

A case of convulsion: Brugada syndrome

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A healthy 28-year-old man presented with multiple brief episodes of convulsion. He was found to have ventricular fibrillation which required defibrillation in the emergency department. After exclusion of organic heart diseases, Brugada syndrome was diagnosed and required implantable cardioverter defibrillator (ICD) implantation. The case emphasized the recognition of malignant cardiac dysrhythmias as a cause for seizures. The management in the emergency department was discussed. (*Hong Kong j.emerg.med.* 2002;9:105-109)

Keywords: Brugada syndrome, idiopathic ventricular fibrillation, seizure

Case

A 28-year-old man was reported to be semiconscious and had a convulsion lasting 1 minute. Ambulance staff arrived and witnessed another generalised convulsion lasting less than 1 minute.

On arrival at the Accident and Emergency department, he was fully conscious. His vital signs were stable. He had retrograde amnesia and could not remember how he was sent to the hospital. Examination of the chest and heart were normal. Neurological examination was unremarkable. Abdominal examination was normal. He was put on cardiac monitoring in the resuscitation room.

ECG (at 23:50) showed sinus rhythm, RBBB, ST elevation in V1-V2, the QTc 0.408 sec and the axis is 70 deg. (Figure 1)

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Blood investigations included arterial blood gas which was normal with no metabolic or electrolyte disturbances. Chest X-ray performed showed normal cardiothoracic ratio.

Cardiac monitor initially showed occasional polymorphic ventricular ectopics which later changed to ventricular fibrillation (VF), as documented in Figure 2.

During the VF, he had starring eyes and tonic twitching of limbs with loss of consciousness. Defibrillation with 200J was required to return his rhythm to sinus.

A repeat ECG showed a RBBB pattern with persistent ST elevation V1 to V2.

His past health, family and medication history were unremarkable.

Patient was admitted to Coronary Care Unit (CCU) for investigation of his cardiac syncope. The RBBB and ST elevation in V1 to V2 were the only peculiar findings in his ECG. Blood tests performed in CCU were all normal. Toxicology screening and viral study for myocarditis were negative. Echocardiogram revealed a normal heart with no structural abnormalities and good left and right ventricular ejection fractions. Cardiac

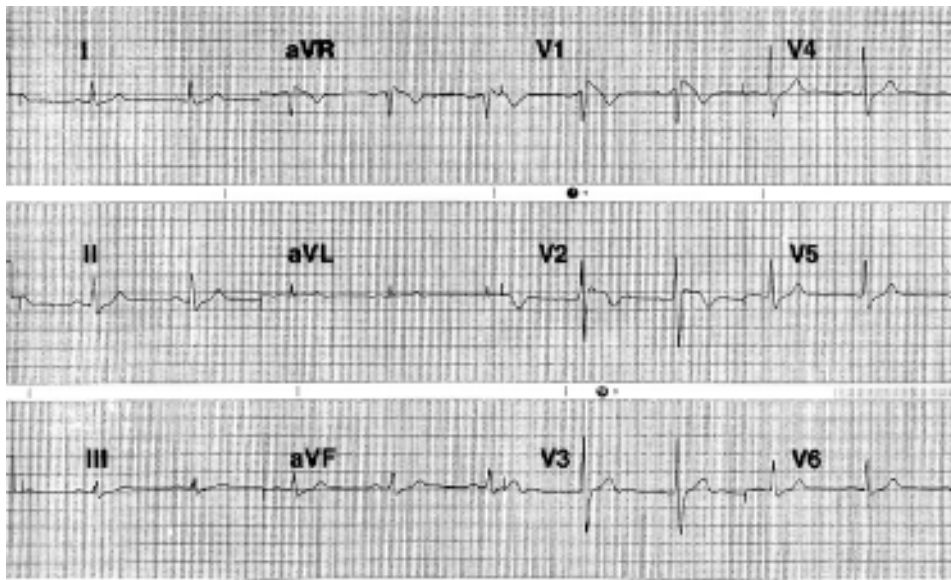


Figure 1. Initial ECG.

