The Value of Cardiac Magnetic Resonance Imaging in the Diagnosis of Isolated Non-Compaction of the Left Ventricle

Offer Amir MD[1], Reynolds M. Delgado III MD FACC[2], Biswajit Kar MD FACC[1] and Frank W. Smart MD FACC[3]

[1]Department of Cardiology, Lady Davis Carmel Medical Center, Haifa, Israel
[2]Department of Cardiology, Texas Heart Institute, St. Luke's Episcopal Hospital, Houston, TX and [3]Department of Cardiology, Morristown Memorial Hospital, NJ, USA

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Non-compaction is a rare etiology of heart failure that occurs as a result of the arrest of the normal compaction of the myocardium mesh-work during embryonic life. In rare situations it is an isolated abnormality of the left ventricle, termed "isolated non-compaction" [1]. The prevalence of INC was estimated to be 0.05% of all patients who were referred for an echocardiogram in a large medical center during a 10 year period. Familial cluster occurred in 12–50% of the cases and association with other congenital cardiomyopathies and syndromes was described [2]. Echocardiogram is the most common tool used for the diagnosis of INC. However, in the case series presented below of four patients who presented with different clinical symptoms of INC in a tertiary hospital, echocardiogram missed the diagnosis of INC in all four.

PATIENT DESCRIPTIONS

PATIENT 1

A 28 year old man was referred to the heart failure clinic for evaluation of exertional dyspnea and paroxysmal nocturnal dyspnea of several months duration. On clinical examination he had overt fluid overload. A complete workup was done, including electrocardiogram which revealed sinus rhythm with intraventricular conduction defect, coronary angiogram that showed normal epicardial coronaries, and echocardiography that demonstrated an enlarged heart with diffuse hypokinesis of both ventricles. The patient was diagnosed with "idiopathic dilated cardiomyopathy." Later on, cardiac magnetic resonance imaging was performed, demonstrating excessive trabeculations in the apex and lateral wall that was diagnostic of INC.

INC = isolated non-compaction

PATIENT 2

A 22 year old female patient was admitted for an electrophysiological study. The patient carried a diagnosis of dilated cardiomyopathy secondary to "viral myocarditis" since the age of 3 months. She was functioning fairly well with no heart failure symptoms during daily activities. She had chronic atrial fibrillation. Her coronary angiogram was normal. Repeated cardiac echocardiograms revealed diffuse hypokinesis and dilated heart. During her electrophysiological study, the patient had inducible sustained monomorphic ventricular tachycardia, which was converted with difficulty to sinus rhythm. She eventually underwent a heart transplant. Prior to her heart transplantation the patient had a cardiac MRI that showed significant trabeculations in the apex and lateral wall of the left ventricle, compatible with INC of the left ventricle.

PATIENT 3

A 31 year old man was referred to the heart failure clinic for evaluation of recurrent episodes of supraventricular tachycardia. Two years previously he was diagnosed with "idiopathic dilated cardiomyopathy." The patient had no other medical issues and no significant medical family history. His electrocardiogram showed sinus rhythm and abnormal repolarization changes in the chest leads. His cardiac echocardiogram revealed diffuse hypokinesis and enlarged heart. Coronary angiogram was normal and MRI of the heart demonstrated thick coarse trabeculations in the apex and the lateral wall, and a diagnosis of INC of the left ventricle was made.

PATIENT 4

A 42 year old man was admitted with numbness of his right arm and leg that resolved after several hours. He had been diagnosed with mild hypertension several years earlier, and blood pressure was effectively controlled with medications. The patient had two prior episodes of syncope. Despite this, he engaged in daily physical activity and denied any complaints of dyspnea or chest discomfort. His family history was unrevealing. No volume overload was found on physical examination. As part of the workup of an acute neurological event, an echocardiogram was done, demonstrating severe systolic dysfunction with bi-ventricular enlargement. A single non-obstructing lesion in the right coronary artery was found

on coronary angiogram and a diagnosis of "idiopathic dilated cardiomyopathy" was made. Later, cardiac MRI demonstrated findings compatible with INC.

COMMENT

As our case series shows, although INC is rare it is an important entity to diagnose. In general, patients with INC have a poor prognosis [2]. Neurological manifestation, ventricular tachycardia and sudden cardiac deaths are common. Consequently, INC patients often require more aggressive and more specific therapy, such as anti-arrhythmic devices. Moreover, correct diagnosis of INC is also crucial for the patient's relatives as it may represent a familial form of the disease [3].

Our patient series represents the clinical spectrum of INC: overt heart failure, atrial and ventricular arrhythmias, acute neurological events, and abnormal electrocardiograms. Still, in all the patients, the echocardiograms, performed in tertiary hospital centers, failed to detect the trabeculations and a diagnosis of INC was missed with an incorrect diagnosis of "idiopathic dilated cardiomyopathy" (patients 1, 3 and 4) and "viral myocarditis" (patient 2).

The echocardiogram is considered to be the diagnostic test of choice for non-compaction [1]. However, echocardiography is limited and has its pitfalls in identifying trabeculations as INC and differentiating it from apical hypertrophic cardiomyopathy, ventricular thrombus, false tendon, arrhythmogenic right ventricular cardiomyopathy, prominent normal ventricular trabeculations and double-chambered right ventricle [1].

A cardiac MRI can produce high quality cardiac images in any plane without limitations of acoustic windows and therefore may correctly diagnose patients with INC who could otherwise be missed by the echocardiogram. Moreover, the MRI can measure the non-compacted/compacted myocardium thickness ratio. Using MRI, a ratio > 2.3 in end-diastole has been proposed to diagnose INC, yielding a sensitivity of 86% and a specificity of 99% [1,4,5].

In summary, we believe that once clinical suspicion for INC arises, additional means of imaging such as contrast ventriculography, ultrafast computed tomography and mainly cardiac MRI are often needed.

Correspondence:
Dr. O. Amir
Dept. of Cardiology, Lady Davis Carmel Medical Center, 7 Michal St., Haifa 34674, Israel
Phone: (972-4) 8250-285
Fax: (972-4) 862-5056
email: offeram@012.net.il

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