Incidental Findings of Brugada Syndrome Type-1 and Epsilon-Like Pattern in Otherwise Healthy Man: a Case Report

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Abstract

Background: Brugada Syndrome (BrS) and arrhythmogenic right ventricle dysplasia (ARVD) are rare cardiomyopathies predisposing to sudden cardiac death (SCD). Comprehending the electrocardiographic features of these cardiomyopathies are crucial especially in emergency settings.

Case presentation: A 25-year old medical student presented with no complaints, but had episodes of syncope, chest pain, and palpitations of unknown origin 10 years ago. The initial assessment showed stable hemodynamics. During examination, the ECG demonstrated incomplete right bundle branch block, Brugada-type 1 pattern, with signs of Epsilon wave. The following year, assessment of the ECG was repeated and findings were found suggestive of Brugada syndrome, although his echocardiography showed no structural abnormality. According to ESC guidelines, asymptomatic Brugada patients should undergo electrophysiology study.

Conclusion: Careful follow-up with electrophysiology study is recommended for this patient in order to identify the likelihood of true Brugada and suitability for radiofrequency ablation or implantation of implantable cardioverter defibrillator (ICD).

1. Background

Brugada syndrome (BrS) and arrhythmogenic right ventricle dysplasia (ARVD) are rare genetic arrhythmogenic disorders predisposing to sudden cardiac death (SCD) due to pulseless ventricular tachycardia (VT) or ventricular fibrillation (VF).[1,2] Given its profound consequences, comprehending the electrocardiographic features of these rare cardiomyopathies is crucial, especially in emergency settings. Although clinically some similarities have been found,[3] currently, understanding of such an overlap is still limited. Thus, in the following case, we report incidental ECG findings of Brugada syndrome and Epsilon wave in a 25-year old patient without any symptoms.

2. Case Presentation

A 25-year old medical student during his clinical clerkship was instructed for electrocardiography examination. He had no complaints, but had a history of syncope episodes, chest pain, and palpitations of unknown origin 10 years ago. The initial assessment revealed blood pressure of 110/70 mmHg and pulse of 68 beats/min. During examination, the ECG showed incomplete right bundle branch block, Brugada-type 1 pattern, with signs of Epsilon wave (Figure 1). However, he refused to take further examinations. One year later, assessment of the ECG was repeated, and findings were suggestive of Brugada syndrome. Thus, to increase the sensitivity, the lead was placed in the third and second intercostal spaces (Figure 2). This modification further unmasked the ECG abnormalities suggestive of Brugada syndrome. Echocardiography was the performed, revealing unremarkable results with EDD 42, ESD 29, EF 60%, global normokinetic with good valves, TAPSE 2.4, no RV dilatation, and regional dyskinesia in RV suggesting no arrhythmogenic right ventricular dysplasia.

3. Discussion

This case report identified incidental findings of type-1 Brugada syndrome with “Epsilon-like” pattern in otherwise healthy patient. According to a consensus report in 2005, there are three types of abnormal ECG repolarization patterns (type 1-3) for BrS, of which only type 1 (characterized by ST-segment elevations in the right precordial leads) is diagnostic. The current 2012 ACC/AHA ECG criteria, however, only recognized 2 ECG patterns: pattern 1 (the coved pattern), which is identical to type 1, and pattern 2 (the saddle-back pattern), which combines pattern 2 and 3 of the former consensus. Pattern 1 is characterized by the coved-shaped ST segment elevation (STE) > 2 mm, J-point elevation, and a gradually descending ST segment, which terminates with a negative T-wave in the right precordial leads (V1, V2 and V3) with or without a class I antiarrhythmic drug challenge such as flecainide. Meanwhile, pattern 2 is characterized by a saddleback morphology with a minimum 2 mm J-point elevation along with ST segment elevation of at least 1 mm that includes ECG repolarization types 2 and 3.[3-5] In this patient, his ECG findings revealed a fragmented QRS complex with incomplete right bundle branch block (RSr’), a coved-shaped ST elevation pattern with J-point elevation of 1 mm, a gradually descending ST segment, and T inversion in the anterior precordial leads, indicating a type-1 Brugada pattern. Placement of the right precordial leads in a superior position (up to the second intercostal space above normal) in this patient could increase the sensitivity of the ECG for detecting the Brugada phenotype.[5] The presence of either pattern advocates SCD prevention using an implantable cardioverter defibrillator (ICD).[5-7] In this case, we also observed an Epsilon wave, which is pathognomonic for arrhythmogenic right ventricle dysplasia (ARVD). Arrhythmogenic right ventricle dysplasia is an inherited heart muscle disease that predominantly affects the right ventricle, characterized by the replacement of myocardium with fibrofatty tissues. Clinically, ARVD presents with ventricular electric instability with VT or VF that may lead to sudden cardiac death, primarily in young people and athletes. ARVD and BrS are distinct clinical entities with respects to both clinical presentation and genetic predisposition. In contrast to BrS, imaging techniques such as echocardiography in patient with ARVD shows right ventricular morphological and functional changes. Previous studies have found some crossover has been reported between these conditions, and some even reported that BrS can be considered as a form of ARVD. It is possible, given the fact that in early stage of ARVD, fatty deposits could be so minimal that it is undetectable.[2] In this case, we could not detect any structural heart disease with echocardiography, such that we diagnosed this patient with Brugada syndrome with “Epsilon-like pattern”.

