Lack of MR late-enhancement in left ventricular non-compaction in infants and young children

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Abstract

Objective: Noncompaction cardiomyopathy or left ventricular noncompaction is a rare disease that is probably underdiagnosed. The diagnosis is reached by echocardiography, although MRI provides additional morphological and functional information. Late MRI enhancement is a hallmark of the disease that reflects fibrosis or infarction of affected areas in adults and older children. We aimed to review the presence of late enhancement in left ventricular noncompaction in infants and young children.

Material and methods: We found five very young patients (mean age, 29.4 months; range 1 month to 5 years) with left ventricular noncompaction in our cardiac MRI database. We reviewed the morphological and functional findings, including late enhancement after the administration of contrast material.

Results: All patients had been previously diagnosed by echocardiography. At MRI, the morphological findings and the ratio of noncompacted myocardium to compacted myocardium were compatible with left ventricular noncompaction. None of the cases showed late enhancement after the administration of contrast material.

Conclusions: Unlike in adults and older children, none of the infants and young children we studied had late enhancement. This finding might reflect the natural history of the disease, with subendocardial fibrosis developing over time.

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Palabras clave

Miocardiopatía; Imagen por resonancia magnética cardíaca; Ausencia de realce tardío por resonancia magnética en la no compactación del ventrículo izquierdo en lactantes y niños pequeños

Resumen

Objetivo: La miocardiopatía no compactada o la no compactación del ventrículo izquierdo (NCVI) es una enfermedad rara y probablemente infradiagnosticada. El diagnóstico es ecográfico,
Introduction

Left ventricular non-compaction (LVNC) is a rare congenital heart disease which was first described in 1990. It is one of the “non-classified cardiomyopathies” listed in the World Health Organization’s International Classification of Disease. This myocardial anomaly is frequently under-diagnosed or misidentified as hypertrophic or dilated cardiomyopathy. Early treatment and diagnosis are important because ventricular disfunctions, ventricular arrhythmias or systemic embolisms occur in most patients at some point during the course of the disease. The imaging modality used for diagnosis is ultrasound. Magnetic resonance (MR) imaging correlates well with ultrasound results and can provide additional findings based on the signal intensity and the extent of myocardial contrast enhancement. Inversion-recovery gradient echo sequences with late enhancement (LE-MRI) show abnormal contrast media uptake by the pathological myocardium after the signal from normal myocardium is suppressed. The anomalies that produce these MR findings may trigger lethal arrhythmias or indicate poor function; therefore, this technique may greatly contribute to the care of patients with LVNC.

Material and methods

We searched for “non-compaction” in the cardiac MR database of our hospital and identified six cases of LVNC. One patient was excluded from the study as it was a complex congenital heart disease (tricuspid atresia, ventricular septal defect (VSD) and systemic-pulmonary shunt). In the five remaining cases, LVNC was diagnosed by ultrasound, and a cardiac MR scan was requested as a complementary exam. All studies were performed in an intera 1.5 T magnet (Philips Medical Systems, Best, Netherlands) using a flexible multi-element or specific cardiac coil, depending on the patient’s size. Turbo spin echo sequences were carried out with black blood imaging (repetition time [TR]/echo time [TE] 500/24 ms; echo train 19) and a balanced fast field echo (B-FFE) (TR: 3.4 ms, TE: 1.7 ms, flip angle [FA]: 70°) in the axial, sagittal and coronal planes. The B-FFE sequences were also performed in the cardiac short-axis, 4-chamber and 3-chamber views. Other additional planes were obtained when necessary. The post-contrast late enhancement sequences (TR: 4.2 ms, TE: 1.3 ms, FA: 15°, inversion time [TI]: 200-350 ms and SPIR (spectral inversion recovery) were performed 15 minutes after injection of 0.2 mmol/kg of gadopentetate dimeglumine (Magnevist, Schering, Berlin, Germany). No patient presented clinical suspicion of renal insufficiency, and there were no complications during or after the procedure. To obtain correct cancellation of the normal myocardial signal, the optimal TI was determined using the Look-Locker test sequence.

In patients with abnormal trabeculation of the left ventricular wall, LVNC was diagnosed when the ratio of non-compacted to compacted myocardium was greater than 2. Standard nomenclature was used for myocardial segmentation by topographic cardiac image. Left ventricle ejection fraction was calculated using the Simpson method in all cases. Late enhancement was evaluated as “present” or “absent”.

Results

Five patients (2 males and 3 females) were included in the study, with ages ranging from one month to 5 years (mean 29.4 months). The MR diagnosis agreed with the ultrasound diagnosis. All patients presented with prominent trabeculations and deep intertrabecular recesses in the left ventricle wall. The morphological and functional findings are summarized in table 1.

Both the black-blood and B-FFE sequences showed a left ventricular wall with deep trabeculations in the areas identified as LVNC (fig. 1). Cardiac function was abnormal in three patients, with the ejection fraction ranging from 18-53% (mean 39%). One of the patients (Case 3) (fig. 2) exhibited clear functional alteration and had a seriously affected, dilated lateral wall of the left ventricle (LV). One patient exhibited mild mitral insufficiency, one had moderate mitral insufficiency and another had moderate mitral insufficiency and stenosis. One patient had coarctation...
of the aorta with a bicuspid aortic valve. Segments 10, 11, 12, 15, 16 and 17 (the apex, inferior apex and posterior, inferomedial and inferolateral walls) were affected in all patients. Diffuse disease was found in only one patient. Two patients presented a septal impairment (including the patient with diffuse disease). Late enhancement was not detected in any case.

