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Case report

Ventricular tachycardia in a patient with cardiac haemochromatosis and normal left-ventricular ejection fraction

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Summary

We describe a rare case of ventricular arrhythmia in a patient with cardiac haemochromatosis related to iron overload in the myocardium and without evidence of systolic dysfunction or heart failure. This case stresses the utility of cardiac magnetic resonance imaging for the early identification of iron overload in the heart and starting appropriate treatment.

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Keywords: iron overload, cardiac arrhythmia, magnetic resonance imaging

Introduction

The deposition of iron in the myocardium can lead to heart failure and conduction disturbances. We describe the rare case of ventricular arrhythmia in a patient with cardiac haemochromatosis and normal ventricular ejection fraction

Case presentation

A 54 year-old man diagnosed of hereditary haemochromatosis, presented to the emergency department with sustained palpitations for approximately two hours. On physical examination, the systolic blood pressure was 120 mmHg and his heart rate was 225 bpm with peripheral pulse oximetry around 95%. No murmurs, rales or gallops were appreciated on cardiac examination.

His ECG (Fig. 1) showed a wide QRS complex tachycardia with a right bundle branch block and a frequency around 225 bpm. Following amiodarone administration, the tachycardia terminated with ECG showing sinus rhythm of 72 bpm (Fig. 2). Blood test revealed normal markers of myocardial necrosis levels and transferring saturation was 42.68% and serum ferritin was 767 ng/ml.

A transthoracic echocardiogram showed a normal biventricular ejection fraction with mild mitral and tricuspid regurgitation and normal diastolic function. To rule out coronary disease, a coronary angiography was performed. In this study, there were no significant coronary stenosis.

Cardiac magnetic resonance was performed in order to visualize cardiac haemochromatosis. The gradient echo sequences showed decreased focal signal in the mid infero-lateral segments of the left ventricle (Fig. 3) and a notably decreased liver signal. The functional sequences confirmed the presence of normal ventricular volumes and biventricular ejection fraction (left-ventricular end-diastolic and end-systolic volumes of 82.8 and 35.6 ml/m^2 and left-ventricular ejection fraction of 57%).

An electrophysiological study demonstrated normal atrioventricular conduction (AH 113 ms; HV 44 ms) and no accessory pathways. An aggressive ventricular tachycardia induction protocol was performed with drive trains of 8 paced beats (cycle lengths; 600 ms – 500 ms – 400 ms) with single, double and triple extrastimulus from

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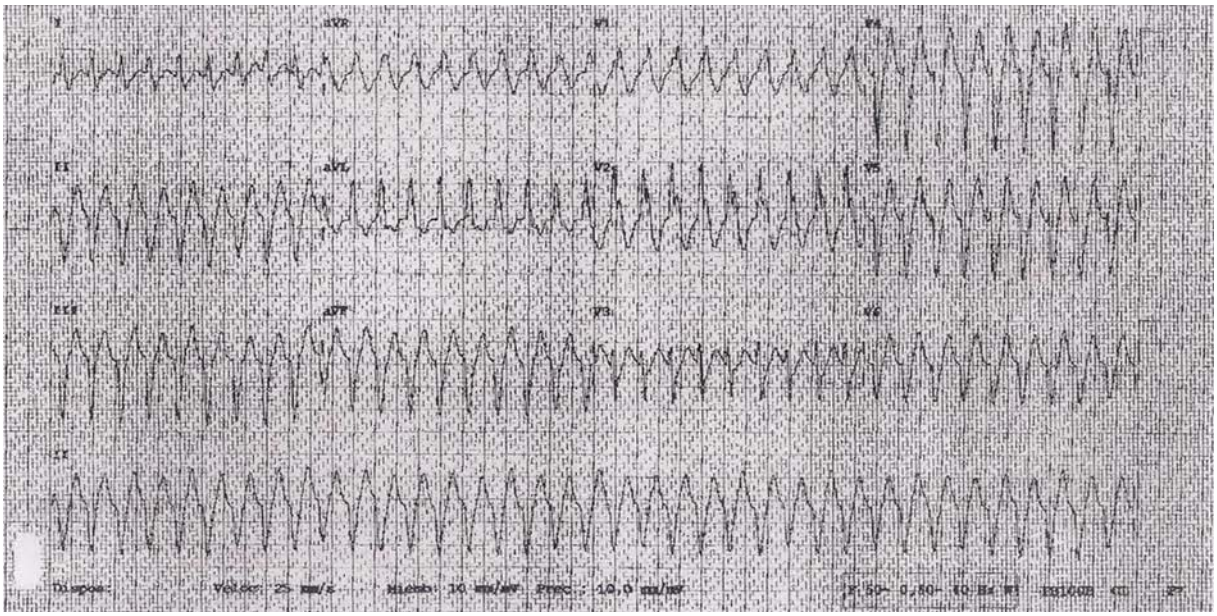

Figure 1. 12-lead electrocardiogram. Ventricular tachycardia.

