

Case Report

Left Ventricular Apical Aneurysm and Ventricular Tachycardia in a Patient With Bicuspid Aortic Valve and Hypertrophic Cardiomyopathy

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ABSTRACT

A 36-year-old man, who had a history of aortic valve replacement 8 years previously because of severe aortic stenosis and bicuspid aortic valve, presented to the emergency department with a hemodynamically unstable ventricular tachycardia. Echocardiography showed an asymmetrical left ventricular hypertrophy and a normal functioning prosthetic valve with a negligible transvalvular gradient and no evidence of patient-prosthetic mismatch.

Cardiac magnetic resonance imaging revealed left ventricular hypertrophy with an apical aneurysm, diffuse late gadolinium enhancement, and a midcavitary obstruction, all in favor of hypertrophic cardiomyopathy.

Ventricular tachycardia ablation was done via the trans-septal approach, and an implantable cardioverter-defibrillator was inserted. (*Iranian Heart Journal 2018; 19(4): 58-61*)

KEYWORDS: Hypertrophic cardiomyopathy, Aortic stenosis, Bicuspid aortic valve, Ventricular tachycardia ablation, Cardiac magnetic resonance imaging

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Hypertrophic cardiomyopathy (HCM), a complex heart disease with an autosomal dominant hereditary pattern, is manifested with a wide spectrum of morphological and clinical findings. The prevalence of this disease is about 0.2%, and it exhibits a very diverse pattern of clinical progress. Some patients are asymptomatic in the course of their life, while others suffer sudden cardiac death.¹⁻³

Bicuspid aortic valve (BAV) is considered the most prevalent congenital heart defect in adults, with a prevalence rate of about 0.5% to 2%. A

previous study reported a prevalence rate of about 0.9% for BAV coinciding with HCM, similar to the general population.⁴

We herein describe a male patient with a history of aortic valve replacement due to severe aortic stenosis and BAV, who presented with ventricular tachycardia (VT). Based on the findings of cardiac magnetic resonance imaging (CMR), HCM was considered for the patient.

Case Presentation

Our patient was a 36-year-old man, who had undergone aortic valve replacement 8 years

previously because of symptomatic severe valvular aortic stenosis and BAV, without concomitant sub-or supra-valvular stenosis. There was no suspicion of HCM at the time of surgery and in the routine postoperative follow-ups.

The patient presented to the emergency department with palpitations and cold sweating

in a hemodynamically compromised state. Electrocardiography (ECG) showed a wide QRS tachycardia with a right bundle branch block pattern, transition in lead V₂, and a superior axis, indicating VT with a left ventricular (LV) origin (Fig. 1).

