CASE REPORT

Mid-ventricular Hypertrophic Obstructive Cardiomyopathy with Apical Aneurysm Complicated with Syncope by Sustained Monomorphic Ventricular Tachycardia

Andrés Ricardo Pérez-Riera, M.D., Ph.D.,* Raimundo Barbosa-Barros, M.D.,†
Augusto Armando de Lucca, Jr M.D.,* Mujimbi Jose Viana, M.D.,* and Luiz
Carlos de Abreu, Ph.D.*

From the *ABC Faculty of Medicine – ABC Foundation, Santo André, São Paulo, Brazil; and †Coronary Center of the Messejana Hospital Dr. Carlos Alberto Studart Gomes, Fortaleza, CE, Brazil

Mid-ventricular hypertrophic obstructive cardiomyopathy with secondary formation of apical aneurysm is a rare variant of hypertrophic cardiomyopathy. They have a unique behavior because unlike other variants it causes sustained monomorphic ventricular tachycardia, which makes it particularly severe.

Ann Noninvasive Electrocardiol 2016;21(6):618–621

mid-ventricular hypertrophic obstructive cardiomyopathy; apical aneurysm; sustained monomorphic ventricular tachycardia

ID: ILP, Caucasian, male, 53 years old. **Main complaint:** sudden discomfort and syncope. **History of current disease:** Approximately 1 hour before, he started with profuse diaphoresis, dyspnea at rest, followed by syncope. Following electrical cardioversion, the patient himself reported being followed by a cardiologist and knowing to be a carrier of familial hypertrophic cardiomyopathy (HCM). He also recounted a son dying suddenly at age 18, during cycling. He is using β -blockers. **Physical:** cold sweating, unconscious, and dyspnea. Tachycardic regular cardiac rhythm (heart rate: 185 bpm), without cannon a waves, changes in beat-to-beat systolic blood pressure or variations in the first heart sound intensity. Blood pressure: 80 $\times$ 60 mmHg. Peripheral pulses present without edema. Bilateral crackles in both lungs.

Figure 1 shows the admission tracing. After reversion, it was decided for an implantable cardioverter defibrillator (ICD). The ECG in Figure 2 was performed after the electrical cardioversion. Figure 3 shows the echocardiographic image.

Figure 4 shows the ventriculography.

DISCUSSION

Mid-ventricular hypertrophic obstructive cardiomyopathy (MVHOCM) is a rare variant of HCM that occurs in 1% of patients carriers of this entity.¹ It could be complicated by apical aneurysm, as in this case.² It is characterized by asymmetrical hypertrophy in the middle part of the septum (mid-cavity obstruction), causing high intraventricular pressure gradients between the middle and low part of the left ventricle (LV) cavity. The patients complicated with apical aneurysm by LV necrosis are a significant clinical subset that is under identified and potentially fatal because it causes a tendency to sustained monomorphic ventricular tachycardia (SMVT). In the pathogenesis of the apical myocardial necrosis has been suggested to be secondary to increase in postload and to apical pressure augmentation, microcirculation disease associated with a decrease in coronary flow reserve, decrease in coronary perfusion pressure, spasm, and coronary lumen narrowing.³ The mechanical

Address for correspondence: Andrés Ricardo Pérez-Riera, Rua Sebastião Afonso, 885 CEP 04417-100 – Santo André, São Paulo/SP, Brazil. Tel.: (55) 11 5621-2390; Fax: (55) 11 5621-2390; E-mail: riera@uol.com.br

© 2016 Wiley Periodicals, Inc.
DOI: 10.1111/anec.12377

Figure 1. ECG diagnosis: Wide sustained QRS complex tachycardia, heart rate = 187 bpm, QRS axis in the right superior quadrant “no man’s land or Northwest axis” ($\approx -175^\circ$), absence of fusion and/or capture beats, R wave in V_1 , and QS pattern from V_2 to V_6 . Conclusion: VT originated from apical focus.

