Case Report

Multiple Cardiac Arrhythmias Detected by a Dual Chamber Implantable Cardioverter Defibrillator in a Patient with Hypertrophic Obstructive Cardiomyopathy

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We present the case of a patient with hypertrophic obstructive cardiomyopathy (HOCM), with multiple risk factors for sudden cardiac death (SCD), who received a dual chamber implantable cardioverter-defibrillator (ICD). Within three weeks of implantation the patient experienced multiple ICD discharges. A review of the ICD strips, with a careful evaluation of the electrograms from the atrial (A) and ventricular (V) channels, revealed multiple simultaneous cardiac rhythm disturbances. This case highlights the importance of carefully risk stratifying these patients for SCD and the utility of a dual chamber ICD.

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Hypertrophic obstructive cardiomyopathy (HOCM) is a genetically determined myocardial disease with a diverse natural history[1-3]. Patients with HOCM are at high risk of sudden cardiac death (SCD)[4-8]. Risk factors predisposing to SCD, include: a family history of SCD, syncope, severe and diffuse left ventricular hypertrophy (LVH), patients who exhibit myocardial ischemia associated with syncope, cardiac arrest survivors, inducible ventricular tachycardia during invasive electrophysiologic evaluation, and those patients who have exhibited sustained or non-sustained ventricular arrhythmias[9-18]. However, the clinical value of NSVT as a marker of risk is limited by a relatively modest positive predictive value[16]. In addition to ventricular tachycardia (VT), these patients experience a variety of atrial arrhythmias, most commonly atrial fibrillation[11]. We present here the case of a 51-year-old man with HOCM who was deemed at high risk for SCD and received a dual chamber implantable cardioverter defibrillator (ICD).

Case report

A 51-year-old man with a known history of HOCM presented to the emergency room complaining of substernal chest pain of several hours duration associated with palpitations and lightheadedness. The patient was admitted for the management of an acute coronary syndrome. On physical examination the patient was noted to have a hyperdynamic and laterally displaced point of maximal impulse with a systolic murmur heard most prominently at the base. The 12-lead electrocardiogram showed sinus rhythm, left ventricular hypertrophy, left atrial hypertrophy and pseudoinfarct pattern in leads V_1-V_6. The patient's chest discomfort was relieved with nitroglycerin and he was scheduled for a coronary angiogram. Telemetry monitoring revealed frequent ventricular ectopy (>10/h) with runs of polymorphic NSVT (3-5 beat runs). Further work up revealed extreme septal hypertrophy (37 mm, normal 6-11 mm), a significant LV outflow tract gradient (>100 mm Hg), normal epicardial coronary arteries with

evidence of myocardial bridging in the mid-section of the left anterior descending artery and reversible anterior/ anteroseptal ischemia during radionuclide stress testing. This patient had severe diastolic dysfunction and a limited exercise tolerance. This patient's CHF failed to respond to medical therapy including beta-blockers, calcium channel blockers and disopyramide therapy. With a failure to respond to medical therapy and a persistence of a severe LV outflow tract obstruction and multiple risk factors predisposing to SCD, it was decided to implant a dual chamber ICD.

Three weeks following surgery the patient experienced two episodes of dizziness each followed by an ICD discharge. Interrogation of the ICD revealed two episodes of rapid ventricular tachycardia (VT). The timing of the events correlated with the patient's symptoms. Both episodes were properly detected and converted to sinus rhythm after a 28.5 J shock. The second episode revealed atrial fibrillation, characterized by rapid atrial activity in the atrial channel, which organized to atrial flutter. These atrial arrhythmias occurred while the patient was experiencing VT. Comparison of this episode to the first ICD discharge helps to further clarify the arrhythmias detected in the second episode. The atrial electrograms in figure 1, Panel A revealed an atrial rate of 60 bpm (cycle length of 1000 ms). Inter-dispersed between the atrial activations are broader low signals reflecting far field sensing of ventricular depolarization. The ventricular channel revealed a VT at a rate of 240 beats per minute (cycle length of 250 ms) with clear A-V dissociation. There is no evidence of retrograde VA conduction. The second example revealed an initial rapid and irregular atrial rhythm, consistent with atrial fibrillation, which later organizes to a regular rhythm (cycle length of 220 ms). The regular atrial arrhythmia is consistent with an atrial flutter. Interspersed throughout the atrial electrogram channel are additional low amplitude signals, identical to the far-field activations seen in the first strip. The ventricular channel revealed a VT with a cycle length of 200 ms (300 bpm), which is identical to the far field activations noted on the atrial channel. The 28.5 J shock was appropriately delivered by the ICD, successfully converting both the atrial and ventricular tachyarrhythmias to sinus rhythm. In