There are many risk factors that have been associated with higher likelihood of developing VT/VF in BrS. These include male gender, occurrence of syncope, genetic mutation in SCN5A, the presence of a spontaneous type 1 Brugada pattern, early repolarization pattern in inferolateral leads, S-wave in lead I, T-wave alternans, fragmented QRS morphology, burden of Brugada ECG pattern on Holter monitoring, augmented ST elevation during exercise recovery, an abbreviated ventricular refractory period of <200 ms, activation-recovery interval prolongations, and inducible arrhythmias observed during programmed electrical stimulation.[1,8,9] Of these, the risk factors identified in this patient include male gender, history of syncope, presence of spontaneous type-1 Brugada pattern, and fragmented QRS morphology, which increased the likelihood of developing Brugada syndrome.

Since the pathophysiological basis of BrS lie in the decreased magnitude of inward currents, pharmacological agents that act to increase the inward currents or reduce the outward currents can restore the balance. Interventional alternatives include implantable cardiac defibrillator (ICD) insertion and radiofrequency ablation. ICD insertion appears to be safe in the long term and reduces cardiovascular mortality but not morbidity including in BrS patients. However, its use is not without significant morbidity, as complications such as lead failure and infections could occur. Moreover, the quality of life could be affected from inappropriate shocks, frequently prior to the presence of supraventricular arrhythmias. In other cases, radiofrequency ablation can be used to successfully prevent VT/VF.[10-12] In this case, the patient received no further examination nor intervention due to cathlab shutdown and restrictions during the COVID-19 pandemic. However, according to ESC guidelines, it is recommended that this asymptomatic patient should undergo electrophysiology study to observe the likelihood of true Brugada and suitability for radiofrequency ablation or implantation of ICD.[13]

4. Conclusion

Brugada syndrome and arrhythmogenic right ventricle dysplasia are inherited primary arrhythmia syndromes which predisposes to sudden cardiac death. As we further recommend in this case, asymptomatic Brugada patients should undergo electrophysiology study to observe the likelihood of true Brugada and suitability for radiofrequency ablation or implantation of ICD.

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Abbreviations

ARVD	arrhythmogenic right ventricular dysplasia
BrS	Brugada syndrome
ICD	implantable cardioverter defibrillator
SCD	sudden cardiac death
VF	ventricular fibrillation
VT	ventricular tachycardia

Declarations

Funding

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Conflicts of interest

None

Ethics approval and consent to participate

Patient in the case report has agreed and fulfilled Ethics Committee Faculty of Medicine, Universitas Indonesia, Jakarta, Indonesia

Consent for publication

Informed consent of patient's data was obtained from the patient's guardian

Availability of data and material

Not applicable

Code availibility

Not applicable

Authors' contributions

BM has contributed in design of the work, interpretation of the patient's data, and editing the figures. VJD, RP and SNS have contributed in elaborating the discussions. All authors have read and approved the manuscript.

