

Systemic lupus erythematosus and transient left ventricular noncompaction

Cristina Ferreira Rabelo¹, Fabio Batistella¹, Ronaldo Nunes Godinho¹,
Tiago Santini Machado², Flavio José Petersen Velho², Henrique Luiz Staub¹

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ABSTRACT

Objectives: Myocarditis is a rare but serious manifestation of patients with systemic lupus erythematosus (SLE). Left ventricular noncompaction (LVNC) is a disorder of myocardial morphogenesis frequently associated to neuromuscular diseases. Hypertrabeculation, a cardinal echocardiographic feature of LVNC, might represent a morphological expression of a number of morbidities, nevertheless. The relationship of LVNC with connective tissue disorders such as SLE is unknown. We aim to present a case of a patient with SLE who recently showed features compatible with an atypical LVNC. **Methods:** To report a case of a young female with a 10-year history of SLE who developed haematological disease activity and cardiac failure. **Results:** Ecocardiography showed hypertrabeculation/noncompaction, a very low ejection fraction and pulmonary hypertension. Clinical and echocardiographic features reverted with standard treatment for SLE activity and cardiac insufficiency. **Conclusion:** The transitory aspect of the cardiomyopathy made unlikely a “true” LVNC for this patient, but she might have presented a lupus myocarditis with “LVNC-like” features. The occurrence of hypertrabeculated myocardium in patients with SLE warrants further studies.

Keywords: Systemic lupus erythematosus; left ventricular noncompaction; cardiomyopathy.

INTRODUCTION

Myocarditis is an uncommon, but life-threatening, feature of patients with systemic lupus erythematosus (SLE)¹. Left ventricular noncompaction (LVNC) has been a matter of interest for the last years. An arrest of normal endomyocardial morphogenesis supposedly leads to LVNC. Heart failure, systemic embolism and arrhythmias are cardinal findings, and echocardiography characteristically reveals prominent ventricular trabeculations. LVNC occurs isolately or in association with neuromuscular disorders², but is very rare in SLE. We herein report a case of a patient with SLE and a transient cardiomyopathy which resembled LVNC.

CLINICAL CASE

The patient, a 30-year-old black female, was diagnosed SLE in 2002 (polyarthritis, photosensitivity, leukopenia, hemolytic anemia, pleuropericarditis, positive test for antinuclear, anti-DNA and anticardiolipin antibodies). For 5 years she was treated with variable doses of prednisone, low-dose aspirin, chloroquine and, eventually, intravenous (IV) methylprednisolone (MP) pulsetherapy. A routine echocardiography carried out in September 2009 was unremarkable. Maintenance therapy with leflunomide and low-dose prednisone were utilized up to September 2011, when she developed progressive dyspnea. The thorax radiogram revealed an augmented cardiac area. The echocardiography proceeded on systole showed, at this time, an excentric hypertrophy of the left ventricle, diffuse hypokinesia, and a considerable systolic dysfunction. Left ventricular hypertrabeculation/noncompaction within the apex, and also in the anterior and lateral walls, were seen. Deep intertrabecular recesses were documented (Figure 1). The relationship of non-compacted to compac-

1. Rheumatology Department, Sao Lucas Hospital of Pontifical Catholic University of Rio Grande do Sul (PUCRS), Porto Alegre, Brazil

2. Cardiology Department, Sao Lucas Hospital of Pontifical Catholic University of Rio Grande do Sul (PUCRS), Porto Alegre, Brazil

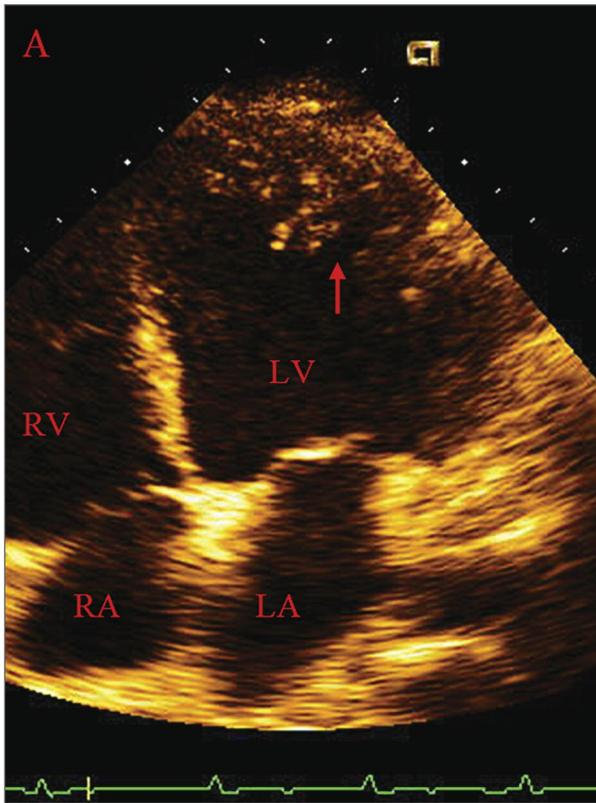


FIGURE 1. Four chamber view echocardiography carried out on systole showing excentric hypertrophy of the left ventricle (LV). Prominent ventricular trabeculations and deep intertrabecular recesses are seen (red arrow). RV: right ventricle; LA: left atrium; RA: right atrium.

