Status Epilepticus as an Initial Presentation of Brugada Syndrome: A Case Report

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Patients presenting with convulsions are sometimes incorrectly treated for epilepsy, as these symptoms may be manifestations of underlying cardiac disease. Brugada syndrome, associated with an elevated risk of developing fatal arrhythmic events, is a rare disorder characterized by a unique electrocardiographic pattern. The typical clinical presentation mainly involves syncope of unclear cause or sudden death. Seizures are uncommon clinical manifestations of Brugada syndrome, and reports of status epilepticus as initial presentation of Brugada syndrome are extremely rare. In this case report, we present a male patient with a typical pattern on 12-lead electrocardiography, following resuscitation for generalized convulsive status epilepticus.

Key Words: Brugada syndrome, status epilepticus
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Status epilepticus is usually defined as continuous seizure activity lasting 30 minutes or more, or intermittent seizure activity lasting 30 minutes or more, during which time consciousness is not regained [1]. This condition may be attributed to many causes, including cerebral hypoxic damage [2]. Patients presenting with convulsions are sometimes mistakenly treated for epilepsy, although these symptoms may be a manifestation of hypotension caused by bradycardia [3]. These symptoms may also be seen in patients with hemodynamically compromising ventricular tachycardia (VT) or ventricular fibrillation (VF), presumably resulting from cerebral hypoperfusion [4].

Brugada syndrome, which is unrelated to structural cardiac abnormality, is a rare electrical disorder characterized by a unique electrocardiographic pattern of right bundle branch block, and ST-segment elevation in right precordial leads (V1 to V3). There is a high risk of developing fatal arrhythmic events such as VT or VF. The clinical presentations are mostly syncope of unclear cause or sudden death. Seizure is an uncommon clinical presentation of Brugada syndrome, reported in only a few articles [4,5], and none of which note status epilepticus as the initial manifestation of the syndrome. In this article, we describe a male patient who presented with status epilepticus and typical pattern on 12–lead electrocardiography (ECG) after resuscitation, demonstrating generalized convulsive status epilepticus as the initial manifestation of Brugada syndrome.

Case Presentation

A 41-year-old male patient was brought to the local emergency department (ED) for generalized tonic-clonic movements. According to his co-workers, the patient was talking to them without any apparent discomfort, before starting to exhibit tonic-clonic movements that lasted for more than half an hour before arrival to the ED. During resuscitation, VF in ECG records (Figure 1) with seizure activities persisted until direct current (DC) shock was applied and sinus rhythm was restored. The patient was then transferred to the intensive care unit at another hospital as soon as vital signs were stabilized and maintained with medication. Initial physical examination was normal except

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for Glasgow Coma Scale (GCS) status of E1VEM2. Neurologic examination revealed positive light reflex, negative Doll's eye sign, hyperreflexic deep tendon reflex, and positive Babinski sign. The complete blood count and chemistry panel were within reference ranges, except for glutamate-oxalacetate transaminase and glutamate-pyruvate transaminase, which were 94 IU/L and 136 IU/L, respectively. Creatinine phosphokinase and creatinine phosphokinase isoenzyme MB were 378 IU/L and 65 IU/L, respectively. Troponin I was initially 0.123 ng/mL and subsequently peaked at 0.995 ng/mL. Sinus tachycardia with V1~V3 RSR' pattern and ST-segment elevation (Figure 2) was found on ECG records. Echocardiogram was normal. Chest X-ray revealed no cardiopulmonary abnormality, and brain computerized tomography showed diffuse brain edema with blurring sulci and no obvious local hypodense or hyperdense lesions. Electroencephalography several days later revealed generalized continuous voltage suppression, which was compatible with hypoxic encephalopathy. Supportive treatment was arranged for the patient, including tracheostomy, stress ulceration prevention and rehabilitation.

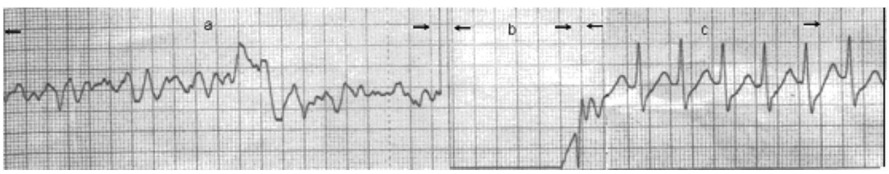

Figure 1.
a: Electrocardiography recording during the status epilepticus showing ventricular fibrillation.
b: Period of direct current shock.
c: Sinus rhythm was restored after direct current shock was applied.

