Myocardial noncompaction recognized following a transient ischemic attack

Geçici iskemik atak sonrası saptanan süngerimsi miyokart (noncompaction)

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Myocardial noncompaction is a rare type of congenital cardiomyopathy characterized by excessively prominent trabeculations in one or more segments of the ventricles and deep intertrabecular recesses in ventricular walls. A 25-year-old male patient presented to the neurology department with complaints of weakness in the left extremities. A mild loss of muscle strength was detected on neurological examination. With a preliminary diagnosis of acute cerebrovascular event, treatment with aspirin and enoxaparin was instituted, which improved his complaints within two hours. Electrocardiography showed sinus rhythm, left ventricular hypertrophy, and loss of R-wave progression in the precordial leads. Transthoracic echocardiography showed apical hypokinesia, marked left ventricular hypertrophy, and normal left ventricular diameters. There were numerous trabeculations in the apex, apical, lateral, and inferior walls, and deep intertrabecular recesses. Color Doppler showed blood flow into the intertrabecular recesses. He also had mild mitral regurgitation and diastolic dysfunction of restrictive type. He was scheduled for outpatient follow-up on aspirin and warfarin treatment.

Key words: Cardiomyopathies/congenital; echocardiography, heart ventricles/abnormalities; intracranial embolism/etiology.

Myocardial noncompaction (MN) is a rare type of congenital cardiomyopathy characterized by excessively prominent trabeculations and deep intertrabecular recesses in ventricular walls associated with arrest in intrauterine myocardial growth.[1,2] Left ventricular apical segment is mostly involved. There has been a recent increase in the incidence of isolated left ventricular MN. Diagnosis is generally based on symptoms and findings due to heart failure, arrhythmias and systemic embolism. Myocardial noncompaction is one of the major factors of mortality and morbidity.[2] Patients with cerebral embolism particularly associated with MN are rarely seen.[1]


Anahtar sözcükler: Kardiomyopati/doğuştan; ekokardiografı; kalp ventrikülü/anormallik; intrakraniyal emboli/etyoloji.

In this paper we presented a case diagnosed with isolated left ventricular MN during investigation of the etiology of cerebral embolism.

CASE REPORT

A 25-year-old male patient with no previous symptoms visited neurology clinic with complaints of weakness in the left extremities and was referred to the cardiology clinic. Following detection of mild loss of muscle strength in the left upper and lower extremities (4/5 forme-fruste paralysis) during the neurological examination, the patient was followed-up with the preliminary diagnosis of acute cerebrovascular event and...
received a treatment schedule of aspirin 300 mg, 1x1 and subcutaneous enoxaparin 8000 IU, 1x1. Findings of cranial computed tomography and carotid Doppler ultrasonography were found to be normal. The patient whose complaints improved after two hours was referred to the cardiology clinic. His blood pressure was 85/60 mmHg and pulse was 75/min. Physical examination did not reveal any significant pathology. His medical history did not reveal any complaint apart from dyspnea following excessive effort. The family history of the patient who had a 10 pack-year smoking history was also uneventful. Electrocardiography showed sinus rhythm, left ventricular hypertrophy, and loss of R-wave progression in the precordial leads. Transthoracic echocardiography was performed using the 1.5-3.5 MHz multi-frequency probes by GE Vivid 3 Pro. Transthoracic echocardiography showed apical hypokinesia and marked left ventricular hypertrophy (IVS 2.0 cm; PW 1.6 cm) in the patient with ejection fraction of 51%. Diameters of left ventricles were within normal limits (diastolic diameter 4.7 cm; systolic diameter 3.6 cm), while there were numerous trabeculations and deep intertrabecular recesses (non-compaction) in the apex, apical, lateral, and inferior walls (Figure 1,2). Color Doppler showed blood flow into the intertrabecular recesses. A mild mitral regurgitation and diastolic dysfunction of the restrictive type were detected, whereas no pathology was found in left spaces. A 24-hour Holter monitor revealed a mean heart rate 61/min, 490 multifocal ventricular extrasystoles, a bilateral ventricular beat, and 110 supraventricular beats including aberrant beats. The patient was scheduled for outpatient follow-up on aspirin and warfarin treatment.

