An overlooked diagnosis on transthoracic echocardiography: apical hypertrophic cardiomyopathy

Transtorasik ekokardiyografide gözden kaçan bir tanı: Apikal hipertrofik kardiyomiyopati

Ömer Yiğiner, M.D., Bekir Yılmaz Cingözbay, M.D., Ömer Uz, M.D., Bekir Sıtkı Cebeci, M.D.

Department of Cardiology, Haydarpaşa Training Hospital, Gülhane Military Medical Academy, İstanbul

Apical hypertrophic cardiomyopathy (HCM) may be overlooked during routine echocardiography. A 54-year-old male patient with dyslipidemia and positive family history presented with atypical chest pain. The 12-lead electrocardiogram showed increased QRS voltage and deep T-wave inversions in precordial leads. Transthoracic echocardiography performed with poor echocardiographic windows demonstrated normal findings. Coronary angiography showed normal coronary arteries. Left ventriculography revealed a spade-like deformity (ace of spades), typical for apical HCM. Cardiac magnetic resonance imaging confirmed the diagnosis.

Key words: Cardiomyopathy, hypertrophic; catheterization; echocardiography; electrocardiography; magnetic resonance imaging.

Apikal hipertrofik kardiyomiyopati rutin ekokardiyografi sırasında gözden kaçabilir. Dislipidemisi ve pozitif aile öyküsü olan 54 yaşında erkek hasta atipik göğüs ağrısıyla başvurdu. On iki derivasyonlu elektrokardiyografide prekordiyal artmış QRS voltajı ve derin T-dalga negatiflikleri izlendi. Yetersiz eko penceresi nedeniyle sınırlı olarak yapılan transtorasik ekokardiyografide anormal bulguya rastlanmadı. Koroner anjiyografide koroner arterler normal izlendi. Sol ventrikülografide ise, apikal hipertrofik kardiyomiyopati için tipik olan maça ası görüntüsü saptandı. Tanı kardiyak manyetik rezonans görüntüleme ile doğrulandı.

Anahtar sözcükler: Kardiyomiyopati, hipertrofik; kateterizasyon; ekokardiyografi; elektrokardiyografi; manyetik rezonans görüntüleme.

Apical hypertrophic cardiomyopathy (HCM) is an uncommon disease which is inherited autosomal dominantly in about 50% of the cases.[1] In Japan, this apical variant accounts for about 25% of HCM patients, compared to 1-2% in other parts of the world.[1,2] Although the tool of choice is the echocardiographic examination for the diagnosis of HCM, apical HCM may be occasionally overlooked during routine echocardiography.[3]

CASE REPORT

A 54-year-old male having cardiovascular risk factors of dyslipidemia and positive family history was admitted to our department with atypical chest pain. The 12-lead electrocardiogram (ECG) showed increased QRS voltage and deep T-wave inversions in precordial leads (Fig. 1). Clinical examination was unremarkable. Limited transthoracic echocardiographic examination due to poor echocardiographic windows demonstrated normal findings with an ejection fraction of 64%. We decided to perform coronary angiography in order to clarify the findings on his electrocardiogram. Angiography showed normal coronary arteries without any stenosis in the coronary tree. Left ventriculography revealed a spade-like deformity (ace of spades), typical for apical HCM at diastole and marked obliteration of the left ventricular apex at systole (Fig. 2). Cardiac magnetic resonance imaging (MRI) also demonstrated marked thickening of the left ventricular apex, confirming apical HCM (Fig. 3).

DISCUSSION

Patients with apical HCM may have little or no symptoms.[4] Large negative T-waves and increased QRS

Received: March 23, 2009 *Accepted:* April 16, 2009

Correspondence: Dr. Ömer Yiğiner. GATA Haydarpaşa Eğitim Hastanesi, Kardiyoloji Kliniği, 34668 İstanbul, Turkey
Tel: +90 312 - 325 89 74 e-mail: oyiginer@yahoo.com

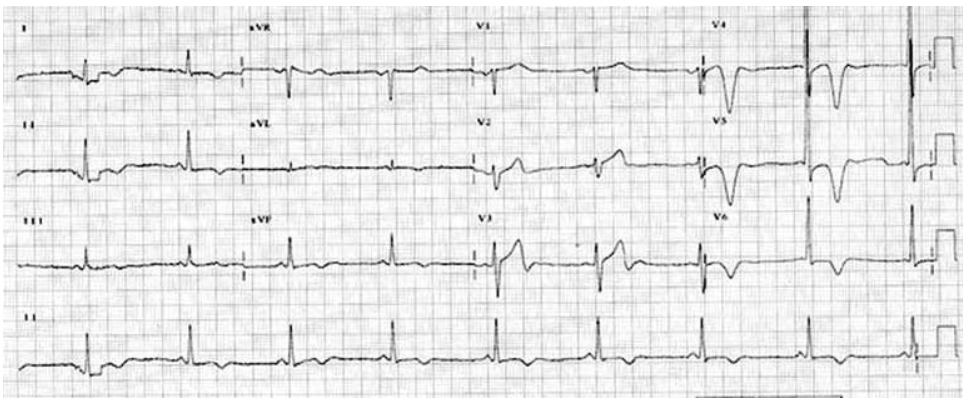

Figure 1. The electrocardiogram showing increased QRS voltage and deep T-wave inversions in the leads V3-6.

