WHICH IS YOUR DIAGNOSIS?

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A 42-year-old female patient, weighting 58 kg, 1.60 m in height, 74 bpm, blood pressure 140/80 mmHg, presenting a long history of abnormal findings on electrocardiograms and a familial history of heart disease has been referred to the Service of Radiology and Diagnostic Imaging at Hospital Pró-Cardíaco for undergoing a heart magnetic resonance imaging (MRI).

At physical examination, extrasystoles or arrhythmias have not been detected. An electrocardiogram (ECG) has demonstrated sinusal bradycardia, left ventricular hypertrophy and negative T wave. Echocardiogram (ECHO) has showed that the left ventricular (LV) global systolic function was normal (Simpson ejection fraction = 78%).

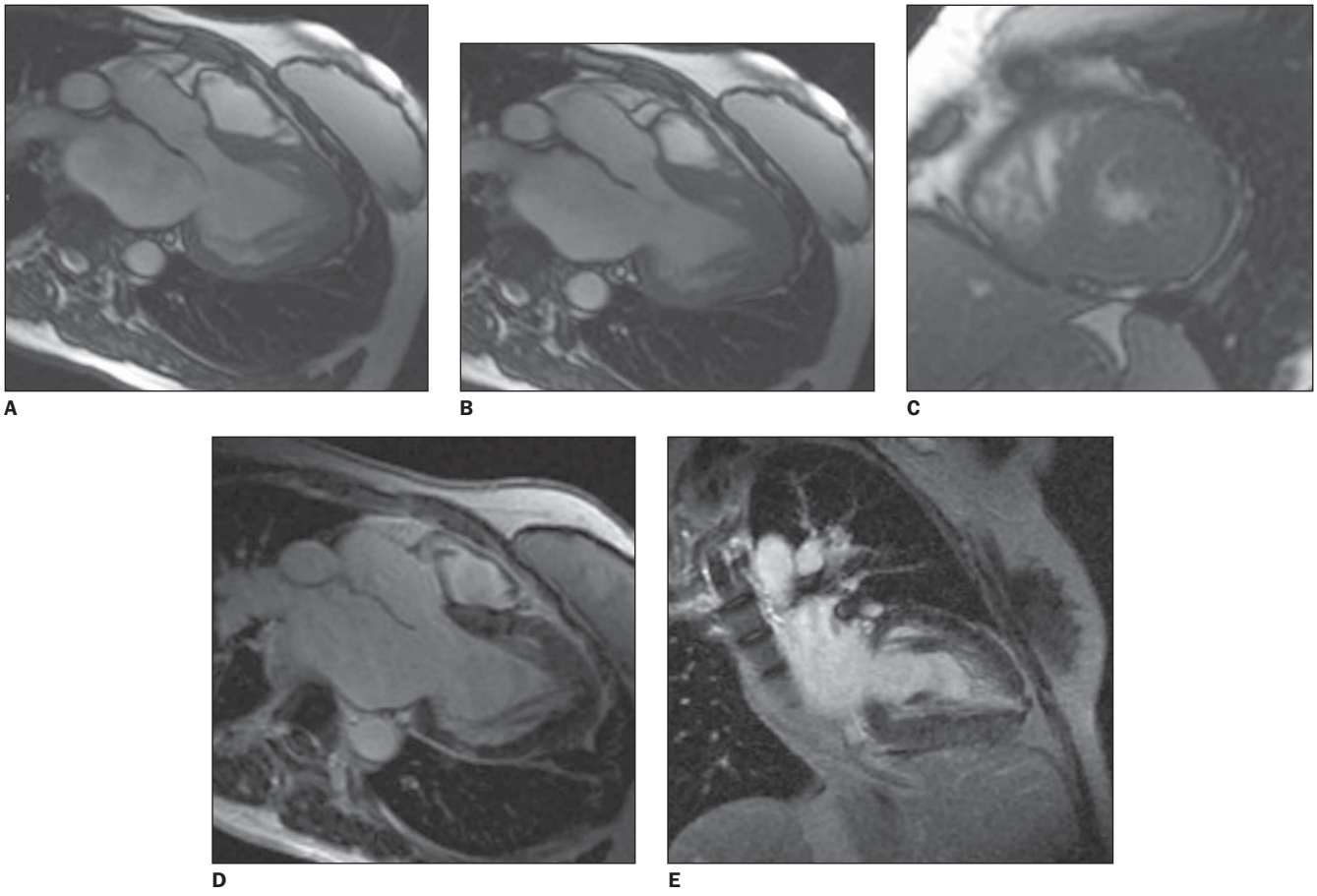


Figure 1. Images acquisition with ECG-gating, in cine Fiesta sequence (SSFP), T1FGRET and T1FGRIR, outflow tract plane diastole (**A**) and systole (**B**); short-axis (**C**); delayed enhancement – outflow tract (**D**) and two LV chambers (**E**).

Images description

Figure 1 – Acquisition with ECG-gating, in cine Fiesta sequence (SSFP), T1FGRET and T1FGRIR, outflow tract plane diastole (**A**) and systole (**B**); short-axis (**C**); delayed enhancement – outflow tract (**D**) and two LV chambers (**E**). Observe a mild concentric hypertrophy, with an increase of the myocardial thickness predominating in the apical and anteroseptal segments, associated with a moderate decrease the LV end relaxation and accentuated endomyocardial trabeculation.

Diagnosis: Apical hypertrophic cardiomyopathy (AHCM).

