Markedly High B-type Natriuretic Peptide Level in a Patient with Duchenne Muscular Dystrophy and Left Ventricular Non-Compaction

Ikuo Misumi¹, Yasuto Nishida², Tsuyoshi Honda¹, Hiroyumi Kurokawa¹, Hisayo Yasuda³, Koichi Kaikita³, Seiji Hokimoto³ and Hisao Ogawa³

Abstract

A boy with Duchenne muscular dystrophy was admitted to our hospital due to a transient loss of consciousness. Transthoracic echocardiography revealed left ventricular (LV) dilatation and diffuse hypokinesis of the LV wall. The LV wall was thin, and both non-compaction of the LV wall and marked thinning of the posterior LV wall resulting from a lesion were observed. The plasma B-type natriuretic peptide (BNP) level ultimately increased to 7,795 pg/mL, and the patient died of cardiac arrest following ventricular tachycardia. Severe heart failure, a critical condition, and thinning of the LV wall may have contributed to the markedly high plasma BNP level in this case.

Key words: Duchenne muscular dystrophy, high BNP level, heart failure, left ventricular noncompaction

(DOI: 10.2169/internalmedicine.54.3960)

Introduction

Duchenne muscular dystrophy (DMD), an inherited X-linked disease associated with the absence of dystrophin (1), is characterized by progressive muscle weakness and fatal cardiac involvement similar to that observed in dilated cardiomyopathy (DCM) (2). The plasma B-type natriuretic peptide (BNP) level increases in association with the progression of cardiac dysfunction, although it is lower than that seen in cases of DCM with similar cardiac dysfunction (3, 4). In the present case report, we describe the case of a patient with DMD with a markedly increased plasma BNP level who died of left ventricular (LV) failure.

Case Report

A teenage boy with DMD who had been followed up for nine months and relied on a power wheelchair was admitted to our hospital due to a transient loss of consciousness. A physical examination showed a BMI of 30.8 (height: 155 cm and weight: 73.9 kg), blood pressure of 123/73 mmHg and pulse rate of 136/min. Pulse oximetry revealed an arterial oxygen saturation of 99% and blood sampling disclosed mild anemia (hemoglobin level, 11.3 g/dL), liver injury (aspartate aminotransferase level, 47 IU/L; alanine aminotransferase level, 56 IU/L; and lactate dehydrogenase level, 396 IU/L), a high creatine kinase level (827 IU/L), a high creatine kinase level (827 IU/L) and a high plasma BNP level (3,003 pg/mL). A chest radiograph demonstrated a cardiothoracic ratio of 66%, without pulmonary congestion (Fig. 1, left). In addition, 12-lead electrocardiography (ECG) showed left atrial overload, typical tall R waves in the right precordial leads and Q waves in the lateral leads (Fig. 1, right), and continuous bedside ECG monitoring revealed nonsustained ventricular tachycardia, suggesting that the faintness may have been caused by ventricular arrhythmia. Furthermore, transthoracic echocardiography demonstrated LV dilatation and diffuse hypokinesis of the LV wall (LV end-diastolic dimension, 69 mm; LV end-systolic dimension, 63 mm; and ejection fraction, 20%). The LV wall was thin, with non-compaction, demonstrating hypertrabeculation and marked thinning of the posterior LV
Figure 1. A chest radiograph (left panel) showing a cardiothoracic ratio of 66%, without pulmonary congestion. A 12-lead electrocardiogram (right panel) showing left atrial overload, tall R waves in the right precordial leads and Q waves in the lateral leads.

Figure 2. Apical four-chamber view in diastole (left panel) and systole (right panel) showing left ventricular (LV) non-compaction of the LV wall, with hypertrabeculation (right panel, arrow) and marked thinning of the posterior LV wall.

wall as a result of a lesion (Fig. 2) (5). The thickness of the interventricular septum and posterior wall was 7 and 3 mm, respectively (Fig. 3), while the left atrial dimension was 33 mm. Moreover, continuous-wave Doppler echocardiography revealed a pressure gradient of tricuspid regurgitation of 15 mmHg. The patient did not undergo a myocardial biopsy or cardiac MRI.

As the patient’s family did not wish for aggressive treatment and the ventricular arrhythmia may have been caused by the heart failure, he was medically treated with diuretic
agents and carperitide infusion. However, the plasma BNP level eventually increased to 7,795 pg/mL (Fig. 4), and he died of cardiac arrest after the onset of ventricular tachycardia.

**Discussion**

**Diffuse LV hypokinesis in this case**

Myocardial involvement of DMD is known to be initially confined to the posterior wall, after which it extends to the whole ventricle. In the present case, echocardiography showed marked LV enlargement and diffuse LV hypokinesis, indicating whole LV involvement of the DMD.

**Diagnostic criteria for LV non-compaction**

Findings of a ratio of >2.0 between the thickness of the non-compacted and compacted myocardial layers in systole is considered diagnostic (5) and compatible with that observed in the present case. Moreover, the LV thinning noted in the present case may have been related to the incomplete compaction of the LV posterior wall.

**Low plasma BNP level in DMD**

In cases of DMD, the plasma BNP level remains normal until the onset of advanced cardiac dysfunction and is lower than that seen in cases of DCM with similar cardiac dysfunction (3, 4). There are reports that obesity, physical inactivity and myocardial fibrosis contribute to low plasma BNP levels (6). The present patient was obese and physically inactive due to muscular weakness. Although a myocardial biopsy and cardiac MRI can be used to detect myocardial fibrosis (7), our patient did not undergo these examinations.

We previously reported the plasma BNP levels in 16 cases of DMD. In that report, plasma BNP levels of more than 200 pg/mL were observed in only two patients with marked LV dilation who died of heart failure. However, the degree of elevation of the plasma BNP levels was moderate (349 pg/mL and 1,116 pg/mL, respectively) (8). As far as we investigated, there have been no reports of markedly high plasma BNP levels in patients with DMD.

**Markedly high plasma BNP level in this case**

In the present case, the symptoms were present even at rest, and transthoracic echocardiography showed marked LV

---

**Figure 3.** Apical long-axis view showing a thickness of the interventricular septum and posterior wall of 7 and 3 mm, respectively.

**Figure 4.** Time course of the plasma BNP level in this case, showing that the plasma BNP level increased to 7,795 pg/mL one week before the patient’s death.
enlargement as well as severe LV systolic dysfunction, indicating the end stage of heart failure; the patient subsequently died of ventricular tachycardia and severe heart failure. There are reports that the plasma BNP level increases considerably as a result of a high LV filling pressure (9), high pulmonary capillary wedge pressure (10) and sudden cardiac death (11). These conditions may have caused marked elevation of the plasma BNP level in the current case. Moreover, in the present case, high LV wall stress due to the markedly thin LV wall may have contributed to the high plasma BNP level (12). This case suggests that a marked increase in the BNP level during heart failure treatment may be a sign of a catastrophic cardiac and/or hemodynamic state that requires additional treatment, such as an implantable LV assist device.

We herein reported a rare case of DMD in which the plasma BNP level was markedly high, likely due to severe LV dysfunction, enlargement, a high filling pressure and wall stress.

The authors state that they have no Conflict of Interest (COI).

References


© 2015 The Japanese Society of Internal Medicine
http://www.naika.or.jp/imonline/index.html