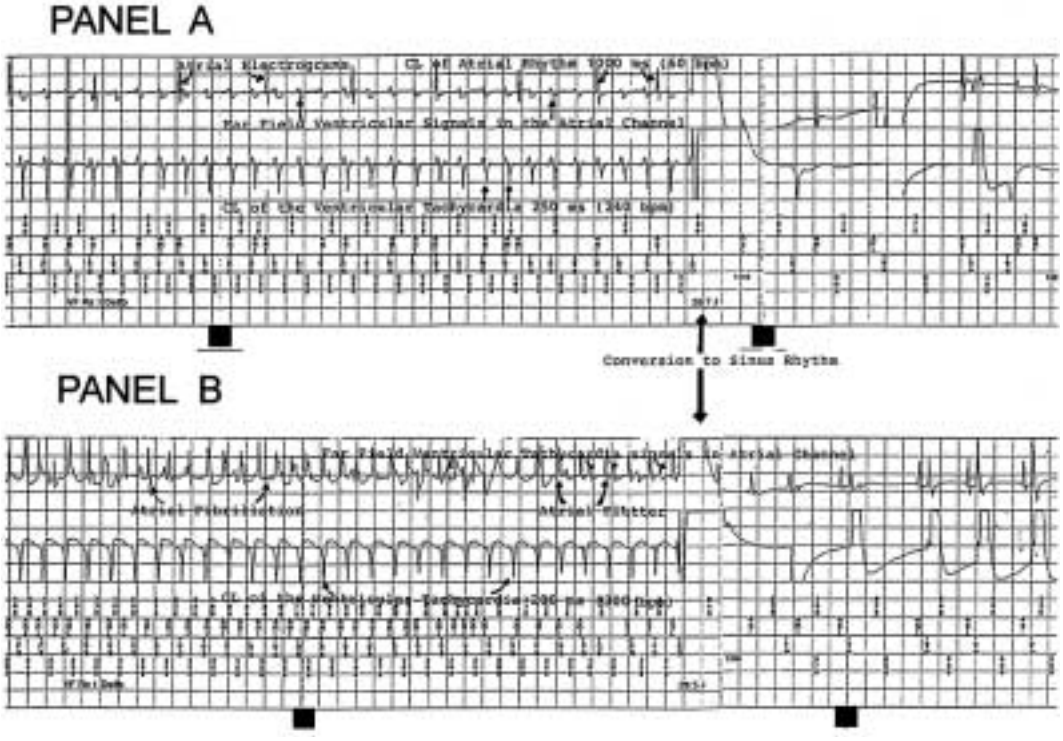

Figure 1. Panel A. Ventricular tachycardia detected and appropriately converted by a 28.7 J shock to sinus rhythm. Clear evidence of AV dissociation with the atrial channel (top) exhibiting an atrial cycle length of 1000 ms with a ventricular tachycardia cycle length of 250 ms.
Panel B. Atrial fibrillation organizing to atrial flutter occurring simultaneously along with a ventricular tachycardia appropriately detected and converted to sinus rhythm by a 28.5 J shock.

addition, the morphology of the electrograms in panel A and B are identical and distinctly different to the ventricular electrograms, during sinus rhythm. Lastly, the ventricular electrograms are very regular distinguishing atrial fibrillation with abberancy from two simultaneously occurring arrhythmias.

Discussion

This case report illustrates the importance of carefully screening patients with HOCM for their potential risk of SCD. Patients with a HOCM who present with complaints of altered consciousness should be carefully screened for potential risk factors predicting a significant risk of SCD. The work-up should include extensive telemetry or ambulatory holter monitoring (48-72 hours), exercise testing with hemodynamic monitoring and radionuclide imaging, an evaluation of LV systolic function, and a cardiac catheterization and invasive electrophysiologic evaluation to determine the extent of the left ventricle outflow tract obstruction and propensity for inducible VT, respectively. It should be noted that the role of the invasive electrophysiologic study in risk stratifying patients with HOCM has been controversial[13]. Early experience was associated with a higher risk of complications in this subset of patients. Subsequent data has shown that in experienced hands there is no increased risk in performing this procedure on this patient population[13].

Patients with HOCM experience a variety of atrial and ventricular arrhythmias. Atrial fibrillation occurs in 10-15% of patients with HOCM[11]. The sudden onset of any supraventricular tachycardia can be associated with hypotension, which may precipitate syncope and potentially lead to ventricular arrhythmias and sudden cardiac death. This is more prevalent in a patient with a severe left ventricle outflow tract obstruction, myocardial bridging and impaired diastolic function.

In this patient, who experienced shocks early after the implantation of the dual chamber ICD, the electrogram analysis revealed multiple arrhythmias detected simultaneously. The first episode detected was a primary ventricular arrhythmia. The second VT detected either occurred simultaneously with atrial fibrillation or was caused by atrial fibrillation with a rapid ventricular response. We speculate that the VT occurred due to a transient elevation in filling pressures prompting the onset of atrial fibrillation with a rapid ventricular response. The rapid ventricular rate may have prompted ischemia and a resultant ventricular tachyarrhythmia. The patient was started on amiodarone therapy.

