Apical Hypertrophic Cardiomyopathy Associated with High Degree Atrioventricular Block: Case Report

Yüksek Dereceli Atriyoventriküler Blokla Gelen Apikal Hipertrofik Kardiyomiyopati

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ABSTRACT Apical hypertrophic cardiomyopathy (HCM) is a specific variant of HCM. Aryhythmias are markers that predict poor prognosis. In the present report we describe a 54-year-old female patient who was admitted to our hospital with dizziness and syncope. Electrocardiography showed high degree atrioventricular AV block. Apical hypertrophy was detected on the transthorasic echocardiography and spades like configuration on the left ventriculagraphy. To the our knowledge, this is one of the scarce cases of apical hypertrophic cardiomyopathy (Yamaguchi disease) with high degree atrioventricular block treated with a permanent cardiac pacemaker implantation.

Key Words: Cardiomyopathy, hypertrophic, atrioventricular block

ÖZET Apikal hipertrofik kardiyomiyopati (HKM) HKM'nin spesifik bir alt tipidir. Aritmilerin varlığı hastada kötü prognoz belirtisidir. Bu olgu sunumunda, hastanemize baş dönmesi ve bayılma şikâyetiyle başvuran 54 yaşında kadın hastadan bahsedilmektedir. Elektrokardiyografide yüksek dereceli atriyoventriküler (AV) blok izlenmiştir. Transtorasik ekokardiyografide hipertrofi ve ventrikülografide maça ası görünümü tespit edilmiştir. Bu olgu, kardiyak kalıcı pil ile tedavi edilmiş apikal HKM (Yamaguchi hastalığı) yüksek dereceli AV blok birlikteliğine verilebilecek az sayıdaki örneklerden bir tanesidir.

Anahtar Kelimeler: Hipertrofik kardiyomiyopati, atriyoventriküler blok

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Apical hypertrophic cardiomyopathy (HCM) is a specific variant of HCM. Arrhythmias are markers of this disease that predict poor prognosis. We report a female patient presenting with high degree AV block and in whom apical HCM was incidentally found. Our present report is one of the scarce cases of apical HCM (Yamaguchi disease) with high degree atrioventricular block treated with a permanent cardiac pacemaker implantation.

CASE REPORT

A 54-year-old female patient was admitted to our hospital with dizziness and syncope. Her past medical history revealed hypertension and hyperlipidemia. The patient had no complaint of angina, dyspnea or palpitation. She was treated with candesartan and rosuvastatin.

Her physical examination was normal, blood pressure and heart rate were 120/80 mmHg and 57 bpm, respectively. Electrocardiography (ECG) demonstrated a high degree AV block, left ventricular hypertrophy criteria and negative T waves at all derivations (Figure 1). ECG findings were consistent with atypical Wenckebach secondary AV block with 6:5 AV conduction. On 2-dimensional echocardiography (ECO), an apical 4-chamber view of the left ventricle (LV) revealed hypertrophy of the apex. The thickness of the LV apex was 2.1 cm, the interventricular septum and posterior wall thicknesses were 1.0 and 0.9 cm, respectively and no significant gradient was detected in the LV cavity (Figure 2). The coronary angiography was normal and an ace-of-spades like configuration was demonstrated on the left ventriculogram in right anterior oblique position (RAO) (Figure 3). The patient was treated with a DDR permanent cardiac pacemaker implantation. The written informed consent was taken from the patient.

DISCUSSION

In this article we describe an apical HCM patient admitted to our hospital with high degree AV block (atypical Wenckebach secondary AV block with 6:5 AV conduction) treated with a permanent cardiac pacemaker implantation.

HCM is defined as a significant myocardial hypertrophy in the absence of an identifiable cause. Apical HCM which has been first described in Japan by Yamaguchi is one of the HCM subtypes.[1] Various mutations at cardiac muscle proteins like myosin heavy chain, cardiac troponin T, tropomyosin, myosin binding protein C have been defined. It constitutes 25% of cases of HCM in Japan; however with an only 1% to 2% of incidence in the HCM patients in the non-Japanese population. It is characterized by hypertrophy that is confined to the apex which causes a ace of spades like configuration in RAO on the left ventriculogramphy. Giant negative T waves and tall R waves in the left precordial leads are the ECG hallmarks of the Japanese form of apical hypertrophy as seen on the ECG in our patient. ECO demonstrates localized apical hypertrophy with at least 15 mm apical wall thickness or a ratio of maximal apical to posterobasal thickness greater than 1.5.[2-4] Arrhythmias are markers of this disease that predict poor prognosis and sudden cardiac death is the most leading cause

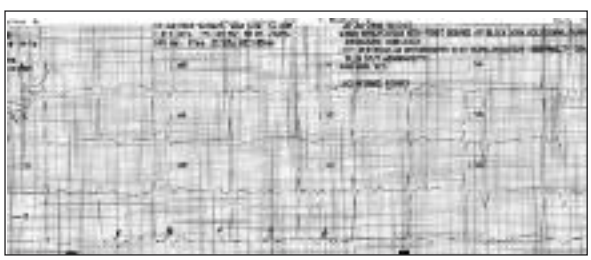

FIGURE 1: Electrocardiography demonstrated third degree atrioventriculer block (arrow), left ventricle hypertrophy criteria and negative T waves at all derivations.

