Massive Electrical Storm at Disease Onset in a Patient with Brugada Syndrome

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Patient: Male, 49
Final Diagnosis: —
Symptoms: —
Medication: —
Clinical Procedure: —
Specialty: —

Objective: Rare disease
Background: Brugada syndrome (BrS) is a genetic arrhythmogenic disease characterized by ST-segment elevations in the right precordial leads of the electrocardiogram (ECG). These ECG changes may be concealed and BrS may present with electrical storm characterized by recurrent ventricular tachycardia and fibrillation.

Case Report: A 49-year-old previously healthy man was admitted with electrical storm. The patient received direct current (DC) cardioversion shocks and only after intravenous lidocaine did the electrical storm slowly subside with a total of 255 DC shocks administered during the first 24 h after admission. He fully recovered and received an implantable cardioverter-defibrillator. Subsequent drug challenge with flecainide revealed type 1 BrS.

Conclusions: Massive electrical storm can be the first symptom of BrS and the diagnostic ECG changes may be concealed at presentation. Although hundreds of DC shocks may be required during initial treatment, full recovery can be achieved.

MeSH Keywords: Brugada Syndrome • Defibrillators • Tachycardia, Ventricular

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Background

Brugada syndrome (BrS) is a genetic arrhythmogenic disease with an estimated prevalence of 1–5/10 000 in Europe, characterized by ST-segment elevations in the right precordial leads of the electrocardiogram (ECG) and increased risk of sudden death due to ventricular tachycardia (VT) or fibrillation (VF) [1]. Three (type 1, 2 and 3) abnormal ECG repolarization patterns, of which only type 1 is diagnostic, are associated with BrS but these patterns may be dynamic or concealed, and unmasked by exposure to various drugs (e.g., sodium channel blockers including class 1A or 1C antiarrhythmic agents) [2,3]. Indeed, challenge with class I drugs is used as a diagnostic test in subjects with suspected BrS and normal or non-diagnostic ECG (3). Electrical storm characterized by recurrent VT or VF (usually defined as ≥3 separate episodes separated by >5 min in 24 h) is known to occur in patients with BrS [4,5]. Electrical storm is most frequently seen in the acute phase of myocardial infarction and in patients with implantable cardioverter-defibrillators (ICDs) and/or genetic arrhythmia syndromes, including BrS. Reports have indicated that 25% of individuals with BrS will experience VF or sudden cardiac death and although electrical storm is not a rare event in patients with BrS, no specific clinical, laboratory, or electrophysiologic characteristics have been demonstrated in BrS subjects with ICD with electrical storm vs. without electrical storm [6]. The genetic basis of BrS includes loss-of-function mutations in genes for cardiac ion channels, and the effect of reduced inward positive (Na+, Ca++) current on the potassium transient outward current (Ito) is thought to contribute to VT/VF by a mechanism of phase-2 reentry [1].

Electrical storm in a previously healthy adult with non-diagnostic ECG at presentation is a rare and dramatic clinical entity. We present such a case to show that hundreds of direct current (DC) cardioversions may be required at presentation, as well as emphasizing the value of subsequent diagnostic drug challenge.

Case Report

A 49-year-old previously healthy man with an unremarkable family history and without medication or notable exposures was admitted to a local hospital after 3 days of afebrile digestion and general malaise that culminated with recurrent syncope. At presentation the patient repeatedly lost consciousness due to episodes of VT/VF that were terminated by external DC defibrillation. Twelve-lead ECG was considered to show no significant ST-segment elevation, and PQ and QT intervals were normal (Figure 1A). Laboratory test results were unremarkable, including normal plasma levels of potassium, magnesium, calcium, and C-reactive protein, and only a small increase of plasma troponin I levels to 1080 ng/l (reference <40 ng/l). Hereafter, the patient remained in a state of electrical storm with incessant polymorphic VT/VF episodes that mostly required DC defibrillation. Between VT/VF episodes, circulatory parameters normalized and prolonged chest compressions were not needed. Transthoracic echocardiography during brief periods of sinus rhythm showed a structurally normal heart with minor global dyskinesia and a left ventricular ejection fraction of 50%. After prompt intubation and intravenous amiodarone, the patient was transferred to a tertiary center where acute coronary angiography was normal. Temporary transvenous overdrive pacing, repeated intravenous amiodarone, isoproterenol, magnesium sulphate, and metoprolol provided no benefit and he received 85 DC defibrillations during the first 2 h after transfer. Intravenous lidocaine (class 1B agent) was commenced and thereafter the electrical storm only slowly subsided, with absence of arrhythmia after 24 h and a total of 255 DC defibrillation shocks. The patient was subsequently extubated and displayed no sequelae after the electrical storm. He was treated with oral mexiletine (class 1B agent) and received an ICD. Eight months later, the patient has been well and without arrhythmias. One month after discharge, mexiletine was discontinued and drug challenge with flecainide (class 1C agent) demonstrated development of a diagnostic type 1 BrS pattern in the ECG with >0.2 mV coved-type ST-segment elevation in the ECG (Figure 1B and 1C). At present, cardiac magnetic resonance imaging has not been performed and family screening and genetic tests for BrS are ongoing.

Discussion

An ICD is the only proven therapy for prevention of sudden cardiac death in symptomatic individuals with BrS, but in these patients quinidine (a class 1A antiarrhythmic agent that also inhibits potassium channels, including Ito and the rapid [I-Kr] component of the delayed potassium rectifier current) may prevent VF and suppress spontaneous ventricular arrhythmias, and isoproterenol (which increases the ICaL current) have been effective for treatment of electrical storms [1,7–9]. Moreover, isolated reports are available on potentially favorable effects of mexiletine, cilostazol, and sotalol in subjects with BrS [7]. Specifically, in the limited case series of Miyazaki et al., 1 individual received mexiletine after having experienced VF 3 times within 1 month after ICD implantation and was thereafter free of arrhythmia for 16 months [10].

Conclusions

In our patient, mexiletine was administered as ultimum refugium in the emergency setting at a time when the diagnosis was uncertain. Intravenous quinidine was not available. Indeed, the
combined effects of drugs (amiodarone, magnesium sulphate, and metoprolol) that were first administered and transvenous overdrive pacing may have contributed to the apparent efficacy of mexiletine in our case. Notwithstanding these considerations, the case exemplifies the treacherous nature of BrS with massive electrical storm as the presenting symptom in a young and otherwise healthy adult, the usefulness of diagnostic drug challenge, and the potential for full recovery despite an excessive number of external DC shocks during the first 24 h, which to our knowledge is unprecedented in the literature.

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Figure 1. (A) Resting ECG. (B) ECG prior to flecainide test. L1 – Lead V1 lifted to intercostal space 3 (sensing ECG electrode position), L2 – Lead V2 lifted to intercostal space 3. L1½ – Placed between L1 and L2 at sternum (left intercostal space 3). (C) ECG during flecainide test.