**Discussion**

LVNC is a rare congenital cardiopathy that was reported for the first time by Chin et al. in 1990. The anatomic characteristic of this myocardial disease is the presence of deep trabeculations in the left ventricular wall that communicate with the ventricular cavity. It is among the “unclassified cardiomyopathies” in the International Classification of Diseases from the World Health Organization. Terms such as “spongiform cardiomyopathy” and “hypertrabeculation of the left ventricle” are no longer used. Recently, the condition was included as a genetic cardiomyopathy in the American Heart Association’s classification.

Prominent trabeculation is a normal finding in the right ventricle. It is believed that LVNC results from an inappropriate cessation of endomyocardial morphogenesis, which normally occurs between the 5th and 8th week of fetal life and is characterized by gradual compaction of the myocardium and development of the trabecular spaces into capillaries and the coronary circulation. LVNC usually affects the left ventricle but can extend to the right ventricular wall in 20% of cases. Although it was first described in the pediatric population, most of the subsequently reported patients were adults. LVNC seems to be a genetically heterogeneous disease; for instance, a familial variant of the disease is associated with craniofacial anomalies and Wolf-Parkinson-White syndrome.

The clinical presentation is similar to that of other cardiomyopathies and includes systolic and diastolic left ventricular dysfunction, arterial embolism and tachyarrhythmia. Some patients have been described as having a remitting-relapsing clinical course with transitory deteriorations and improvements. Left ventricular dysfunction is seen 10 years after diagnosis in 90% of patients, and mortality as high as 80% has been described at 6 years after dysfunction is diagnosed.

LVNC is frequently under-diagnosed, or misdiagnosed as hypertrophic or dilated cardiomyopathy. It is probably a more common disease than formerly believed. The diagnosis criteria are based on ultrasound findings. Jenni et al. defined four diagnostic criteria: 1) absence of coexistent cardiopathy; 2) multiple trabeculations and deep intertrabecular recesses; 3) communication between the recesses and the ventricular cavity as demonstrated by a Doppler exam; and 4) a non-compacted layer to compacted layer thickness ratio higher than 2 during systole. Pignatelli et al. recommended a ratio of 1.4 for pediatric patients and did not require the absence of other cardiopathy as a requisite for LVNC diagnosis. All our five patients exhibited ratios higher than 2. One patient with complex congenital cardiopathy was excluded due to difficulty in interpreting
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The most frequently described locations for LVNC are at the apex and medioventricular regions, which is in agreement with our series; however, LVNC has also been described in both ventricles and in the interventricular septum.

MR is a complementary diagnostic technique that correlates well with ultrasound findings. As it is accurate assessing cardiac function, and provides a better identification of certain cardiac segments, MR is a useful tool in the diagnostic work-up of these patients. The non-compacted to compacted layer ratio threshold (higher than 2) has been reported using MR data. Computed tomography has also been used in pediatric patients, but the use of ionizing radiation, lower performance as a functional study, and lower capacity for characterizing myocardial tissue and detecting fibrosis make this technique less suitable for the non-compacted cardiomyopathy evaluation. Assessment of subendocardial fibrosis using late enhancement on contrast-enhanced MR and its correlation with pathological findings have been described in the adult population. Dodd et al. found late enhancement in segments that did not meet LVNC criteria, suggesting that the use of morphological criteria alone may result in underestimation of disease extent. In a recent study, Calvillo et al. only saw late enhancement in one of their patients who also presented a diffuse coronary disease during cardiac angiography. Junga et al. found a correlation between positron emission tomography perfusion data and non-compacted areas seen on MR images in children between 10 to 15 years old, but in this study, late enhancement by MR was not assessed. As far as we know, only one case of late enhancement in a pediatric patient has been published; this study showed extensive subendocardial uptake in the hypertrabeculated

Figure 1 Case no. 4. A five-year-old girl with apical, basal and lateral involvement (arrows). A) Balanced fast field echo [B-FFE]) in four-chamber view. B) B-FFE sequence in three-chamber view. C) Gradient echo sequences with inversion recovery and late enhancement in the short-axis view. Contrast is seen within the ventricles, but myocardium enhancement is not observed.

Figure 2 Case no. 3. A 37-month-old boy with apical, basal and lateral involvement. A) A turbo spin echo sequence in the sagittal plane. B) A gradient echo sequence with inversion recovery and late enhancement shows severe involvement with deep recesses in the lateral wall (arrows) with cardiac dilatation and no evidence of late enhancement.
segments in a 12-year-old child. In our series, we did not find significant enhancement in any of our cases. The oldest patient was a 5-year-old girl. These data might suggest that late enhancement and subendocardial fibrosis may represent evolving sequelae of the non-compacted ventricular wall, leading to dysfunction and perfusion impairment. That could be the reason explaining why late enhancement and fibrosis have not showed up in very young children.

This study presents several limitations. Because this is a rare disease, the series is small (five patients) and retrospective. We have no radiological-pathological correlation. Moreover, the late-enhancement sequences are limited in younger children due to the small size of the heart and lack of patient cooperation (keeping still, performing breath-holds, etc.). The optimal time during the heart and lack of patient cooperation (keeping still, performing breath-holds, etc.). The optimal time during the sequence for assessment of late enhancement is controversial, and when performed at any time between 5 and 30 minutes after the injection, there was no significant change in results. We waited 15 min to avoid excessive concentrations of contrast in the ventricular cavity. Nevertheless, to go in depth into this topic, we refer the reader to other publications. We believe that to confirm the presence or absence of late enhancement and fibrosis in infants and young children with LVNC, larger series correlating findings on radiology and pathology are essential. New studies in the pediatric population are needed to establish the exact role for late enhancement in LVNC.

Conflict of interest

The authors declare there no conflict of interest exists.

Authors

Carlos Marín Rodríguez: study concept and design, writing and revision.
Silvia Ossaba Vélez: study concept and design, writing and revision.
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