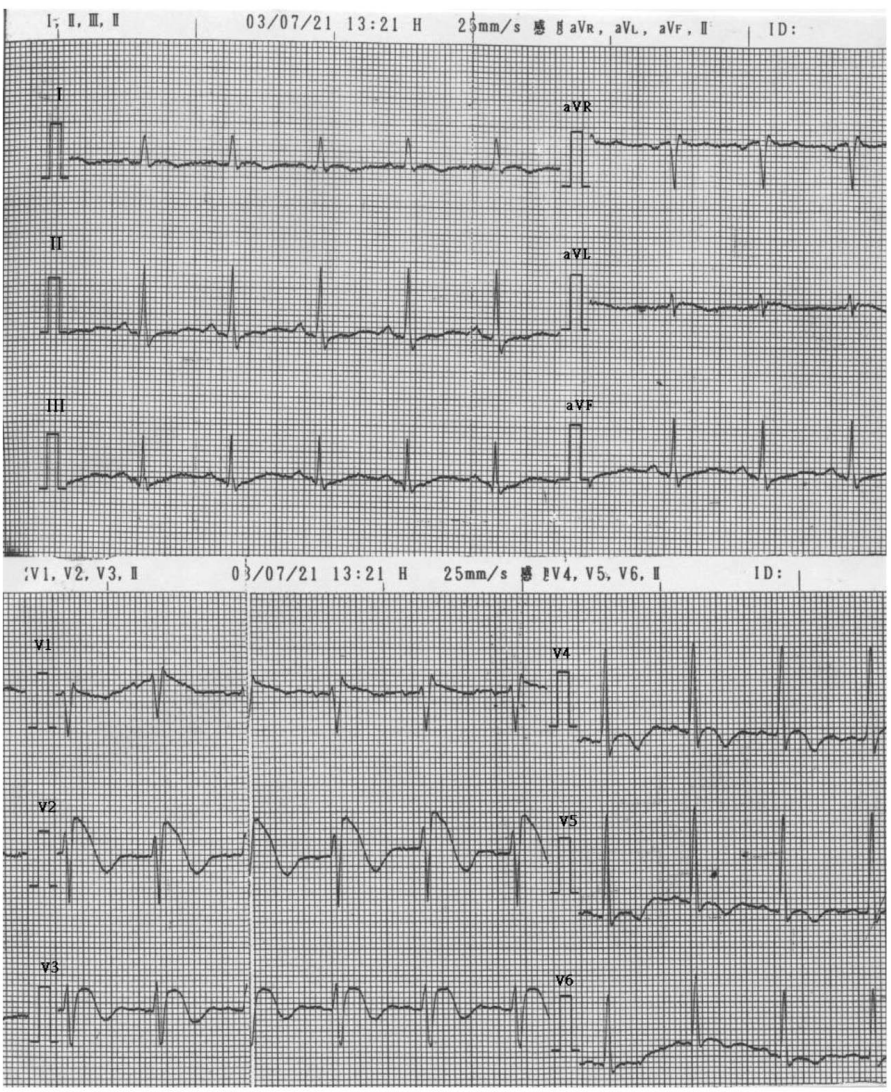

Figure 2. *12-lead electrocardiography showing a pattern of right bundle branch block and ST-segment elevation in leads V1 through V3, the typical characteristics of Brugada syndrome.*

Three weeks later, the patient was transferred to the rehabilitation ward, with the diagnosis of hypoxic encephalopathy, Brugada syndrome, and respiratory failure post tracheostomy. On admission to the rehabilitation ward, neurologic examination was the same as previously, with the exception of mildly improved GCS score of E1VTM3 and positive Doll's eye sign. Physical examination was entirely normal. Laboratory evaluations remained within reference ranges. Repeat chest plain film revealed no active cardiopulmonary lesions. Serial ECGs continued to show RSR' pattern with ST-segment elevation in V1 through V3. There were no other abnormalities on Holter monitoring.