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Figures

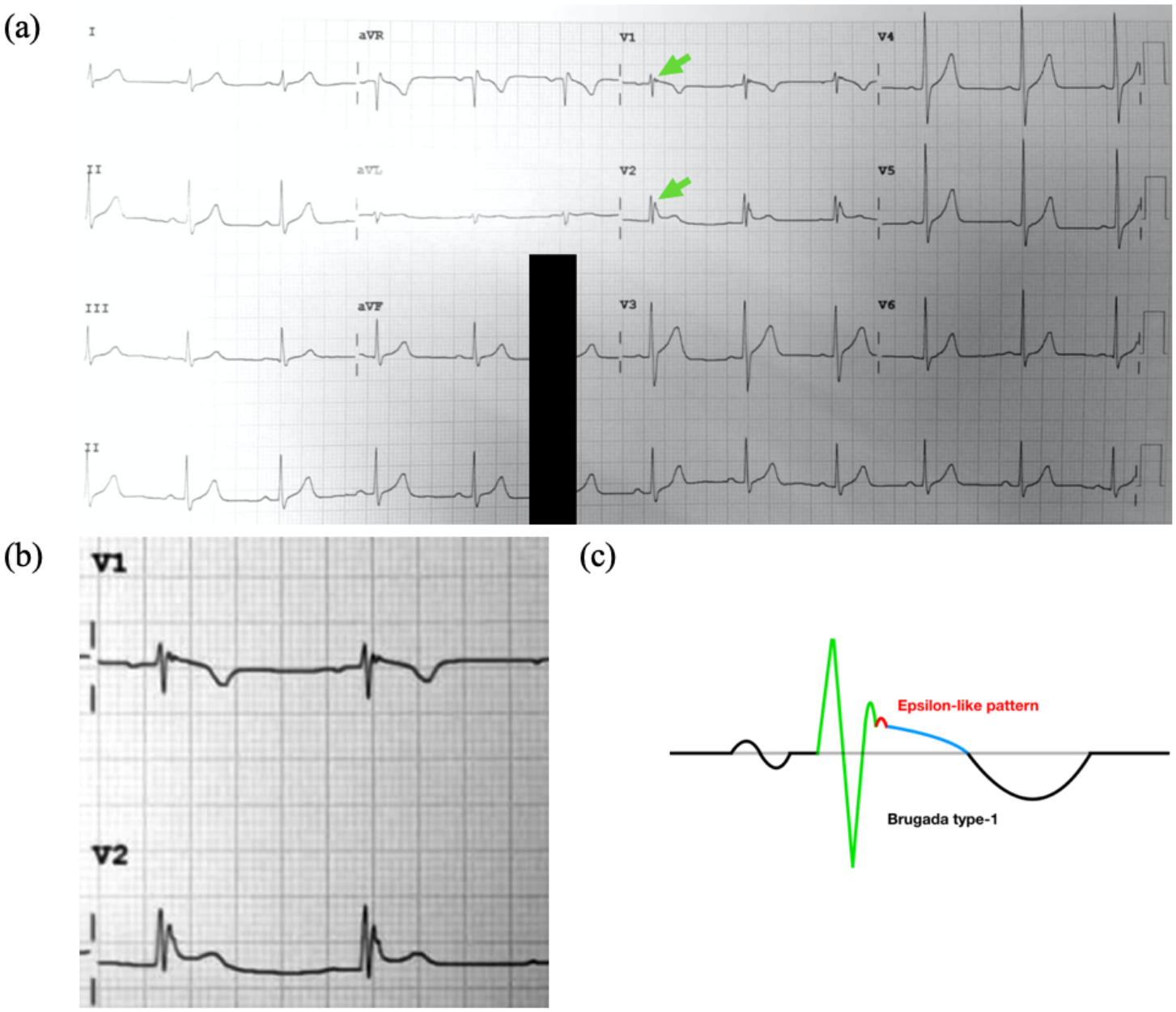
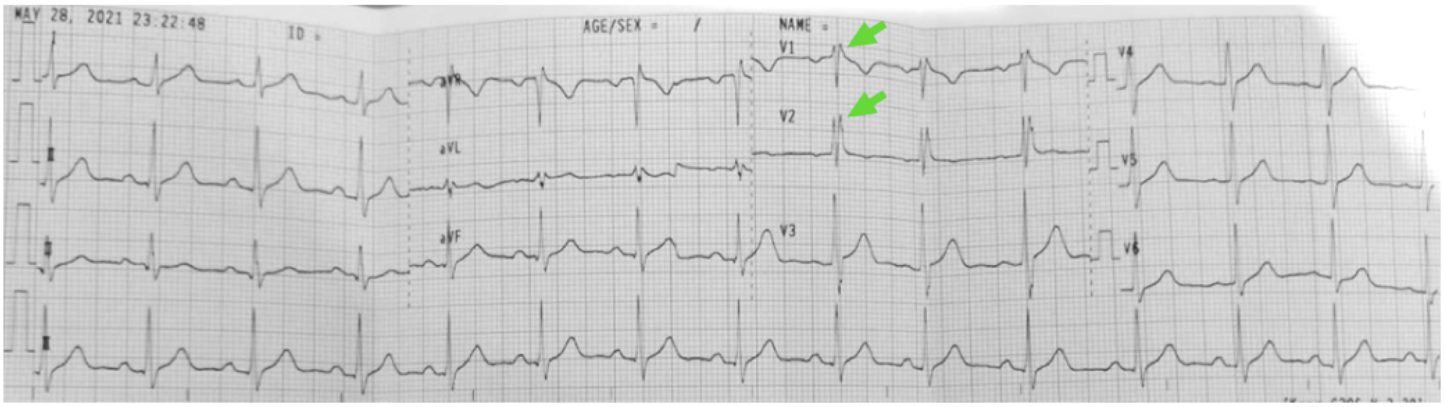


