

Japanese type cardiomyopathy without deep negative T-waves and with findings of preexcitation on ECG

EKG'de derin negatif T dalgaları izlenmeyen, preeksitasyon bulguları izlenen Japon tipi kardiyomiyopati

Dear Editor,

We present an apical cardiomyopathy case with normal T waves and findings of preexcitation on electrocardiogram (ECG). Apical type hypertrophic cardiomyopathy (HCM) is a specific variant of HCM. This disease is first described in Japan where the disease is more prevalent than in Western countries. It is generally accepted as a subgroup of nonobstructive HCM, but in some cases cavitory gradients may be detected. The disease is diagnosed with deep negative T waves ECG, spade-like cavity shape on left ventriculography and apical hypertrophy on echocardiography.

A Caucasian male patient presented to our outpatient clinic with the complaint of chest pain developing on exercise. Physical examination was normal, blood pressure measured as 110/80 mmHg and pulse rate was 74/min. Blood tests showed no anomaly. Electrocardiography showed sinus rhythm with incomplete right bundle branch block., T wave was positive in all derivations except for aVR.

D1, D2, aVF, V3, V4, V5 derivations showed short PR interval (0.12 msec) and delta wave representing preexcitation (Fig. 1, 2). Chest X-Ray revealed normal cardiac silhouette. Echocardiography showed normal cavity size in basal ventricular segments. Apical wall thickness was 28 mm (Fig. 3).

Left ventricular systolic function was normal (EF 65%). There was no valve pathology. Mid ventricular pulsed Doppler showed mild obstruction ($V_{max}=1.52$ m/second, $P_{max}=9.24$ mmHg) Aortic Doppler study showed normal flow (1.57 m/second).

Magnetic resonance imaging also correlated well with the echocardiographic findings (Fig. 4, 5). Exercise ECG test showed no sign

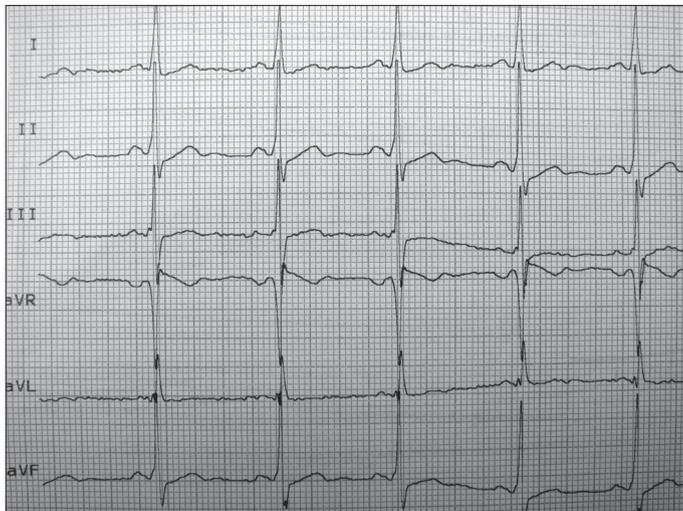


Figure 1. Short PR interval and delta waves are seen in leads D1-2 and aVF

of ischemia and the patient achieved 12 METs. The 24-hour Holter monitoring revealed very rare atrial extrasystoles (15-20 in total). There were no family history of cardiomyopathy and sudden death. Syncope or near syncope had not occurred. Moderate dose metoprolol (50 mg daily) was started and a follow-up program was planned. Coronary angiography was not planned.

In a study conducted by Morimoto et al. (1), spade-like configuration on ventriculography representing severe obstruction of left ventricular apex was shown in 43.9% of patients with giant negative T waves on ECG. Oval shape without left ventricular deformity was seen in 42.1% of patients. Spade like configuration was seen in 7% of patients with asymmetrical septal hypertrophy without showing giant negative T waves (1). As a result, giant negative T waves represented apical HCM less than in half of patients. Although giant negative T waves show apical HCM at higher rates, regression of T waves was reported in two cases. After 10 years of follow-up, one of the patients developed

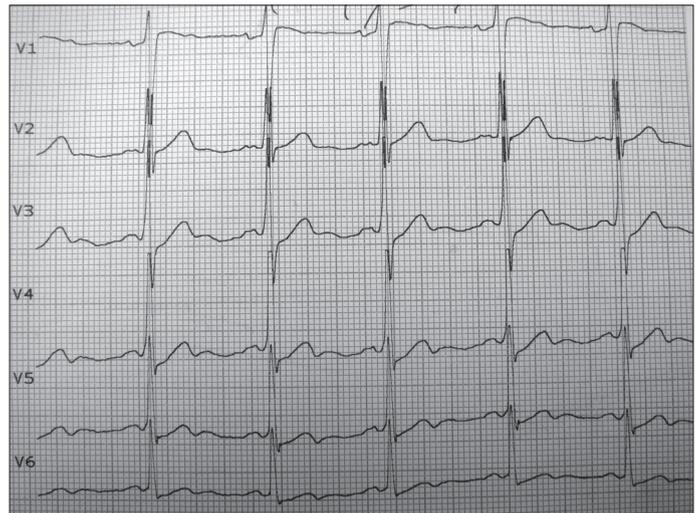


Figure 2. Short PR interval and delta waves are evident in leads V3-4-5. Positive T waves and U waves are also seen in precordial derivations



