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Delayed Enhancement Cardiac MRI in Isolated Noncompaction of the Left Ventricular Myocardium in a Child

— A Case Report —

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Isolated noncompaction of the ventricular myocardium (INVM) was diagnosed with delayed enhancement cardiac magnetic resonance imaging (MRI) in a 12-year-old boy, who developed dyspnea and syncope while running. Chest radiograph showed no marked cardiomegaly, but revealed bilateral consolidation caused by aspiration pneumonia. Laboratory findings showed plasma level of brain natriuretic peptide (BNP) of 768 pg/dl. Echocardiography showed a slightly thickened myocardium, but the trabecular meshwork region was unclear. He was given a diagnosis of unknown heart failure. His dyspnea and cyanosis improved in response to inotropic agents, oxygen and steroid therapy. However, the plasma BNP levels could not be decreased to normal. Cardiac MRI was performed and delayed-enhancement demonstrated hyperenhancement of prominent trabeculation in the lateral and apical regions of the left ventricle, suggesting fibrosis. The patient was given a diagnosis of INVM, but his status was New York Heart Association Class I heart failure. The diagnosis of INVM in children is difficult because heart failure symptoms are present in only 30% of the cases at diagnosis. Delayed-enhancement MRI is a more precise method of assessment. (*Circ J* 2008; **72:** 676–678)

Key Words: Delayed enhancement cardiac MRI; Isolated noncompaction; Pediatrics; Ventricular myocardium

Isolated noncompaction of the ventricular myocardium (INVM) is a rare congenital disorder, categorized as unclassified cardiomyopathy by the World Health Organization[1] Its pathogenesis is yet to be elucidated, but there is speculation that arrested normal endomyocardial embryogenesis is the basis, and it may occur in isolation or in association with other congenital heart anomalies[2] Patients with INVM have reduced systolic function, systemic embolism, arrhythmia and a poor prognosis for heart failure. The mechanism of the progressive ventricular failure in cases of INVM is still unknown. Characteristic echocardiographic findings are multiple, prominent myocardial trabeculations and deep intertrabecular recesses communicating with the ventricular cavity[3–5]

Case Report

A 12-year-old boy presented with worsening dyspnea and cyanosis, which developed when he was running. In the emergency department he was clearly conscious on arrival, but vomited many times after syncope. The patient was referred to us because of cyanosis after vomiting. Upon admission, physical examination revealed a body temperature of 38.2°C, and mild cyanosis. The liver was not palpable below the right costal margin. The patient's blood pressure was 105/69 mmHg. Laboratory findings included a white blood cell (WBC) count of 16,900/μl, hemoglobin of 15.8 g/dl, platelet count of $28.2\times10^4/\mu$l, lactate dehydrogenase (LDH) of 1,295 U/L, aspartate aminotransferase (AST) of 345 U/L, alanine aminotransferase (ALT) of 100 U/L, creatine kinase (CK) of 2,074 U/L, C-reactive protein (CRP) of 3.12 mg/dl, and a plasma brain natriuretic peptide (BNP) of 768 pg/dl. Blood gases in the radial artery were pH 7.484, PCO_2 31.4 mmHg, PO_2 48 mmHg, HCO_3 23.6 mmol/L, and BE 0 mmol. Chest radiography showed no marked cardiomegaly, but revealed to bilateral consolidation. Electrocardiography (ECG) revealed left axis deviation and slight ST elevation in leads V_{1-3}, but retrospectively, ECG had revealed normal sinus rhythm when he was screened for cardiovascular disease. Two-dimensional echocardiography showed no left ventricular (LV) dilatation (35.0 mm of end-diastolic dimension), trivial mitral regurgitation, and the systolic LV fractional shortening was reduced by 25% on M-mode echocardiography. A slightly thickened myocardium was also observed, but the trabecular meshwork region was unclear. The presence of LV thrombi could not be discounted. He was given a diagnosis of unknown heart failure with aspiration pneumonia because no bacterial infection was revealed. His dyspnea and cyanosis improved in response to inotropic agents, oxygen therapy and steroid therapy, and the WBC, LDH, AST, ALT, CK, CRP decreased to normal levels 1 month later; however, the plasma BNP level remained elevated, so we performed a hemodynamic study, myocardial scintigraphy and cardiac magnetic resonance imaging (MRI) to diagnose the cause of the heart failure.

