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Case report

Changes in N-terminal pro-B-type natriuretic peptide in a neonate with symptomatic isolated left ventricular noncompaction

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= Abstract =

We describe here our experience with a neonate presenting with cyanosis, grunting, and cardiomegaly, who was diagnosed with isolated left ventricular noncompaction (IVNC) by echocardiography. The patient had high levels of N-terminal pro-B-type natriuretic peptide (NT pro-BNP) and symptoms of heart failure including poor feeding and tachypnea. During the period in which NT pro-BNP levels steadily increased, the patient suffered sudden cardiac arrest despite heart failure management. Following cardiopulmonary resuscitation, cardiac arrest was resolved, NT pro-BNP levels decreased, and all symptoms showed improvement. We consider that assessment of NT pro-BNP with cardiac functional analysis using echocardiography could help in the prediction of disease progress in IVNC. (Korean J Pediatr 2009;52:129-132)

Key Words : Newborn, Myocardium, Natriuretic peptide, Brain

Introduction

Isolated left ventricular noncompaction (IVNC), also known as spongy myocardium, is a rare congenital cardiomyopathy. This cardiomyopathy results from an arrest of the normal process of myocardial compaction in intrauterine endomyocardial morphogenesis[1]. Since the first report for young patients in 1990[2], it has been subsequently described in adults[3,4] and neonate[5]. Echocardiography is the gold standard method for the diagnosis of IVNC. This is characterized by excessively prominent trabecular meshwork and deep intertrabecular recesses in the absence of other structural heart diseases. In spite of intensive care, mortality is more than 50% and perinatal mortality is also high. We report a case of IVNC in a neonate.

Case report

A baby boy was born at 41 gestational weeks and 1 day from a 33 year old mother and a 39 father at a local obstetric clinic by vaginal delivery on 3,560 g. The Apgar score was 8/1 min and 9/5 min. On family history, his father had stroke, but his mother and one sister were healthy.

On the 2nd day of life, he was transferred to our hospital because of cyanosis and grunting.

On physical examination of admission day, his blood pressure was 74/47 mmHg and heart rate was 152 beat per minute with regular rhythm. He had tachypnea (76/min) and mild chest retraction, but aeration was good. Oxygen saturation in room air condition was 95%. Serologic laboratory findings showed elevated N-terminal pro B-type natriuretic peptide (NT pro-BNP, 8,875 pg/mL, Fig. 1A) and creatinine kinase MB (CK-MB, 12.6 ng/mL). Cardiac troponin I was not elevated at that time (0.12 μg/mL, Fig. 1B). The chest X-ray showed cardiomegaly with cardiothoracic ratio 0.73 (Fig. 2).

A 12-lead electrocardiogram finding was unremarkable. The echocardiogram revealed an asymmetric thickness of septum and the left ventricular myocardium and prominent trabeculations with deep intertrabecular recesses (Fig. 3). And color Doppler analysis revealed typical forward and reverse direct blood flow from the ventricular cavity into the spaces between the prominent trabeculations throughout the cardiac cycle. There was also demonstrated an enlarged left atrium and left ventricle with impaired systolic function

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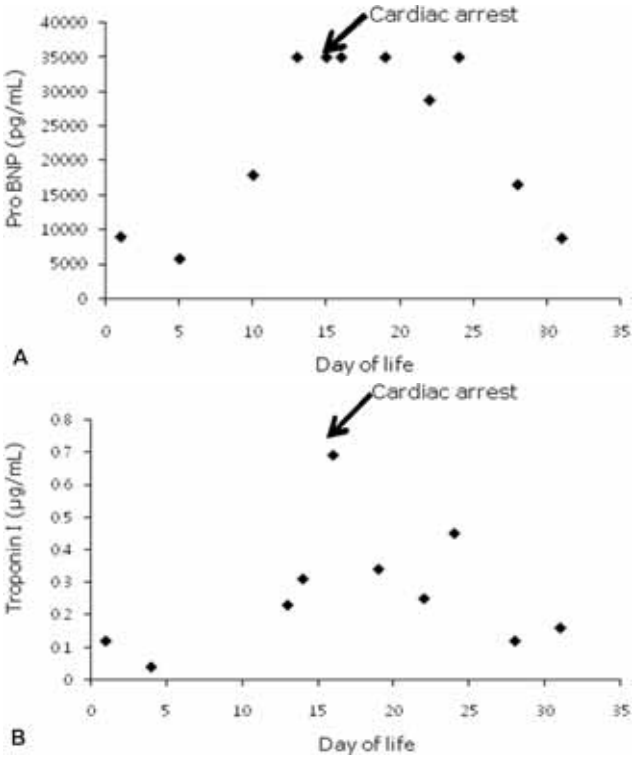

Fig. 1. The plasma levels of NT pro-BNP (A) and cardiac troponin-I (B) according to dates of examination. At 15 days of life, the patient underwent sudden onset apnea and cardiac arrest with no arrhythmia and responded to resuscitative efforts. Black arrow : event day of cardiac arrest.

