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Left ventricular hypertrophy: hypertensive or hypertrophic cardiomyopathy? What a dilemma! A case report

Hipertrofia ventricular izquierda: ¿cardiopatía hipertensiva o miocardiopatía hipertrófica? ¡Vaya dilema! Reporte de caso

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Abstract

In the presence of the left ventricle hypertrophy (LVH), the differential diagnosis with hypertrophic cardiomyopathy (HCM) or some phenocopy must be always considered, which can be easily suspected when the hypertrophy is markedly asymmetric. However, when the hypertrophy is homogeneous, especially if the patient has concomitant hypertension, it may be a challenge to distinguish between hypertensive and HCM, although some clinical features may help us to suspect it. In addition, patients with HCM may present with exertional angina due to microcirculation involvement in the setting of the hypertrophy itself or dynamic obstruction in the left ventricular outflow tract, but in some cases, the presence of concomitant coronary artery disease must be suspected as the cause of angina, especially if the patient has an intermediate or high-risk probability of having ischemic heart disease. We present the case of a 46-year-old Afro-American man with poorly controlled hypertension who was found to have severe LVH, and who presented with symptoms of exertional angina during follow-up. We will review the clinical features that can help us in the differential diagnosis in this context.

Keywords: *Left ventricular hypertrophy. Hypertensive cardiomyopathy. Hypertrophic cardiomyopathy. Case report.*

Resumen

Ante la presencia de hipertrofia del ventrículo izquierdo (HVI), siempre se debe considerar el diagnóstico diferencial con la miocardiopatía hipertrófica (MCH) o alguna fenocopia, que puede sospecharse fácilmente cuando la hipertrofia es marcadamente asimétrica. Además, los pacientes con MCH pueden presentar angina de esfuerzo debido a la afectación de la microcirculación en el contexto de la propia hipertrofia o si ésta condiciona obstrucción dinámica al tracto de salida del ventrículo izquierdo, pero en algunos casos debe sospecharse la presencia de enfermedad coronaria concomitante como causa de la angina, especialmente si el paciente tiene una probabilidad de riesgo intermedio o alto de padecer cardiopatía isquémica. Presentamos el caso de un varón de 46 años de afroamericana con hipertensión arterial mal controlada a quien se le detectó una HVI severa, y que durante el seguimiento presentó síntomas de angina de esfuerzo. Revisaremos las características clínicas que nos pueden ayudar en el diagnóstico diferencial en este contexto.

Palabras clave: *Hipertrofia ventricular izquierda. Miocardiopatía hipertrófica. Cardiopatía isquémica. Reporte de caso.*

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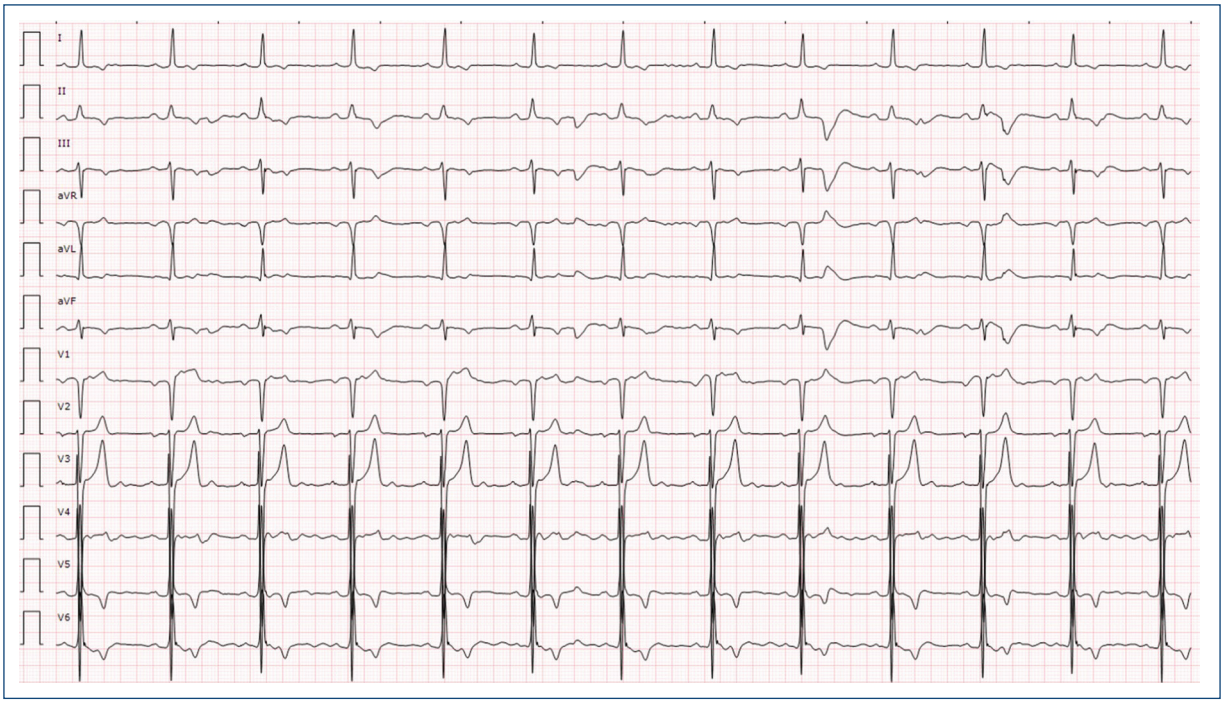

