

Silent and Malignant Early Repolarization Syndrome Mimicking Hyper-Acute ST Elevation Myocardial Infarction

Weng-Chio Tam,^{1,2} Ming-Hsiung Hsieh,^{1,2} Yung-Kuo Lin^{1,2} and Jong-Shiuan Yeh^{1,2}

A 55-year-old male with underlying type 2 diabetes mellitus and hypertension presented at our emergency department with ventricular fibrillation-related cardiac arrest. Hyper-acute ST elevation myocardial infarction was the preliminary diagnosis by 12-lead electrocardiography, which simultaneously showed J point ST elevation and tall T waves. However, the echocardiography showed concentric left ventricle hypertrophy and preserved left ventricular systolic function with no regional wall motion abnormalities, and coronary angiography did not show any critical coronary artery lesion. Malignant early repolarization syndrome was diagnosed, and an implantable cardioverter defibrillator was implanted. Early repolarization syndrome is associated with J point elevation, and more involved leads and an increased J point elevation amplitude can increase the risk of arrhythmogenicity. In summary, we report a case with asymptomatic type 3 early repolarization syndrome-induced idiopathic ventricular fibrillation mimicking hyper-acute ST elevation myocardial infarction.

Key Words: Early repolarization • Idiopathic ventricular fibrillation • J wave syndrome

INTRODUCTION

Herein, we have presented the case of a patient with asymptomatic type 3 early repolarization syndrome with the initial presentation of ventricular fibrillation and sudden cardiac arrest. An electrocardiography (ECG) pattern mimicking hyper-acute ST elevation myocardial infarction was the initial presentation, and acute myocardial infarction and obvious structural heart disease were excluded by use of coronary angiography and echocardiography. To the best of our knowledge, this is

the first case report of its type in Taiwan. For an asymptomatic early repolarization pattern, distinguishing malignant ECG abnormalities from normal variants may be more important than natural history with regards to prognostic significance. Early screening, such as electrophysiological studies with programmed electrical stimulation to evaluate ventricular arrhythmia may be necessary to predict the risk in patients with a potentially malignant early repolarization pattern.

CASE REPORT

A 55-year-old male presented with a medical history of type 2 diabetes mellitus and hypertension but denied any syncope or family history of sudden cardiac death. He was sent to our emergency department due to sudden cardiac arrest at home. His vital signs recovered after several cycles of cardiopulmonary resuscitation and cardio-version. The patient's initial 12-lead ECG showed atrial fibrillation with ST elevation in V3-6 (Figure 1A). A

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¹Division of Cardiovascular Medicine, Department of Internal Medicine, Wanfang Hospital Taipei Medical University; ²Department of Internal Medicine, School of Medicine, College of Medicine, Taipei Medical University, Taipei, Taiwan.

Address correspondence and reprint requests to: Dr. Jong-Shiuan Yeh, Division of Cardiovascular Medicine, Wanfang Hospital, Department of Internal Medicine, Taipei Medical University, No. 111, Section 3, Hsing-Long Road, Taipei 116, Taiwan. Tel: 886-2-2930-7930; Fax: 886-2-2933-9378; E-mail: yvesyeh@gmail.com

second ECG was performed 5 minutes later, which showed J point ST elevation and tall T waves at precordial leads with reciprocal changes at leads I and aVL (Figure 1B). Hyper-acute ST elevation myocardial infarction was then suspected. However, emergency coronary angiography showed an absence of significant coronary artery lesions, and laboratory data revealed that electrolytes were within normal range. Slightly elevated creatine kinase-MB (15.9 ng/ml) and troponin-I (0.51 ng/ml) levels were noted initially, however they did not increase further during follow-up. Echocardiography showed concentric left ventricle hypertrophy (septal wall: 14 mm, posterior wall: 14 mm) with preserved left ventricle systolic function. A 24-hour Holter-ECG monitor revealed sinus rhythm without any sustained or non-sustained ventricular arrhythmia. Ventricular fibrillation (VF) was assessed by analyzing automated external defibrillator records (Figure 1C). Early repolarization syndrome-induced idiopathic VF was diagnosed because of J the point-like ST elevation, and the absence of significant stenosis of the coronary arteries and other structural heart diseases. An implantable cardioverter defibrillator was implanted for secondary prevention and he was discharged uneventfully. No recurrence of ventricular arrhythmia was noted during follow-up.

