

Overlook Could be Catastrophic; Mid-Ventricular Obstructive Hypertrophic Cardiomyopathy Associated with an Apical Aneurysm: Case Report

Gözden Kaçırma Ölümcül Olabilir; Apikal Anevrizma ile İlişkili Ventrikül Ortası Darlığı Olan Hipertrofik Kardiyomiyopati

Erkan BAYSAL,^a
Barış YAYLAK,^a
Bernas ALTINTAŞ,^a
Önder BİLGE^a

^aDepartment of Cardiology,
Diyarbakır Gazi Yaşargil Training and
Research Hospital,
Diyarbakır

Geliş Tarihi/Received: 12.12.2015
Kabul Tarihi/Accepted: 10.07.2016

Yazışma Adresi/Correspondence:
Erkan BAYSAL
Diyarbakır Gazi Yaşargil Training and
Research Hospital,
Department of Cardiology, Diyarbakır,
TURKEY/ TÜRKİYE
dr.erkans.baysal@hotmail.com

ABSTRACT Mid-ventricular obstructive hypertrophic cardiomyopathy (MVOHCM) is a rare type of cardiomyopathy that can be accompanied by apical aneurysm. MVOHCM was demonstrated to have caused cardiac arrhythmias and sudden cardiac death. The identification of high-risk patients with hypertrophic cardiomyopathy (HCM) for primary prevention of sudden cardiac death (SCD) remains a challenging issue, since major risk factors sometimes lack specificity. We report the case of a patient who presented with ventricular fibrillation, ST-segment elevation on electrocardiography, and cardiac-enzyme elevation, in the presence of normal coronary arteries. Continuous-wave Doppler echocardiography revealed a pressure gradient between the apical and basal chambers of the left ventricle.

Key Words: Death, sudden, cardiac; cardiomyopathy, hypertrophic; stroke

ÖZET Ventrikül ortası darlığı olan hipertrofik kardiyomiyopati (VODOHK) apikal anevrizmanın eşlik ettiği nadir bir kardiyomiyopati tipidir. VODOHK'in kardiyak aritmilerin ve ani kardiyak ölümün sebebi olduğu gösterilmiştir. Majör risk faktörlerinin zayıf özgünlüğünden dolayı hipertrofik kardiyomiyopati hastalarda ani kardiyak ölümün birincil korumasında yüksek riskli hastaların belirlenmesi tartışmalı bir konu olarak devam etmektedir. Biz bu vakada ventriküler fibrilasyon ile başvuran, elektrokardiyogramda ST-segment elevasyonu olan, troponin yüksekliği saptanan ve normal koroner arterler tespit edilen bir hastayı derledik. Devamlı-dalga doppler ekokardiyografide sol ventrikül bazal ve apikal bölümleri arasında basınç gradiyenti izlendi.

Anahtar Kelimeler: Ölüm, ani, kardiyak; kardiyomiyopati, hipertrofik; inme

Türkiye Klinikleri J Case Rep 2016;24(4):326-8

Mid-ventricular obstructive hypertrophic cardiomyopathy (MVOHCM) is a rare type of cardiomyopathy, associated with apical aneurysm formation in some cases.¹⁻⁴ This type of hypertrophic cardiomyopathy (HCM) is strongly related to cardiac death associated with cardiac arrhythmias.^{5,6} Herein, we describe a case of MVOHCM in association with stroke and sudden cardiac death.

CASE REPORT

A 50 years-old man presented with ventricular fibrillation has been admitted to another hospital. After successful defibrillation in that hospital, he was transferred to the coronary care unit of our hospital because of ad-

doi: 10.5336/caserep.2015-48968

Copyright © 2016 by Türkiye Klinikleri

vanced investigation. There was family history of sudden death. For the previous 3 years, he had experienced left hemiplegia due to stroke, for which he had been prescribed warfarin. At the time of initial medical treatment, he had been diagnosed hypertrophic obstructive cardiomyopathy in the mid portion of the interventricular septum.

After successful defibrillation, the patient's electrocardiogram showed obvious ST segment elevation in leads V3 through V6 (Figure 1). Laboratory findings are normal. Chest radiography did not show cardiomegaly or pulmonary congestion. We performed coronary angiography and found normal coronary arteries. Echocardiography disclosed mid-ventricular obstruction during systole, and an apical aneurysm. There was a insignificant pressure gradient (25 mmHg) between the basal and apical sites of the left ventricle (LV) at rest, Using continuous wave Doppler echocardiography, the pressure gradient was measured as 40 mmHg after valsalva maneuver. There was paradoxical jet flow from the apical aneurysm to the left ventricular outflow tract during early diastole (Figure 2).

