

Left ventricular “noncompaction” with hypothyroidism and sensorineural hearing loss

Hipotiroidi ve sensörinöral işitme kaybı ile birlikte olan sol ventrikül kökenli “noncompaction”

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Introduction

Ventricular myocardial “noncompaction” (MN), which is also called honeycomb or spongy myocardium, is a rare type of cardiomyopathy characterized by a hypertrophic left ventricle with deep trabeculations and poor systolic function, with or without associated left ventricle dilatation (1-3). It has been thought that the basic morphogenetic abnormality may be the arrest of normal compaction of the loose interwoven mesh of myocardial fibers during the myocardial morphogenesis (4). The disease was first described among children, but subsequently adult cases were also reported. It is more commonly observed in the left ventricle, although involvement of the right ventricle has been reported (3). Mortality and morbidity rates are high due to cardiac failure, ventricular arrhythmias and systemic emboli (2). There are ventricular noncompaction cases reported in our country (5-7). Cardiac and less commonly non-cardiac anomalies accompanying left ventricular MN have also been reported (8). In our case, ventricular MN is accompanied by hypothyroidism and bilateral sensorineural hearing loss, which has not been reported in the literature.

Case

A 19-year-old female patient presented to a clinic with complaints of abdominal distention and bilateral leg edema that had been present for the preceding one year. The treatment she received led to a regression of her complaints. She was admitted to our hospital with increase in her complaints in the last 2-3 months accompanied by dyspnea on exertion and at rest, lack of appetite, and weakness. Her medical history revealed bilateral sensorineural hearing loss diagnosed when she was 8-year-old.

Her physical examination revealed a blood pressure of 90/60 mmHg, regular pulse of 90/min and the respiration rate of 20/min. The sclera was subicteric and conjunctiva was pale. Cardiovascular system examination revealed increased left ventricular activity and 3/6 systolic murmur on the left side of sternum and in the mitral listening area. There were ascites and pretibial (+1) edema.

Laboratory analyses revealed that the hemoglobin level was 11.4 gr/dl and leukocyte and platelet levels were in normal ranges. Biochemical analyses demonstrate normal electrolyte and liver function test results. Thyroid hormone analysis showed that the TSH level was 11.56 uIU/ml (normal range 0.5-4.95). Thyroid autoantibodies were found to be negative. Thyroid ultrasonography revealed that the gland was in normal size and no nodular appearance was noted. Abdominal ultrasonography showed large liver size (17 cm) and congested appearance.

Electrocardiography showed left bundle branch block. Cardiothoracic ratio was increased. Transthoracic echocardiography showed enlarged left ventricle (diastolic inner diameter: 6.5 cm), global hypokinesia (EF: 35%) and paradoxal movement in the septum. Moderate tricuspid regurgitation (regurgitation gradient of 36mmHg), right atrial dilatation and mild-to-moderate mitral and pulmonary insufficiency were recorded. Deep trabeculations observed in the apex and lateral wall of the left ventricle were considered consistent with ‘noncompaction’ (Fig. 1). The size and wall structure of right ventricle were normal. Angiotensin converting enzyme inhibitor, spironolactone, furosemide, low-dose carvedilol, aspirin and L-thyroxin were started and the patient was followed-up.

Discussion

Myocardial “noncompaction” is a rare myocardial disease that represents an arrest of myocardial morphogenesis (3,9). This disorder was firstly described in 1932 in an autopsy of a newborn with aortic atresia and coronary-ventricle fistula (8). The first case of MN not accompanied by another morphologic cardiac anomaly was described by echocardiography in 1984 (10). The typical characteristics of this disorder are prominent ventricular trabeculations and deep intertrabecular recesses associated with ventricular gap (2). Through a careful analysis by echocardiography, abnormal trabeculations may be detected. Determining the blood flow between trabeculations by color Doppler is considered as a pathognomonic finding for diagnosis (1). There is no intracardiac shunt. Other techniques used

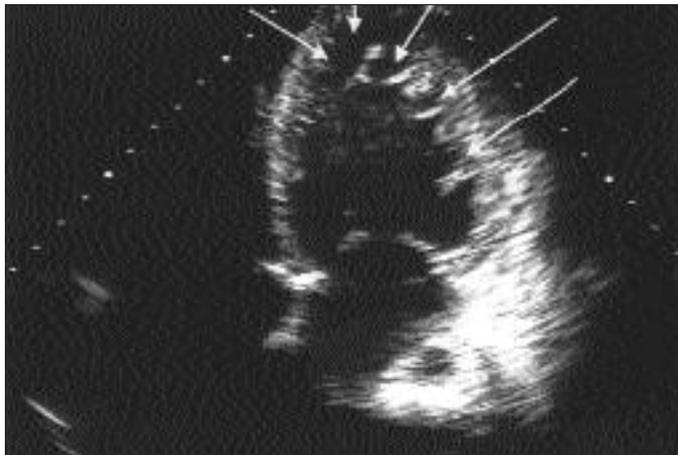


Figure 1. Two-dimensional echocardiographic view of deep trabeculations in left ventricular apex and lateral wall

for diagnosis are computerized tomography, magnetic resonance imaging and ventriculography (8,10). Diagnosis of ventricular MN in adult patients may be late due to late beginning of complaints. Although the apical area of the left ventricle is commonly involved, both ventricles may be involved and familial forms have also been identified besides isolated forms (2,7, 9).

In our case, the left ventricular apex and posterolateral wall showed characteristic morphological changes of ventricular MN. We considered our case as isolated ventricular MN because no signs of noncompaction on echocardiographic examination were found in parents of our patient. The patients commonly had findings related with cardiac failure, ventricular arrhythmias, and systemic emboli (2,9). It has been reported that the diastolic dysfunction seen in these patients may be related with the restriction in filling of ventricles developing as a consequence of abnormal ventricular relaxation and extreme intracavitary trabeculations. However, the real factor in the development of diastolic and systolic dysfunction has not been elucidated yet (11). Although myocardial “noncompaction” exists at birth, ventricle dysfunction occurs in older ages (4). Arrhythmias may also be seen and are very important for the prognosis of the patient (12). The prognosis is also poor in symptomatic patients (8). Our patient had symptoms of cardiac failure and echocardiographic parameters that indicate ventricular arrhythmias and diastolic dysfunction.

There are reports on association of left ventricular MN with other morphological cardiac anomalies, such as ventricular septal defects, pulmonary stenosis, and atrial septal defects (8). Non-cardiac abnormalities consisting of neurological, facial, hematological, endocrine, dermatological and skeletal anomalies have also been described as isolated case reports (8). Our patient, in addition to left ventricular MN, had congenital hypothyroidism and bilateral sensorineural hearing loss. In the

literature, only one case of MN with hypothyroidism has been reported (13). Additionally, literature review revealed also two cases of MN with hearing loss (13,14). Our case is the first case of ventricular MN in the literature, in which both bilateral congenital sensorineural hearing loss and hypothyroidism are seen.

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