Several months following the implantation of the ICD, evaluation of the left ventricle outflow tract revealed a modest reduction in the gradient (85 mm Hg) with an increase in exercise tolerance and no further episodes of ventricular or atrial arrhythmias. The role of a dual chamber ICD is multiple in these patients. The ICD offers dual chamber pacing which may reduce the left ventricle outflow gradient by allowing maximal dosing of negative ionotropic medications required to enhance diastolic filling. Theoretically, the creation of a paced left bundle branch block with paradoxical septal motion may reduce the left ventricle outflow tract gradient. In addition, the ICD is invaluable in aborting sudden cardiac death. Lastly, the emergence and refinement of dual chamber devices with more sophisticated discrimination algorithms minimizes inappropriate discharges without sacrificing the efficacy in appropriately detecting and treating potentially lethal ventricular arrhythmias.

Conclusion

Patients with HOCM are at risk for SCD, highlighting the importance of careful risk stratification. These patients experience a variety of atrial and ventricular arrhythmias. This report illustrates the multiplicity of rhythm disturbances, which can occur in a patient with HOCM, the importance of the ICD in aborting SCD and the importance of a dual chamber device with a sophisticated discrimination algorithm in properly distinguishing an atrial tachycardia from a coexistent ventricular tachycardia.

References

1. Jeschke B, Uhl K, Weist B, et al: A high risk phenotype of hypertrophic cardiomyopathy associated with a compound phenotype of two mutated beta myosin heavy chains genes. Hum Genet 1998; 102: 299-304.
2. Lankford EB, Sweeney HL, Epstein ND, et al: Abnormal contractile properties of muscle fibers expressing mutations in beta-myosin in- patients with hypertrophic cardiomyopathy. J Clin Invest 1995; 95: 1409-1414.
3. Cuda G, Fananpazir L, Epstein ND, et al: The in vitro motility activity of beta-cardiac myosin depends on the nature of the beta-myosin heavy-chain gene mutation in hypertrophic cardiomyopathy. J Muscle Res Cell Motil 1997; 18: 1-19.
4. Maron, BJ, Lipson LC, Roberst WC, et al: Malignant hypertrophic cardiomyopathy: Identification of a subgroups of families with unusually frequent premature deaths. Am J Cardiol 1978; 14: 1133-1140.

5. McKenna WJ, Deabfield JE: Hypertrophic cardiomyopathy: An important cause of sudden cardiac death. Arch Dis Child 1984; 59: 971-975.
6. Maron BJ, Roberts WC, Epstein SE: Sudden death in hypertrophic cardiomyopathy: a profile of 78 patients. Circulation 1982; 65: 1388-1394.
7. Maron BJ, Epstein SE, Roberts WC: Causes of sudden death in competitive athletes. J Am Coll Cardiol 1986; 7: 204-214.
8. Maron BJ, Fananapazir L: Sudden cardiac death in hypertrophic cardiomyopathy. Circulation 1992; 85: I57-I63.
9. Savage DD, Sedes T, Maron BJ, et al: Prevalence of arrhythmias during 24-hour electrocardiographic monitoring and exercise testing in-patients with obstructive and non-obstructive hypertrophic cardiomyopathy. Circulation 1979; 59: 866-875.
10. Bjarnason I, Hadarson T, Jonsson S: Cardiac arrhythmias in hypertrophic cardiomyopathy. Br Heart J 1982; 48: 198-203.
11. Stafford WJ, Trohman RG, Bilsker M, et al: Cardiac arrest in adolescents with atrial fibrillation and hypertrophic cardiomyopathy. J Am Coll Cardiol 1986; 7: 701-704.
12. Fananapazir L, Epstein SE: VT and sudden death in HCM patients. Circulation 1989; 80: 1923-1934.
13. Fananapazir L, Chang AC, Epstein SE, et al: Prognostic determinants in hypertrophic cardiomyopathy: Prospective evaluation of a therapeutic strategy based on clinical, holter, hemodynamics, and electrophysiologic findings. Circulation 1992; 86: 730-740.
14. Dilsizian V, Bonow RO, Epstein SE, et al: Myocardial ischemia is a frequent cause of cardiac arrest and syncope in young patients with hypertrophic cardiomyopathy. Circulation 1990; 82: 35A.
15. Anderson KP, Stinson EB, Derby GC, et al: Vulnerability of patients with hypertrophic cardiomyopathy to ventricular arrhythmia induction in the operating room. Am J Cardiol 1983; 51: 811-815.
16. Spirito P, Rapezzi C, et al: Prognosis of asymptomatic patient with hypertrophic cardiomyopathy and nonsustained ventricular tachycardia. Circulation 1994; 90: 2743-2747.
17. McKenna W, Elliot P: Arrhythmia, sudden death and clinical risk stratification in hypertrophic cardiomyopathy, in cardiac electrophysiology. From cell to bedside, 3rd ed, Zipes D, Jalife J; W.B. Saunders Company, 2000, pp 555-562
18. Spirito P, Marion BJ: Relation between extent of left ventricular hypertrophy and occurrence of sudden cardiac death in hypertrophic cardiomyopathy. J Am Coll Cardiol 1990; 15: 1521-1526.