Figure 3. Apical 4-chamber window shows apical thickening of myocardium and normal thickness at basal myocardial segments

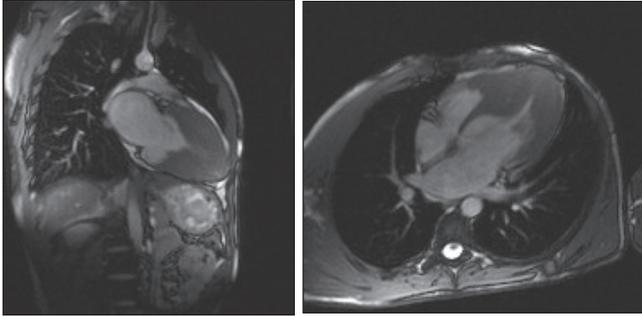


Figure 4 and 5. Transverse frontal cross-sectional magnetic resonance images. Normal basal and thickened apical segments

coronary artery disease and T waves normalized. In the other case, dilated cardiomyopathy developed and T waves regressed (2). Amplitude of giant T waves is far more correlated with the thickness of all myocardium than the thickness of sole apical myocardium (3). Horita at all. (4) reported that decreasing in the amplitude or disappearing T wave negativity is correlated with decreasing systolic function. In literature, some cases with hypertrophy restricted only by apical lateral, apical anterior or apical septum were reported. In these cases, spade-like ventriculogram do not occur (5).

Deep negative T waves may not be seen in all cases. T wave negativity may decrease or disappear with progression of disease. These changes may be early findings of other concomitant diseases (i.e coronary artery disease). On the other hand, as shown in our case, apical HCM may show normal T waves without concomitant diseases. As far as we explored, this is the first case report of Yamaguchi Syndrome without deep negative T waves and with findings of preexcitation in literature.

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A newborn with pentalogy of Cantrell and pulmonary hypoplasia

Bir yenidoğanda Cantrell pentalojisi ve pulmoner hipoplazi

To the Editors,

Ectopia cordis (EC) is rare, often fatal abnormality characterized by complete or partial displacement of heart out of thoracic cavity and occurs in 5.5-7.9 per 1 million live births (1). Four types of EC were defined: thoracic, abdominal, thoraco-abdominal and cervical (1, 2). If EC associated with deficiency of anterior diaphragm, midline supraumbilical abdominal wall defect, defect in diaphragmatic pericardium, various congenital intracardiac abnormalities, and defect of lower sternum, called pentalogy of Cantrell (PC) with an estimated incidence of 1/65.000- 1/200.000 live births (3). Chromosomal abnormalities, cleft lip-palate, pulmonary hypoplasia (PH), in some patients cranial, gastrointestinal, and renal abnormalities were reported in association (1, 2). We present a newborn with PC and PH.

Term female baby was born by vaginal delivery with irregular antenatal follow-up, with no history of drug abuse and consanguinity. She was transferred to our hospital two hours after birth because of extrathoracic heart. On the physical examination down slanted palpebral fissures, hypertelorism, micrognathia, short neck, extrathoracic heart with visible pulsations, supraumbilical abdominal wall defect, omphalocele between sternum and umbilicus were observed (Fig. 1). Echocardiography revealed four chambers; atrioventricular cavities on the left side were hypoplastic while atrioventricular cavities on the right side were dilated. There was single cardiac outlet [truncus arteriosus (TA)] originating from the ventricle on the right side, secundum atrial septal defect (ASD) and ventricular septal defect (VSD). No other congenital cranial, renal or gastrointestinal anomalies were present. Chromosomal analysis was 46, XX. She died on the 5th day before performing surgical procedure.

Autopsy findings; 6x4x2 cm heart mass with four chambers apex lying caudally was visible in front of the sternum. Left atrioventricular-atrioventricular valvular hypoplasia, ASD, VSD, TA originating from right ventricle, absent pericardium (naked heart), coarse trabeculae of right ventricle (L-loop position), four cusps of truncal valve were observed. Pulmonary veins meet inferior vena cava (VC) and inferior- superior VC drain to right atrium. Omphalocele was 5x5 cm, sternum was 3cm, xiphoid process was not present and both lungs were hypoplastic (2 lobes on the right, 3 lobes on the left with primitive upper lobe).

Toyama (4) made classification of PC. In class 1, cases have all five defects. Class 2 patients show four defects including intracardiac-



Figure 1. Dysmorphic face, ectopia cordis, lower sternal and supraumbilical abdominal wall defects of the patient