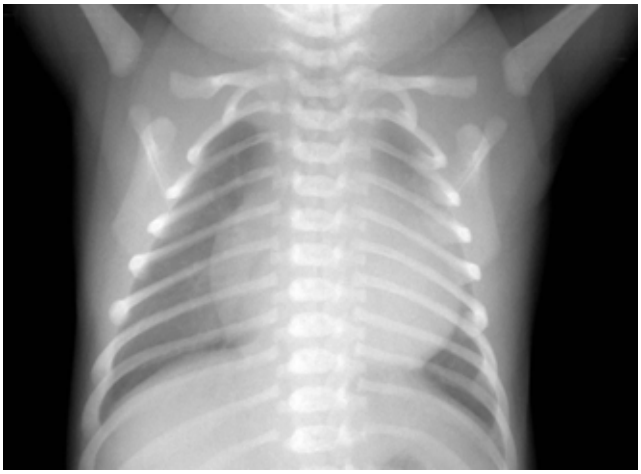

Fig. 2. On admission, chest radiography demonstrated cardiomegaly and clear lung fields (CT ratio: 0.73).

(fraction shortening, FS, 10.8%) and grade I mitral regurgitation. Other cardiac malformation was not showed. We had diagnosed the IVNC with heart failure. Continuous electrocardiographic monitoring and treatment for heart failure using inotropics and diuretics were started.

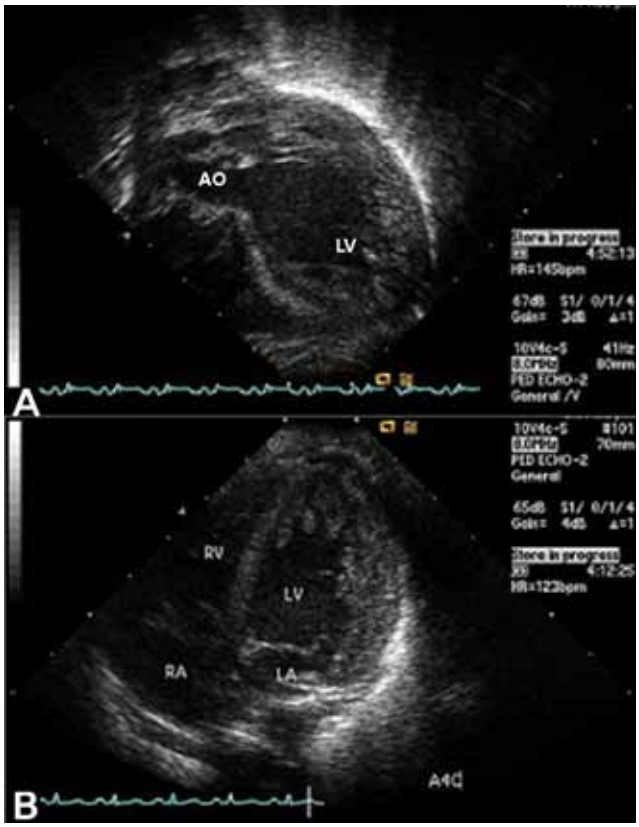

Fig. 3. Findings of echocardiographic examination at subxiphoid long axis view (A) and apical 4-chamber view (B). Prominent trabeculations and intertrabecular recesses were observed at the left ventricular apex and midventricular segment of the inferior and lateral walls. The non-compacted areas were associated with a wall motion abnormality (hypokinesis). The right atrial and ventricular sizes and morphology, as well as the function of the aortic, tricuspid and pulmonary valves were normal. There was no pericardial effusion.

On the 5th day of life, the systolic function (FS 21.5%) was improved at follow-up echocardiography and NT pro-BNP was decreased to 5,680 pg/mL. Follow-up check of NT pro-BNP was performed on the 10th and 13th day. NT pro-BNP was increased to 17,855 pg/mL and 35,000 pg/mL, respectively, but general condition did not showed any interval changes.

On the 15 th day of life, sudden onset apnea and cardiac arrest with no arrhythmia were developed. Full cardiopulmonary resuscitation was done and he responded to resuscitative efforts. However, cardiac function grew worse to FS 6% at follow up echocardiography. At the first cardiac arrest, NT pro-BNP and troponin I were increased to 35,000 pg/mL and 0.69 μg/mL, respectively (Fig. 1). One week has passed and follow-up NT pro-BNP and troponin I were decreased to 16,462 pg/mL and 0.12 μg/mL, respectively

(Fig. 1). In the two weeks following, NT pro-BNP was decreased to 8,671 pg/mL and cardiac arrest did not occur anymore. His parents wanted discharge in spite of the risk of sudden death. One week after discharge, the patient was taken to the emergency department with sudden onset cyanosis after feeding. At the emergency department, he was apneic, cyanotic and had no palpable pulse. He did not respond to full resuscitative efforts and was pronounced dead. An autopsy was recommended but could not be performed due to the refusal by the parents.

