

Complicated Hypertrophic Cardiomyopathy of Complete Ventricular Atrio Block: About a Case in a 42-Year-Old Patient

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Received: March 28, 2022

Published: April 11, 2022

Abstract

Hypertrophic Cardiomyopathy (MHC) complicated atrioventricular block (AVM) is very rare. The clinical case we report involves a 42-year-old patient with a family history of MHC who experienced syncope of effort. The Electrocardiogram (ECG) shows a complete ventricular atrio block. The biological assessment in search of a metabolic or infectious cause of THE BAV is without particularity. Echocardiography revealed a predominant asymmetric hypertrophy to the septal wall without obstruction of the left ventricle. The management consisted of an implementation of a pacemaker with a good evolution. The treatment of MHC is based on the prevention of sudden death by the Implant Automatic Defibrillator (ICD).

Keywords: Hypertrophic cardiomyopathy; Ventricular atrio block; Echocardiography.

Introduction

Hypertrophic Cardiomyopathy (MHC) is a genetic disease with autosomal dominant inheritance for which complete atrioventricular block is a rare complication. MHC is one of the most common causes of sudden death in young people. Doppler echocardiography and cardiac MRI are the gold standard tests for the diagnosis of MHC. Genetic screening for MHC allows the detection of subjects carrying a mutation on the genes responsible for the disease, even before the development of symptoms [1]. Medical treatment is essentially symptomatic. Only the implantable defibrillator can prevent sudden death which is related to ventricular fibrillation to the effort and not to the obstruction.

We report the case of a patient with complicated non-obstructive MHC of complete atrioventricular block.

Clinical Case

Patient aged 42 years without cardiovascular risk factors, nor notion of drug intake and having a family history of MHC in three of his brothers with a notion of sudden death in one of them, admitted for management of a syncope of effort. The initial electrocardiogram (ECG) records a BBG and BAV 1st degree (Figures 1, 2). Echocardiography showed a predominant concentric hypertrophy at the interventricular septum (SIV) of 20.2 mm in diastole (Figure 1), a fraction of ejection of the Left Ventricle (LVEF) at 52.1% (Figure 2). During hospitalization, the evolution was marked by the occurrence of several epi-

sodes of syncope with the ECG the appearance of a complete ventricular atrio block requiring the emergency of an electro-systolic training probe followed by a pacemaker implantation.

Comment

Hypertrophic Cardiomyopathy (MHC) is an asymmetric hypertrophy of the left ventricle with or without obstruction that preferentially affects the interventricular septum that exceeds 15 mm thick. It is a genetic disease with autosomal dominant inheritance involving several genes. It is called familial hypertrophic cardiomyopathy in case of a family history of MHC. Its prevalence is 0.2% and affects both men and women with a male predominance. Patients with MHC are mostly asymptomatic but sometimes they may experience symptoms such as exercise dyspnea, palpitations and exertion syncope. The latter is of very bad prognosis. In more than 50% of cases the Electrocardiogram (ECG) is abnormal and shows hypertrophy of the left ventricle (HVG) or fine and deep q waves of pseudo necrosis. The holter ECG can record in 25% of cases bursts of unsupported ventricular tachycardia whose presence evokes a risk of sudden death. Cardiac Doppler ultrasound confirms the diagnosis by showing an HVG of more than 15 mm or 13 mm in case of familial MHC and asymmetrical character.

Unlike ventricular and supraventricular rhythm disorders, the occurrence of a complete atrioventricular block (BAV) requiring a pacemaker is a rare complication. Our patient benefited from a biological assessment that turned out to be normal

