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CASE REPORT

Cardiomyopathy mimicking left ventricular noncompaction in a patient with lupus nephritis

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Abstract A 37-year-old female patient was admitted with exertional dyspnea. Her serum creatinine was 2.4 mg/dL and anti-nuclear antibody was positive in a titer of 1/320. Renal biopsy revealed diffuse proliferative lupus nephritis. Echocardiography and cardiac magnetic resonance (MR) imaging showed increased apical trabeculations compatible with left ventricular noncompaction (LVNC), which is a rare genetic cardiomyopathy. The patient expressed a marked improvement in exertional dyspnea after the immune-suppressive treatment for systemic lupus erythematosus (SLE). Control echocardiography revealed a significant increase of ejection fraction. SLE may cause a kind of cardiomyopathy with high resemblance to LVNC. Discrimination of these two similar clinical entities is important because SLE-induced cardiomyopathy is potentially reversible after the immune-suppressive treatment for SLE.

Keywords Left ventricular noncompaction · Systemic lupus erythematosus · Cardiomyopathy · Lupus nephritis

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Introduction

Left ventricular noncompaction (LVNC) is a rare genetic cardiomyopathy. It is thought to be due to the failure of condensation of the myocardial meshwork of fibers during intrauterine life, leading to persistence of ventricular trabeculations [1, 2]. The major clinical manifestations of LVNC are heart failure, systemic thromboembolism, ventricular arrhythmias, conduction disorders, and neurologic abnormalities. LVNC may be associated with several disorders, especially with neuromuscular disorders [3, 4]. However, LVNC associated with systemic lupus erythematosus (SLE) has not been described previously. Herein, we present the first report of SLE-induced cardiomyopathy with high resemblance to LVNC.

Case presentation

A 37-year-old female patient was admitted with exertional dyspnea. Her baseline biochemical values are presented in Table 1. Hair loss, photosensitivity, and oral aphthous ulcers were present. Upon physical examination including the neuromuscular system, no other pathological finding was found. When the family history was investigated, no specific disease or syndrome was found in the family

Table 1 Results of the initial laboratory tests

	Values	Reference values
Creatinine	2.4 mg/dL	0.7–1.4
Calcium	8.3 mg/dL	8.5–10.5
Phosphorus	3.7 mg/dL	2.7–4.5
ALP	195 U/L	30–135
AST	39 U/L	5–42
ALT	54 U/L	5–45
LDH	227 U/L	0–247
GGT	124 U/L	5–85
Total bilirubin	0.33 mg/dL	0.2–1.0
Total cholesterol	212 mg/dL	130–200
HDL	46 mg/dL	>40
LDL	140 mg/dL	100–130
Triglyceride	203 mg/dL	60–150
Total protein	6.2 g/dL	6.0–8.0
Albumin	3.1 g/dL	3.2–5.5
ESR	31 mm/h	0–20
CRP	0.4 mg/dL	0–0.8
WBC	4100/μL	4000–11000
Hb	10.8 g/dL	12–18
MCV	90 fL	80–100
Platelet	196000/μL	150000–400000

ALP alkaline phosphatase, *AST* aspartate aminotransferase, *ALT* alanine aminotransferase, *LDH* lactate dehydrogenase, *GGT* gamma glutamyl transferase, *HDL* high-density lipoprotein, *LDL* low-density lipoprotein, *ESR* erythrocyte sedimentation rate, *CRP* C-reactive protein, *WBC* white blood cells, *Hb* hemoglobin, *MCV* mean corpuscular volume

history. Anti-nuclear antibody was positive in a titer of 1/320. Because her serum creatinine was 2.4 mg/dL and pyuria, hematuria, and proteinuria (3.84 g/day) were present, with the pre-diagnosis of lupus nephritis, renal biopsy was performed. A full-house pattern was observed in immunofluorescence studies and biopsy was compatible with diffuse proliferative lupus nephritis. Anticardiolipin IgM/IgG and lupus anticoagulant tests were all negative. For the investigation of exertional dyspnea, echocardiography was performed. The ejection fraction was 35 %, and pericardial effusion of 9 mm in diameter was present. Mitral regurgitation 2+ was found, together with an increased pulmonary artery pressure of 52 mmHg. Increased apical trabeculations were also detected (Figs. 1 and 2). Cardiac magnetic resonance (MR) imaging was consistent with LVNC, and the ratio of noncompacted to compacted myocardial thickness was 2 (Fig. 3). Pulse intravenous methylprednisolone 1000 mg/day in three consecutive days and pulse intravenous cyclophosphamide 750 mg/month was started for the treatment of lupus nephritis. The patient was followed with maintenance

