

Noncompaction of the Myocardium, Chagas' Disease and Dysfunction - A Case Report

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We report the association between heart disease associated with noncompaction of the left ventricular myocardium (NCLVM) and chronic Chagas' heart disease (CCHD) in a patient with heart failure, ischemic stroke and cardiac arrhythmia. Images typical of NCLVM and CCHD were documented by cardiac magnetic resonance imaging (CMRI).

Introduction

Persistence of noncompaction of the left ventricular myocardium (NCLVM) and chronic Chagas' heart disease (CCHD) are described as causes of ventricular dysfunction. NCLVM is characterized by persistence of trabeculations, which are deep intertrabecular recesses associated with the presence of predominantly endocardial fibrosis^{1,2}. On the other hand, CCHD is related to a long disease course, ventricular dysfunction and fibrosis in different stages (endocardial, mesocardial, epicardial or transmural fibrosis) in the basal region or left ventricular (LV) apex³. We report the association between these two causes of heart disease in a patient with ventricular dysfunction.

Case report

AJF, a 65-year-old Caucasian male was admitted with complaints of palpitations, leg edema, exercise intolerance and progressive dyspnea for the past six months. He reported an acute ischemic stroke confirmed by computed tomography. His family history was negative for cardiovascular diseases. Previous Chagas' Disease (CD) infection was confirmed by serologic tests. Physical examination showed blood pressure of 100/60 mmHg, irregular pulse of 70 bpm, and a grade III/VI mitral regurgitation murmur. Electrocardiogram showed typical atrial flutter with controlled ventricular rate, normal

QRS duration, and no ventricular hypertrophy. Chest radiography revealed enlarged cardiac silhouette. In order to evaluate the severity of the ventricular dysfunction, the presence of myocardial fibrosis and mural thrombi caused by CCHD, cardiac magnetic resonance imaging (CMRI) was performed. Dynamic imaging (cine) by the use of the steady state free precession (SSFP) sequence showed akinesia in LV apex. Ejection fraction was estimated at 40%. There was evidence of images of apical transmural delayed enhancement and visible trabeculations at a ratio of 6:1 of the normal myocardial thickness at diastole, predominately in lateral wall and consistent with the diagnoses of CCHD and NCLVM, respectively. (Figure 1). Electrophysiological study for ablation treatment of the atrial flutter was successfully performed. No complex ventricular arrhythmias were induced at programmed ventricular stimulation. Sinus node recovery time was 4.4 seconds, consistent with sinus node dysfunction (SND) (normal up to 1.5 s). HV interval was 60 ms (35 to 55 ms). Intracardiac two-dimensional echocardiography was used to guide the positioning of catheters in the valve annulus and cavotricuspid isthmus; it recorded extensive NCLVM in the lateral wall. Doppler identified flow across the trabeculations (Figure 2). Coronary angiography performed to rule out any critical lesion subject to reperfusion that could justify the ventricular dysfunction showed no coronary artery disease. Dual-chamber pacemaker implantation was chosen for the treatment of the sinus node dysfunction. During the clinical follow-up, the symptoms of heart failure significantly improved.

Discussion

This case report is the first description of the association between NCLVM and heart disease induced by CD. NCLVM was demonstrated by the presence of multiple deep trabecular recesses in the endocardial side of the left ventricular cavity, whereas CCHD was characterized by serologic diagnosis and presence of segmental akinesia associated with transmural delayed enhancement in LV apex.

NCLVM is a morphogenetic disorder of the endocardium with a high mortality rate. This disorder represents an interruption in the normal process of myocardial compaction during the 5th to 8th week of development and results in the persistence of multiple prominent ventricular trabeculations. Association of mutations in gene G 4.5 (in 6% of the cases), CSX, FKBP12, Peg1, muscular and mitochondrial abnormalities have been reported⁴ before. Typically, NCLVM involves the LV; however, the right ventricle may also be affected. Predominance of the male gender (55.6%) and family occurrence may be identified^{5,6}. The clinical presentation is

Key words

Noncompaction of the ventricular myocardium; Chagas' Disease; ventricular dysfunction left.