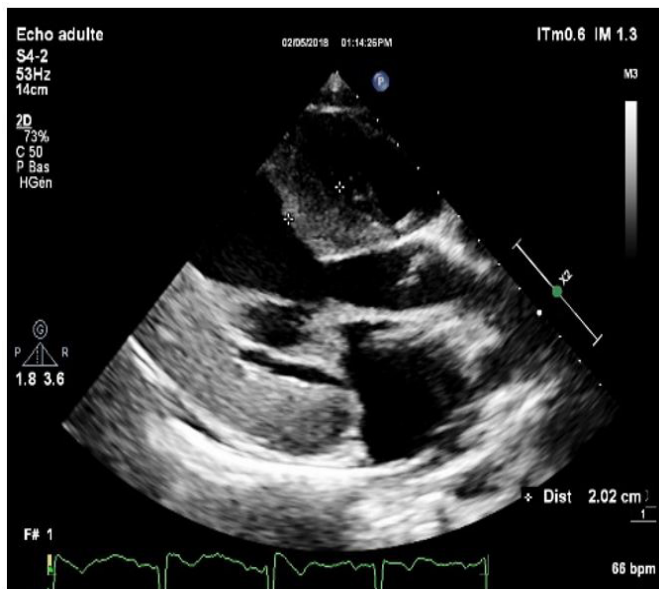


Figure 1: Broad Axis Parasternal Section Showing SIV Thickness.

which allowed us to eliminate a metabolic or infectious cause of BAV. On the other hand, certain elements of orientation of this clinical case such as the family history of MHC, the absence of high blood pressure and the age of the patient could suggest a family MHC. Some authors recommend genetic testing in patients with MHC and their relatives, but their use in the management of this patient has been limited by cost. Medical treatment for MHC is symptomatic and uses first-line beta-blockers. Studies by Fananapazir L and Jeanrenaud X have shown that implanting a double-chamber pacemaker would reduce the severity of the gradient of the left ventricular flow pathways and relieve symptoms during short- and medium-term follow-up periods [2,3]. Cases of complete BAV requiring permanent pacemaker implantation have been observed following surgery (5%) and after alcoholization (20%) for septal reduction in patients who remain symptomatic despite optimal medical treatment. The risk of developing a complete BAV is low (less than 5%) if the surface ECG shows no conductive disorder. However, it is very high if the patient is already a carrier of a BBG because in 50% of cases septal alcoholization is complicated by a BBD promoting the occurrence of a complete BAV [4]. Thus, this patient benefited from pacemaker implantation and monitoring for MHC in accordance

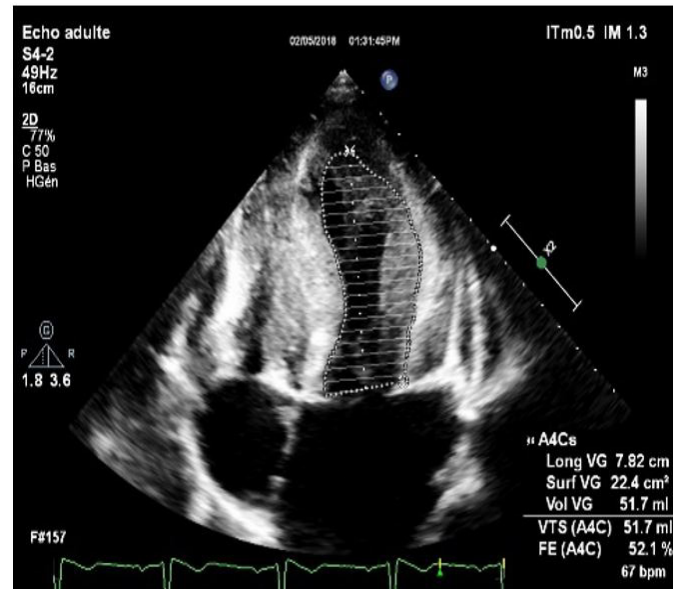


Figure 1: Apical section 4 cavities showing LVEF.

with current recommendations.

Conclusion

Familial MHC is the most common monogenic cardiovascular disease and has a very visible risk of sudden death. However, irreversible complete BAV remains a very rare complication. Management by surgical myectomy and alcohol ablation is generally used in patients who remain symptomatic despite optimal well-conducted medical treatment and a gradient greater than or equal to 50 mmHg at Doppler echocardiography.

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