CASE REPORT

Brugada Syndrome: An Electrical Storm without Warning

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ABSTRACT

Brugada Syndrome (BrS) is an inherited channelopathy causing sodium channel dysfunction in cardiac myocyte. These patients are prone to develop Ventricular Fibrillation (VF) or polymorphic Ventricular Tachycardia (VT). Next to coronary artery disease and cardiomyopathies, BrS is an important cause of sudden cardiac death. We report here a case of 22-year unmarried female with "unexplained" cardiac arrest without prior history of syncope and family history of sudden cardiac death. She was resuscitated out of hospital in some local dispensary in rural settings and after prolonged hypoxia with its neurological sequelae, she eventually died. BrS should be considered in differentials of unexplained cardiac arrest even in patients without family history of sudden cardiac death. First episode of VT/VF in BrS patients can be life threatening. Only prompt cardioversion / defibrillation can save life. We have suggested some measures to identify patients of BrS.

Key Words: *Brugada syndrome. Brugada phenocopy. Ventricular fibrillation. Brugada ECG pattern. Sudden cardiac death.*

INTRODUCTION

In 1992, Pedro and Joseph Brugada described a new syndrome characterized by syncope and/or sudden death due to VT/VF in young and otherwise healthy adults, with a structurally normal heart and a characteristic electrocardiogram (ECG) pattern of Right Bundle Branch Block (RBBB) with ST segment elevation in leads V1 to V3.[1] In the new ECG criteria, only 2 ECG patterns are considered: pattern 1 is identical to classic type 1 of other consensus (coved pattern) and pattern 2 that joins patterns 2 and 3 of previous consensus (saddle-back pattern).[2] Pattern 1 is highly suggestive of BrS as compared to pattern 2 which may need to be investigated with sodium channel blocking agents before diagnosing BrS.

Diagnosis of BrS requires ECG manifestation and any one of clinical criteria that include history of syncope, family history of sudden cardiac death, the presence of pattern 1 in other family members or documented VT/VF. BrS is 8 - 10 times more prevalent in men than in women. An estimated 4% of all sudden deaths and at least 20% of sudden deaths in patients with structurally normal hearts are due to this syndrome.[3] BrS is due to mutations in the cardiac sodium channel gene. Over 160 different mutations in SCN5A gene have been described so far and at least 50% are spontaneous mutations, but familial clustering and autosomal dominant inheritance has been demonstrated.[4]

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CASE REPORT

A 22-year unmarried female became unconscious suddenly. She did not have history of palpitations and syncope in past. There was no history of sudden death in family. She was not taking any medicines. She was resuscitated in a local dispensary. Medical record and treatment given there were not available. She was transferred to our facility after about 12 hours since the indexed event with suspicion of suicidal attempt.

On admission, she was deeply comatosed with shallow breathing efforts. Her BP was 75/48 mmHg. Oxygen saturation was 95%. Pulse rate was 64/minute. Her brain stem reflexes were intact. There were no localizing neurological signs. Respiratory and precordial examinations were unremarkable. ECG showed broad QRS complexes (Figure 1) with ST segment elevation and T-wave inversion in all leads except aVR. Arterial blood gas analysis showed pH=6.8, pCO_2=49 and HCO_3=8.2 mmol/L. SGOT=118, SGPT=99, K^+=9.4 mmol/L, Blood sugar=181 mg/dl, Creatinine=1.67 mg/dl, and Troponin-I=5.02 ng/ml. Echocardiography revealed moderate left ventricular systolic dysfunction with global hypokinesia and ejection fraction of 35 - 40%. Chest X-ray was normal.

