CASE REPORTS

Noncompaction of ventricular myocardium involving both ventricles

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KEYWORDS
Noncompaction of ventricular myocardium; Cardiomyopathy; Heart failure

Abstract  Aim: We aimed to present a case with ventricular myocardial noncompaction involving both ventricles.
Methods and results: Noncompaction of ventricle is a rare and unclassified congenital cardiac malformation is due to an arrest in intrauterine endomyocardial morphogenesis. We presented a ventricular myocardial noncompaction case involving both left and right ventricles. The physical examination of this case is consistent with mitral regurgitation and the echocardiographic findings are consistent with noncompaction of ventricular myocardium involving both ventricles with left ventricular systolic failure.
Conclusion: Transthoracic echocardiography is a useful clinical tool for diagnosing noncompaction of both the right and left ventricular myocardium. The LVNC definition can also be utilized for RVNC, which this diagnosis has never been reported in a Turkish patient.
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Introduction

Left ventricular noncompaction (LVNC) is a rare congenital anomaly both in children and adults.1,2 Some authors regarded this pathology as a subvariety of the dilated cardiomyopathy (CMP) syndrome.3 This unclassified cardiomyopathy results from an arrest in intrauterine endomyocardial morphogenesis, and is diagnosed either sporadically or with a familial tendency due to chromosomal anomalies4–11 and the age of onset varies widely.11,12 Its common clinical presentations involve heart failure, ventricular tachyarrhythmia and thromboembolic events. In the Australian Childhood

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Cardiomyopathy trial, they have found a male predominance and 9.2% of patients with unclassified cardiomyopathy have LVNC.\textsuperscript{13}

The diagnosis is based on Jenni and Stöllberger’s LVNC/left ventricular hypertrabeculation definition.\textsuperscript{2,14} We present a patient with myocardial non-compaction of both ventricles. The diffuse right ventricular (RV) involvement is based on Jenni and Stöllberger’s definition (RVNC).

Case report and discussion

A 26-year-old male was referred to our clinic for the evaluation of mild effort dyspnea. Physical examination revealed a blood pressure of 120/70 mmHg with a regular pulse of 75/min. Cardiac auscultation revealed normal first and second heart sounds without any third and fourth heart sounds and a systolic murmur at the fourth intercostal space without any other pathologic physical findings. ECG was consistent with sinus rhythm and marked both left and right ventricular hypertrophy by voltage criteria with ST and T wave strain pattern.

Transthoracic echocardiography (TTE) was performed with a Vingmed System V (GE, Horton, Norway) and a 2.5-MHz probe according to American Society of Echocardiography (ASE) guidelines.\textsuperscript{15} The TTE was consistent with a mild dilated left ventricular (LV) chamber and diffuse LV hypokinesis with severe hypertrophy of both septal (IVS) and lateral LV walls. The RV wall thickness was 18.6 mm (Fig. 1). The trabeculations with deep fissures and grooves were located in the LV inferior, apical, posterolateral and septal (IVS) walls. A bright echo reflectance was detected at the basal septal and lateral walls of both the LV and RV in the apical four-chamber view (Fig. 2). Blood flow was detected inside these deep fissures with color flow Doppler. The IVS thickness was thicker at certain sites (29.3 mm) than the posterior (12 mm) and lateral (12 mm) LV walls. The LVEF and RVEF were measured as 35—40% and 55%, respectively. The Doppler examination of the mitral valve was consistent with mild degree mitral regurgitation. The pulmonary flow, which was recorded from the parasternal short axis at the aortic level, was also assessed normal (0.97 m/s) without any gradient. We suggested cardiac catheterization to rule out the pulmonary hypertension but the patient refused this invasive procedure. There weren’t any indirect TTE findings consistent with pulmonary hypertension. The patient complaints regressed with medical congestive heart failure therapy (digitalis, ACE inhibitor, diuretic) and regressed from NYHA class III to NYHA class II. The echocardiographic examination of family members showed normal echocardiographic examination for the parents and first degree relatives (uncle, aunt and brother). Medical treatment is one of the treatment modalities, which is effective in terms of relieving the heart failure symptoms besides implantable cardiac defibrillator as well as cardiac transplantation.\textsuperscript{16—18} The prognosis

Figure 1  Apical four-chamber view showing noncompaction of LV with diffuse RV involvement. LV = Left ventricle, RV = right ventricle.
of LVNC differs according to authors, which we still follow our case for two years after the diagnosis.16,17

Conclusion

Transthoracic echocardiography is a useful clinical tool for diagnosing noncompaction of the ventricular myocardium. Jenni and Stöllberger’s definition for the LVNC can also be utilized for the RVNC.

References


Figure 2  Apical four-chamber view showing severe thick RV walls with bright echo reflectance. LV = left ventricle, RV = right ventricle.
Systemic lupus erythematosus: An unusual cause of cardiac tamponade in a young man

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Abstract Although pericarditis and pericardial effusion are common cardiac complications of systemic lupus erythematosus (SLE), cardiac tamponade is a very rare initial manifestation of this disease. We describe a case of a young male patient in whom cardiac tamponade secondary to a loculated pericardial effusion was the presenting symptom of SLE. © 2005 The European Society of Cardiology. Published by Elsevier Ltd. All rights reserved.

Introduction

Systemic lupus erythematosus (SLE) is a connective tissue disorder which often involves the heart, mostly the pericardium usually manifests as diffuse pericardial effusion.1 It is mostly of a mild degree and more common in elderly.2 Cardiac involvement as the initial presentation of SLE has been reported in a few patients and cardiac tamponade of the disease is rare as the first manifestation.3 SLE related cardiac tamponade has generally a benign evolution with proper treatment.4 Although pericardiocentesis associated with anti-inflammatory drugs is the treatment of choice, surgery is indicated in some cases. In this report, we describe a young male in whom cardiac tamponade secondary to localized pericardial effusion was surgically treated and the diagnosis of SLE was established.