Discussion

Typical noncompaction of the ventricular myocardium involves the left ventricular (LV) myocardium[2, 3], whereas the right ventricular (RV) myocardium may be additionally involved in a proportion of cases. Although noncompacted myocardium was originally associated with other congenital cardiac malformations: obstructive lesions of the RV or LV outflow tract and congenital coronary anomalies[6, 7], noncompaction of the LV myocardium can be isolated.

Noncompaction of the ventricular myocardium has been categorized as unclassified cardiomyopathy by the World Health Organization[8]. Its prevalence was 0.05% in an adult referral population and 6.9% of all patients with primary cardiomyopathy in a childhood population-based study[9].

The pathogenetic pathway leading to the isolated form has not been documented yet.

There have been reports on families with several inheritance form of isolated IVNC[10, 11]. In Korea, familial IVNC in asymptomatic phase has been also reported[12]. Familial forms have been observed in 40-50% of IVNC patients in the pediatric population[13].

The clinical manifestations of isolated IVNC are not specific and are highly variable from the asymptomatic status to severe cardiac dysfunction following by death. The major clinical manifestations are heart failure due to systolic and diastolic dysfunction, tachyarrhythmias like as atrial fibrillation, paroxysmal supraventricular tachycardia and ventricular tachyarrhythmias, conduction abnormalities or thromboembolic events[14-16]. The age at onset of symptoms varies widely and deteriorations of left systolic function were observed predominantly in adults and in 60% of pediatric cases.

Two-dimensional echocardiography with color and Doppler studies is the standard diagnostic procedure for IVNC[13], which show excessively prominent trabeculations and multiple deep intertrabecular recesses in the ventricular cavity and the absence of any coexisting cardiac anomalies.

Despite an increasing interest in IVNC, there is no specific treatment for IVNC, especially in asymptomatic patients. In general, treatment depends on the patient's symptoms and complication. The prognosis of IVNC is poor and is associated with high mortality and morbidity due to progressive heart failure[2].

Early detection and treatment of left ventricular dysfunction is very important before irreversible cardiac damage. To do this, a sensitive evaluation of cardiac function is required. In clinical practice, there are noninvasive techniques like echocardiography, magnetic resonance imaging or computed tomography. Brain natriuretic peptide (BNP) is a natriuretic hormone that is present in the heart, particularly in the ventricles. BNP has diuretic, natriuretic and hypotensive effects. It inhibits the renin-angiotensin system, endothelin secretion and sympathetic activity[17]. Therefore, the release of BNP is increased in heart failure although false positive BNP results can be developed by the age. In normal subjects, the plasma concentrations of BNP and NT pro-BNP are similar. However, in patients with left ventricular dysfunction, plasma NT pro-BNP level rises higher than that of BNP[18].

Serial check of NT pro-BNP was performed in our patients. During the period that NT pro-BNP was steadily increased, the patient had sudden cardiac arrest and severe deterioration of general condition. In the following period after NT pro-BNP was decreased , cardiac arrest didnt occur any more and deteriorated symptoms were improved.

In addition to echocardiographic imaging with tissue Doppler imaging, initial screening and serial follow-up with NT pro-BNP can be useful for prediction of disease progress in patients with IVNC. In addition, because of the risk of familial occurrence, screening of first-degree relatives should be performed by echocardiography to identify asymptomatic patients[4].

한 글 요 약

신생아기에 발견된 단독 심실 비치밀화증 1예에서 관찰된 NT pro-BNP의 변화

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저자들은 청색증과 끙끙거림, 심비대로 전원되어 심장 초음파

검사에서 좌심실에 발생한 단독 심실 비치밀화증을 가진 신생아 1예를 경험하였다. 환자에서 측정한 NT pro-BNP는 높았고, 수유 곤란과 빈호흡을 동반한 심부전 증상이 있었다. 심부전증에 대한 치료에도 불구하고 주기적으로 시행한 NT pro-BNP가 더 높아지면서 환자에게서 급성 심정지가 발생하였고, 심폐소생술 이후 NT pro-BNP가 다시 낮아지면서 심부전 등의 증상은 호전을 보였다. 심초음파 검사와 함께 NT pro-BNP를 주기적으로 검사해 보는 것이 병의 경과를 예측하는데 도움이 될 수 있을 것이다.

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