Figure 2. ECG diagnosis: Sinus rhythm, HR: 61 bpm, P wave duration ($P = 145$ ms), P axis ($+60^\circ$), augmented P-terminal forces (PTF- V_1): left atrial enlargement, PR interval (260 ms): first degree AV block. QRS axis $+5^\circ$, QRS duration 115 ms, and fQRS. The ST segment and T wave are in an opposite direction to the preceding QRS complexes in left leads: strain pattern.

compression of the coronary arteries may lead to acute infarction by spasm and microcirculation disease as a consequence of myocardial wall stress during systole in an acute fashion,⁴ causing a tendency to microthrombi and plaque rupture that may lead to apical aneurysm formation. The β -blockers are the drugs of choice. Because of the hemodynamic instability, we opted for secondary prevention implanting an ICD.

The SMVT has reentry mechanism, within the aneurysm or micro-reentry at the neck of it. Radiofrequency catheter ablation (RFCA) looking for the entrainment may remove the ventricular tachycardia (VT) reentry circuit. The ICD implant should be followed by the addition of pharmacological treatment or RFCA. This procedure is capable of abolishing the SMVT.⁵ Kono et al.⁶ presented a patient with MVHOCM associated to

Figure 3. Echocardiographic image in apical two-chamber view. Mid-left ventricular narrowing in the mid-septum. Turbulent flow high velocity within the middle section of the LV chamber. Mid-LV gradient = 91 mmHg at rest. Diastolic thickness of intraventricular septum with 24 mm and diastolic posterior wall thickness with 31 mm. Apical aneurysm observed only with contrast (4.4 cm). Conclusion: MVHOCM complicated with apical aneurysm. LV: left ventricle.

Figure 4. Left ventriculogram with hourglass aspect of the LV and large aneurysmal dyskinetic sac with stasis of contrast at the LV apex. Mid-ventricular obstruction (arrow) with 91 mmHg of gradient.

apical aneurysm and drug-refractory SMVT. In the electrophysiology study, polymorphic VT/ventricular fibrillation was induced. The patient underwent ICD implant associated with RFCA.

Minami et al.⁷ investigated the prevalence, clinical features, and prognosis of MVHOCM. The population of the study included 490 patients with HCM. The diagnosis of MVHOCM (mid-cavitary gradient ≥ 30 mmHg) was observed in 46 patients. This group was more symptomatic and with a greater tendency to sudden cardiac death (SCD). The formation of apical aneurysm was a marker of fatal arrhythmias. The following risk markers for SCD are present in our case: wall thickness > 30 mm; resting mid-ventricular gradient > 30 mmHg⁸; positive family history of SCD in young first-degree relatives; recording of syncope related to the events; and ECG with QRS fragmentation (fQRS).⁸ It is defined as narrow QRS complexes (< 120 ms) with multiple notches in R and/or S waves: ≥ 4 spikes in a single lead or ≥ 8 spikes in right precordial leads (V_1 – V_3).^{9–11} Also, fQRS is defined as the presence of one or more additional R' wave in two contiguous leads, corresponding to a major coronary artery territory of ECG.¹² The presence of fQRS points out scarred myocardium and it constitutes a noninvasive marker of fatal events.¹³ It has been described in coronary artery disease,¹⁴ dilated cardiomyopathy,¹⁵ HCM,¹⁶ arrhythmogenic right ventricular dysplasia/cardiomyopathy,¹⁷ cardiac sarcoidosis,¹⁸ Brugada syndrome,¹⁹ and acquired long QT syndrome.²⁰ In this last case, it is a marker of the appearance of Torsade de Pointes.²¹

CONCLUSION

MVHOCM, when in association with apical aneurysm, frequently complicates with the SMVT, unlike other forms of HCM, the VTs of which are usually not sustained. This difference makes the mid-ventricular obstructive form more severe, which indicates the need of implanting automatic ICD as a secondary prevention for SCD, in association to β -blockers. The latter are aimed at decreasing the number of shocks by the device.

Acknowledgements: The authors do not report any conflict of interest regarding this work.

REFERENCES

1. Harada K, Shimizu T, Sugishita Y, et al. Hypertrophic cardiomyopathy with midventricular obstruction and apical aneurysm: A case report. *Jpn Circ J* 2001;65:915–919.