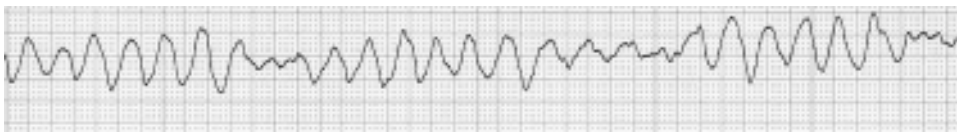


Figure 2. Ventricular fibrillation as documented on cardiac monitor.

catheterisation showed normal study. An Electrophysiological study performance (EPS) revealed inducible VF that required defibrillation to restore sinus rhythm. The flecainide test was used to unmask Brugada syndrome. Patient had ICD implantation and discharged home.

His brothers and sisters were informed of the diagnosis. Since the inheritance is autosomal dominant they were invited to undergo screening. Such a screening is essential to make a diagnosis of Brugada syndrome and could potentially prevent them from dying of dysrhythmias.

Discussions

This patient presented with recurrent 'convulsion' initially. There are many causes for such a presentation.

History, physical examination and ECG constitute the core of the workup for patients presenting with syncope (combined diagnostic yield, 50%). These will provide a diagnosis or determine whether diagnostic testing is necessary in most patients. In subjects in whom heart disease is suspected or known, further cardiac testing, including echocardiography, stress testing, Holter monitoring or intracardiac electrophysiologic studies (diagnostic yield 5% to 35%) may be necessary.²

This patient was put on cardiac monitoring and was closely observed in the resuscitation room. His ventricular fibrillation rhythm was revealed to be the cause of his myotonic twitching of the limbs and transient loss of consciousness. Close monitoring is a useful way to elucidate cardiac dysrhythmias. Even though such rhythms may not appear during patient's stay at the Emergency Department but certainly there is a need to look out for them.

Electrolytes, glucose levels and toxic causes should be screened at the Emergency Department. They can also cause both neurologic and cardiac symptoms.

The final diagnosis of Brugada syndrome turned out to be an increasingly recognised dysrhythmic functional cardiac disease that emergency physicians should be aware of. This syndrome is in fact not uncommon in Hong Kong.

Brugada syndrome

Historical discovery of the syndrome

ECG pattern of RBBB with ST elevation in leads V1 to V3 were found to associate with sudden death. In 1992, Brugada and Brugada were the first to speculate the syndrome to be a distinct clinical and ECG syndrome of functional cardiac origin.³ The notion was further substantiated by the recognition that ECG abnormalities were that inconsistently present and altered autonomic tone and antiarrhythmic drugs modulate the extent of ST-segment elevation.⁴

Diagnostic criteria for the syndrome includes:

1. $>$ or $=$ 0.1mV ST-segment elevation in leads V1, V2 and V3 (Refer to patient's initial ECG)
2. absence of organic heart disease (absence of ischemia, electrolyte or metabolic disorders, pulmonary or inflammatory diseases or abnormalities of central or peripheral nervous system.)
3. dynamic nature of the ST segment, both spontaneously in time and under the influence of pharmacological agents (Sodium channel blockers and autonomic modulators)
4. malignant family history in the case of an asymptomatic individual.³

Other ECG features suggesting of the syndrome:⁷

- ST-segment elevations were designated either as coved or saddleback. (Figure 3) It is not unique or highly specific for the Brugada Syndrome and can be found in a variety of clinical settings. (Table 1)

Diagnosis and management

In many patients with the syndrome, the ECG manifestations may show dynamic changes or transiently normalize, leading to underdiagnosis of the syndrome. Sodium channel blockers (procainamide, ajmalin, or flecainide in therapeutic dosages) can unmask the ST segment elevation in many patients with concealed or intermittent forms of the syndrome with high sensitivity and specificity.⁹ Electrophysiological study (EPS) with sodium channel blockers challenge in asymptomatic family members are recommended. The occurrence of $>$ or $=$ 0.1mV ST-segment elevation in leads V1, V2, and V3 is regarded as a positive response.