Patient history revealed no systemic disease or complaints of syncope, convulsion, or chest pain. No drug history or allergic reaction and no surgery or admission records were found. The patient never smoked nor consumed alcohol. There had been no family history of syncope, convulsion, heart disease or sudden death.

The patient continued in rehabilitation programs, which included passive range of motion, pressure sore and thrombosis prevention, oral hygiene education and aspiration pneumonia prevention, and contracture prevention by orthoses. The patient did not receive implantable cardioverter-defibrillator (ICD) implantation subsequently, and is currently in a vegetative state.

Discussion

In the present case, the patient had been in perfect health when he suddenly presented in status epilepticus. ECG on arrival revealed ventricular fibrillation, and sequential series revealed Brugada-type patterns. These signs, combined with normal echocardiogram, were supportive evidence for the diagnosis of Brugada syndrome. Although we cannot exclude the possibility of myocardial infarction because of the elevation in cardiac enzyme (the peak level of troponin I: 0.995 ng/mL), this was most likely the result of electrical injury, secondary to DC shock. Furthermore, the typical ECG pattern persisted, although cardiac enzyme levels had returned to normal. Hence, Brugada syndrome was identified as the most likely cause.

Status epilepticus is a medical emergency requiring urgent management to prevent brain damage and systemic complications. Failure to diagnose and treat it adequately results in significant morbidity and mortality. The search for the cause is essential, as this will determine subsequent therapy. Causes of status epilepticus are numerous and may be categorized as acute or chronic [1]. Although hypoxia is well known to be an acute cause, the processes leading to hypoxia are difficult to study. Ventricular tachyarrhythmias are uncommon causes of convulsions in adults. The underlying mechanism has been proposed by several studies, which suggest that hypoperfusion resulting from ventricular tachyarrhythmias is the cause of the convulsions [4,6]. As such, it could be further inferred that, if the arrhythmic event is of sufficient duration, status epilepticus may occur. Therefore, we believe the reported patient is a case of Brugada syndrome with an initial presentation of generalized convulsive status epilepticus.

Classically, patients with Brugada syndrome generally have no evident systemic diseases in their past history. All too often, syncope or sudden cardiac death is the only symptom in these patients. In some cases, sudden death is the first symptom of the disease [7]. To our knowledge, this case is the first that demonstrates status epilepticus as the initial symptom of Brugada syndrome. Therapy of status epilepticus involves management of precipitating causes and prevention of seizure recurrence [8]. Afflicted patients can be prevented from having recurrence, as long as they receive adequate therapy. Consequently, Brugada syndrome should not be omitted in the causes of status epilepticus, especially when typical treatments for status epilepticus are ineffective.

At present, ICD implantation is the most effective method in symptomatic patients with Brugada syndrome [4,9]. However, controversy exists in the treatment of asymptomatic patients. Asymptomatic patients should undergo electrophysiologic study, and if inducible, ICD implantation is recommended [9]. Agents that boost the calcium current, such as isoproterenol, and some class IA agents, such as quinidine and tedisamil, have been shown to be effective in normalizing ST-segment elevation in patients with Brugada syndrome, and in controlling electrical storms. None of these, however, have demonstrated long-term efficacy in the prevention of sudden death [9].

The purpose of our presentation was to demonstrate that Brugada syndrome could be one of the causes of status epilepticus. The rarity of status epilepticus as the initial manifestation of Brugada syndrome makes the presentation particularly interesting.

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Brugada症候群以癲癇重積狀態為初始表現

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病人以全身抽搐為臨床表現時，有時會被不適當地以癲癇來治療，儘管原因可能是心臟疾病。**Brugada** 症候群 具有發展致命性心律不整的高危險性，是一種罕見的疾病，具特有的心電圖特徵。此症候群的臨床表現多是不明原因的昏厥或猝死。癲癇是此症候群不常見的臨床表現，而癲癇重積狀態更是極少。本篇文章報告一位男性在急救後具有此症候群典型的心電圖特徵，他是以癲癇重積狀態為初始表現。

關鍵詞：Brugada症候群，癲癇重積狀態

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