voltage in leads V3-6 are characteristic ECG findings for apical HCM.[5] Although echocardiography is the tool of choice for the diagnosis of all types of HCM, apical HCM may be overlooked in patients with poor echocardiographic windows, as in our case.[3] Cardiac MRI may be used especially if the apex is difficult to visualize on transthoracic echocardiography.[6] On catheterization, apical HCM has a distinctive left ventricular appearance, "ace of spades", with obliteration of the apex during systole.[5,7] Apical HCM generally has a more benign course compared to other variants of HCM. Although it usually does not lead to outflow tract obstruction like other variants, it may cause arrhythmias.[1] Therefore, it may be necessary to identify patients at risk for sudden death. In an asymptomatic patient with no evidence for myocardial ischemia or significant arrhythmia, no specific therapy is needed.[1]

In conclusion, a routine echocardiogram without contrast may not exclude apical HCM in patients with unexplained repolarization abnormalities on precor-

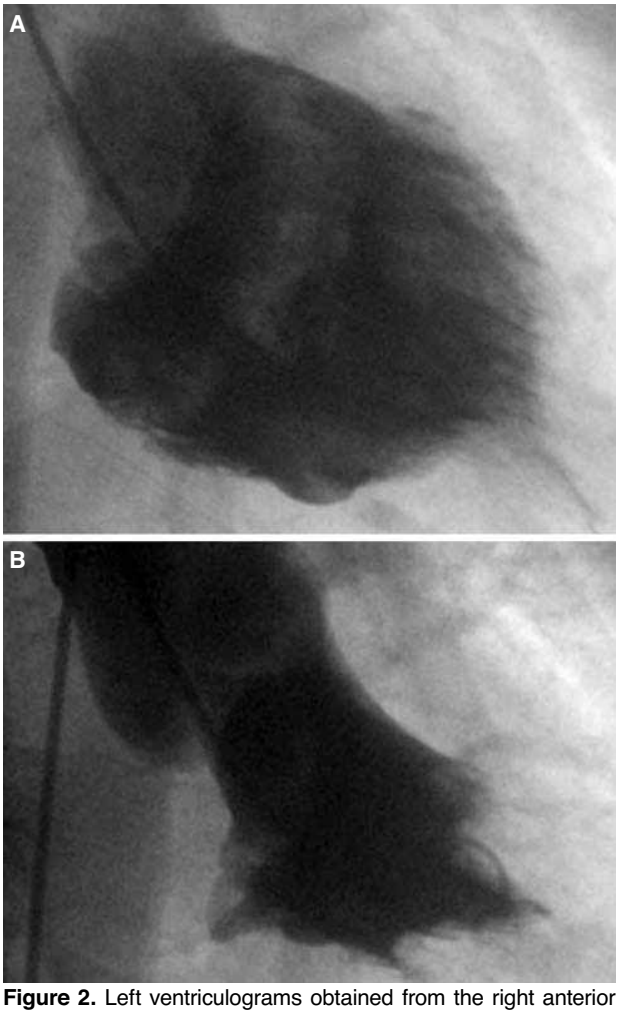

Figure 2. Left ventriculograms obtained from the right anterior oblique projection. **(A)** The typical appearance of "ace of spades" at diastole and **(B)** marked obliteration of the left ventricular apex at systole.

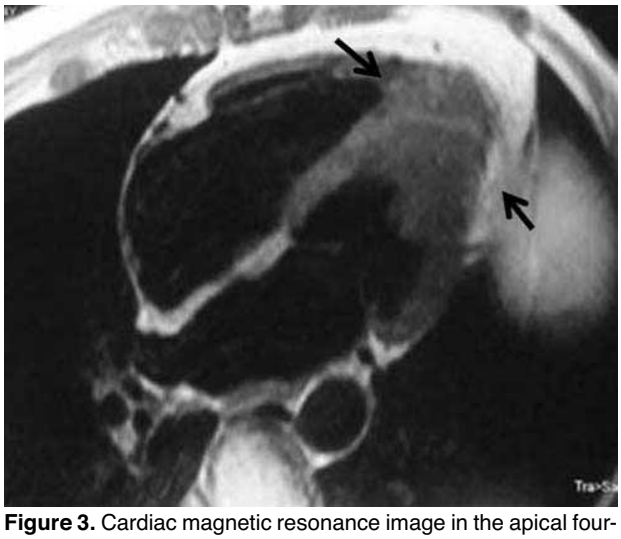

Figure 3. Cardiac magnetic resonance image in the apical four-chamber view showing marked thickening of the left ventricular apex.

dial leads. Further imaging with cardiac MRI or contrast echocardiography may be required.

REFERENCES

1. Ting P, Gunasegaran K, Teo WS. Electrocardiographical case. Asymptomatic patient with deep T-wave inversions. Singapore Med J 2007;48:586-8.
2. Cam FS, Güray M. Hypertrophic cardiomyopathy: pathological features and molecular pathogenesis. Anadolu Kardiyol Derg 2004;4:327-30.
3. Moon JC, Fisher NG, McKenna WJ, Pennell DJ. Detection of apical hypertrophic cardiomyopathy by cardiovascular magnetic resonance in patients with non-diagnostic echocardiography. Heart 2004; 90:645-9.
4. Sakamoto T. Apical hypertrophic cardiomyopathy (apical hypertrophy): an overview. J Cardiol 2001;37 Suppl 1:161-78.
5. Saygı S, Türk U, Özdoğan Ö, Zoghi M. Apical hypertrophic cardiomyopathy coexistent with a myocardial bridging. Anadolu Kardiyol Derg 2004;4:370-1.
6. Gaudio C, Pelliccia F, Tanzilli G, Mazzarotto P, Cianfrocca C, Marino B. Magnetic resonance imaging for assessment of apical hypertrophy in hypertrophic cardiomyopathy. Clin Cardiol 1992;15:164-8.
7. Yüksel UC, Kursaklıoğlu H, Çelik T. Apical hypertrophic cardiomyopathy with giant left atrium. Arq Bras Cardiol 2007;88:e47.