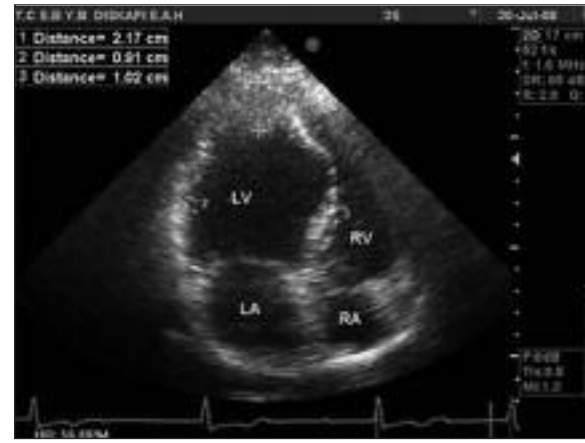

FIGURE 2: On 2-dimensional echocardiography, an apical 4-chamber view of the left ventricle (LV) revealed hypertrophy of the apex.

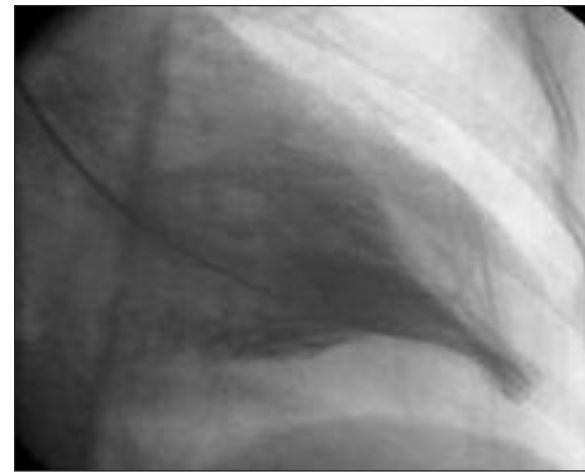

FIGURE 3: Ace-of-spades like configuration was demonstrated on the left ventriculography in right anterior oblique position (RAO).

of death in young athletes particularly.[5,6] But in this type of HCM prognosis is rather good and severe arrhythmias, sudden cardiac death are less common than the other types.[7,8] First degree AV block is the most seen block type on the ECG.[9] Pathophysiology of the AV block is not completely understood. Kaneshige et al observed degeneration and fibrous replacement of the AV conduction system in cats with AV block. Also chondrometaplastic and osseus lesions surrounding the central fibrous body have been found compressing the conduction fibers.[10]

Complete AV block with other types of hypertrophic cardiomyopathy has been declared by many authors.[11,12] We preferred to introduce this case as one of the scarce cases of apical HCM (Yamaguchi disease) with high degree atrioventricular block treated with a permanent cardiac pacemaker implantation.

REFERENCES

1. Ishiwata S, Nishiyama S, Nakanishi S, Nishimura S, Yanagishita Y, Kato K, et al. Natural history of 82 patients with hypertrophic cardiomyopathy: follow-up for over ten years. J Cardiol 1991;21(1):61-73.
2. Yamaguchi H, Ishimura T, Nishiyama S, Nagasaki F, Nakanishi S, Takatsu F, et al. Hypertrophic nonobstructive cardiomyopathy with giant negative T waves (apical hypertrophy): ventriculographic and echocardiographic features in 30 patients. Am J Cardiol 1979;44(3):401-12.
3. Bayrak F, Kahveci G, Mutlu B, Değertekin M, Başaran Y, Demirtaş E. [Demographic, clinical and echocardiographic features of Turkish hypertrophic cardiomyopathy patients]. Turkiye Klinikleri J Cardiovasc Sci 2007;19(1):7-11.
4. Tezcan H, Fak AS, Okucu M, Oktay A. [Apical hypertrophic cardiomyopathy: case report]. Turk Kardiyol Dern Arş 1996;24(9):565-7.
5. Carter D, Pokroy R, Barenboim E, Azaria B, Goldstein L. Apical hypertrophic cardiomyopathy and arrhythmia in military pilots. Aviat Space Environ Med 2006;77(4):459-61.
6. Ridjab D, Koch M, Zabel M, Schultheiss HP, Morguet AJ. Cardiac arrest and ventricular tachycardia in Japanese-type apical hypertrophic cardiomyopathy. Cardiology 2007;107 (2):81-6.
7. Webb JG, Sasson Z, Rakowski H, Liu P, Wigle ED. Apical hypertrophic cardiomyopathy: clinical follow-up and diagnostic correlates. J Am Coll Cardiol 1990;15(1):83-90.
8. Zack PM, Blausey WL. Symptomatic Japanese-type apical hypertrophic cardiomyopathy in a non-Asian patient: favorable response to treatment with verapamil. Am Heart J 1994;128 (3):613-6.
9. Reddy SVK, Satish OS, Patnaik AN, Dubey BK, Rao M, Azam W, et al. Apical hypertrophic cardiomyopathy: Clinico-angiographic profile. Indian Heart Journal 2001;53(5):Article No. 240.
10. Kaneshige T, Machida N, Itoh H, Yamane Y. The anatomical basis of complete atrioventricular block in cats with hypertrophic cardiomyopathy. J Comp Pathol 2006;135(1): 25-31.
11. Wang DW, Deng YB. Hypertrophic cardiomyopathy complicated by severe bradycardias: a pedigree report. Clin Cardiol 2002;25(2):76-80.
12. Doven O, Cicek D, Pekdemir H, Camsari A, Parmaksiz T, Cin GV, et al. Abnormal His-Purkinje system conduction leading to complete atrioventricular block in patients with hypertrophic cardiomyopathy: a report of 3 cases. Jpn Heart J 2004;45(2):347-52.