Case Report

Appropriate Management of Syncope in a Patient with Hypertrophic Cardiomyopathy: Rationale Behind Long-Term Cardiac Rhythm Monitoring

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Key words:
Syncope, monitoring, device.

We describe the case of a patient with hypertrophic cardiomyopathy who had recurrent syncopal episodes, the cause of which remained unexplained despite a thorough evaluation. Two years after his first evaluation, an implantable loop recorder revealed asymptomatic episodes of advanced second degree atrioventricular block while the patient was awake. Although a permanent pacemaker was implanted, the patient continued to suffer syncopal episodes, during which the pacemaker recorded episodes of ventricular tachycardia. Accordingly, the device was upgraded to an implantable cardioverter-defibrillator.

Manuscript received:
November 13, 2008;
Accepted:
February 10, 2009.

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In patients with hypertrophic cardiomyopathy (HCM) symptoms of impaired consciousness (syncope and presyncope) occur in approximately 15-25%, while according to accumulating data syncope is associated with a substantial increase in the risk of sudden cardiac death.[1-3] Here we describe a case where an implantable loop recorder revealed the underlying arrhythmia in a patient with HCM who suffered repeated and unexplained episodes of syncope.

Case presentation

A 64-year-old male smoker, with arterial hypertension and hypercholesterolaemia, was admitted to our department after 3 episodes of syncope while seated during the previous 2 months, without any injuries. The initial evaluation (medical history, physical examination, blood pressure measurement in both supine and upright position, standard ECG) failed to demonstrate the cause of syncope. The patient, who showed no symptoms of heart failure (New York Heart Association functional Class I) was thus referred for a more thorough evaluation. The echocardiographic examination demonstrated the existence of HCM with asymmetric septal hypertrophy (septal wall thickness during diastole 18 mm) and systolic anterior motion of the mitral valve, while no left ventricular (LV) outflow tract gradient was evident either at rest or during provocative manoeuvres. Ventricular systolic function was normal (LV ejection fraction 65%). A tilt test and carotid sinus massage were also performed without any pathological findings. Both carotid and vertebrobasilar ultrasound were negative, as was the brain computed tomography scan, and the neurological evaluation was normal. Ambulatory ECG monitoring detected an episode of non-sustained ventricular tachycardia (5 beats at a rate of 150 beats/min) as well as one episode of atrial tachycardia at a rate of 180 beats/min that lasted for 5 seconds. During the electrophysiological study the only pathological finding was an episode of inducible non-sustained ventricular tachycardia (NSVT)

at a rate of 130 beats/min and lasting for 3 seconds, with no findings of sinus or atrioventricular (AV) node dysfunction. Myocardial perfusion was assessed by single photon emission computed tomography with TL-201, which revealed reversible ischaemia of the anterior septal and inferior walls of the LV. The coronary angiogram revealed no significant coronary artery stenosis (stenotic lesions less than 40%). Based on the above findings we decided to keep the patient under regular observation, but without proceeding to any therapeutic interventions for syncope.

A year later the patient experienced a new episode of syncope while seated, without injury, and despite thorough reappraisal (tilt test, carotid sinus massage, electrophysiological study, 48-hour Holter monitoring) no pathological finding emerged. At that point, an implantable loop recorder (ILR) (Reveal® Plus Model 9526, Medtronic, Minneapolis MN, USA) was implanted and 12 months later, although the patient remained asymptomatic, the device recorded episodes of advanced second degree AV block while the patient was awake, lasting for 4 seconds (Figure 1). Based on that finding, we decided to implant a permanent DDDR pacemaker (Guidant INSIGNIA I Plus DR, St. Paul MN, USA). However, as the patient was asymptomatic during these periods of AV block we were not entirely convinced that the syncopal episodes could be attributed to that conduction disturbance; thus, we decided to implant a device with the facility to monitor and store high quality intracardiac electrograms, effectively continuing our cardiac rhythm monitoring. Three years later a new episode of syncope with injury took place, during which the pacemaker's Holter detected 3 self-limited episodes of ventricular tachycardia (which were not pacemaker-induced) at a rate of 200 beats/min, lasting for 4, 6 and 11 seconds (Figure 2). On the basis of this new evidence, the pacemaker was replaced with an implantable cardioverter-defibrillator (ICD) device and since then the patient has remained asymptomatic.