In the case of inducible VF, an ICD is the only effective treatment. In asymptomatic individuals with a malignant family history, the threshold for ICD implantation should be low, regardless of electrophysiology results.⁵

Prevalence of the Brugada syndrome

In Japan, the prevalence of ECGs compatible with the syndrome is 0.05%.⁶ The typical coved pattern of the Brugada syndrome (0.1%) was found in Europe. This foresees a major public health problem. Eighty percent of these patients could have inducible ventricular arrhythmias and ICD is the only effective treatment.⁷

It was reported a 0.05% prevalence of the typical coved pattern⁸ While a more recent study prevalence of saddleback ST-segment elevation (6%) and to typical coved ST-segment elevation (0.1%).⁷

The Hermida et al study⁷ did not find an increased mortality with a midterm follow-up. Sensitisation by a sodium channel blocker test could be proposed in the healthy population but needs to be further evaluated. Prophylactic ICD is the issue being further investigated.

Since the first local case was reported in April 1999, there were over 30 patients in Hong Kong identified with Brugada syndrome by end of 2000. Preliminary data showed a male predominance, one third had either cardiac arrest or inducible polymorphic VT/VF.¹⁰

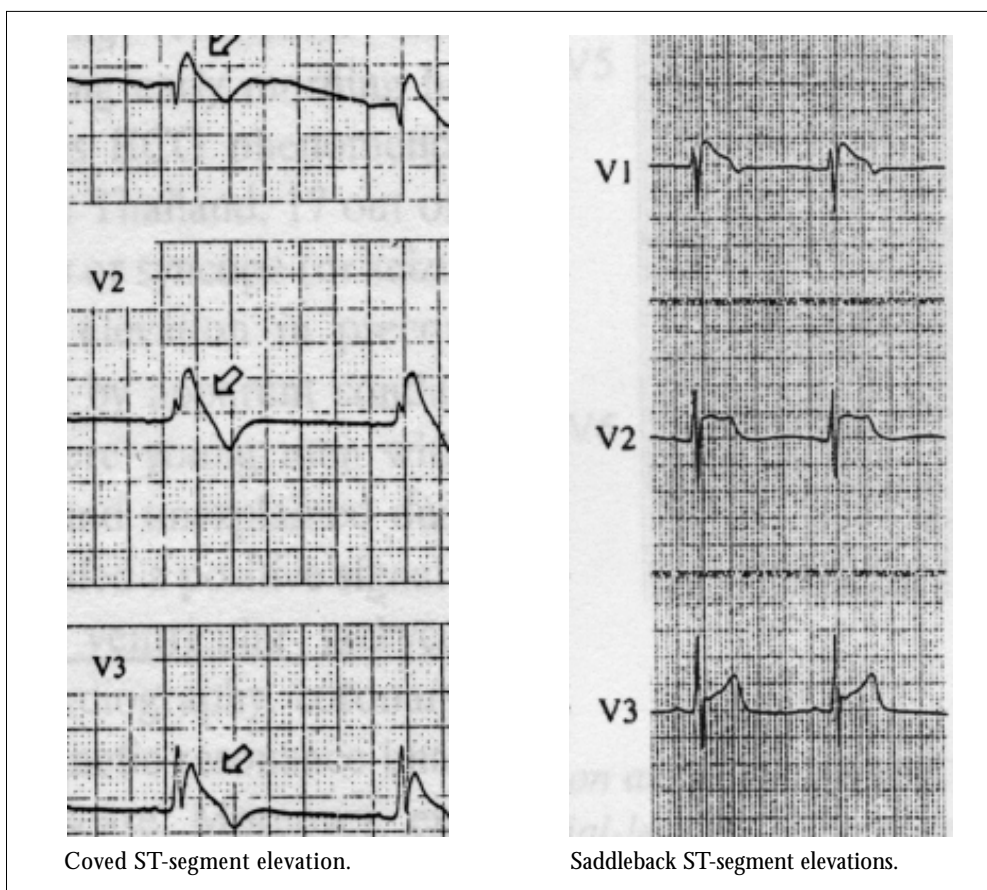


Figure 3. ECG features that are suggestive of Brugada syndrome.

Table 1. Differential diagnosis of ST segment elevation in Right Precordial Leads.¹¹

- Arrhythmogenic RV dysplasia
- Right or left bundle branch block
- Left ventricular hypertrophy
- Left ventricular aneurysm
- Acute myocarditis
- Right ventricular infarction
- Pulmonary embolism
- Hyperkalaemia
- Hypercalcaemia
- Various central and autonomic nervous system abnormalities
- Friedreich ataxia
- Duchenne muscular dystrophy
- Thiamine deficiency
- Cocaine intoxication

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