Figure 1

(a) Initial ECG showed fragmented QRS complex with incomplete right bundle branch block (RSr'), coved-shaped ST elevation pattern with J-point elevation of 1 mm, gradually descending ST segment, and T inversion in the anterior precordial leads indicated by the green arrow. **(b)** Epsilon wave could be observed after the incomplete RBBB, suggesting the presence of arrhythmogenic right ventricular dysplasia. **(c)** Illustration of the ECG in this patient; green color showed incomplete RBBB, red color showed Epsilon-like pattern and blue color showed characteristic of Brugada syndrome.

(a)



(b)

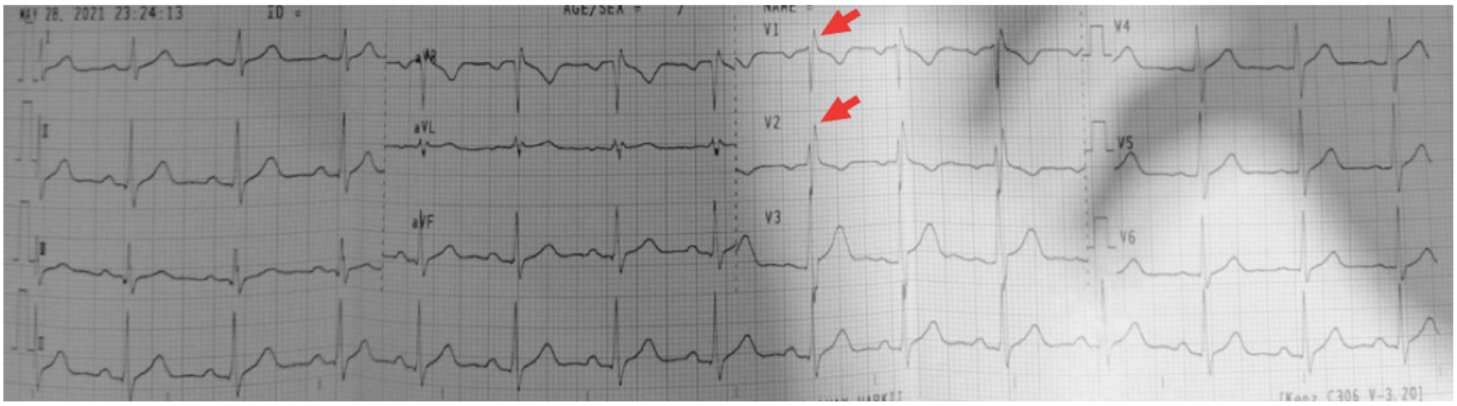


Figure 2

The following year, the ECG was repeated. (a) Incomplete right bundle branch block with downward ST-segment and T inversion could be observed (indicated by green arrow). **(b)** To increase the sensitivity, the lead was placed in the third and second intercostal spaces (indicated by red arrows).

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- CAREchecklistEnglish2013.pdf