DISCUSSION

An early repolarization pattern is a common electrocardiographic finding first described in 1936 by Shipley et al.¹ It is frequently observed in young individuals and is considered to have a benign clinical course and normal variants.² However, Gussak et al.³ reported a potentially dangerous role in arrhythmogenicity. They also reported that some idiopathic ventricular arrhythmias may be related to an early repolarization pattern in healthy young adults without structural heart disease. Supporting this hypothesis, Haissagure et al.⁴ reported that patients with a history of idiopathic VF have a higher prevalence of early repolarization. These findings changed the traditional thinking about an early repolarization pattern from being a benign clinical course to a risk factor for malignant ventricular arrhythmia.

An early repolarization pattern in ECG consists of J point elevation, a J wave with or without ST segment

elevation and a notch or slur of the terminal part of the QRS complex. Early repolarization syndrome is classified into three categories which are characterized by the location of the early repolarization pattern. Type 1 mainly manifests in the lateral precordial leads, often occurs in healthy athletes, and is associated with a lowest risk of arrhythmia. Type 2 simultaneously manifests in the inferior and inferior-lateral leads, and is associated with a moderate risk. An early repolarization pattern existing in inferior, lateral and right precordial leads is categorized as Type 3, which has been reported to carry the highest risk of malignant ventricular arrhythmia and electrical storm.⁵ The early diagnosis and screening of early repolarization syndrome still remains a challenge, because J wave amplitude and the early repolarization pattern can change and fluctuate dynamically and dramatically in various situations such as exercise, taking drugs and rapid atrial pacing.⁶ Acute myocardial infarction and pericarditis should be differentiated from early repolarization syndrome by the aid of physical examination, specific electrocardiographic, echocardiographic manifestation and images of coronary angiography. After structural etiologies have been excluded, some specific early repolarization patterns in ECG can also help to evaluate risk stratification. Elevated J point amplitude, convex upward J wave, horizontal ST segment, lambda-wave ST shape and left precordial QRS notching pattern have been reported to be associated with malignant ventricular tachyarrhythmia.⁷ The diagnostic algorithm of early repolarization syndrome has been summarized in Figure 2. In our case, both of global distribution of early repolarization pattern, obvious (> 2 mm) J point amplitude elevation (Figure 1B arrow) and convex upward ST segment elevation in inferior and lateral leads (Figure 1A arrow) were suggestive of potential risk of ventricular tachyarrhythmia.

The treatment of early repolarization syndrome mainly focuses on secondary prevention. Implantable cardioverter defibrillator implantation is indicated to prevent sudden cardiac death in patients with early repolarization syndrome and documented ventricular fibrillation. Quinidine or amiodarone are also effective to prevent the recurrence of ventricular arrhythmia and to reduce the number of shocks, particularly in an electrical storm setting.⁸ Currently, there are no definitive guidelines for the primary prevention of ventricular

