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Hypertrophic Cardiomyopathy and Ventricular Arrhythmia

Z. Bennoui

Cardiology Department, Tipaza, Algeria

Abstract

Hypertrophic cardiomyopathy (HCM) is the most common genetic cardiac disease. Its clinical presentation is heterogeneous, ranging from asymptomatic patients to sudden death in young individuals. This complication is unpredictable, and no preventive medical treatment has proven effective. We report the case of a 32-year-old individual who was admitted for poorly tolerated ventricular tachycardia (VT), which was resolved with external electrical shock. We diagnosed localized hypertrophic cardiomyopathy, confirmed by MRI. This paper outlines the patient's medical journey, highlighting the rhythm complications experienced and the various therapeutic options available.

Keywords : Hypertrophic Cardiomyopathy (HCM), Ventricular Tachycardia (VT), Magnetic Resonance Imaging (MRI), Preventive Medical Treatment, Therapeutic Options.

Introduction

HCM is characterized by a thickening of the myocardium, most commonly predominant at the level of the interventricular septum, without any identifiable clinical cause (such as severe hypertension or significant aortic stenosis). Echocardiography (or MRI) shows thickening of a myocardial wall (≥15 mm if it's a sporadic case; ≥13 mm in a familial context). Asymmetric septal thickening is the most frequent and suggestive of a sarcomeric origin (1, 2). The diagnosis of HCM can be considered in various circumstances: cardiac symptoms (palpitations, dyspnea, precordial pain), discovery of a heart murmur on auscultation, routine assessment in an athlete, upon finding a complication (embolism, arrhythmia, sudden death), or during screening in an asymptomatic relative. Electrocardiographic anomalies may also suggest HCM (pseudo-necrotic Q waves, repolarization abnormalities).

Observation

We report the case of a 32-year-old patient who was examined in neurology for a syncope three months ago, with normal findings in all evaluations. He had a family history of sudden death (brothers and cousins). This patient presented with sudden-onset palpitations lasting about an hour. Upon examination, the patient was in hypotensive collapse with a regular tachycardia. The ECG indicated ventricular tachycardia (VT) at 200 beats per minute. The diagnosis of VT with right bundle branch block pattern and left axis deviation was based on these criteria: absence of RS complex in precordial leads, initial R wave in AVR, RS interval greater than 100 ms, and initial R wave greater than 40 ms. The diagnosis of poorly tolerated ventricular tachycardia was confirmed (Figure 1), and the patient underwent electrical cardioversion, which restored normal sinus rhythm (Figure 2). The patient was given continuous infusion of amiodarone for 24 hours, followed by oral amiodarone and bisoprolol. Blood tests were

normal (complete blood count, serum electrolytes, renal function, blood glucose, TSH, and phosphocalcium profile). Cardiac echodoppler revealed a non-obstructive hypertrophic cardiomyopathy (HCM) with left ventricular hypertrophy at 17 mm, good systolic and diastolic function, non-dilated left atrium, no mitral regurgitation, non-dilated right heart chambers, and normal right ventricular systolic and diastolic function, without pericardial effusion (Figure 3). Cardiac MRI confirmed the diagnosis of HCM localized at the inferolateral wall of the left ventricle, measuring 19.7 mm, non-obstructive, with preserved ejection fraction, non-dilated left atrium, and no systolic anterior motion (SAM) of the mitral valve (Figure 4). Based on the ACC/AHA and ESC guidelines, the indication for a dual-chamber implantable cardioverter-defibrillator (ICD) was established for secondary prevention.

