Case report

Ventricular tachycardia on hemochromatosis in a young subject: about a case.

ABSTRACT

Background Hemochromatosis is a hereditary disease characterized by excessive iron accumulation in tissues, particularly affecting the liver, but also the heart. In young patients, this iron overload can induce serious cardiac complications, including ventricular arrhythmias.

Case presentation We present the case of a 16-year-old patient, diagnosed with hepatic hemochromatosis at the age of 10, without cardiological follow-up. He was admitted for palpitations and malaise. His EKG revealed ventricular tachycardia (VT), and echocardiography showed non-obstructive hypertrophic cardiomyopathy. The patient was treated with Cordarone to stabilize his heart rhythm, and implantable automatic defibrillator (ICD) implantation was considered.

Conclusion This case highlights the importance of cardiological follow-up in patients with hemochromatosis to prevent serious complications, such as VT. Prompt management with antiarrhythmic therapy and implantation of an ICD are crucial measures to improve prognosis.

Keywords*: Ventricular tachycardia, hemochromatosis, palpitations, case report*

1. INTRODUCTION

Hemochromatosis is an inherited disorder characterized by iron overload due to excessive intestinal absorption. Although hepatic manifestations are the most commonly described, this accumulation of iron also affects other organs, including the heart, joints and pancreas. Cardiac iron overload, also known as cardiac hemochromatosis, is a dreaded complication, as it can lead to cardiomyopathy, heart failure and rhythm disorders, including malignant ventricular arrhythmias such as ventricular tachycardia (VT) and ventricular fibrillation.

The cardiac involvement of hemochromatosis can manifest as dilated or hypertrophic cardiomyopathy, the latter being less frequent but just as dangerous. Excessive iron accumulation in the myocardium leads to myocardial fibrosis, which constitutes an arrhythmogenic substrate, increasing the risk of VT. In young patients, this complication can occur insidiously, making rigorous screening and cardiological follow-up essential.

We report here the case of a 16-year-old patient, diagnosed with hepatic hemochromatosis since the age of 10, with no notable cardiac history, who presented with symptomatic VT. This case highlights the importance of cardiological monitoring in hemochromatosis, as well as the challenges of managing arrhythmic complications in this young population.

2. CASE PRESENTATION

A 16-year-old male diagnosed with hepatic hemochromatosis at the age of 10. Although regularly monitored for liver function, he had never had a cardiological evaluation.

On admission, he reported frequent palpitations and intense fatigue. He also presented with several episodes of syncope.

The patient was admitted to the emergency department for palpitations and syncope. Physical examination revealed regular ventricular tachycardia with a rate of 170 beats per minute, correct blood pressure of 123/72 mmhg and no clinical signs of heart failure.

The EKG (figure1) revealed a regular monomorphic VT with large complexes, suggesting a threatening ventricular arrhythmia that had required rapid stabilization with Cordarone 300 mg followed by a maintenance dose of 900 mg/24h.

An EKG (figure 2) was repeated 30 min after the loading dose of cordarone, showing a return to sinus rhythm.

Transthoracic echocardiogram (TTE) (figure 3) revealed non-obstructive hypertrophic cardiomyopathy with a thickened interventricular septum of 19 mm. No evidence of outflow tract obstruction was detected.

As the patient stabilized, a cardiac MRI was performed as part of the etiological investigation, showing myocardial iron overload, confirmed by a marked reduction in T2*, indicating diffuse myocardial fibrosis.

The serum ferritin level was elevated to 2200 ng/mL and transferrin saturation reached i95%, confirming iron overload.