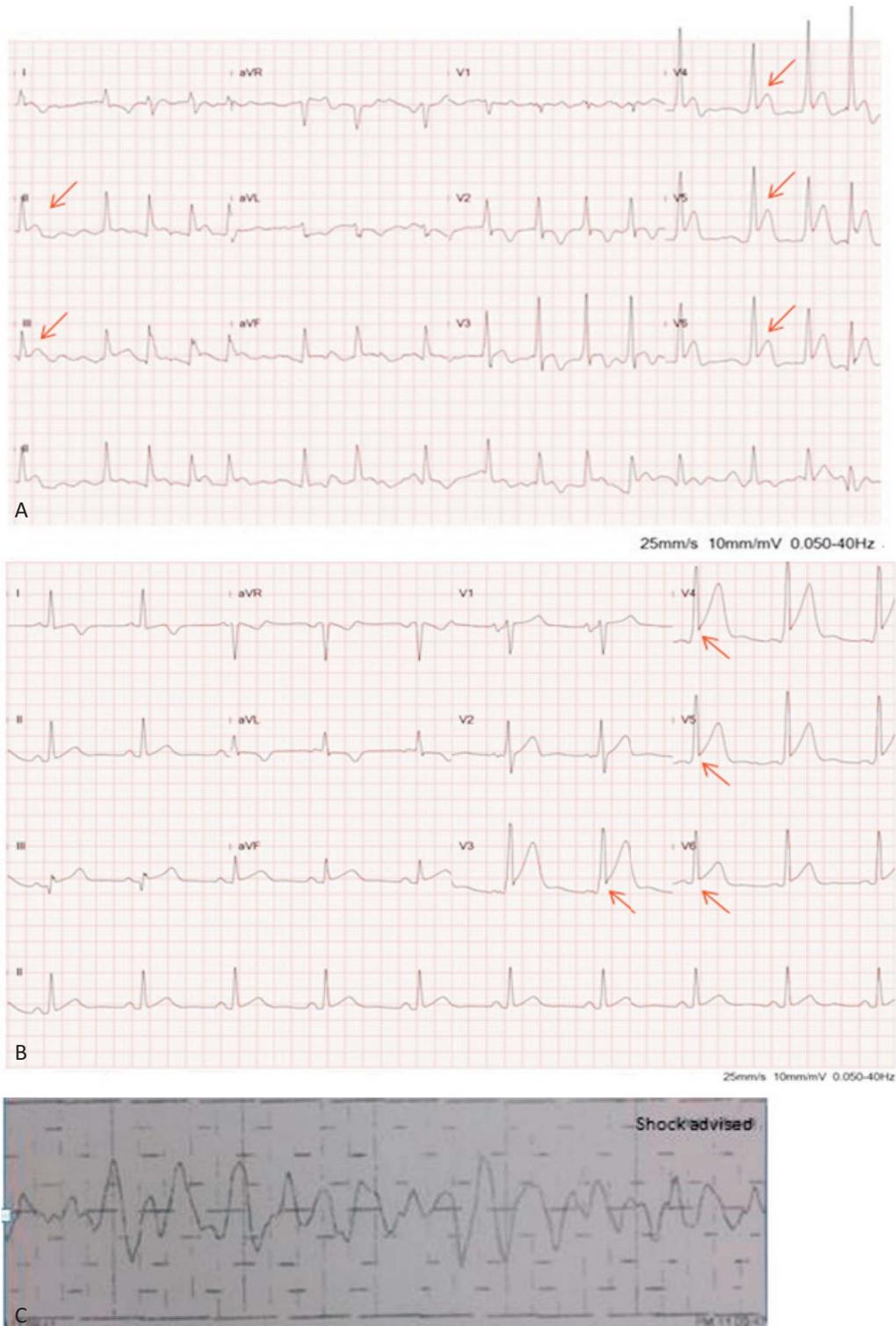


Figure 1. A 12-lead electrocardiogram (ECG) showed atrial fibrillation with moderate ventricular response and new onset of convex upward ST segment elevation in inferior and lateral leads (arrow) (A). A 12-lead electrocardiogram (ECG) showed sinus rhythm with type 3 early repolarization pattern which consisted of J point ST elevation in inferior (II, III, aVF), inferior lateral (I, V5-6) and anterior precordial leads (V1-4), especially in precordial leads (arrow). Tall T waves in all precordial leads (V1-6) with some reciprocal changes in leads I, aVL were also revealed (B). Electrocardiogram tracing of ventricular tachyarrhythmias from automated external defibrillator (C).

