Combined Finding of Left Ventricular Non-Compaction and Dilated Cardiomyopathy

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Background

Left ventricular non-compaction (LVNC) or spongy left ventricular (LV) myocardium is a disorder of endomyocardial morphogenesis that results in multiple trabeculations in the LV myocardium [1]. Numerous, excessively prominent LV trabeculae and deep inter-trabecular recesses together create a spongiform appearance. According to current literature, LVNC in adults is rare [1], although associated with a poor prognosis [1]. Several studies have reported on highly symptomatic cases with a high incidence of ventricular arrhythmia [1] and progressive heart failure [2].

Recent evidences suggest an overlap between LVNC and idiopathic dilated cardiomyopathy (IDC) [2] and apical hypertrophic cardiomyopathy [2].

Case Report

We report the case of a 30-year-old man, referred for dyspnoea since a couple of years. He was a smoker, obese, without history of heart disease; he was receiving drug therapy with bisoprolol, enalapril, simvastatin and diuretics.

Arterial blood pressure at admission was 100/80 mmHg, and physical examination was unremarkable. Resting ECG showed sinus rhythm (66 bpm) and diffuse negative T waves (Figure 1). Chest radiography showed no sign of pulmonary congestion, with an enlarged cardiac transverse diameter. Cholesterol, troponin and creatinine levels were normal.

Echocardiography showed a severe systolic dysfunction (25% LV ejection fraction). Coronary angiography was not able to show any sign of significant coronary atherosclerosis, with no need of coronary angioplasty. At LV angiography, enlarged diameters and severe hypokinesis were detectable; diffuse irregular trabecles were also detectable in LV angiogram, mimicking features typical of spongy LV myocardium (Figure 2).

New York Heart Association class was I-II at discharge; 6-month follow up was uneventful.

Discussion

To our knowledge, this is one of the first cases reporting LV angiography images showing the concomitant finding of IDC and LV spongy myocardium in a young man. LVNC is believed to be the

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result of an arrest in endocardial morphogenesis [2]. Severity ranges from moderately abnormal ventricular trabeculations in association with normal LV function to profoundly abnormal, loosely compacted trabeculations in association with poor LV function [1]. Clinical manifestations of LVNC are symptoms associated with depressed LV systolic function, ventricular arrhythmias, and systemic embolization [2].

In a symptomatic patient, however, differentiation of LVNC from other cardiomyopathies may be difficult. Several cases of IDC have been shown to be LVNC at autopsy [2]. Because the natural histories of the two conditions differ, it is important to differentiate them. However, with a persisting disease state, spongy left ventricle has a tendency to dilate and becomes more spherical [2]. This structural change does not have a specific cause and occurs terminally in any form of chronic myocardial dysfunction.

Misdiagnosis due to technical difficulties may have led to a significant underestimation of LVNC prevalence in the heart failure population [6], this in keeping with recent reviews which noted an increased incidence of LVNC with improving cardiac imaging [2].

Recent evidences suggest an overlap between LVNC and apical hypertrophic cardiomyopathy [7] or IDC [6] as reported in our patient. The presence of IDC without LVNC in relatives of LVNC patients has shown that the phenotype of familial LVNC can overlap with that seen in familial IDC [6].

These data seem to support the hypothesis of a clinical and genetic overlap between LVNC and other cardiomyopathies [6]. However, this issue needs further investigations.

Limitations

This is a case report with first diagnostic suspicion of LVNC initially based on angiogram imaging; definitive diagnosis needs to be confirmed by echo-Doppler imaging.

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