Figure 1. ECG showing significant signs of the left ventricle hypertrophy and systolic overload.

Introduction

Hypertrophic cardiomyopathy (HCM) presents with marked left ventricular hypertrophy (LVH), frequently asymmetric, but sometimes it can also be homogeneous, which can be difficult to diagnose, especially in hypertensive patients. Furthermore, patients with HCM may present with exertional angina due to the pathophysiology of the cardiomyopathy itself, but in some cases the presence of concomitant coronary artery disease should be suspected, especially if the patient has asociated cardiovascular risk factors.

Case report

A 46-year-old man from Senegal with a personal history of hypertension (HT) secondary to primary hyperaldosteronism, with poor control and multiple associated vascular complications (retinopathy, nephropathy, and ischemic stroke), was studied previously due to an ECG (Fig. 1) with significant signs of the left ventricle hypertrophy (LVH) and systolic overload, showing severe LV hypertrophy in the echocardiogram, as well as rheumatic involvement of the aortic valve without functional repercussion. As a family history, highlights sudden death in two of his four brothers, at the ages of 21 and 29 years, respectively, as well as of his father at 52 years of age.

Due to frequent travel to his native country, he was lost to follow-up without being able to complete the study. A few years later, follow-up was resumed performing transthoracic and transesophageal echocardiogram (Fig. 2) which again described the presence of severe concentric LVH of up to 23-25 mm with involvement of the right ventricle (RV), with a non-dilated LV, normal biventricular systolic function, without dynamic obstruction of the LV outflow tract, as well as significant aortic regurgitation due to rheumatic involvement of the aortic valve (trivalve), with no other remarkable findings. Given the presence of such severe LVH and the family history of sudden death, it was decided to complete the study with magnetic cardiac magnetic resonance (CMR), which showed diffuse late enhancement uptake with subendocardial pre-dominance.

With these findings and his family history of sudden death, after ruling out other phenocopies such as amyloidosis, a cardiac scintigraphy with pyrophosphates was also performed without evidence of uptake suggestive of transthyretin amyloidosis. Furthermore, the CMR did not meet the criteria for amyloidosis, and the patient also did not present involvement of other organs that would support the diagnosis, beyond renal failure that

