Sudden Death Due to Atrial Fibrillation in Hypertrophic Cardiomyopathy: A Predictable Event in a Young Patient

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FAVALE, S., ET AL.: Sudden Death Due to Atrial Fibrillation in Hypertrophic Cardiomyopathy: A Predictable Event in a Young Patient. This case refers to a 39-year-old woman with hypertrophic cardiomyopathy (HCM) and family history of sudden death (SD). In 1985, high rate atrial stimulation induced VF. In 1996 an ICD was implanted and she remained without arrhythmic events until November 2000 when the device reported one episode of atrial fibrillation degenerating into VF and terminated by the ICD. The VF induction mechanism recorded by the ICD was similar to that observed in 1985. The high incidence of atrial tachyarrhythmias in HCM renders cases like this at higher risk of SD. The predictive role of incremental atrial stimulation merits highlighting in future studies. (PACE 2003; 26[Pt. I]:637–639)

sudden death, hypertrophic cardiomyopathy, implantable cardioverter defibrillator, ventricular fibrillation

Introduction

Hypertrophic cardiomyopathy (HCM) is a genetically determined myocardial disease.1 The spectrum of clinical phenotypes and natural history includes a group at high risk for sudden death (SD).1,2–7 Clinical studies have recently focused on the risk stratification and appropriateness of preventive measures.8,9 However, limitations of these studies are related to low disease prevalence and the fact that patients at risk for SD are often young and asymptomatic with a low incidence of unpredictable, lethal events.9 Therefore, even in a single patient, long-term follow-up may help to clarify some aspects of the natural history of disease and of the SD mechanisms in a patient with HCM.

This case refers to a 39-year-old woman with hypertrophic obstructive cardiomyopathy (HOCM) who has been followed since 1977. She was first referred for cardiological evaluation at age 16 years due to atypical chest pain. Her father died (age of 50 years) as did two sisters (age 2 and 9 years) and two brothers (age 16 and 17 years).

A HOCM with a septum thickness of 15 mm and a conduction of 1:1 during atrial pacing up to 200 beats/min were observed. At this atrial rate the AH interval passed from 55 to 160 ms.

Eight years later (1985) at further evaluation the septum thickness was 18 mm. Holter monitoring elicited rare monomorphic premature ventricular beats (10 in 24 hours). Transesophageal atrial stimulation (stimuli with 9.9 duration and 20-mA current intensity) showed 1:1 atrioventricular (AV) conduction with narrow QRS up to a rate of 190 beats/min with an increase of the stimulus to R interval from 120 to 200 ms. Five seconds after the onset of atrial stimulation at 200 beats/min a premature ventricular 3-beat run was followed by ventricular fibrillation (VF). Prior to VF an increasing ST depression was observed.

Amiodarone (600 mg/day) or verapamil (120 mg three times a day) treatment prevented VF induction. After withdrawal of either drug, VF was induced again by transesophageal atrial pacing at 190 beats/min.10

The patient remained asymptomatic while taking verapamil until 1996, when she was submitted to a new evaluation, including a programmed ventricular stimulation which induced a polymorphic, sustained, syncopal ventricular tachycardia (VT). Therefore, a single chamber cardioverter defibrillator (model Medtronic 7221, Minneapolis, MN, USA) was implanted in a subcutaneous left pectoral pocket.

The patient was followed and the device interrogated every 6 months. No events were reported by the device for 4 years and she remained asymptomatic, autonomously suspending drug therapy. At the last interrogation (November 2000) the device reported one event. The episode interval plot (Fig. 1) and the electrogram (EGM) marker strip (Fig. 2) showed atrial fibrillation (AF) with RR intervals ranging between 500 and 270 ms followed by VF. The diagnosis of AF, given the absence of atrial EGMs in the single chamber device, was supported by the analysis of the RR interval and of the
morphology of the six ventricular EGMs preceding VF which were similar to those recorded during sinus rhythm. AF and VF were terminated by the implantable cardioverter defibrillator (ICD) with a 32-J shock. While sleeping (4:27 AM) the patient did not sense the ICD intervention.

SD occurs in young, asymptomatic patients with HCM, and has a reported annual mortality rate ranging from 4% to 6% percent. Secondary prevention with an ICD in patients who have had episodes of sustained VT or resuscitated VF is mandatory due to the high incidence of life-threatening arrhythmias. The indication for ICD is related to low risk/benefit and favorable cost effectiveness ratios achieved by the latest generation of pectorally implantable devices in high risk patients.

The role of ICD for primary prevention of SD in patients who have not had prior cardiac arrest or spontaneous sustained VT needs to be defined. A retrospective, multicenter study evaluated the arrhythmic events of HCM patients with an ICD implanted solely for primary prevention due to one or more of the following SD risk factors: syncope, a family history of SD, nonsustained VT, sustained VT induced during electrophysiological testing and a left wall thickness of ≥30 mm. The reported 5% annual rate of appropriate ICD discharges for high rate VT or VF is similar to the rate of SD in selected high risk patients with HCM. The case described in this article presents two interesting aspects related to primary prevention. The first is patient risk profile, the second the mechanism by which aborted SD occurred. In this case the risk profile included a family history of SD and an interventricular septum thickness of 18 mm. A family history of SD at young age, defined as at least two SDs in primary, related, family members, is considered an indicator of high risk. A prophylactic implantation of ICD in these patients may be considered although no data are reported on its preventive efficacy: a better quality-of-life due to the patients’ greater sense of security may also be expected. The wall thickness is another strong independent predictor of SD. At present, malignant family history and wall thickness would be
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References