Implantation of a cardioverter-defibrillator was recommended. After treatment with an oral β -blocker and implantation of cardioverter-defibrillator the patient experienced no episodes of ventricular tachycardia; but his electrocardiographic and wall-motion abnormalities persisted during the 3-month follow-up period.

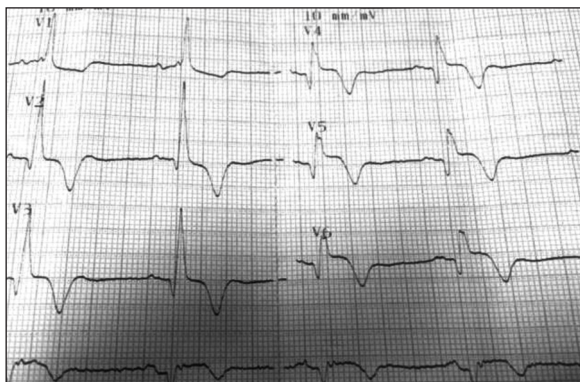


FIGURE 1: ECG showed obvious ST segment elevation in leads V3 through V6.

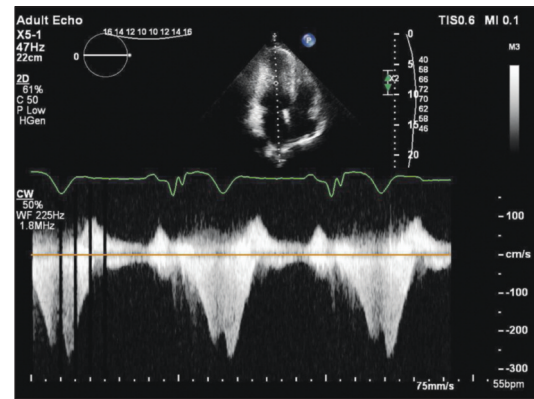


FIGURE 2: There was paradoxical jet flow from the apical aneurysm to the left ventricular outflow tract during early diastole.

DISCUSSION

MVOHCM is a rare form of hypertrophic cardiomyopathy, characterized by the presence of a pressure gradient between the apical and basal chambers of the left ventricle.^{1,4,5,7-10} It is frequently associated with an apical aneurysm without significant atherosclerotic coronary artery disease. The mechanisms responsible for apical aneurysm formation are not well understood. But it has been suggested that apical aneurysm may be secondary to the increased afterload and high apical pressure arising from the midventricular obstruction seen in the degenerative process of hypertrophic cardiomyopathy.^{2,11} Other possible causes of aneurysm formation are small vessel disease with decreased coronary flow reserve, coronary stenosis due to an increased wall stress in the hypertrophied myocardial segment, decreased coronary perfusion pressure due to the mid-ventricular obstruction, coronary spasm, and decreased capillary/myocardial fiber ratio.¹²

Approximately 2% of patients with hypertrophic cardiomyopathy present with apical LV aneurysms. Incidence of apical thrombus is unknown in HCM, although embolic events could be first presentation in patients with HCM, such as our case.

The clinical course of our HCM patients with LV apical aneurysms was variable but overall proved to be largely unfavorable. Large apical aneurysm in patients with HCM to likely represent

an arrhythmogenic substrate for the generation of malignant ventricular tachyarrhythmias.

Generally, beta blockers are the first choice of treatment for patients with subaortic obstructive hypertrophic cardiomyopathy, but the optimal treatment for MVOHCM has not yet been established. Dualchamber pacing and percutaneous myocardial ablation have been proposed as non-surgical treatments, but their long-term prognosis and procedural safety await further observation with a large patient population.^{13,15} The surgical treatment of midventricular hypertrophic obstructive cardiomyopathy is described in literature. Kunkala et al. described the results of a transapical approach, noting that this option allows an excellent approach for myectomy, as well as for the re-

lief of the intraventricular gradient and associated symptoms without any complications related to the apical incision were observed with a five-year survival similar to that expected in the general population (95% vs. 97%).¹⁶ Another surgical option is successful septal myectomy plus excision of the aneurysm.