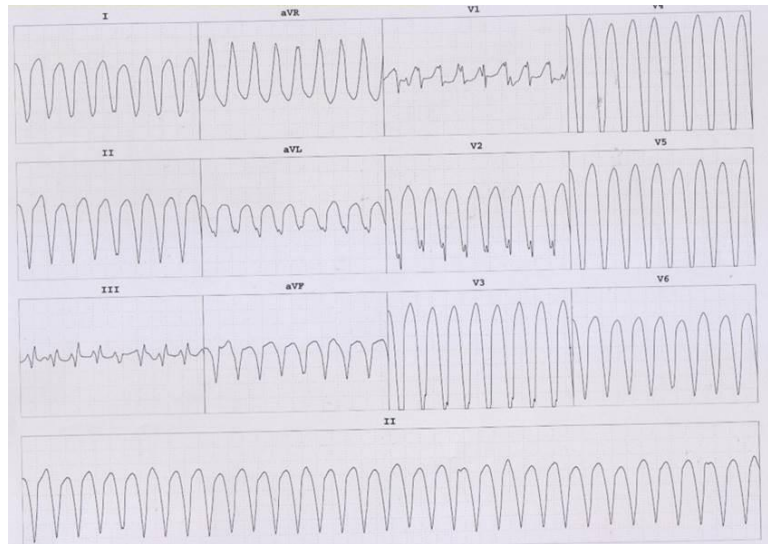
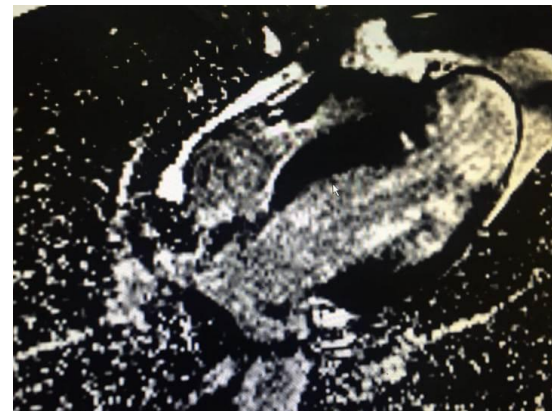
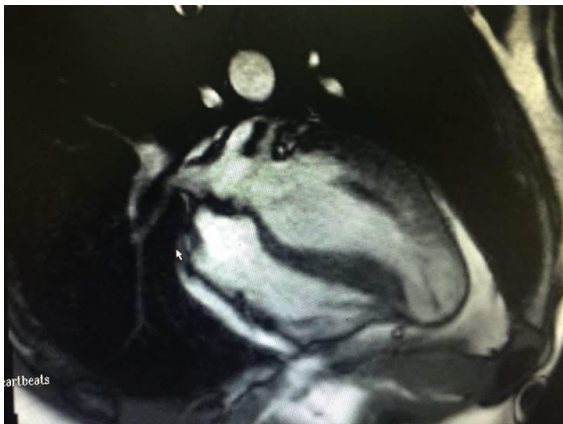


Figure 1. Ventricular tachycardia with a right bundle branch block pattern, transition in lead V₂, and a superior axis

QRS-synchronized electrical cardioversion (100 J) was performed in the emergency department, resulting in the termination of the arrhythmia. A 12-lead ECG revealed left ventricular hypertrophy (LVH) in a normal sinus rhythm state.

Echocardiography, conducted by a cardiology assistant, showed LVH with a normal

functional prosthetic valve and no significant valvular gradient. Because of the discrepant findings in echocardiography, CMR was performed with a 1.5-Tesla Aventis device (Siemens) and showed LVH with an apical aneurysm, diffuse late gadolinium enhancement, and a midcavitary gradient. All the findings were in favor of HCM (Fig. 2 & 3).



Figures 2 and 3. Left ventricular apical aneurysm with late gadolinium enhancement

Given the patient's history of a sustained monomorphic VT, electrophysiological study and mapping was done via the trans-septal approach because of the prosthetic aortic valve. Substrate mapping during the normal sinus rhythm state revealed a large low-voltage apical area, and activation mapping after VT induction showed a reentrant VT around the apical aneurysm. Radiofrequency ablation with a cooled-tip catheter (30 W and 43°C) was performed, and the apical aneurysm was isolated (Fig. 4).



Figure 4. Apical aneurysm ventricular tachycardia ablation using a 3D NavX system

At the end of the procedure, the VT was non-inducible and because of the high risk features of the patient, an implantable cardioverter-defibrillator was inserted.

DISCUSSION

The coincidence of BAV and HCM was first described by Brown in 1990 in 4 adult patients.^{4,5} The coincidence of HCM with other

cardiomyopathies such as LV noncompaction has also been previously reported in some studies.⁶

Echocardiography remains the gold standard in the examination of patients with HCM; however, echocardiography has its own shortcomings when it comes to the anterolateral segments of the LV, papillary muscles, some portions of the right ventricle, and the apex.^{7,8} CMR using gadolinium can not only reveal areas of fibrosis but also better clarify the points missed on echocardiography.