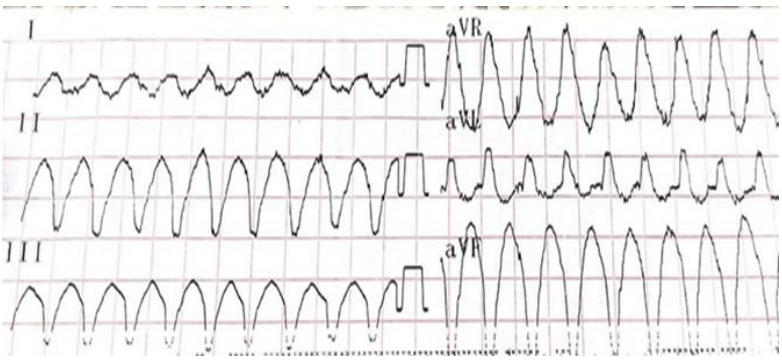
Discussion

HCM is a primary myocardial disease that can present with various clinical and anatomical features. It is defined by myocardial hypertrophy exceeding 15 mm in at least one myocardial segment in individuals without a family history, and more than 13 mm in those with a family history of HCM (1). It is the leading cause of sudden death (SD) in young individuals. The annual incidence of sudden death exceeds 4% in high-risk individuals (2), most commonly due to ventricular tachycardia (VT) or ventricular fibrillation (VF). The most frequent etiology is sarcomeric cardiomyopathies with autosomal dominant inheritance (3). Risk stratification for sudden death is evaluated through two complementary approaches: the traditional approach based on five major and minor risk factors (Tables 1,2), and the ESC-recommended approach, which uses a risk calculator derived from 7 parameters (Figures 5,6). American and European guidelines classify patients differently regarding the indication for implanting an ICD for primary prevention. For secondary prevention, in those who have already experienced a recovered SD or sustained VT, all guidelines agree on the indication for ICD implantation, as the risk of recurrent sudden death is more than 10% per year (4,5) (Figure 7). Late gadolinium enhancement (LGE) on cardiac MRI is a marker of fibrosis, but its presence is common in HCM. It has been shown that extensive LGE, covering more than 15% of myocardial mass, doubles the risk of sudden death (8,9).

Conclusion

HCM is a high-risk genetic cardiomyopathy for sudden death, with ventricular arrhythmia as its primary mechanism, which is not controllable by antiarrhythmic medications. The need to stratify the risk of sudden death for primary prevention with ICD implantation, and the selection of symptomatic patients for secondary prevention, remain the best strategies for preventing arrhythmic complications.

Family follow-up with systematic screening is crucial for early diagnosis and appropriate management.