2. Tse HF, Ho HH. Sudden cardiac death caused by hypertrophic cardiomyopathy associated with midventricular obstruction and apical aneurysm. *Heart* 2003;89:178.
3. Tomochika Y, Tanaka N, Wasaki Y, et al. Assessment of flow profile of left anterior descending coronary artery in hypertrophic cardiomyopathy by transesophageal pulsed Doppler echocardiography. *Am J Cardiol* 1993;72: 1425–1430.
4. Mohiddin SA, Fananapazir L. Myocardial bridging in adult and pediatric patients with hypertrophic cardiomyopathy is not associated with poor outcome. *J Am Coll Cardiol* 2004;43:1133.
5. Lim KK, Maron BJ, Knight BP. Successful catheter ablation of hemodynamically unstable monomorphic ventricular tachycardia in a patient with hypertrophic cardiomyopathy and apical aneurysm. *J Cardiovasc Electrophysiol* 2009;20:445–447.
6. Kono K, Higashi T, Hara K, et al. Mid-ventricular obstructive hypertrophic cardiomyopathy associated with an apical aneurysm and sustained ventricular tachycardia: A case report. *J Cardiol* 2001;38:343–349.
7. Minami Y, Kajimoto K, Terajima Y, et al. Clinical implications of midventricular obstruction in patients with hypertrophic cardiomyopathy. *J Am Coll Cardiol* 2011;57: 2346–2355.
8. Femenía F, Arce M, Arrieta M, et al. Surface fragmented QRS in a patient with hypertrophic cardiomyopathy and malignant arrhythmias: Is there an association? *J Cardiovasc Dis Res* 2012;3:32–35.
9. Morita H, Kusano KF, Miura D, et al. Fragmented QRS as a marker of conduction abnormality and a predictor of prognosis of Brugada syndrome. *Circulation* 2008;118: 1697–1704.
10. Zhang L, Liu L, Kowey PR, et al. The electrocardiographic manifestations of arrhythmogenic right ventricular dysplasia. *Curr Cardiol Rev* 2014;10:237–245.
11. Schuller JL, Olson MD, Zipse MM, et al. Electrocardiographic characteristics in patients with pulmonary sarcoidosis indicating cardiac involvement. *J Cardiovasc Electrophysiol* 2011;22:1243–1248.
12. Das MK, Khan B, Jacob S, et al. Significance of a fragmented QRS complex versus a Q wave in patients with coronary artery disease. *Circulation* 2006;113: 2495–2501.
13. Das MK, Zipes DP. Fragmented QRS: A predictor of mortality and sudden cardiac death. *Heart Rhythm* 2009; 6(3 Suppl):S8–S14.
14. Das MK, Maskoun W, Shen C, et al. Fragmented QRS on twelve-lead electrocardiogram predicts arrhythmic events in patients with ischemic and nonischemic cardiomyopathy. *Heart Rhythm* 2010;7:74–80.
15. Tigen K, Karaahmet T, Gurel E, et al. The utility of fragmented QRS complexes to predict significant intraventricular dyssynchrony in nonischemic dilated cardiomyopathy patients with a narrow QRS interval. *Can J Cardiol* 2009;25:517–522.
16. Kang KW, Janardhan AH, Jung KT, et al. Fragmented QRS as a candidate marker for high-risk assessment in hypertrophic cardiomyopathy. *Heart Rhythm* 2014;11:1433–1440.
17. Peters S, Trümmel M, Koehler B. QRS fragmentation in standard ECG as a diagnostic marker of arrhythmogenic right ventricular dysplasia-cardiomyopathy. *Heart Rhythm* 2008;5:1417–1421.
18. Homsí M, Alsayed L, Safadi B, et al. Fragmented QRS complexes on 12-lead ECG: A marker of cardiac sarcoidosis as detected by gadolinium cardiac magnetic resonance imaging. *Ann Noninvasive Electrocardiol* 2009;14: 319–326.
19. Morita H, Zipes DP, Wu J. Brugada syndrome: Insights of ST elevation, arrhythmogenicity, and risk stratification from experimental observations. *Heart Rhythm* 2009; 6(11 Suppl):S34–S43.
20. Moss AJ. Fragmented QRS: The new high-risk kid on the block in acquired long QT syndrome. *Heart Rhythm* 2010;7:1815–1816.
21. Haraoka K, Morita H, Saito Y, et al. Fragmented QRS is associated with torsades de pointes in patients with acquired long QT syndrome. *Heart Rhythm* 2010;7: 1808–1814.