Necrosis can be found in half of the patients suffering from LVH as a consequence of aortic stenosis or arterial hypertension.⁹ Diffuse fibrosis with an apical aneurysm is seldom a finding associated with BAV and aortic stenosis, and it strongly suggests HCM.

Late gadolinium enhancement in CMR can predict a high risk for arrhythmias.⁹ LV apical aneurysms also constitute a high risk factor for arrhythmias.

In our patient, in spite of arrhythmia ablation, we implanted a cardioverter-defibrillator because of the high-risk features for sudden cardiac death.¹⁰⁻¹³

REFERENCES

1. B.J. Maron, J.M. Gardin, J.M. Flack, S.S. Gidding, T.T. Kurosaki, D.E. Bild Prevalence of hypertrophic cardiomyopathy in a general population of young adults. Echocardiographic analysis of 4111 subjects in the CARDIA Study. Coronary Artery Risk Development in (Young) Adults. *Circulation*, 92 (1995), pp. 785-789
2. B.J. Maron, S.R. Ommen, C. Semsarian, P. Spirito, I. Olivetto, M.S. Maron Hypertrophic cardiomyopathy: present and future, with translation into contemporary cardiovascular medicine *J Am Coll Cardiol*, 64 (2014), pp. 83-99
3. J. Somerville, L. McDonald Congenital anomalies in the heart with hypertrophic cardiomyopathy *Br Heart J*, 30 (1968), pp. 713-722

4. Branislav Obžut, Peter Blaško, Martin Porzer
Coincidence of bicuspid aortic valve presence and hypertrophic cardiomyopathy, and significance of magnetic resonance in its diagnostics *Cor et Vasa*, Volume 55, Issue 3, June 2013, Pages e271-e276
5. Padang R1, Gersh BJ1, Ommen SR1, Geske JB2. Heart Lung Circ. Prevalence and Impact of Coexistent Bicuspid Aortic Valve in Hypertrophic Cardiomyopathy. 2018 Jan;27(1):33-40. doi: 10.1016/j.hlc.2017.01.020. Epub 2017 Mar 8
6. Zahra Alizadeh-Sani, MD, Shabnam Madadi, MD, Anita Sadeghpour, MD, Zahra Khajali, MD, Pedram Golnari, MD, and Majid Kiavar, MD Cardiac MRI in a Patient with Coincident Left Ventricular Non-Compaction and Hypertrophic Cardiomyopathy *J Tehran Heart Cent*. 2011 Nov; 6(4): 214–216. Published online 2011 Nov 30.
7. A. Rudolph, H. Abdel-Aty, S. Bohl, et al. Noninvasive detection of fibrosis applying contrast-enhanced cardiac magnetic resonance in different forms of left ventricular hypertrophy relation to remodeling, *Journal of the American College of Cardiology*, 53 (2009), pp. 284-2913.
8. SJ Naeini, M Parsaee, S Madadi, Z Hosseini, Is There Concordance Between CMR and Echocardiography in Assessing Aortic Stenosis Severity? *IRANIAN HEART JOURNAL* 17 (2), 38-43
9. Majid Kyavar, Somayeh Mohammadi, Shabnam Madadi Relationship between syncope and sudden cardiac death in patients with hypertrophic cardiomyopathy and left ventricular mass index calculated by cardiac MRI, *IRANIAN HEART JOURNAL (IHJ) SPRING 2015*, Volume 16, Number 1; Page(s) 12 To 19.
10. S.C. Siu, C.K. Silversides, Bicuspid aortic valve disease *J Am Coll Cardiol*, 55 (2010), pp. 2789-2800
11. V.B. Hebl, W.R. Miranda, K.C. Ong, D.O. Hodge, J.M. Bos, F. Gentile, et al. The Natural History of Nonobstructive Hypertrophic Cardiomyopathy *Mayo Clin Proc*, 91 (2016), pp. 279-287.
12. 2017 AHA/ACC/HRS Guideline for Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death, Publication Date: October 30, 2017.