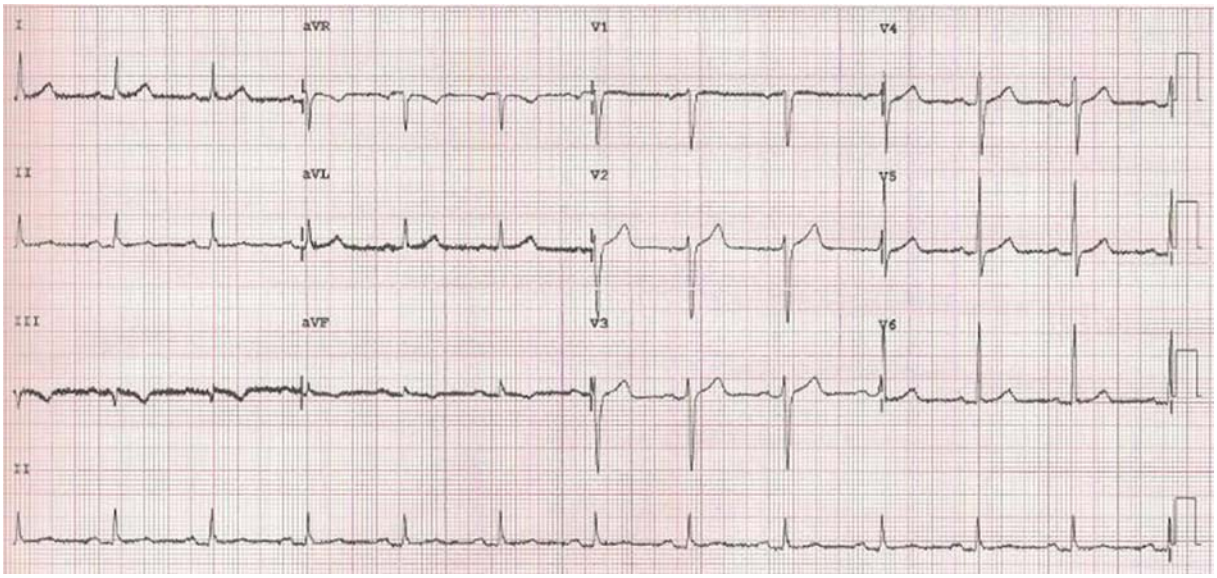

Figure 2. 12-lead electrocardiogram. Sinus Rhythm of 72 bpm. T wave inversion in leads aVF and III.

the right ventricular apex and outflow tract. No ventricular tachycardia was induced during the study.

We found correlation between the electrocardiographic origin of the tachycardia and the presence of iron in the myocardium and after all studies the patient was discharged with a 5 mg daily dose of bisoprolol and started regular phlebotomy treatment.

Discussion

Hereditary haemochromatosis is an autosomal recessive disorder due to mutations of the HFE gene, and characterized by excessive iron deposition in different tissues [1].

Iron loading in the heart usually occurs in the ventricular myocardium earlier than in the atrial myocardium [2], and it is initially characterized by diastolic dysfunction and arrhythmias. Cardiac manifestations due to iron deposition in the myocardium lead over time to a restrictive cardiomyopathy, symptoms of heart failure and conduction abnormalities [3].

The association of cardiac arrhythmia with hereditary haemochromatosis is well documented. The increased incidence of atrial and ventricular tachyarrhythmias in patients with iron overload may be explained due to the fact

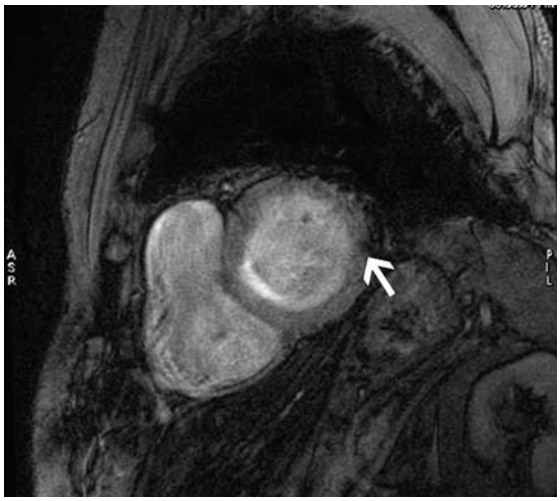

Figure 3. Cardiac Magnetic Resonance. Gradient-echo still. Short-axis view. Decreased focal signal in the mid inferolateral segments of the left ventricle (white arrow).

that iron is probably proarrhythmic by itself and the heterogeneous deposition of iron in the heart, leading to nonhomogeneity in conduction velocity or repolarization [4].

However, it is not well known whether patients with hereditary haemochromatosis present cardiac arrhythmias before signs and symptoms of heart failure occur. In fact, the incidence of ventricular arrhythmias is not statistically increased in asymptomatic patients with hereditary haemochromatosis [5].

Phlebotomy is the treatment of choice in patients with cardiac haemochromatosis who do not have anemia. Therapeutic phlebotomy removes 200–250 mg of iron per 500 ml of blood, and this mobilizes an equal amount of iron stored in the tissues to form haemoglobin. At the beginning phlebotomy is scheduled twice a week to once in two weeks as tolerated. Once a target ferritin level of less than 50 ng/ml is achieved, the frequency of phlebotomy is decreased [6].

We consider that our patient didn't meet high-risk criteria for sudden death (well-tolerated arrhythmia, normal left ventricle ejection fraction and non-inducible arrhythmias in the electrophysiological study) so low-dose beta-blocker therapy and regular phlebotomy treatment was started. After one year follow up, he is asymptomatic without recurrence of the arrhythmia.

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