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Case Report

Clinical usefulness of gated technetium-99m sestamibi myocardial perfusion single-photon emission computed tomography with phase analysis for the management of patients with isolated ventricular noncompaction

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ABSTRACT

Gated Tc-99m sestamibi myocardial perfusion single-photon emission computed tomography (GMPS) with phase analysis provides information on myocardial perfusion, left ventricular (LV) function, and LV dyssynchrony. We present a case of isolated left ventricular noncompaction (IVNC) cardiomyopathy in which GMPS with phase analysis proved to be beneficial and reliable to monitor the long-term response to cardiac resynchronization therapy with defibrillator (CRT-D). The patient was an 84-year-old man with shortness of breath on minimal exertion (New York Heart Association class III) who had severe drug-refractory heart failure with hypotension and ventricular tachycardia. He was diagnosed with IVNC using echocardiography. At baseline, GMPS with phase analysis revealed a reduced ejection fraction (EF, 21%), large perfusion defects in the inferior and inferolateral walls, and severe LV dyssynchrony [histogram bandwidth (HBW) 120°]. Combination therapy with CRT-D and a titrated beta-blocker was initiated to induce LV reverse remodeling and reduce LV dyssynchrony. Two years after CRT-D implantation, GMPS with phase analysis showed marked improvement in LV function and LV dyssynchrony (EF 28%, HBW 36°). This case demonstrates that GMPS with phase analysis is an important and useful modality to evaluate LV function and LV dyssynchrony in IVNC patients undergoing CRT-D. <**Learning objective:** We experienced a rare case of heart failure with isolated left ventricular noncompaction (IVNC) treated with cardiac resynchronization therapy with defibrillator (CRT-D). We demonstrate that gated Tc-99m myocardial perfusion SPECT (GMPS) with phase analysis can simultaneously evaluate myocardial perfusion and left ventricular dyssynchrony to assess the indication and efficacy of CRT-D. This case demonstrates that GMPS with phase analysis is useful to monitor IVNC patients before and after CRT-D.>

Introduction

Isolated left ventricular noncompaction (IVNC) is a rare congenital genetic cardiomyopathy characterized by prominent trabeculations and deep intertrabecular recesses [1]. In patients with IVNC, left ventricular (LV) dyssynchrony is detected between the non-compacted and compacted myocardial layers by echocardiography. Several case reports used echocardiography to demonstrate that cardiac resynchronization therapy (CRT) with defibrillator (CRT-D) causes substantial improvements in clinical symptoms, functional outcome, and LV performance in IVNC patients [2]. However, echocardiography could not demonstrate the advantage in selecting appropriate candidates for CRT/CRT-D beyond current guidelines [3]. On the other hand, gated Tc-99m

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myocardial perfusion single-photon emission computed tomography (GMPS) with phase analysis can simultaneously evaluate myocardial perfusion, LV function, and LV dyssynchrony [4]. It has been demonstrated that phase analysis with GMPS may be superior to the echocardiographic techniques currently available in assessing LV dyssynchrony because of its high repeatability and reproducibility for follow-up protocols [5]. However, in IVNC patients, the potential of GMPS with phase analysis to monitor cardiac alterations before and after CRT/CRT-D has not been fully investigated. Herein, we report the case of a patient with IVNC in whom serial GMPS with phase analysis was performed to assess LV reverse remodeling and LV dyssynchrony over two years before and after CRT-D implantation.

Case report

An 84-year-old man, who had been diagnosed with chronic heart failure (HF), was admitted to our hospital after an exacerbation of congestive HF. He presented with shortness of breath on minimal exertion [New York Heart Association (NYHA) class III] and weight loss since one month. The initial chest X-ray revealed cardiomegaly [cardiothoracic ratio (CTR) of 66.1%, Fig. 1A] with bilateral pleural effusion. The 12-lead electrocardiogram (ECG) showed LV hypertrophy and non-specific intraventricular conduction disturbance with QRS duration of 128 ms (Fig. 1B). Two-dimensional echocardiography revealed reduced ejection fraction (EF, 22%), LV dilatation, and diffuse hypokinesis mimicking dilated cardiomyopathy. Additionally, multiple trabeculations and deep intertrabecular recesses were prominent in the inferior and inferolateral walls, with a thickness ratio of noncompaction over compaction layers (N/C ratio) > 2.0 (Fig. 1C, D) [1]. Coronary angiography was normal, whereas endomyocardial biopsy obtained from the inferior LV wall showed moderate myocyte hypertrophy with mild interstitial fibrosis.