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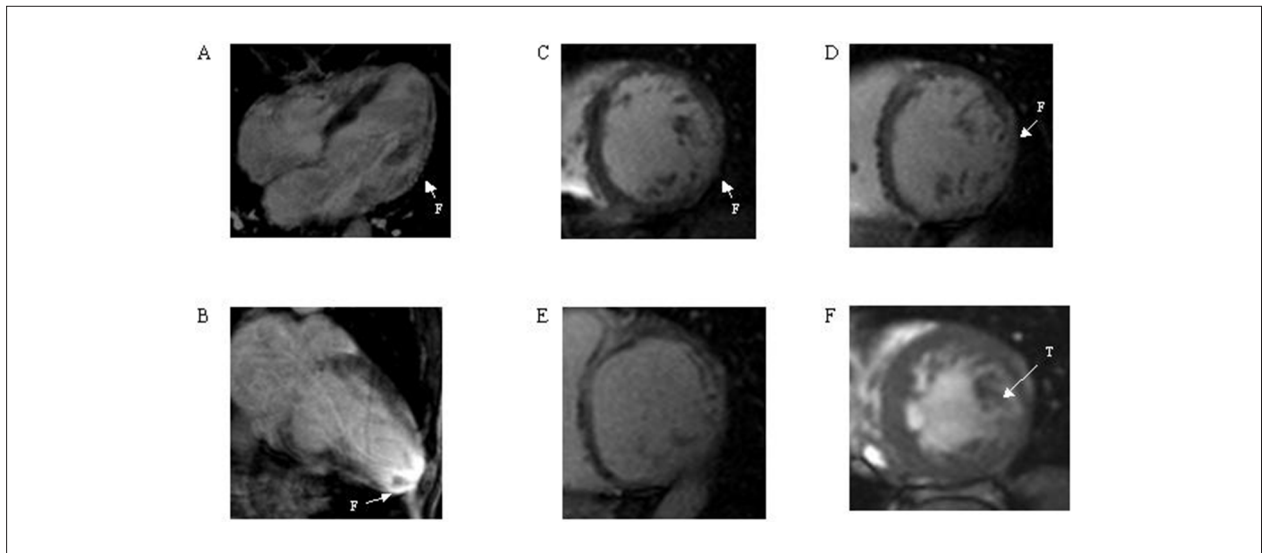


Figure 1 - Cardiac magnetic resonance imaging showing excessive trabeculations (T) consistent with NCLVM, and transmural fibrosis (F) in apical and posterior lateral regions consistent with CCHD. A. Delayed enhancement technique in the 4-chamber view showing transmural delayed enhancement in the apex and in a large portion of the LV lateral wall. Mesocardial enhancement is also identified in the septum. B. Delayed enhancement technique in the 2-chamber view showing transmural delayed enhancement in LV apex. C, D, and E. Delayed enhancement technique in the LV short axis. The three images in this view were obtained by the use of a single shot sequence which, despite providing lower spatial resolution, is very useful to minimize the artifacts related to the difficulty in synchronization due to severe arrhythmia. F. Extensive trabeculation in the LV lateral wall is observed in this image by the use of the SSFP technique.