Discussion

Syncope in patients with HCM can be attributed to two basic mechanisms: arrhythmia and primary haemodynamic disturbances. Paroxysmal atrial fibrillation and ventricular tachycardia are the most common types of syncope-related arrhythmias, while LV outflow tract obstruction, hypotension due to inappropriate vasodilatation, or impaired filling in the setting of reduced preload and diastolic dysfunction are the main haemodynamic mechanisms that can lead to syncope.[4,5] However, in the majority of cases no mechanism is found despite thorough and detailed investigations, such as repeated 24-hour ambulatory ECG, patient-activated monitoring, or invasive electrophysiological studies.[6] Based on the recently published ACC/AHA/ESC 2006

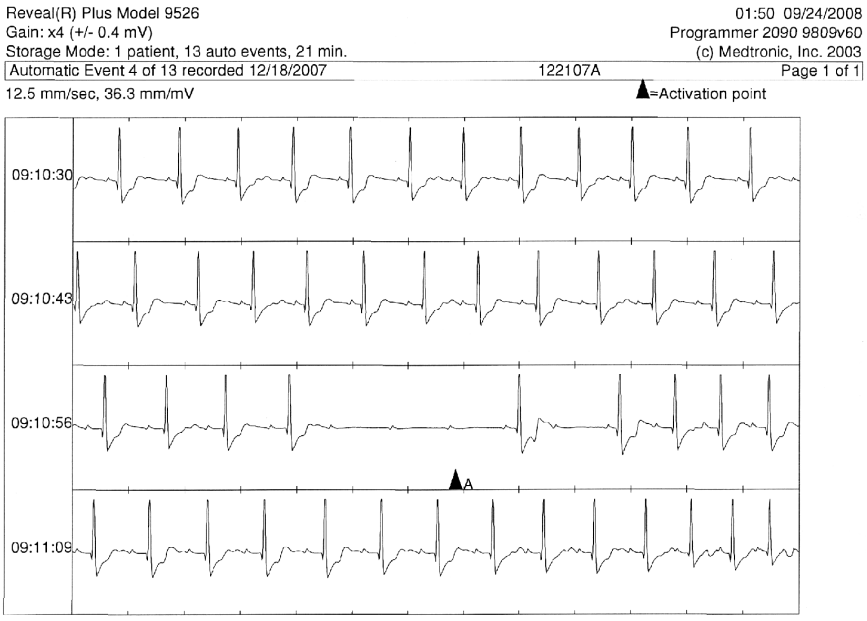

Figure 1. The implantable loop recorder tracing showing a 4-second pause due to high-degree atrioventricular block.

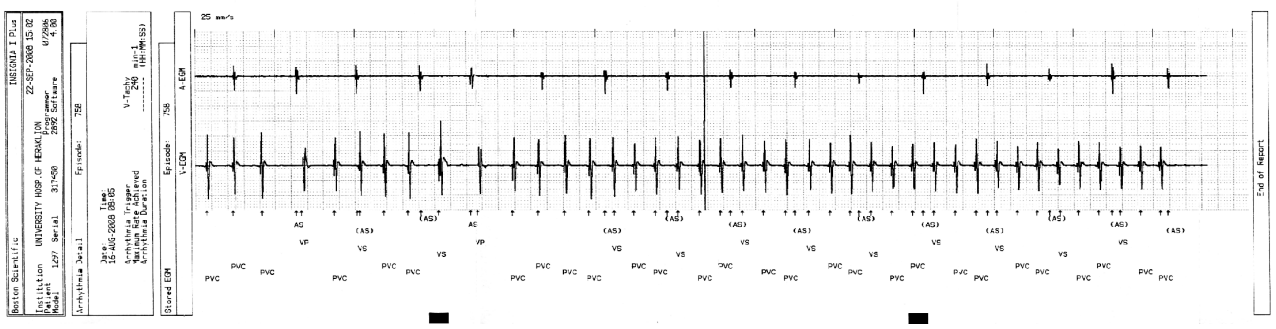

Figure 2. One of the three episodes of ventricular tachycardia recorded by the pacemaker.