COMMENTS

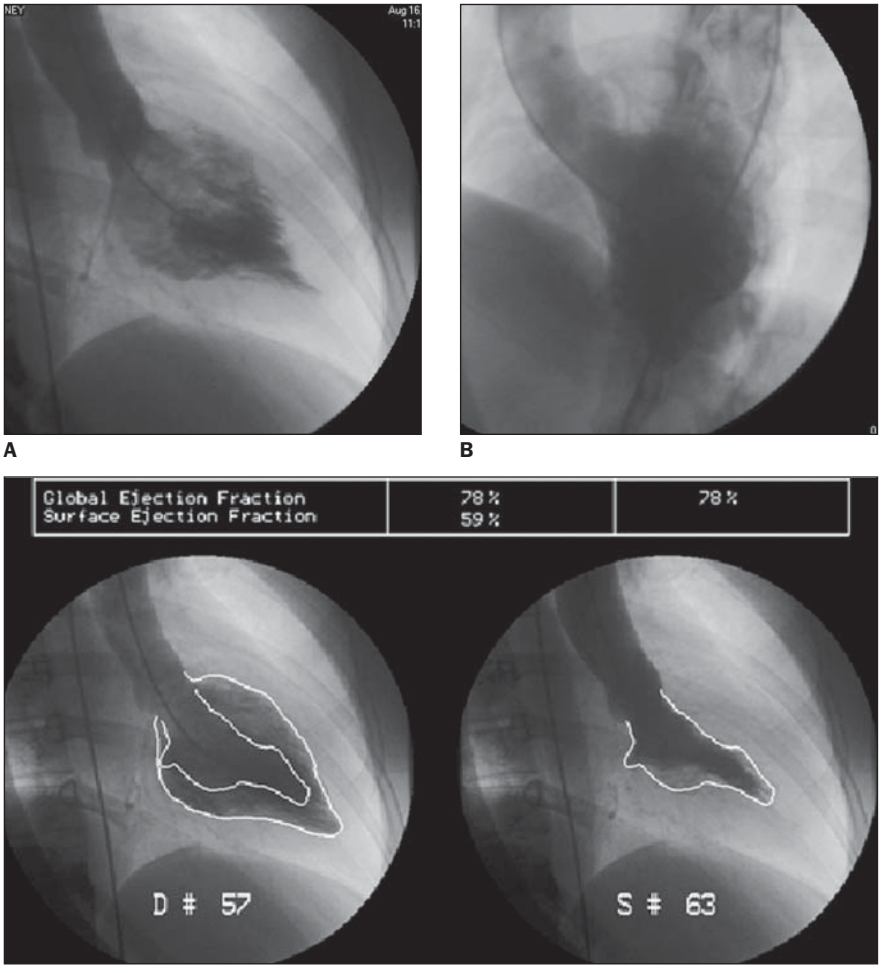
The AHCM is characterized by myocardial hypertrophy, predominantly from the apex to the left ventricle, and was first described in Japan, by Sakamoto *et al*.[1], in 1976, and universally disseminated by Yamaguchi *et al*.[2], in 1979.

The AHCM is a rare form of hypertrophic cardiomyopathy (HCM), with higher predominance amongst male Asians (5:1), with a lower ratio amongst Caucasians (2.5:1)[3]. The incidence ranges between 2% and 10% of patients in the general population, achieving 25% in the Japanese population. In Rio de Janeiro, a study performed by Albanesi Fº *et al*.[4] has found an incidence of 8.34%[3,4].

Despite the good prognosis, it is known that, in some cases, after long term follow-up periods, there is a possibility of evolution with severe arrhythmias, such as fibrilation or atrial flutter and ventricular tachycardia, myocardial infarction (with or without atherosclerosis), with formation of apical aneurysms or associated with severe mitral and/or tricuspid regurgitation and sudden death[4–6].

Typical findings of this disease are: a) large inverted T waves (> 10 mm) at ECG; b) the finding of "suit of spades" at left ventriculography (Figure 2).

From the histological point of view, an extensive disarrangement of myocardial fibers and myofribrillar and myocyte alterations are observed, more restricted to the ventricular end, indistinguishable from other forms of HCM, reinforcing the concept that the apical type is a HCM variant [7,8].



A B C

Figure 2. Catheter ventriculography (A,B,C) clearly demonstrating the typical "suit of spades" configuration (same case).

The echocardiogram has been the first-line method for evaluating patients with HCM, but its limitation in the study of the apex, mainly in the hands of inexperienced professionals, complicates the diagnosis of AHCM. The MRI of the heart should be performed in the suspect of AHCM, since this study minimizes these problems because it is not operator-dependent as much as ECHO, does not present any limitation to the window size, has multiplanar capacity and presents an excellent contrast between soft parts[4,7–9].

In athletes, it seems that exercises would increase the left ventricular (LV) wall stress and work, factors which could affect the development of the localized LV hypertrophy. Little reference is made to this hypothesis in the study of the AHCM, and up to this moment, no relation has been established between the sports practice and the onset of the disease[3,5,8].

The MRI of the heart has been performed for better studying the myocardium thickness, LV function and perfusion and, mainly, the myocardial enhancement, ruling out the likelihood of an associated myocardial fibrosis and, through a non-invasive method, excluding the possibility of endomyocardiofibrosis or amyloidosis.

With the huge development of the non-invasive cardiac study, mainly by means of new methods like MRI and, most recently, CT, we must be careful with these diagnoses, particularly in cases of asymptomatic patients presenting a familial history of heart disease.

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