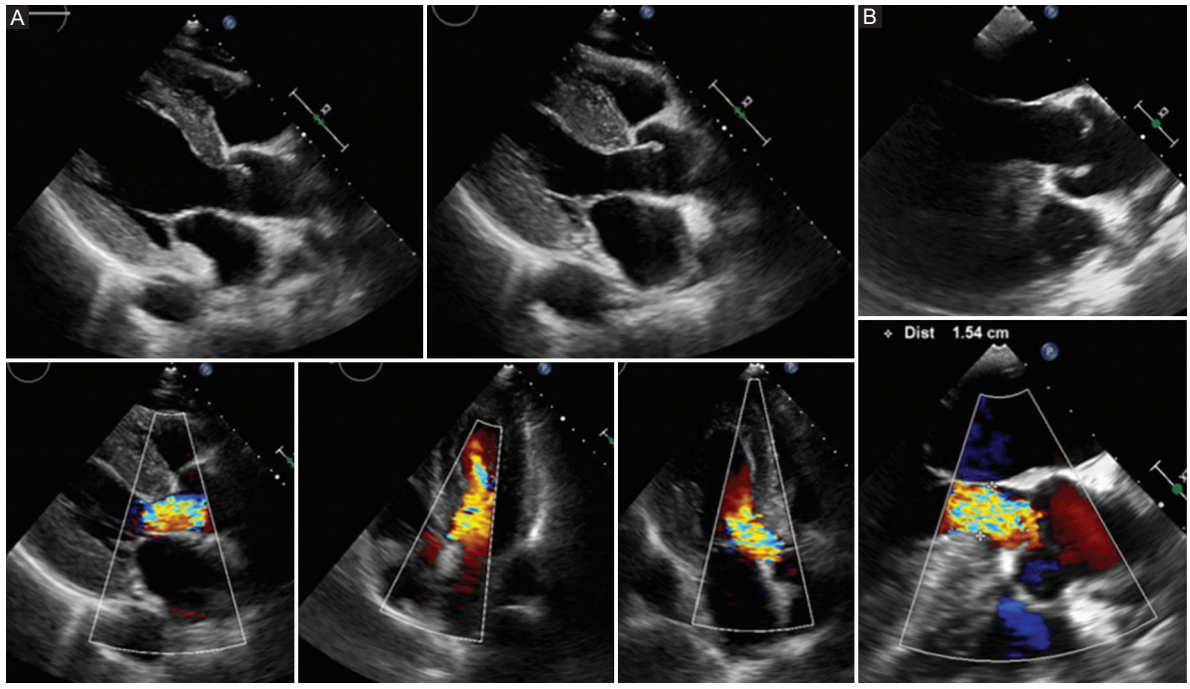

Figure 2. **A:** transthoracic and **B:** transesophageal echocardiogram showing severe concentric LVH of up to 23-25 mm with involvement of the right ventricle, with a nondilated the left ventricle and normal biventricular systolic function, as well as significant aortic regurgitation due to rheumatic involvement of the aortic valve.

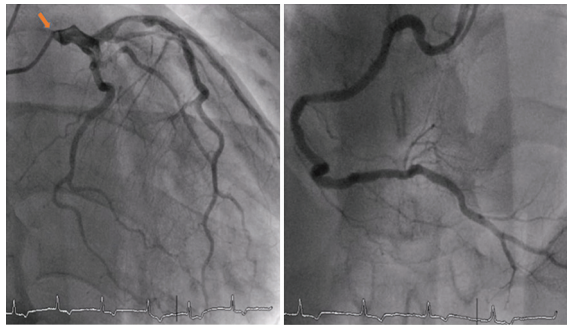

Figure 3. Coronary angiography evidencing critical lesion in the ostium of the left main (orange arrow), with no other coronary stenosis.

we assumed that it was secondary to longstanding severe HT) or fabry disease (a genetic test for alpha-glucosidase was performed, which was negative. In addition, the fibrosis pattern by CMR was not suggestive of fabry disease either). The patient was diagnosed with hypertrophic cardiomyopathy (HCM). A few months later, the patient presented with symptoms consistent with progressive angina on exertion. Although the patient had a HCM that could justify it, it was decided to perform a coronary angiography (Fig. 3) that showed a critical lesion in the ostium of the left main, for which he was preferably referred for cardiac surgery. A double aortocoronary bypass graft with internal mammary artery to anterior descendent, and the left radial artery to first marginal obtuse as well as aortic valve replacement with a mechanical prosthesis were performed. As post-operative complications, the patient presented an episode of atrial fibrillation in the first 24 h that was reversed with amiodarone and dressler's syndrome with severe the left pleural and moderate-severe pericardial effusion that required drainage. The patient presented subsequent slow but favorable evolution with anti-inflammatory treatment with ASA, prednisone, and colchicine.

Since then, the patient showed adequate blood pressure control, but with the persistence of severe LVH several months later. Three years later, the patient presented recurrent syncope as well as the appearance of the left bundle branch block on the ECG (Fig. 4). A 24 hour-ECG recording was performed showing little ventricular extrasystole without other arrhythmias, so an electrophysiological study was performed, which showed normal atrioventricular conduction, but