HCM patients with LV apical aneurysm represent a previously underrecognized but clinically important subset within the broad HCM disease spectrum. In recent study show that MVOHCM patients with an aneurysm had a higher probability to progress to end-stage HCM. Clinical recognition of this phenotype often requires a high index of suspicion. In the present case, overlook of diagnosis led to catastrophic event.

REFERENCES

- Harada K, Shimizu T, Sugishita Y, Yao A, Suzuki J, Takenaka K, et al. Hypertrophic cardiomyopathy with midventricular obstruction and apical aneurysm: a case report. *Jpn Circ J* 2001;65(10):915-9.
- Figali S, Krajcer Z, Edelman S, Leachman RD. Progression of hypertrophic cardiomyopathy into a hypokinetic left ventricle: higher incidence in patients with midventricular obstruction. *J Am Coll Cardiol* 1987;9(2):288-94.
- Webb JG, Sasson Z, Pakowski H, Liu P, Wigle ED. Apical hypertrophic cardiomyopathy: clinical follow-up and diagnostic correlates. *J Am Coll Cardiol* 1990;15(1):83-90.
- Akutsu Y, Shinozuka A, Huang TY, Watanabe T, Yamada T, Yamanaka H, et al. Hypertrophic cardiomyopathy with apical left ventricular aneurysm. *Jpn Circ J* 1998;62(2):127-31.
- Tse HF, Ho HH. Sudden cardiac death caused by hypertrophic cardiomyopathy associated with midventricular obstruction and apical aneurysm. *Heart* 2003 89(2):178.
- Furushima H, Chinushi M, Iijima K, Sanada A, Izumi D, Hosaka Y, et al. Ventricular tachyarrhythmia associated with hypertrophic cardiomyopathy: incidence, prognosis, and relation to type of hypertrophy. *J Cardiovasc Electrophysiol* 2010;21(9):991-9.
- Lin CS, Chen CH, Ding PY. Apical hypertrophic cardiomyopathy mimicking acute myocardial infarction. *Int J Cardiol* 1998;64(3):305-7.
- Ito N, Suzuki M, Enjoji Y, Nakamura M, Namiki A, Hase H, et al. [Hypertrophic cardiomyopathy with mid-ventricular obstruction complicated with apical left ventricular aneurysm and ventricular tachycardia: a case report]. *J Cardiol* 2002;39(4):213-9.
- Sato A, Shiga T, Kawarai H, Matsuda N, Kimura F, Sakai F, et al. Apical hypertrophic cardiomyopathy with aneurysmal degeneration of the apex. *Heart Vessels* 2003;18(1):53-4.
- Tengiz I, Ercan E, Türk UO. Percutaneous myocardial ablation for left mid-ventricular obstructive hypertrophic cardiomyopathy. *Int J Cardiovasc Imaging* 2006;22(1):13-8.
- Matsubara K, Nakamura T, Kuribayashi T, Azuma A, Nakagawa M. Sustained cavity obliteration and apical aneurysm formation in apical hypertrophic cardiomyopathy. *J Am Coll Cardiol* 2003;42(2):288-95.
- Sato Y, Matsumoto N, Matsuo S, Yoda S, Kunimoto S, Saito S. Mid-ventricular hypertrophic obstructive cardiomyopathy presenting with acute myocardial infarction. *Tex Heart Inst J* 2007;34(4):475-8.
- Maron BJ, Nishimura RA, McKenna WJ, Rakowski H, Josephson ME, Kievit RS. Assessment of permanent dual-chamber pacing as a treatment for drug-refractory symptomatic patients with obstructive hypertrophic cardiomyopathy. A randomized, double-blind, crossover study (M-PATHY). *Circulation* 1999;99(22):2927-33.
- Watanabe H, Kibira S, Saito T, Shimizu H, Abe T, Nakajima I, et al. Beneficial effect of dual-chamber pacing for a left mid-ventricular obstruction with apical aneurysm. *Circ J* 2002;66(10):981-4.
- Seggewiss H, Faber L. Percutaneous septal ablation for hypertrophic cardiomyopathy and mid-ventricular obstruction. *Eur J Echocardiogr* 2000;1(4):277-80.
- Kunkala MR, Schaff HV, Nishimura RA, Abel MD, Sorajja P, Dearani JA, et al. Transapical approach to myectomy for midventricular obstruction in hypertrophic cardiomyopathy. *Ann Thorac Surg* 2013;96(2):564-70.