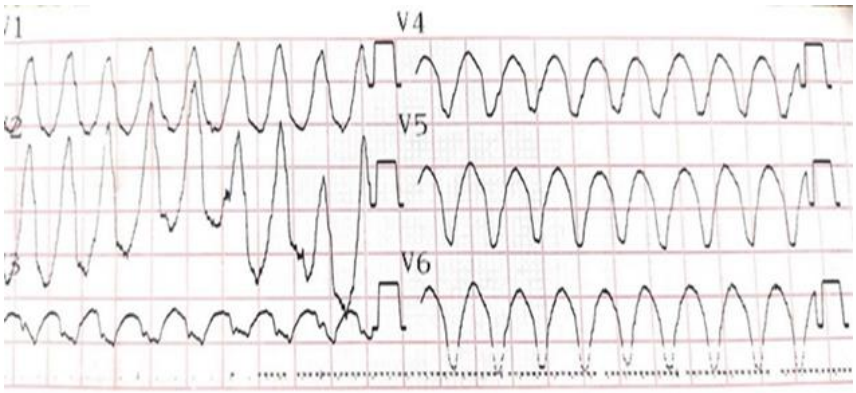


Fig 1 : Tachycardie Ventriculaire

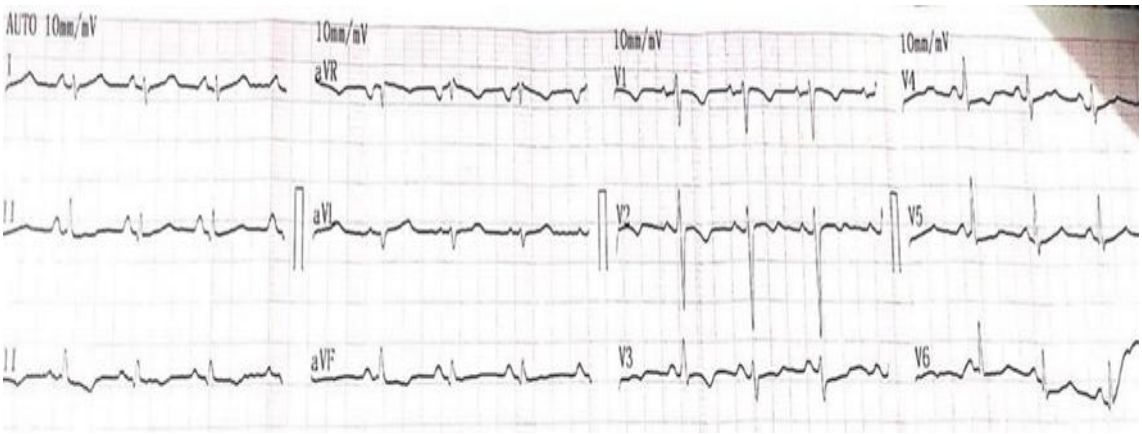


Fig 2 : Rythme Sinusal après CEE

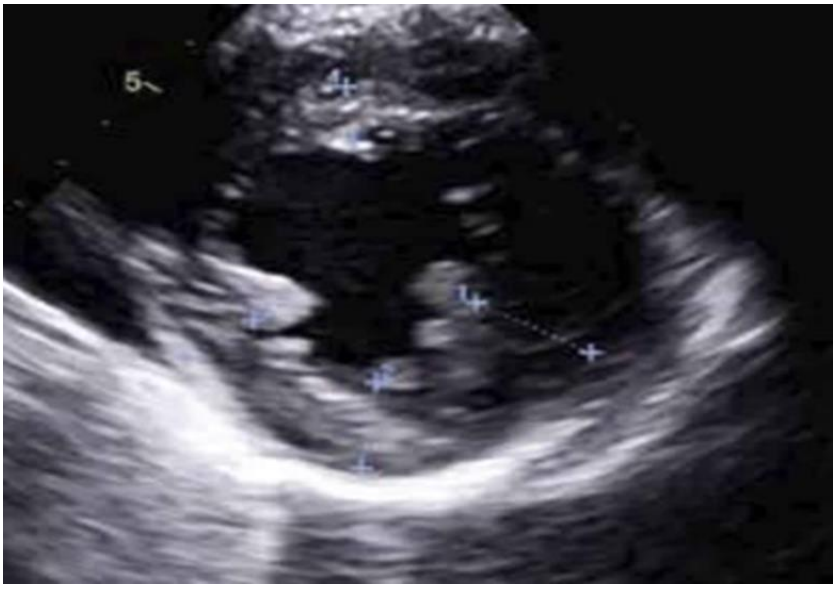


Figure 3: Parasternal short-axis view showing significant inferolateral left ventricular hypertrophy.

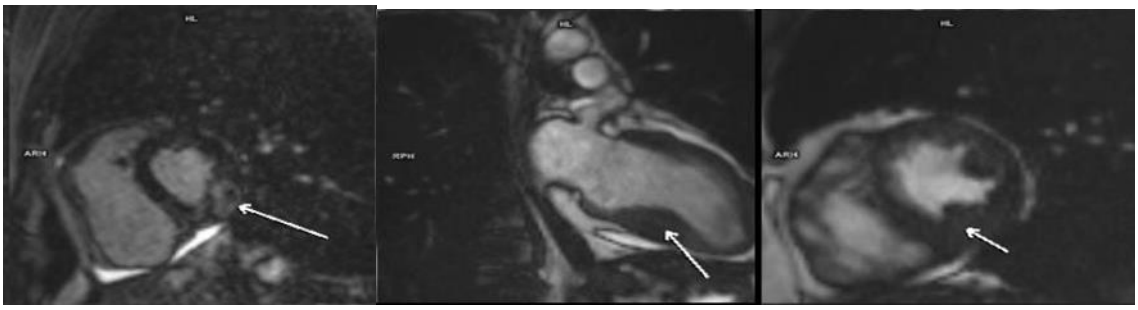


Figure 4: Cardiac MRI showing significant inferolateral hypertrophy of the left ventricle.

Table 1. Major Risk Factors for Sudden Death.

Major Risk Factors for Sudden Death:
• Syncope during exercise or in a young person. History of premature sudden death in the family. • Non-sustained ventricular tachycardia. • Abnormal blood pressure response during exercise stress testing. • Left ventricular wall thickness greater than 30 mm.

Table 2. Minor Risk Factors for Sudden Death.

Minor Risk Factors for Sudden Death:
• Onset of symptoms during childhood. • Documented myocardial ischemia. Isolated subaortic obstruction ≥ 30 mm Hg. • Detection of a so-called high-risk mutation (such as Arg719Gln and Arg403GlnT).

