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Left Ventricle Non Compaction-Like Cardiomyopathy in a Patient with Systemic Lupus Erythematosus

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Case Report

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Abstract: Left ventricular noncompaction (LVNC) is a rare genetic cariomyopathy. 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. 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. 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 Systemic lupus erythematosus (SLE) is unknown. We aim to present a case of a patient with SLE who recently showed features compatible with an atypical LVNC. We report a case of a young female with a 5-year history of SLE who developed haematological disease activity and cardiac failure. Echocardiography showed left ventricle dilation, hypertrabeculation/noncompaction, a very low ejection fraction at 25% and pulmonary hypertension. After treatment, all signs of LVNC disappeared in echocardiography.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: Left ventricle noncompaction, Systemic lupus erythematosus, echocardiography, cardiomyopathy.

INTRODUCTION

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, which are 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 Systemic lupus erythematosus (SLE) is unknown.We aim to present a case of a young patient with SLE who recently showed features compatible with an atypical LVNC.

CASE REPORT

The patient, an 18-year-old young woman, was diagnosed SLE at the age og 13 (polyarthritis, leukopenia, photosensitivity, hemolytic anemia, positive test for antinuclear, anti-DNA and anticardiolipin antibodies). For 5 years she was treated with variable doses of prednisone, low-dose aspirin and chloroquine. However, since about a year, she developped 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 compacted myocardion was > 2. The left atrium was dilated. The systolic pressure of pulmonary artery was 50 mmHg, and the left ventricular ejection fraction (LVEF) was 25%. Anti-DNA antibodies were detected in high titers (1/2000), hemolysis was noted (hemoblobin 8.9 g/dL, positive Coombs test). The renal function was normal.

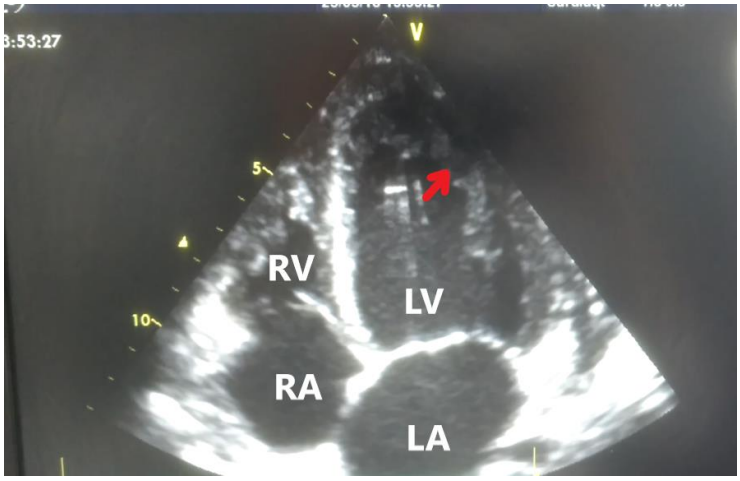

Fig-1: Four chamber view echocardiography showing excentric hypertrophy of the left ventricle (LV). Prominent ventricular trabeculations and deep intertrabecular recesses are seen (red arrow). RV: right ventricle; LA: left atrium; RA: right atrium

The patient was treated with a The patient was treated with a combined pulsetherapy of cyclophosphamide (500 mg) and methylprednisolone (500 mg). Standard therapy for cardiac insufficiency included ramipril, carvedilol, spironolactone and furosemide. Prednisone 60 mg was given daily. Under the treatment, 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: 25 % to control ejection fraction: 45 %) and no signs of hypertrabeculation. At the moment the patient was on daily use of prednisone 20 mg, hydroxychloroquine 400 mg, ramipril 5 mg, spironolactone 25 mg, furosemide 40 mg and carvedilol 12.5 mg.

DISCUSSION

LVNC, also called left ventricular hypertrabeculation, is a rare morphologically distinct primary genetic 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]. While myocarditis, although uncommon, is an established feature in SLE [6], LVNC is yet an unclassified cardiomyopathy. The prevalence of LVNC is often underestimated and patients are thought to have dilated, hypertrophic, or restrictive cardiomyopathies. Out of 3,854 consecutive outpatients who performed echocardiography, the prevalence of LVNC (0.31%) was not out of consideration [7]. Approximately two thirds of the patients with LVNC might present with a neuromuscular disorder, usually Barth syndrome, mitochondrial disorders, or myotonic dystrophies [8]. Patients with such neuromuscular diseases should be promptly screened for cardiomyopathy [9]. Pathogenetic mechanisms linking LVNC and neuromuscular diseases are still unclear [8, 9]. 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 cardiomyopathy [10]. Three other cases of isolated LVNC were recently described in patients who underwent cardiac transplantation [11]. In 2007, LVNC was reported in a patient with Behçet's disease showing multiple thrombus formations [12]. A patient with ankylosing spondylitis was also found to present features of LVNC [13]. 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 ecochardiographic parameters [14]. 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 [15].

CONCLUSION

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 [16,17]. 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 data [18]. Falsely diagnosed LVNC might include apical hypertrophic cardiomyopathy, thrombi and aberrant bands; international standardization of echocardiographic methods for detection of LVNC may be needed [19]. 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.

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