ted myocardium was > 2 . The left atrium was dilated. The systolic pressure of pulmonary artery was 46 mm Hg, and the left ventricular ejection fraction (LVEF) was 25%. Hemolysis was concomitantly present (hemoglobin 9.1 g/dL, positive Coombs test). Anti-DNA antibodies were detected in high titers (1/2560), and Complement depletion was noted (C3 46 mg/dL, C4 8 mg/dL). The renal function was normal. The patient was treated with a combined pulsetherapy of cyclophosphamide (500 mg) and MP (500 mg). Standard therapy for cardiac insufficiency included enapril, carvedilol, spironolactone and furosemide. Aspirin was withdrawn, and oral anticoagulation with warfarin was started. Prednisone 60 mg was given daily. Due to the refractory hemolysis, rituximab (2 grams IV) were infused in January 2012. The more recent echocardiography showed no signs of hypertrabeculation; LVEF was of 57.95%, and the systolic pressure of pulmonary artery was of 26 mmHg. At that moment, the patient

was on daily use of prednisone 20 mg, hydroxychloroquine 400 mg, enalapril 5 mg, spironolactone 25 mg, furosemide 40 mg and carvedilol 12.5 mg. Warfarin therapy was withdrawn.

DISCUSSION

While myocarditis, although uncommon, is an established feature in SLE³, LVNC is yet an unclassified cardiomyopathy. The disorder was proposed as a primary cardiomyopathy of genetic origin in 2006 by the American Heart Association. In 2008 the European Society of Cardiology updated the classification scheme similar to the World Health Organization classification⁴.

Out of 3,854 consecutive outpatients who performed echocardiography, the prevalence of LVNC (0.31%) was not out of consideration⁵. Approximately two thirds of the patients with LVNC might present with a neuromuscular disorder, usually Barth syndrome, mitochondrial disorders, or myotonic dystrophies⁶. Patients with such neuromuscular diseases should be promptly screened for cardiomyopathy⁷. Pathogenetic mechanisms linking LVNC and neuromuscular diseases are still unclear^{6,7}.

The disease could occur isolately, nevertheless. Thirty cases of isolated LVNC were reported in 2009, being the mortality (10%) similar to that of patients with dilated cardiomyopathy⁸. Three other cases of isolated LVNC were recently described in patients who underwent cardiac transplantation⁹.

In 2007, LVNC was reported in a patient with Behçet's disease showing multiple thrombus formations¹⁰. A patient with ankylosing spondylitis was also found to present features of LVNC¹¹. Recently, a cardiomyopathy with high resemblance to LVNC was described in a patient with proliferative lupus nephritis; similarly to the case here reported, immunosuppressive therapy improved clinical and echocardiographic parameters¹².

The current case is probably the second describing features of LVNC in a patient with SLE. Our patient presented a typical SLE characterized by cutaneous, articular, hematological and serosal features. No cardiac complaints were present in the first 9 years of disease. Heart failure and echocardiography features compatible with LVNC appeared in September 2011, in conjunction with hemolysis, Complement depletion, and circulant anti-DNA antibodies. After immunosuppression (including traditional drugs and ritu-

ximab) plus standard therapy for heart failure, SLE activity, heart symptoms and echocardiographic changes all came to normalization.

Although our patient fulfilled classical echocardiographic criteria for LVNC¹³, the reversible pattern of cardiomyopathy turns unlikely that she presented a “true” LVNC. We hypothesize that “LVNC-like” echocardiographic features were representative of a variant of lupus myocarditis with hypertrabeculation. In this case, myocarditis would be part of a wider context of SLE activity, including hematological and serological findings. Even though the patient was concomitantly treated for heart failure, one can not rule out the possibility that immunosuppression was helpful in the control of the cardiomyopathy. Alternatively, the patient could have presented active SLE and, by chance, a separate and atypical myocarditis with features of LVNC.

At times, differentiating some cases of LVNC from normal variant myocardial architecture can be rather difficult. LVNC might represent more a morphologic expression of different underlying morbidities than a distinct cardiomyopathy, according to some group of authors^{14,15}. If these underlying disorders include SLE and other connective tissue disorders, it is an open question. Of importance, the reproducibility of echocardiographic diagnosis of LVNC has been poor, according to recent data¹⁶. Falsely diagnosed LVNC might include apical hypertrophic cardiomyopathy, thrombi and aberrant bands; international standardization of echocardiographic methods for detection of LVNC may be needed¹⁷.

In summary, we here reported a case of a young female with long-standing SLE who recently showed active hematological disease and an atypical and transitory cardiomyopathy resembling LVNC. The relationship of SLE with cardiomyopathies featuring hypertrabeculation warrants further studies.

CORRESPONDENCE TO

Cristina Ferreira Rabelo
Avenida Ipiranga 6690; sala 220
90610-000 Porto Alegre, Brazil
E-mail: cristinarabelo@yahoo.com.br

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