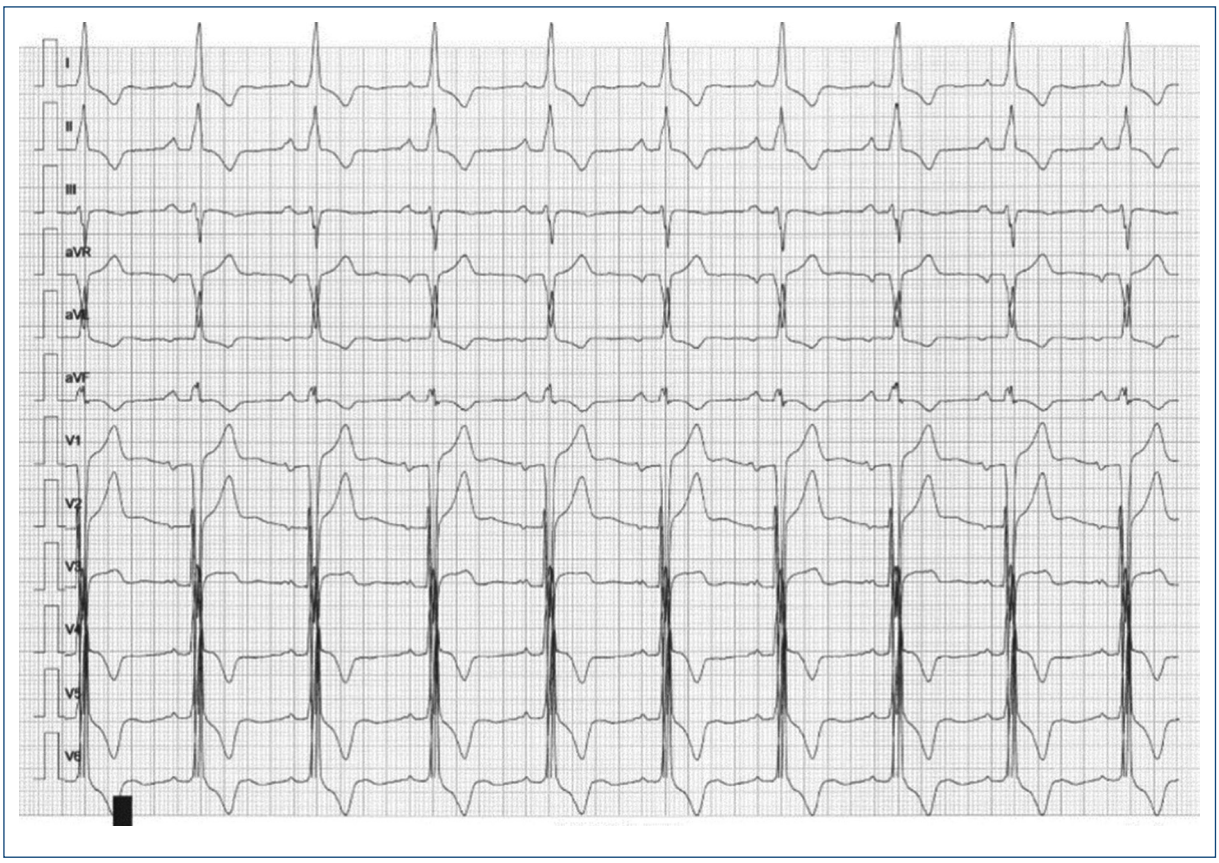

Figure 4. ECG with left bundle branch block.

polymorphic ventricular tachycardia was induced, so it was decided to implant an implantable cardioverter defibrillator. Since device implantation, the patient has had no further syncope and no events or therapies have been recorded on the device.

Discussion

HCM is an inherited cardiomyopathy with an autosomal dominant pattern and a high prevalence, affecting 1:200-1:500 individuals[1]. Most patients are asymptomatic, so it should be suspected in the presence of marked signs of LVH and systolic overload on the ECG, even if the patient has baseline hypertension. The initial study involves performing an imaging test such as 2D echocardiography or CMR which shows a maximal end-diastolic wall thickness of ≥ 15 mm anywhere in the left ventricle, in the absence of another cause of hypertrophy in adults, although lower parietal thicknesses (13-14 mm) can be diagnostic when present in family members of a patient with HCM or in conjunction with a positive genetic test[2-4]. The distribution of parietal thickness is variable, it may be limited to a few segments of the LV or it may affect it more globally, conditioning a concentric LVH, as in the case of our patient, in which case the differential diagnosis with hypertensive heart disease can be a challenge, since in both cases there may be marked ventricular hypertrophy as well as diffuse myocardial fibrosis. Our patient also presented hypertrophy of the RV, which is not affected by hypertension, in addition to the persistence of very severe LVH several months later despite correct blood pressure control, as well as a family history of sudden death, findings that are more compatible with HCM[5-7]. In addition, patients with HCM can present myocardial ischemia due to a mismatch between myocardial oxygen supply and demand. Myocardial hypertrophy, microvascular dysfunction with impaired coronary flow reserve, and medial hypertrophy of the intramural arterioles and their reduced density are common findings[4]. Although the new-onset exertional angina of our patient could be explained by HCM, it was

decided to perform a coronary angiography since he had a moderate pre-test probability of having coronary artery disease, confirming the presence of a critical lesion in the left main, which coexisting with the presence of severe LVH with extensive fibrosis, confers a very poor prognosis with a high probability of adverse cardiovascular events.

Conclusion

In summary, in the presence of concentric LVH, the possibility of an underlying HCM or any other phenocopy must always be suspected, even if the patient has some cause that increases the afterload that may justify it, especially if in the ECG shows marked signs of LVH with systolic overload, and if LVH persists after controlling the possible precipitating cause, in addition to family history of sudden death. It is also important to keep in mind the possibility of underlying ischemic heart disease in a patient with HCM who presents with exertional angina and has a moderate or high probability of having coronary artery disease.

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Conflicts of interest

None.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that no patient data appear in this article. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors declare that no patient data appear in this article. The authors have obtained the informed consent of the patients and/or subjects referred to in the article.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript, nor for the creation of images, graphics, tables, or their corresponding captions.

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