**DISCUSSION**

Myocardial noncompaction, a rare type of congenital cardiomyopathy may present separately or with other congenital malformations. Patients with MN have been reported to present with aortic valve stenosis, bicuspid aortic valve, dysplastic tricuspid valve, Ebstein anomaly, double orifice mitral valve, defect in coronary artery exit pathway, ventricular septal defect and pulmonary stenosis. Involvement of the left ventricle and apical segment in the left ventricle are frequently isolated. Prominent trabecular meshwork and deep perfused intertrabecular recesses are found in the involved segment. The first case of MN was described in 1984 and it has a prevalence of 0.05%. However, prevalen-
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...of the disease is considered to be higher due to recent increases in the number of patients with MN. In particular, apical hypertrophic cardiomyopathy and idiopathic dilated cardiomyopathy should be considered in the differential diagnosis.

Initial symptoms of isolated left ventricular MN are generally associated with left ventricular systolic and diastolic dysfunction. Patients may present with various clinical findings ranging from dyspnea to congestive heart failure. Treatment is similar to standard treatment for heart failure. Patients with MN who underwent biventricular pacing and cardiac transplantation have been reported. Although our patient had mild systolic dysfunction (EF 51%) and restrictive type diastolic dysfunction, his functional capacity was NYHA class I and had no complaints except dyspnea following excessive effort.

In addition to heart failure, conduction disorders are also commonly seen in patients with MN. Ventricular extrasystoles are the most seen conduction disorder. Others include, supraventricular premature beats, atrial fibrillation, supraventricular tachycardia, Wolff-Parkinson-White Syndrome, and complete AV block. Patients who received implantable cardioverter defibrillator due to ventricular tachycardia and ventricular fibrillation have been reported. A 24-hour Holter recording showed 490 multifocal ventricular premature beats and 110 supraventricular premature beats including aberrant beats, whereas malignant ventricular arrhythmia and AV block were not observed.

Systemic embolism is also one of the less commonly encountered complications. Two patients with myocardial infarction due to coronary embolism as well as superior mesenteric artery embolism were reported. Cerebral infarction was also detected in both patients with cerebral embolism. Our patient who did not have cerebral infarction was considered to have transient ischemic attack. The patient’s complaints improved within two hours and did not relapse. Treatment with anticoagulants was initiated as recommended for these patients.

Genetic factors have also been identified in the development of myocardial noncompaction. Xq28 G4.5 mutation, 5q deletion, 1q43 deletion, 11p15 mutation and mutation in the cardiac sarcomere were found in patients with familial inheritance. Genetic analysis was not performed on our patient and the examination of other family members did not reveal MN.

Echocardiography is the primary method used in the diagnosis of myocardial noncompaction. In addition, MN may be diagnosed with ventriculography, computed tomography and magnetic resonance imaging techniques. Diagnostic criteria for echocardiographic assessment are as follows: (i) presence of at least four apparent trabeculations and intertrabecular recesses; (ii) blood flow to intertrabecular recesses from the ventricles observed by color Doppler; (iii) typical double layer structure in the involved ventricular segment and a ratio of more than 2 of the end-systolic non-compacted subendocardial layer to the normal subepicardial layer. There were numerous trabeculations and recesses in the apex, apical, lateral, and inferior walls. Color Doppler also showed blood flow into the intertrabecular recesses. The left ventricle was apparently hypertrophic and the ratio of noncompaction/normal layer was found to be higher than 2. The patient was diagnosed with isolated left ventricular MN due to the absence of any comorbidity.

Although there are many different opinions on the long-term prognosis of the disease, majority of the patients with MN die from heart failure. Alehan reported that five of nine pediatric patients developed heart failure within a 5-year follow-up period, four of these patients were hospitalized, and two of them died. Oechslin et al. also followed 34 adults over 5 years. 12 of the patients died within this period. Sudden death was reported in six of the patients, while four died due to end-stage heart failure. The prognosis is generally worse in asymptomatic patients, in patients with additional congenital anomalies, with ventricular involvement of more than one segment and whose ratio of noncompaction/normal layer was more than three.

In conclusion, MN is a disease with a poor prognosis due to complications such as heart failure, ventricular arrhythmia and systemic embolism; therefore early diagnosis is vital. Late onset of complaints in the elderly may lead to late diagnosis. A careful echocardiographic examination may be helpful in the diagnosis of patients with MN and in initiating treatment during the early stage. A careful echocardiographic examination is critical for the evaluation of patients with central and peripheral embolism of unknown etiology.

REFERENCES