During the acute and sub-acute stages, the standard therapy with a beta-blocker (carvedilol 2.5 mg/day) and angiotensin-converting enzyme inhibitors was limited by symptomatic hypotension and adverse effects. Moreover, ventricular tachycardia and paroxysmal atrial fibrillation were documented by 24-h Holter monitoring. Therefore, CRT-D was considered clinically indicated, based on the current guidelines [6]. Recently it has been reported that myocardial viability is essential for CRT/CRT-D pacing site [7] and can also be evaluated by GMPS [8]. Therefore, GMPS with phase analysis using the quantitative gated single-photon emission computed tomography (SPECT) (QGS) software was performed to assess the LV dyssynchrony and determine the appropriate pacing site. A dose of approximately 740 MBq of Tc-99m sestamibi was administered intravenously with the patient at rest. Resting SPECT image acquisition was initiated 60 min thereafter. GMPS with phase analysis showed severe perfusion defects in the inferior and inferolateral walls (Fig. 2A), increased ventricular volume with reduced systolic function [end-systolic volume (ESV) 155 ml, EF 21%], and marked LV dyssynchrony with histogram bandwidth (HBW) of 120° (Fig. 2B). On M-mode echocardiography, the septal-to-posterior wall motion delay (SPWMD) was 105 ms. He successfully received a CRT-D device

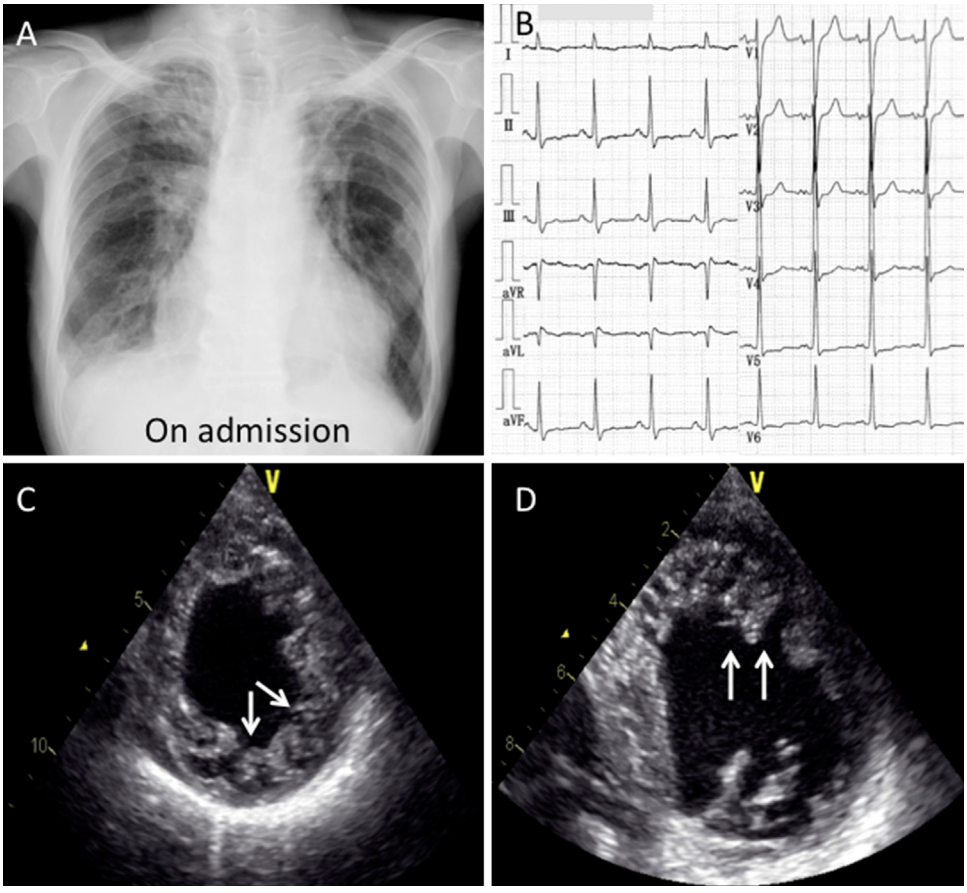

Fig. 1. At the time of admission, chest X-ray showed cardiomegaly with a cardiothoracic ratio (CTR) of 66.1% and lung congestion (A). The 12-lead electrocardiogram revealed a wide QRS duration of 128 ms (B). Two-dimensional echocardiography on the short-axis (C) and apical four-chamber (D) views showed prominent trabeculations and deep recesses in the inferior and inferolateral walls (white arrow). The ratio of noncompacted (N) over compacted (C) layers (N/C > 2.0) was calculated at the end-systole.