guidelines for the management of ventricular arrhythmias and the ACC/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities, our patient (with HCM, syncope and NSVT) would be a candidate for ICD implantation (Class IIa, level of evidence C) without any further investigation.[7-9] However, the findings of the evaluations during both his first and his second admission did not constitute an adequate indication for either a pacemaker or an ICD implantation according to the guidelines in effect during that period.[10]

One year after the insertion of the ILR, the episodes of advanced second degree AV block detected by the device, though not correlated with symptoms, were sufficient to justify the implantation of a permanent pacemaker.[10] Three years later, the patient experienced recurrence of the syncopal symptoms and interrogation of the pacemaker revealed episodes of ventricular tachycardia, which correlated precisely with the occurrence of the symptoms. It was this finding that led us to the decision to upgrade the pacemaker to an ICD, in order to avoid symptoms and improve prognosis. As mentioned above, there are a number of different mechanisms that can provoke an episode of syncope in a patient with HCM. In our patient, who suffered syncopal episodes over a very long period of time, we do not know whether all the episodes can be attributed to the same mechanism. However, the very long-term monitoring, initially by an ILR and subsequently by the pacemaker, led us to the correct therapeutic intervention. Based on this case, we would like to underline the importance of long-term heart rhythm monitoring, even with therapeutic devices such as pacemakers. In similar cases, when the exact nature of a syncopal episode is vague, but implantation of a pacemaker is required, it is crucial that the device incorporates diagnostics with the ability to record cardiac rhythm electrograms and has sufficient memory to store these data.

References

1. Kofflard MJ, Ten Cate FJ, Van der Lee C, Van Domburg RT. Hypertrophic cardiomyopathy in a large community-based population: clinical outcome and identification of risk factors for sudden cardiac death and clinical deterioration. J Am Coll Cardiol. 2003; 41: 987-993.
2. Georgakopoulos D, Tolis V. Hypertrophic cardiomyopathy in children, teenagers and young adults. Hellenic J Cardiol. 2007; 48: 228-233.
3. Manolis A, Chatzis D. Unexplained syncope: diagnostic evaluation in the modern eras. Hellenic J Cardiol. 2004; 45: 100-106.
4. Lynne W, Frenneaux M. Syncope in hypertrophic cardiomyopathy. mechanisms and consequences for treatment. Europace. 2007; 9: 817-822.
5. Kassotis J, Haq S, Mongwa M. Multiple cardiac arrhythmias detected by a dual chamber implantable cardioverter defibrillator in a patient with hypertrophic obstructive cardiomyopathy. Hellenic J Cardiol. 2004; 45: 57-60.
6. Prasad K, Williams L, Campbell R, Elliot PM, McKenna WJ, Frenneaux M. Episodic syncope in hypertrophic cardiomyopathy-evidence for inappropriate vasodilation. Heart. 2008; 94: 1312-1317.
7. Zipes DP, Camm AJ, Borggrefe M, et al. ACC/AHA/ESC 2006 guidelines for management of patients with ventricular arrhythmia and the prevention of sudden cardiac death. Europace. 2006; 8: 746-837.
8. Epstein AE, DiMarco JP, Ellenbogen KA, et al. ACC/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities. J Am Coll Cardiol. 2008; 51: 1-62.
9. Styliadis I, Mantziari A, Gouzoumas N, et al. Indications for permanent pacing and pacing mode prescription from 1989 to 2006. Experience of a single academic centre in northern Greece. Hellenic J Cardiol 2008; 49: 155-162.
10 Gregoratos G, Abrams J, Epstein A, et al. ACC/AHA/NASPE 2002 guideline update for implantation of cardiac pacemakers and antiarrhythmia devices: summary article. Circulation. 2002; 106: 2145-2161.