She was started on infusion of epinephrine titrated to maintain mean arterial pressure around 70 mmHg. Hyperkalemia was corrected with intravenous HCO_3 and glucose-insulin infusion. Intravenous calcium gluconate was given to prevent cardiac arrhythmia. After 2 hours, her K^+ was normalized (4.1 mmol/L). One hour later, she developed VF twice, promptly defibrillated each time. Her breathing efforts after this episode became even shallower. She was then put on artificial positive pressure ventilation. Supportive measures were continued. Metabolic acidosis and electrolyte imbalance were corrected. Epinephrine was continued. On second day SGPT rose to 1806 U/L, suggestive of ischemic

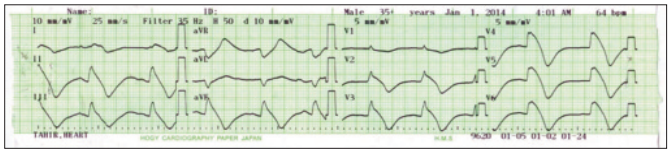

Figure 1: 12-lead ECG on admission. Broad QRS complexes (K^+ = 9.4 mmol/L).

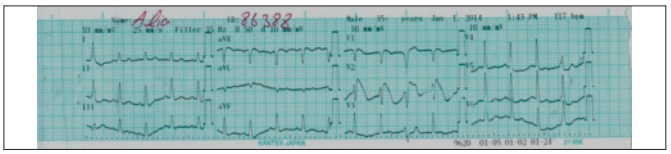

Figure 2: 12-lead ECG recording on second day. Note characteristic type-1 (coved-type) Brugada pattern in V1 and V2. Occasional VPC's are also present.

hepatitis (Shock liver). Routine ECG on second day revealed characteristic type 1 (coved-type) Brugada pattern (Figure 2).

Despite these supportive measures, irreversible neurologic damage that had occurred after prolonged unattended cardiac arrest (presumably due to VT/VF) before admission eventually resulted in her unfortunate death. Available brothers and sisters of deceased were screened with 12-lead ECG but none of them had Brugada pattern.

DISCUSSION

As stated earlier, the presence of Brugada pattern on ECG does not necessarily mean BrS. Type 1 (coved-type) Brugada pattern is more suggestive of BrS as compared to type 2 (saddle-back) Brugada pattern. Clinical correlation is required to diagnose BrS. ECG changes may appear intermittently and in some subjects these can be unmasked only upon challenge with sodium channel blocking agents. To add to complexity, the concept of Brugada Phenocopy (BrP) is emerging. BrP is characterized by ECG patterns that are identical to type 1 or type 2 Brugada patterns despite the absence of the true congenital BrS. BrP is caused by various clinical circumstances including: hyperkalemia, acidosis, adrenal insufficiency, hypothermia, mechanical cardiac compression, myocarditis, pericarditis, and ischemia.[5] But in these cases ECG changes revert after correction / elimination of inciting agent. In this case hyperkalemia, acidosis and mechanical cardiac compression could have caused Brugada like ECG pattern but after correction of these abnormalities, ECG changes should have been reverted which actually did not. Type 1 Brugada pattern with unexplained cardiac arrest is sufficient to make the diagnosis of BrS instead of BrP. Moreover, the patient developed VF twice during hospital stay after correction of acid-base and electrolyte imbalance. Elevation of Troponin and K^+ levels and myocardial dysfunction can be explained by out of hospital prolonged resuscitation.[6-8]

Case reported here would have probably never undergone ECG during her whole life. Her first electrical storm was lethal. So identification of BrS patients is of paramount importance even before they become symptomatic. Only way to identify such patients is 12-lead ECG. In one study, prevalence of Brugada pattern in Pakistani population is 0.8%.[9]

The authors suggest that 12-lead ECG should be included in routine medical evaluation of students seeking admission in higher secondary school level as this syndrome usually manifests in and after 2nd decade of life. All practicing physicians should be familiarized with Brugada pattern. Once Brugada pattern is detected, all family members should be screened with 12-lead ECG. In asymptomatic patients with Brugada pattern on ECG although there is nothing much to offer but close contact with cardiologist may prevent such unfortunate outcomes. Inducible VF on programmed electrical stimulation may be helpful in predicting high risk cases in asymptomatic patients.[10] ICD is reserved only for symptomatic patients. Guidelines do not recommend any specific antiarrhythmic agent for prevention or treatment. Provocative testing with sodium channel blockers is helpful only in diagnosis of BrS in patients with non-diagnostic ECG but not in risk stratification.

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