In the hemodynamic study, the LV pressure was 92/

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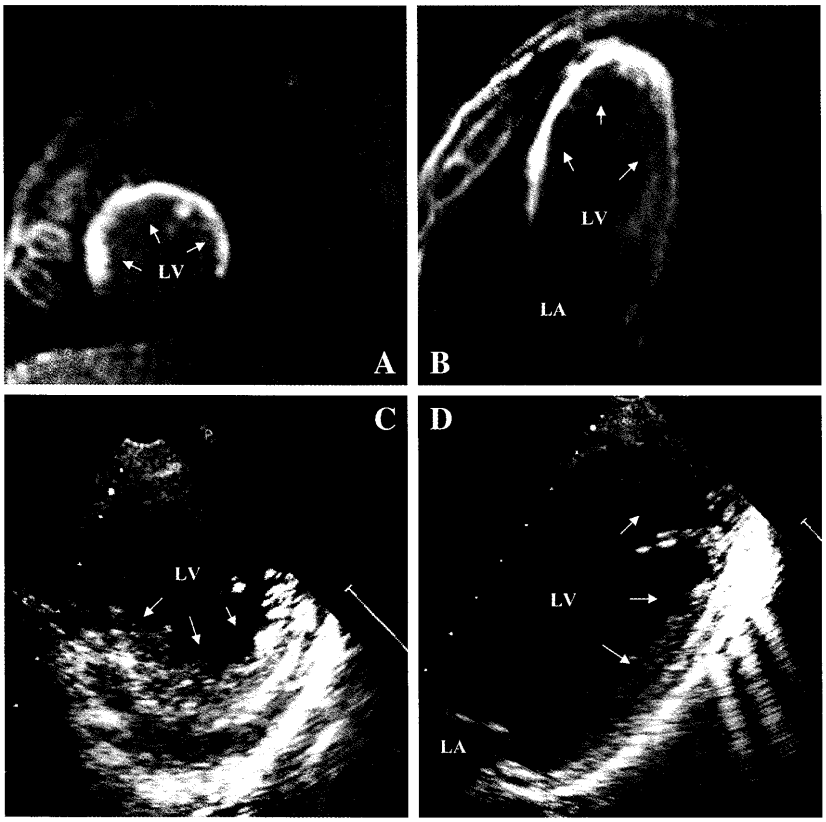

Fig 1. (A,B) Delayed-enhancement magnetic resonance imaging (MRI) 2-chamber view in end-diastole demonstrates hyperenhancement of the trabeculations from the anterolateral to the apex region of the left ventricle (LV) (white arrow). (C,D) Echocardiography in the short-axis view and 2-chamber view demonstrates the trabecular meshwork region from the anterolateral to the apex region of the LV (white arrow) when delayed-enhancement MRI was performed. LA, left atrium.

12 mmHg, the right ventricular pressure was 20/6 mmHg, pulmonary arterial pressure was 18/10 mmHg, and the mean pulmonary capillary wedge pressure was 11 mmHg. LV ejection fraction was 49%. Coronary angiography did not show any occlusive or stenotic lesions. On the left ventriculogram, the trabecular meshwork was suspected from the lateral wall to the apex region. Myocardial thallium-201 scintigraphy at rest showed a low uptake by the apex walls in both the early and delayed phases. Iodine-123 metaiodobenzylguanidine imaging showed a diffuse low uptake and a perfusion defect of the apex walls. MRI was performed with a 1.5-T system (Sigma Excite HD Twin Speed 1.5 T; GE Medical System, Milwaukee, WI, USA). Imaging parameters were as follows: repetition time, 6.6 ms; echo time, 3.1 ms; inversion time, 200 ms to null the signal intensity of normal myocardium. Delayed-enhancement MRI was performed 10 min after infusion of 0.2 mmol/kg injection of gadolinium and hyperenhancement of prominent trabeculation in the lateral and apical regions of the left ventricle was demonstrated, suggesting fibrosis. The patient was given a diagnosis of INVM, but his status was New York Heart Association Class I heart failure. The patient undergoes echocardiography, ECG and chest X-ray every 3 months to monitor ventricular thrombi, arrhythmia and the development of dilated cardiomyopathy.

Discussion

Imaging analyses are important for the diagnosis of INVM because there is no specific histological finding other than fibrosis. Computed tomography and MRI have been reported as useful diagnostic tools, and they may be of value in patients with poor image quality on echocardiography.[6–8] However, echocardiography is considered to be the method of choice for diagnosing INVM, and the promising first-line diagnostic tool,[9] but it must be noted that it is difficult to distinguish normal variants of the physiologically more highly trabeculated right ventricle from pathological noncompaction.[3,5]

In the present case, delayed-enhancement cardiac MRI demonstrated hyperenhancement of the prominent trabeculation in the lateral wall and apex of the left ventricle, suggesting fibrosis, and was very useful for the diagnosis of INVM, which was not initially diagnosed because the trabecular meshwork region was unclear on echocardiography. We could not discern the cause of his syncope because thrombi, arrhythmia, or epilepsy were not present. Jenni et al showed that coronary microcirculatory dysfunction is associated with INVM, causing a decreased coronary flow reserve,[10] and we speculate that in this case syncope was caused by a temporarily low cardiac output related to the coronary microcirculatory dysfunction associated with INVM.

The technique of delayed-enhancement cardiac MRI has been used in several cases of ischemic and non-ischemic cardiomyopathies to detect irreversible myocardial injury (myocardial necrosis or fibrosis).[11] In the present case, the diagnosis of INVM was delayed because of the similarities between INVM and other cardiomyopathies and the examiner's unfamiliarity with its specific diagnostic pattern. The diagnosis of INVM in children is difficult in children, because heart failure symptoms are present in only 30% of the cases at diagnosis. Nearly 90% of pediatric patients developed signs of reduced LV function during a 10-year follow-up period.[9] Jassal et al reported the utility of delayed-enhancement MRI in the pathohistological confirmation of myocardial fibrosis and scarring in the hypertrabeculated myocardium.[12] The imaging technique is simple and pro-

vides much information about the morphology and the tissue character of the LV wall in INVM.

Conclusion

To our knowledge, this is the first reported pediatric case of the use of delayed-enhancement MRI in the pathohistological confirmation of myocardial fibrosis and scarring in the hypertrabeculated myocardium. Delayed enhancement MRI may be an important comprehensive, noninvasive method of evaluating pediatric patients with INVM, because it not only confirms the presence of the recesses and hypertrabeculations in the left ventricle, but also confirms the high risk of such patients for fibrosis within the trabeculations.

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