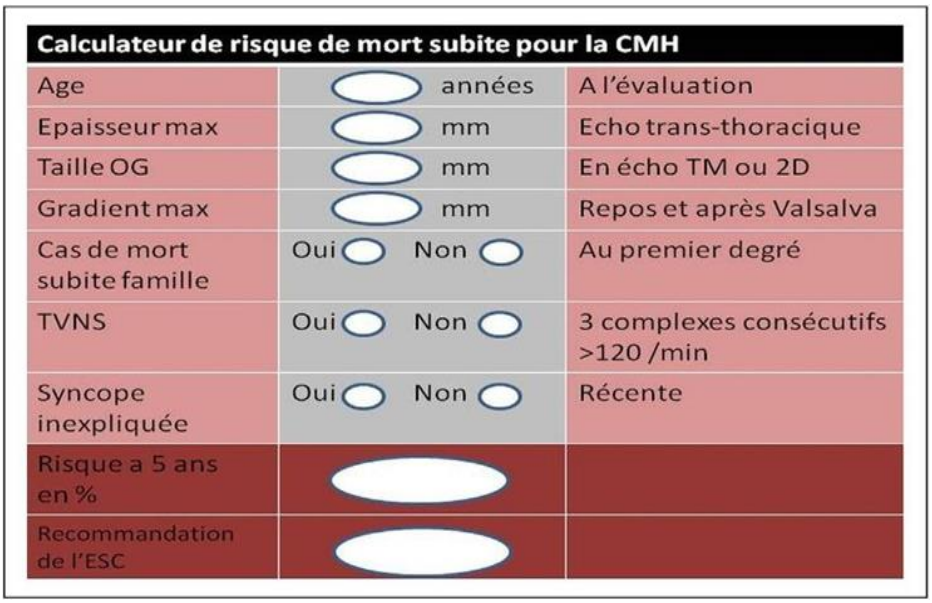


Figure 5: 5-year risk calculator according to the 2014 ESC guidelines.
LA: left atrium; NSVT: non-sustained ventricular tachycardia.

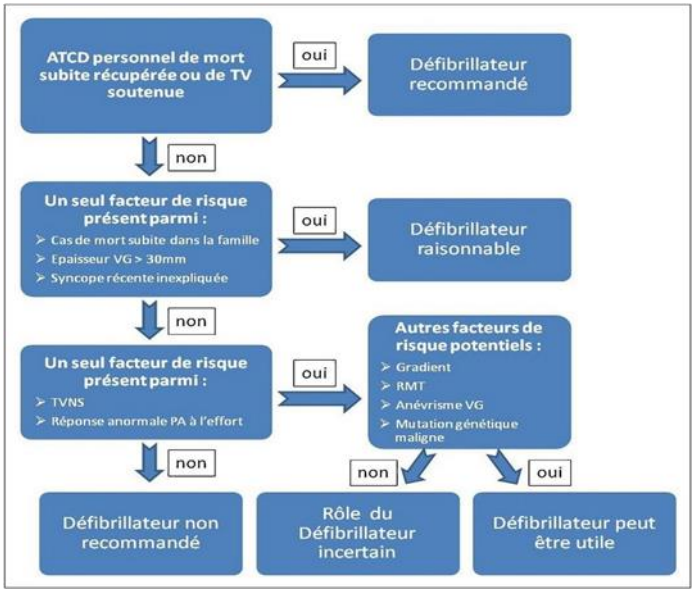


Figure 6: 5-year risk calculator according to the 2014 ESC guidelines.
LA: left atrium; NSVT: non-sustained ventricular tachycardia.

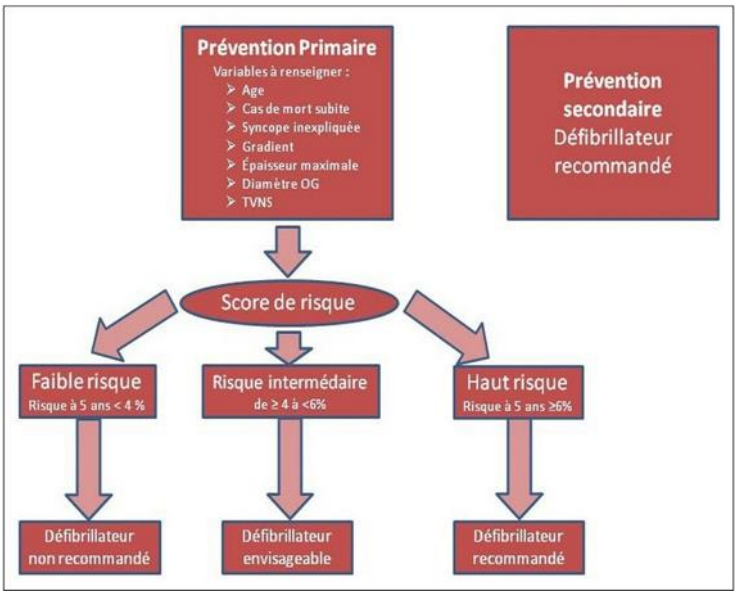


Figure 7: Indication for a defibrillator according to the 2014 ESC guidelines.

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