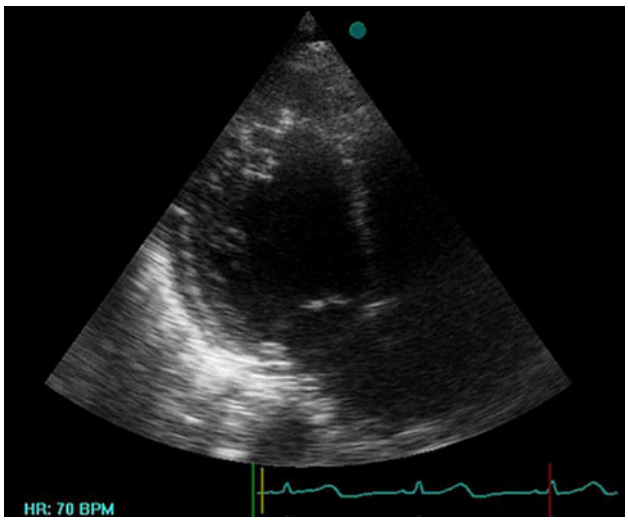

Fig. 1 Echocardiographic images showing increased apical trabeculations with left ventricular noncompaction (sagittal axis)

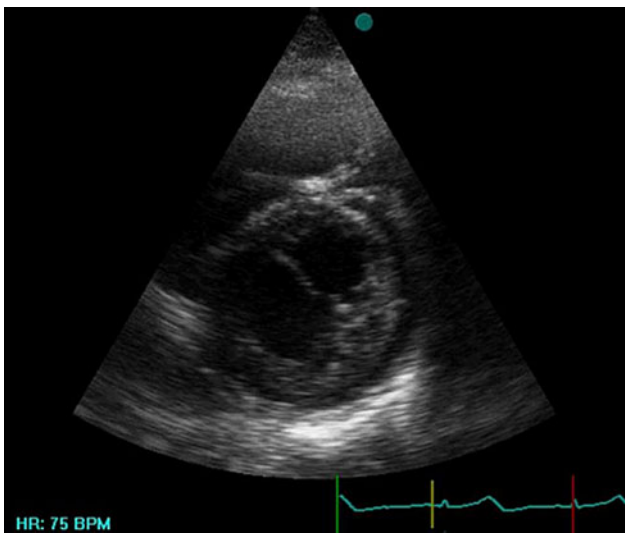

Fig. 2 Echocardiographic images showing increased apical trabeculations with left ventricular noncompaction (short parasternal axis)

steroid treatment with a tapering dose of 1 mg/kg prednisolone. Under the treatment for SLE nephritis, the patient expressed a marked decrease in exertional dyspnea. Echocardiography was repeated in the third month of the treatment and revealed a significant improvement of the ejection fraction (first ejection fraction: 35 % to control ejection fraction: 45 %); however, hypertrabeculation was still present.

