Open Access Journal

International Journal of Medical Research and Pharmaceutical Sciences

December 2014, Volume 1(Issue 1) ISSN: XXX-XXX

ISOLATED LEFT VENTRICULAR NON-COMPACTION: CARDIAC CT FINDINGS

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Keywords:
Noncompaction, left ventricul, cardiac, CT.

Abstract

Ventricular non-compaction also known as spongiform cardiomyopathy, is an uncommon congenital disorder resulting from abnormal embryogenesis of myocardium and endocardium. In this paper, we report cardiac CT findings in the patient of a 15-year-old male with isolated left ventricular noncompaction mimicking mass.

INTRODUCTION

Noncompaction of the ventricular myocardium is a recently recognized genetic cardiomyopathy thought to be caused by arrest of normal embryogenesis of the endocardium and myocardium. It is a rare congenital anomaly and the incidence is reported to be 0.05%. Echocardiography (ECHO-CG) is the diagnostic procedure of choice, but the correct diagnosis is often missed or delayed (1). Negative ECHO-CG and negative heart enzymes is considered a major indication for cardiac-computed tomography (CT) (1,2). The ECHO-CG findings of this disorder have been reported in small series but computed tomography (CT) and magnetic rezonance imaging (MRI) findings have so far been less frequently reported. We describe cardiac CT findings of a patient whom the characteristic myocardial changes of the isolated left ventricular non-compaction (LVNC).

CASE REPORT

A 15-year-old male with history of atypical chest pain was referred to our hospital. Heart enzymes and previously performed ECHO-CG was normal. The patient had no history of coronary artery or congenital heart disease. Also, he has no family history of heart disease or sudden death. He complained of dyspnea during effort that get worse during last year. On admission, his blood pressure was 110/70 mmHg and the heart rate was 70 beats/minute. To rule out coronary artery anomalies and other conditions, cardiac-CT was performed with a 256-slice multidetector CT system (Somatom Flash, Siemens Healthcare, Germany) with prospective electrocardiography (ECG)-gating after bolus-triggered infusion of 40 ml contrast agent (Ultravist; 300 mgI/ml, Bayer, Berlin, Germany). In the contrast enhanced CT study, the coronary arteries were normal, showing no plaques and no stenosis. The left and right atrium and the right ventricle were normal. During diastole, the left ventricle showed deep intramyocardial recesses and prominent trabeculae in continuation with the ventricular lumen in the mid and apical thirds of the interventricular septum. Prominent trabeculations were also found in the region of the lateral and posterior wall **(Figure 1).**

DISCUSSION

Isolated LVNC is a rare disorder of endomyocardial morphogenesis characterized by numerous, excessively prominent ventricular trabeculations and deep intertrabecular recesses. It thought to be caused by arrest of normal embryogenesis of the endocardium and myocardium (1). Noncompaction of the ventricular myocardium is a recently recognized genetic cardiomyopathy (2). The non-compaction cardiomyopathy falls into the group of non-classified cardiomyopathies. Both familial and sporadic forms of isolated LVNC have been described (3). The left ventricle is uniformly affected, but biventricular noncompaction has been reported, with right ventricular noncompaction described in less than one-half of patients (4). In at least one study, approximately 12 % of patients had associated cardiac malformations (5). Sometimes it is associated with other congenital cardiac defects (eg. complex cyanotic heart disease, coronary artery anomalies, obstruction of the right or left ventricular outflow tracts) (5,6).

Clinical manifestations may range from being asymptomatic to presenting with dyspnea, severe systolic dysfunction, malignant arrhythmias, sudden cardiac death or systemic thromboembolism secondary to atrial fibrillation. However, patients with only mild LVNC may be completely asymptomatic (4,7). Traditionally, ECHO-CG has been used to establish a diagnosis of myocardial non-compaction, although CT and MRI provide better visualization of the trabeculations. Also CT enabled quantitative and qualitative assessment of global and regional ventricular function from the same dataset in addition to the morphological assessment for coronary artery disease and general cardiac morphology. Although cardiac MRI is currently considered as the reference standard for morphologic and functional assessment of the heart its use for coronary artery evaluation is limited (1). Symptoms of angina pectoris with negative heart enzymes and negative ECG is considered a major indication for cardiac-CT. As LVNC may have an important impact on morbidity and mortality, early and reliable diagnosis is crucial. The features of LVNC are found predominantly in the apical and the mid ventricular segments of the left ventricle, as confirmed in a larger series (7).

In conclusion, LVNC is a rare cardiomyopathy, but that should always be considered as a possible diagnosis because of its potential complications. ECHO-CG is the standard tool for diagnosis, and cardiac-CT are very useful to confirm or rule out this disease. Further, cardiac-CT imaging allows the simultaneous evaluation of complex intracardiac pathology and ventricular function as well as for coronary artery morphology. The knowledge of imaging findings the ventricular noncompaction will provide convenience in terms of early and differential diagnosis in situtions mimicking mass.

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Figure Legends

Figure 1: Axial CT images demonstrate the prominent left ventricular trabeculations with deep intertrabecular recesses (arrows).