drugs. These medications unmask the ECG manifestations of the disease and can induce ventricular fibrillation or ventricular tachycardia, as occurred in our patient. Our patient has had repeated episodes of vomiting leading to hypokalaemia which was corrected but ECG did not show signs of hypokalaemia. Interestingly initial ECG showed Type 1 abnormality and AF, then the ECG normalized for a brief period, then reverted to Brugada pattern Type 2. After staying in CCU for 48 hours, patient was stable in NSR, was scheduled for ICD implantation in another hospital, but signed DAMA.

Littman et al described a clinical spectrum of the Brugada-type ECG ("Brugada sign") that could be imagined in the form of a pyramid. On the top are patients who demonstrate the sign and who have experienced SCD, who could be termed as those
with the true Brugada syndrome. Below are those who have a sign and who have a family history of SCD, who had an unexplained syncope or are of Southeast Asian ethnicity. These people can be considered as having a latent form of the Brugada syndrome. Below that are patients who have Brugada ECG pattern provoked by right ventricular pathology or sodium channel blockade. The bottom of the pyramid is formed by those having Brugada ECG pattern without any clinical risk factors. The authors recommend that all of the patients in the top two categories should be considered to be high risk and require an ICD placement. Patients in the third category of the pyramid are probably not at an increased risk, but if there is a suspicion of any additional risk factors, they should have EPS, pharmacological testing, genetic testing, and family studies done. Those on the bottom of the pyramid, that is, with a Brugada sign as an incidental finding, no symptoms, and no risk factors, could be labeled as having an asymptomatic Brugada syndrome and should have a good clinical follow-up done. A recent study by the Brugada family discusses the determinants of sudden cardiac death in individuals with Brugada waves in the electrocardiogram who have not experienced previous cardiac arrest.

They studied 547 patients who exhibited Brugada waves in the ECG. The abnormal tracing was present in 391 patients and was precipitated by an antiarrhythmic drug in 156 patients. They concluded after 2 year follow-up that these patients have a high risk of sudden death (8%). Patients who gave a history of syncope or had inducible ventricular arrhythmia were likely to have a poor prognosis. The lowest-risk group of patients had no history of syncope and abnormal ECG only after taking an antiarrhythmic drug and noninducible ventricular arrhythmia.

Each month a new twist is reported, indicating that we have much more to learn about Brugada waves and the Brugada syndrome. For example, which patients should have an internal cardiac defibrillator.

Currently, the only definitive treatment to reduce and prevent mortality from sudden cardiac death in patients with Brugada syndrome is the ICD. However, an ICD may not be practical for young children or adults living in areas of economic hardship. Although currently it is thought that the treatment with anti-dysrhythmic drugs does not effectively prevent new events, there are studies that show promising value of pharmacologic agents. These studies suggest that agents blocking transient outward current, such as quinidine, or those boosting calcium current, such as isoproterenol, may be useful in the prevention of tachydysrhythmias. However, the long-term effects of these agents on overall mortality of patients are unclear at this time. Also, Glatter et al. described a case of a man with Brugada syndrome who had recurrent syncopal events and a family history of SCD who was successfully treated for 13 years with sotalol.

We have previously presented a case of Brugada syndrome in a Burmese adult who had a nocturnal syncope due to polymorphic VT.

References


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