

Ventricular Tachycardia and Atrial Fibrillation in an Adolescent Patient with Congenital Myotonic Dystrophy

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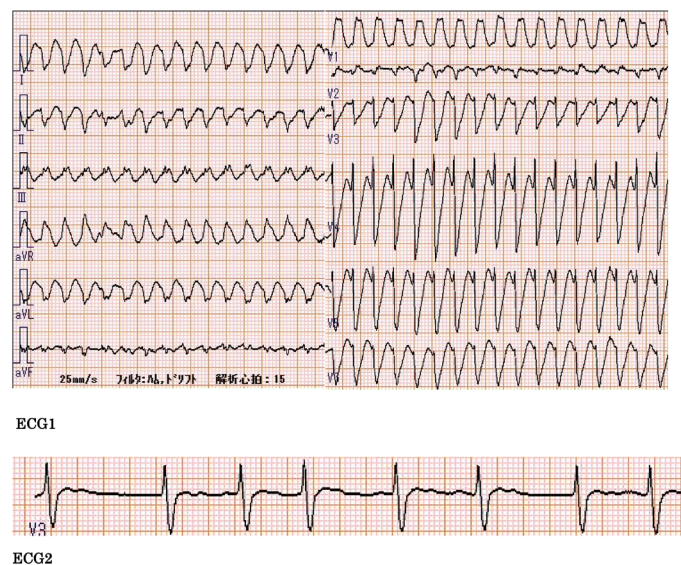


Fig. 1 The ECG records during the arrhythmic events
ECG1; VT (the ventricular rate >250 bpm), ECG2; AFib.

Introduction

Myotonic dystrophy type 1 (MD1) is a multisystem disease caused by mutant CTG expansion. Congenital MD1 (CMD1) is an uncommon phenotype of MD1 and its mortality rate is high up to 40%.¹⁾ The detail of cardiac complications, which are common in classical adult MD1, remains unclear in CMD1. We present an adolescent female who had CMD1 with ventricular

tachycardia (VT) followed by atrial fibrillation (AFib).

Case Report

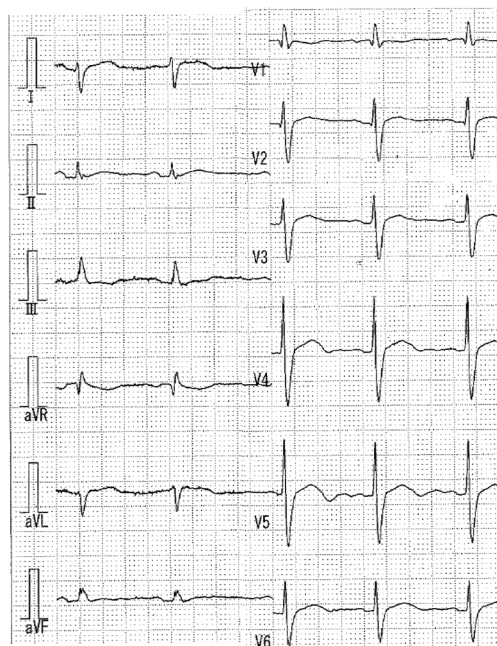
A 13-year-old girl was referred with complaints of general fatigue and palpitation that occurred for the first time after exercise. During physical examinations, she was drowsy, and her systemic systolic blood pressure was very low up to 60 mmHg. The electrocardiography revealed the wide QRS tachycardia (Fig. 1, ECG1), in

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ECG 3

Fig. 2 Nonspecific intraventricular conduction delay was observed on electrocardiogram at elementary school physical examinations, 7 years ago (ECG3)

which a rapid bolus injection of adenosine triphosphate did not make any changes. Even though the atrial response could not be identified in very rapid rhythm, VT was strongly suspected, and a synchronized cardioversion successfully treated the tachycardia. An irregular ventricular rhythm then followed for half a day (Fig. 1, ECG2) and returned to the sinus rhythm spontaneously. We assumed this irregular rhythm was AFib because of being without any distinct P waves. Echocardiography, coronary CT and gadolinium enhanced cardiac MRI were normal. Blood tests revealed the elevation of serum creatine kinase levels (1600 U/L). She also showed foot deformity, facial muscle weakness, intraventricular

conduction delay (Fig. 2), and a history of respiratory difficulty in the neonatal period, which suggested the presence of muscular dystrophy. A genetic examination revealed CTG expansion on the myotonic dystrophic protein kinase gene over 1000 repeats, which gave the definitive diagnosis of CMD1.

Comments

CMD1 demonstrates a unique “biphasic” course, i.e., neonatal symptoms improve in survived neonates, but then adult-type symptoms could become present in later life. In addition, it is unclear from when patients show the adult-type manifestations.²⁾

In our case, the first arrhythmic event happened when she was 13 years old. Though the elementary school physical examinations had revealed her ECG anomaly 7 years ago, the school doctor had followed her up as a normal variant because she had no cardiac problems before this episode.

Since the incidence rate of sudden cardiac death in MD1 patients is reported high,³⁾ the patient's arrhythmic manifestation may worsen in future. We plan to follow her up considering ICD therapy.

Conflict of Interests

The authors have no financial conflicts of interest to disclose concerning the presentation during past three years.

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