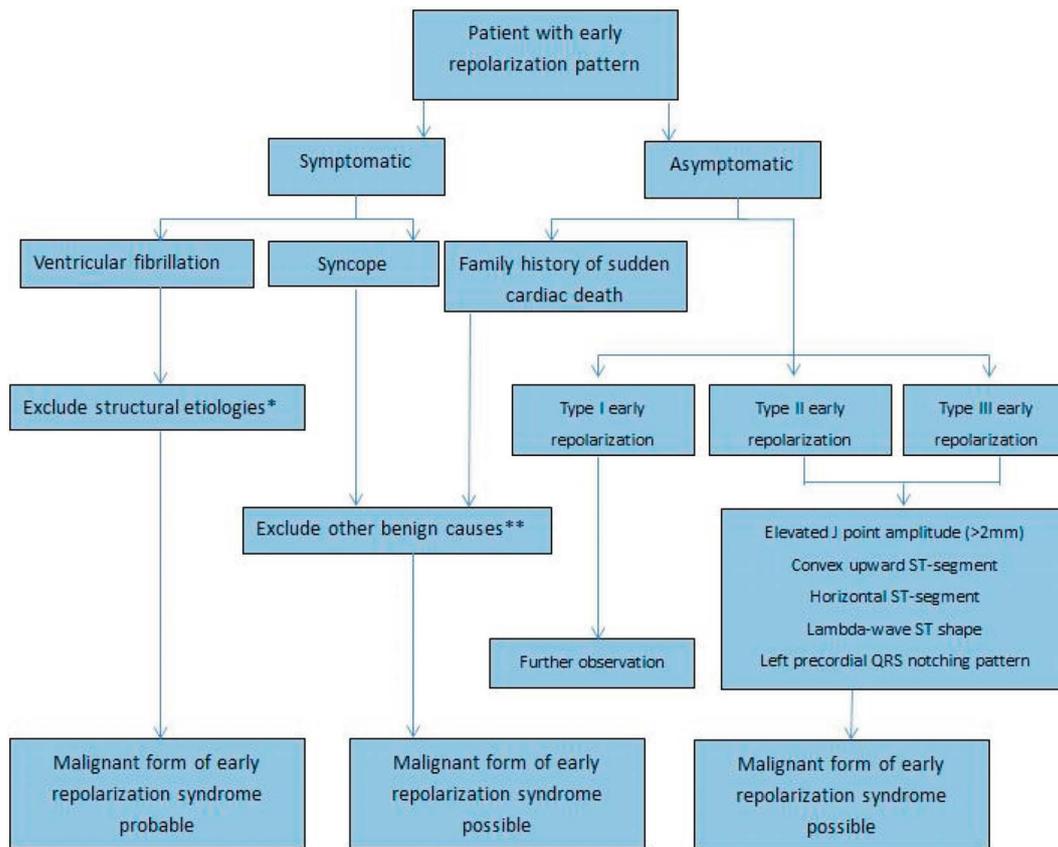


Figure 2. Diagnostic algorithm of early repolarization syndrome. Clinical evaluation including coronary angiography, cardiac magnetic resonance imaging, and signal averaged electrocardiogram is necessary to exclude structural etiologies (*). Clinical evaluation including vasovagal syncope, Head-up tilt test, ambulatory electrocardiogram and electrophysiological study is necessary to exclude other benign causes (**).

arrhythmia in asymptomatic patients with a high risk early repolarization pattern. Some researchers have suggested that implantable cardioverter defibrillator implantation can be considered for such patients,⁹ whereas others have reported controversial results.¹⁰ In these patients, complete work-up such as the head-up tilt test, ambulatory ECG monitoring, and electrophysiological studies with programmed electrical stimulation to induce ventricular arrhythmia may be necessary.⁸

In conclusion, we present a case with asymptomatic type 3 early repolarization syndrome with an initial presentation of VF and sudden cardiac arrest. This is the first such case reported in Taiwan. Despite the initial ECG mimicking hyper-acute ST elevation myocardial infarction, silent and malignant J wave manifestation should be suspected initially. In some asymptomatic subjects with a high risk early repolarization pattern but without syncope or family history of sudden death as with our patient, ECG abnormalities may be more im-

portant than a natural history with regards to the prognostic significance and risk stratification. This case may help to focus attention on asymptomatic and insidious high risk early repolarization syndrome. An early diagnosis and screening may be necessary to predict the risk of malignant ventricular tachyarrhythmia.

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105swf06 Wan Fang hospital Taipei Medical University.

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