Isolated Left Ventricular Noncompaction Mimicking Cystic Mass

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Abstract
Noncompaction of the ventricular myocardium is a recently recognized genetic cardiomyopathy thought to be caused by arrest of normal embryogenesis of the endocardium and myocardium. The diagnosis is usually made by echocardiography. Herein we report the case of a 29-year-old male with isolated left ventricular noncompaction mimicking cystic mass.

Keywords
Cardiomyopathy; Cystic Mass; Echocardiography; Magnetic Resonance Imaging

Özet

Anahtar Kelimeler
Kardiyomiyopati; Kistik Kitle; Ekokardiyografi; Manyetik Rezonans Görüntüleme


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Introduction
Noncompaction of the ventricular myocardium is a congenital cardiomyopathy characterized by deep intertrabecular recesses connected to the ventricular cavity thought to be caused by arrest of normal embryogenesis of the endocardium and myocardium [1, 2]. It may be isolated or associated with other congenital cardiac defects [2, 3]. Echocardiography is the diagnostic procedure, but the diagnosis is often missed or delayed because of poor echo window in assessment of the LV apex, lack of knowledge about this uncommon disease, and its similarity to other diseases of the myocardium and endocardium [1, 2, 4].

Case Report
A 29-year-old male with history of atypical chest pain was referred to our clinic. During his evaluation, intracardiac mass was discovered on a routine echocardiogram. There were no other symptoms of heart failure or coronary artery disease. Patient has no family history of heart disease or sudden death. On admission, his blood pressure was 125/75 mmHg and the heart rate was 68 beats/min. The 12-lead electrocardiography (ECG) showed normal sinus rhythm. Transthoracic echocardiogram and cardiac magnetic resonance imaging (MRI) were performed. Echocardiogram revealed a prominent cystic mass in the apical interventricular septum with normal left ventricular function (LVEF: 60%), and Color Doppler showed no flow within the cystic mass (Figure 1). MRI showed a typical pattern of noncompacted myocardium predominantly of the left ventricle (LV). Ratio of noncompacted to compacted area was 2.7 (pathological >2) (Figure 2). The LV was mildly dilated with preserved systolic function (LVEF: 50%). The ambulatory ECG documented sinus rhythm. There were no atrial fibrillation and ventricular arrhythmias.

Lacking indications for more aggressive therapy, chronic anticoagulation with warfarin was prescribed. The patient was considered for regular follow-up to include assessment of exercise tolerance, measurement of ventricular size and function, and use of continuous ambulatory ECG. First degree relatives were screened with echocardiography.

Discussion
Noncompaction of the ventricular myocardium is a recently recognized genetic cardiomyopathy. Both familial and sporadic forms of isolated left ventricular noncompaction (ILVN) have been described [2, 3]. The median age at diagnosis was 7 years (ranging from 11 months to 22 years) in the first case series of ILVN [2]. But subsequent case reports have described ILVN in adults such as our patient. Men appear to be affected more than women. The prevalence was 0.014% of patients referred to the echocardiography laboratory, but the true prevalence is unclear [2, 3].

Clinical manifestations may range from being asymptomatic to presenting with heart failure, malignant arrhythmias, sudden cardiac death, or systemic thromboembolism [1, 2]. Echocardiography is the method of choice in establishing the diagnosis of ILVN. The echocardiographic characteristics of ventricular noncompaction include, segmental thickening of the LV myocardial wall consisting of 2 layers: a thin, compacted epicardial layer and an extremely thick layer with prominent trabecula-

Figure 1. Apical four chambers (A) and parasternal short axis (B) echocardiograms showing a prominent cystic mass (arrows) in the apical interventricular septum.

Figure 2. Two chamber (A) and short axis (B) steady state free precession (SSFP) MR images showing a typical pattern of noncompacted myocardium predominantly of the left ventricle. Ratio of noncompacted (Y) to compacted (X) area was 2.7.

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Competing interests
The authors declare that they have no competing interests.

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