FIGURE 1. EKG REVEALED SUGGESTING A VENTRICULAR ARRHYTHMIA

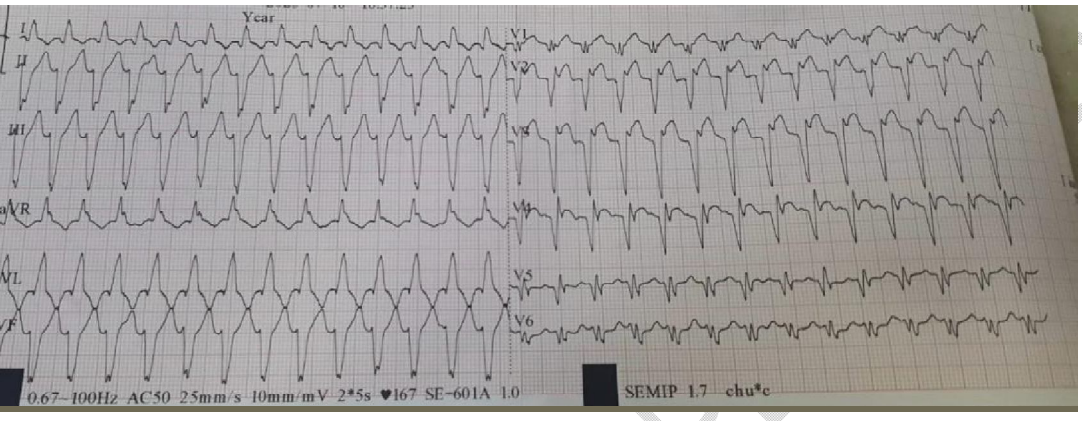


FIGURE 2. EKG SHOWING A RETURN TO SINUS RHYTHM

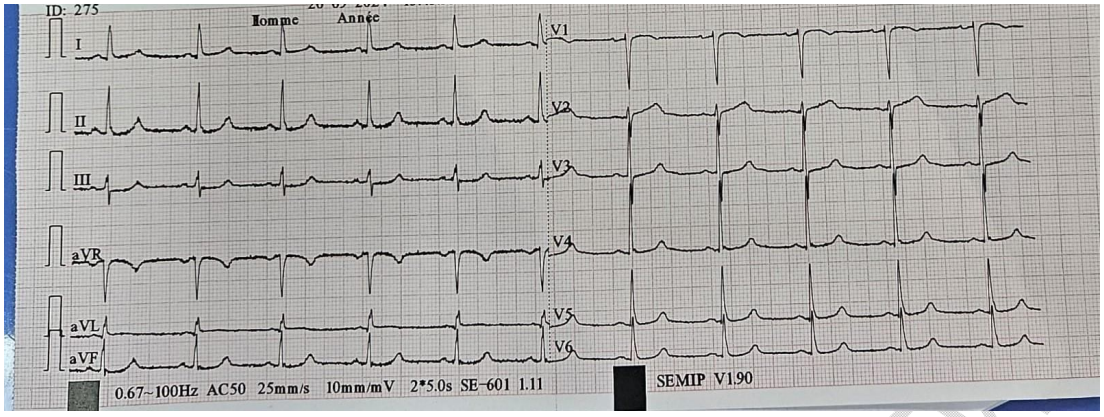
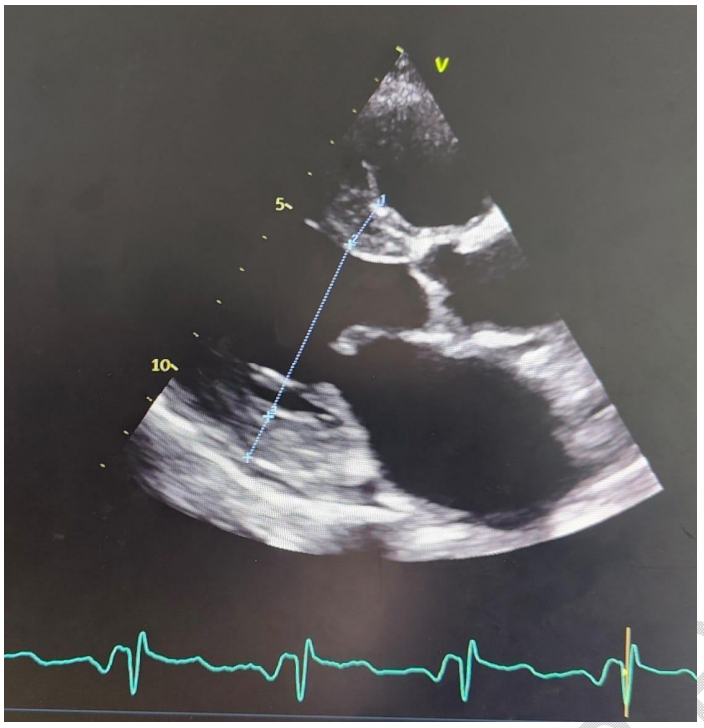


FIGURE 3. TTE REVEALED NON-OBSTRUCTIVE HYPERTROPHIC CARDIOMYOPATHY



3. DISCUSSION

Hemochromatosis primarily affects the liver, but iron can also be deposited in the myocardium, leading to structural and functional abnormalities. The development of cardiomyopathies, whether hypertrophic or dilated, is often a direct consequence of iron overload (1). In our patient, the non-obstructive hypertrophic cardiomyopathy detected by echocardiography and cardiac MRI is typical of the myocardial changes induced by hemochromatosis (2).

Myocardial fibrosis, a common result of excessive iron accumulation, creates an environment conducive to arrhythmias. These areas of fibrosis act as foci of abnormal electrical activity, increasing the risk of severe ventricular arrhythmias, such as VT (3). In this case, the VT observed is probably related to this fibrosis. In addition, non-obstructive hypertrophic cardiomyopathy may contribute to an arrhythmogenic substrate by disrupting myocardial structure and function (4).

The initial management of VT in this patient involved the administration of Cordarone, an antiarrhythmic commonly used to control ventricular arrhythmias. This drug was effective in reducing heart rate and stabilizing the patient. However, given the high risk of recurrence, particularly in the context of underlying cardiomyopathy and iron overload, implantation of an ICD was deemed necessary (5).

ICDs are crucial in preventing recurrence of VT and other life-threatening arrhythmias, such as ventricular fibrillation. Indeed, the presence of fibrosis and myocardial overload increases the likelihood of malignant arrhythmia recurrence (6).

This case highlights the importance of regular cardiological surveillance in patients with hemochromatosis, even in the absence of initial symptoms. Proactive monitoring with non-invasive cardiac imaging tests, such as T2* cardiac MRI, would

enable early detection of myocardial iron overload and optimize therapeutic management (7).

4. CONCLUSION

This case study demonstrates the importance of cardiological follow-up in hemochromatosis, particularly in young patients. Myocardial iron overload, although often initially asymptomatic, can rapidly progress to serious complications such as ventricular tachycardia. The management of this patient with Cordarone, followed by ICD implantation, illustrates the effectiveness of early arrhythmia management in this setting. Multidisciplinary follow-up is essential to improve long-term prognosis.

CONSENT : PATIENT CONSENT WAS OBTAINED PRIOR TO THE STUDY

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