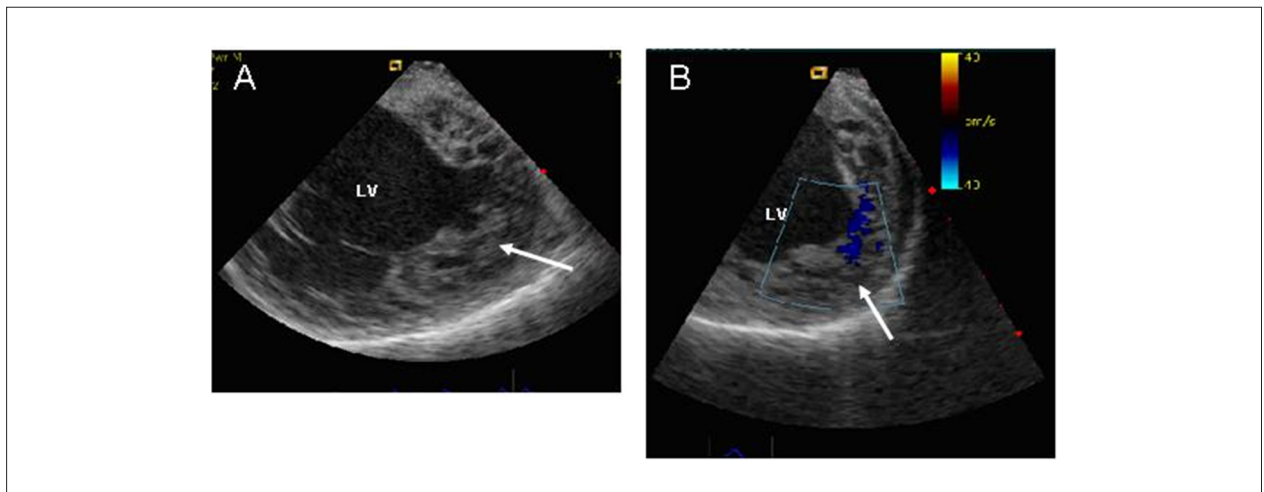


Figure 2 - Image acquired by two-dimensional echocardiography of the LV with intracardiac echocardiography positioned in the right ventricle via a femoral vein approach showing a focal area of NCLVM. (A) Multiple deep intertrabecular recesses of noncompacted myocardium communicating with the ventricular cavity may be identified (arrow). (B) Doppler image showing blood flow (arrow) across those recesses. LV - left ventricle.

variable, ranging from asymptomatic patients to symptoms related to left ventricular systolic or diastolic dysfunction. Embolic events; fatal ventricular arrhythmias; sudden cardiac death; ventricular septal defect; pulmonary valve stenosis or right ventricular hypoplasia; hypoplastic left ventricle; Di George syndrome; hypertrophic heart disease; and congenital adrenal hyperplasia have been reported⁶before. Supraventricular arrhythmia, atrial fibrillation and heart blocks have also been associated with the disease.

Echocardiography is the method of choice for the diagnosis of NCLVM because of easiness to perform, but

other techniques such as CMRI, computed tomography and ventricular angiography may also be useful^{1,4-6}. The echocardiographic diagnostic criteria include the presence of multiple trabeculations associated with multiple deep intertrabecular recesses communicating with the ventricular cavity, and a greater than 2:1 ratio between noncompacted/compacted myocardium in adults (or > 1.4 in the pediatric population)⁶. On CMRI, this ratio should be > 2.3, as measured during diastole, and may be used in cases in which an echocardiographic image is difficult to obtain⁷.

These changes may be found in the LV alone in 78% of

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the patients, and in both ventricles in 22%, but involvement of the right ventricle alone is not likely to happen. LV systolic dysfunction may be found in 83% of the individuals; however, diastolic dysfunction may also be present^{1,6}, as well as thrombus and ventricular hypertrophy. Recent studies have helped characterize the distribution of myocardial fibrosis in NCLVM. These studies demonstrated that three out of 10 patients did not show fibrosis, whereas in the other 7 fibrosis predominated in the endocardial region; only one patient presented with transmural fibrosis in the left ventricular midlateral region. None of them presented with akinesia or apical transmural fibrosis².

Electrocardiographic abnormalities are common in this disease⁶. Treatment of NCLVM depends basically on the presence of complications, on the comorbidities and other associated symptoms. The disease progresses to heart failure, acute embolic events, ventricular tachycardia and death^{1,5}.

CCHD, in turn, is a very important infectious disease in South America and produces contractility changes, fibrosis and abnormal cardiac conduction. The development of ventricular dysfunction, bradyarrhythmias, ventricular tachycardia and sudden cardiac death are important characteristics of the disease. In the typical presentation, systolic ventricular dysfunction is manifested after a long asymptomatic period. The disease may be suspected by ECG because of the typical changes related to infrahisian conduction abnormalities: right bundle branch block, left anterior superior division block.

Sinus node dysfunction is a common presentation of

this disease even in asymptomatic patients in whom ECG abnormalities are frequent^{8,9}.

Characteristically, when assessed by CMRI through the use of the delayed enhancement technique, these patients present with fibrosis in the posterior basal region and LV apex³. The fibrosis process in CCHD is different from that produced by NCLVM.

In the present case, we found myocardial noncompaction characterized by trabeculations and recesses in the LV endocardial wall^{1,10} and also identified transmural delayed enhancement in LV apex associated with akinesia, typical of CCHD³. CMRI in CCHD has the advantage of identifying the extent of hypokinesia and fibrosis typical of the disease, in addition to being a useful method for the identification of other anatomical abnormalities such as NCLVM, and for ruling out thrombi in patients with a difficult acoustic window.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any post-graduation program.

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