Discussion

LVNC, also called left ventricular hypertrabeculation, is a rare morphologically distinct primary genetic

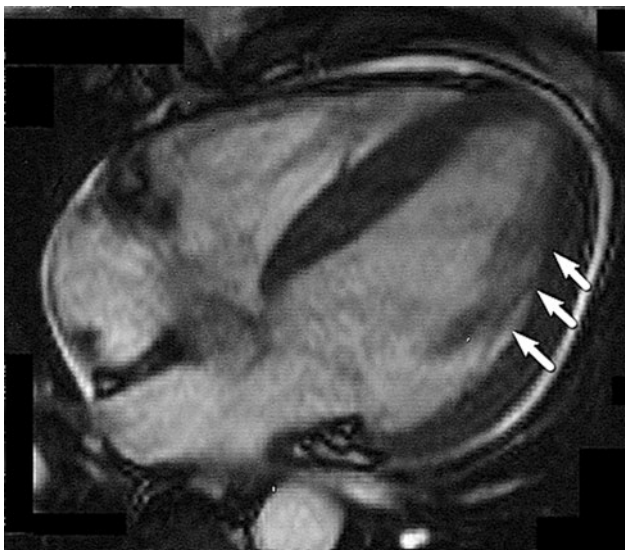

Fig. 3 Cardiac magnetic resonance imaging reflecting increased trabeculations (*white arrows*), with the ratio of noncompacted to compacted myocardial thickness being 2

cardiomyopathy [1]. It is due to the failure of condensation of the myocardial spongy meshwork of fibers and intertrabecular recesses during intrauterine life leading to the persistence of ventricular trabeculations [2]. Histologically, interstitial fibrosis, subendocardial fibroelastosis, and necrotic myocytes within the prominent trabeculations have been described [3, 4]. Both familial (autosomal dominant/X-linked inheritance) and sporadic forms of noncompaction have been described [5]. The prevalence of LVNC is often underestimated and patients are thought to have dilated, hypertrophic, or restrictive cardiomyopathies.

The major clinical manifestations are heart failure, systemic thromboembolism, ventricular arrhythmias, conduction disorders, and neurologic abnormalities [3]. In many cases, LVNC is associated with neuromuscular disorders, including myotonic dystrophies, dystrophinopathies, Charcot–Marie–Tooth disease, Barth syndrome, Friedreich ataxia, and Pompe's disease [6].

The diagnosis of LVNC can be made by echocardiography as well as MR imaging. A ratio of noncompacted to compacted myocardium ≥ 2 was considered as a criteria of LVNC [3, 7]. The affected myocardial segments are located in the apical, mid-lateral, and mid-inferior regions, and are often hypokinetic due to subendocardial hypoperfusion and microcirculatory dysfunction. Abnormalities of the resting electrocardiogram (ECG) include left ventricular hypertrophy, right or left bundle branch block, atrial fibrillation, and repolarization abnormalities [8]. The management of patients with LVNC is similar to that of patients with other cardiomyopathies and include appropriate treatment for heart failure, arrhythmias, and oral anticoagulation to prevent systemic emboli in patients with impaired left ventricular function [9].

The heart is frequently involved in SLE, with a prevalence of >50 % [10]. All three layers of the heart—pericardium, myocardium, and endocardium—may be involved by lupus. Myocarditis is the most characteristic feature of myocardial involvement in SLE. The clinical detection of myocarditis associated with SLE ranges from 3 to 15 % [11]. Signs and symptoms are similar to those of myocarditis due to other causes and they can progress to ventricular dysfunction, dilated cardiomyopathy, and heart failure. There are no typical findings on ECG, and cardiac enzymes may be normal. Recently, MR imaging has been increasingly used for diagnosing myocarditis with high sensitivity, even at the preclinical stage [12]. In our case, no evidence of myocarditis was observed in the cardiac MR imaging.

Importantly, our patient expressed a marked decrease in exertional dyspnea after the immune-suppressive treatment for the lupus nephritis. Echocardiography after treatment revealed a significant improvement of the ejection fraction, which cannot be expected for a case of LVNC, which is a primary genetic cardiomyopathy. Thus, we eventually hypothesize that this was a case of SLE-induced cardiomyopathy resembling LVNC. In a recent report compatible with our hypothesis, a male patient with SLE developed cardiomyopathy with myocardial hypertrabeculation and ventricular functions also improved after the treatment for SLE [13].

In conclusion, SLE may cause a kind of cardiomyopathy with high resemblance to LVNC. Discrimination of these two similar clinical entities is important because SLE-induced cardiomyopathy is potentially reversible after the immune-suppressive treatment for SLE.

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