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Case Report Open Access

A Case of Symptomatic Hypertrophic Cardiomyopathy with Severe Dynamic Gradient

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Abstract

Hypertrophic cardiomyopathy is a disease of the myocardium characterized by the thickening of the heart walls. It is a genetic disorder transmitted by first-degree relatives, caused by mutations in genes that code for sarcomere protein. A 77-year-old woman presented to the emergency department and complained of continuous vertigos and presyncope with subsequent fall to the floor with a concussive traumatic brain injury. The ECG observed signs of left ventricular hypertrophy. Blood exams highlighted a slight increase of troponin T (25 ng/L). The ultrosonography instead demonstrated a pronounced left ventricular hypertrophy with a 240 mmHg dynamic gradient, a moderate mitral regurgitation secondary to SAM.

Lastly, ECG Holter excluded significant arrhythmias (1 couple of ventricular contractions). In the ward, the echocardiographic pattern was repeated and confirmed the clinical picture of a pronounced ventricular hypertrophy at the expense of the IVS (VTD 45 ml), a diastolic dysfunction Grade I with anterior systolic movement of the mitral flap valves with a dynamic obstruction to the left ventricular outflow (DP max 260 mmHg). Moreover, a moderate-severe mitral insufficiency was reported. Vital signs were stable.

The antihypertensive therapy was enhanced with added ACE inhibitors, loop diuretics, beta blockers and the disopyramide administration started. The patient was discharged from the hospital and was scheduled a cardiologic echocardiographic follow up in order to evaluate the effectiveness of the therapy. This study has demonstrated the association between the clinical symptom, the clinical objectivity and the echocardiographic anomalies typical of the obstructive hypertrophic cardiomyopathy. Severe blood pressure values of dynamic obstruction to the left ventricular outflow have been recorded and which are not currently known by the scientific literature. Our purpose will be to evaluate the clinical and echocardiographic follow up and the potential response to the pharmacological therapy prescribed.

Keywords: Hypertrophic cardiomyopathy; Echocardiogram; Dynamic gradient

Introduction

Hypertrophic cardiomyopathy is a disease of the myocardium characterized by the thickening of the heart walls. It is a genetic disorder transmitted by first-degree relatives, caused by mutations in genes that code for sarcomere proteins with an incidence of 1:500 in general adult population. More than 450 mutations are currently known in 13 genes associated with the hypertrophic cardiomyopathy.

The principal genes concerned are those encoding the heavy chain of myosin, the protein C binding myosin and troponin T. Since it is a genetic disorder passed on from parent to child, it is recommended that first-degree relatives undergo cardiac screening periodically. Most of the patients affected by HCM is asymptomatic or mildly symptomatic. Some patients develop symptoms such as chest pain, breathing difficulties, palpitations, presyncope and/or syncope [1-5].

The main causes of such symptoms are the diastolic dysfunction, myocardial ischemia, arrhythmias and - in this case - the so-called left ventricular outflow tract obstruction. Such circumstance occurring under resting conditions in about 30% of patients and in a higher percentage during the physical activity, is characterized by the thickening of the interventricular wall protruding into the left ventricular outflow that provokes a stenosis defined subaortic (just below the aortic valve).

The systolic blood flow, due to the Venturi effect, drags behind the anterior mitral leaflet causing the so-called systolic anterior motion (or SAM), resulting in the obstruction of the arterial outflow and in the mitral insufficiency [6]. The objective examination during the auscultation often reveals the presence of a meso-systolic murmur accentuated by Valsalva maneuver. There are different exams able to establish the diagnosis of the disease and its severity: the 12-lead ECG is the first exam that detects the presence of potential arrhythmias and other anomalies secondary to the cardiac hypertrophy. The echocardiogram is the principal exam for the diagnosis and detects the potential left ventricular outflow tract obstruction (Figures 1-5).

In most cases the disease has a benign natural history. Some patients can develop symptoms that may require a pharmacological intervention and, in the most severe cases, invasive treatments such as the myotomy-myectomy procedure or the alcohol septal ablation. In those patients who are at high-risk of dangerous ventricular arrhythmias, the automatic defibrillator may be appropriate as a

precautionary measure. A smaller portion of patients is eligible for cardiac transplantation [7,8].

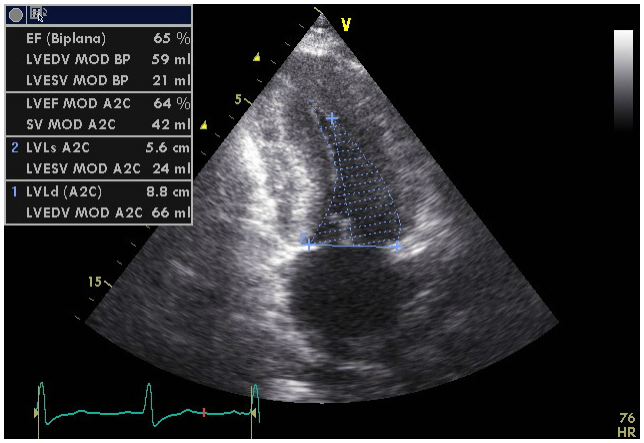

Figure 1: Left ventricular cavity systolic-function.

In selected cases, the cardiac catheterization is necessary for an invasive measurement of pressures inside the heart.

The drugs normally used for the hypertrophic cardiomyopathy are beta-blockers, calcium-antagonists (verapamil, diltiazem) and disopyramide. Some patients may need diuretics. A small portion of patients with left ventricular outflow tract obstruction is still very symptomatic, despite the optimal medical therapy for which the myectomy or the septal ablation may be appropriate.

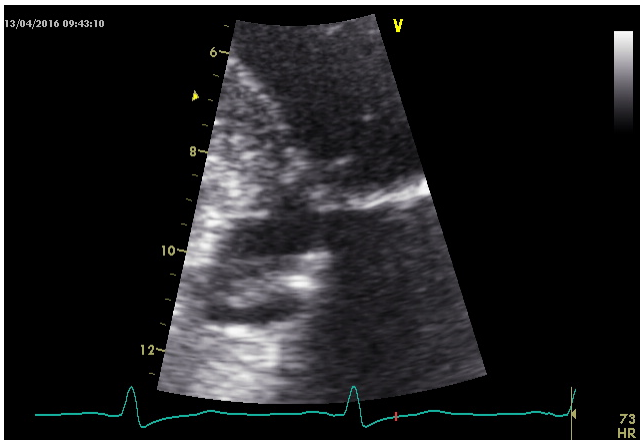

Figure 2: Left ventricular outflow obstruction due to septal hypertrophy and SAM.

Purpose

Identification among the patients presenting to the emergency department of the associations between the typical symptoms, the objectivity and potential echocardiographic anomalies typical of the hypertrophic cardiomyopathy.

Materials and Methods

A 77-year-old woman presented to the emergency department and complained of continuous vertigos and presyncope with subsequent fall to the floor with a concussive traumatic brain injury with lacerated and contused wounds treated with three nylon 3.0 stitches. The patient informed to suffer from arterial hypertension, Parkinsonism and anxiety-depressive disorder. Her case history also reveals a computed tomography coronary dated 2011 with a coronary tree free from atheromatous lesions [9-13].

Pharmacological treatment made with quetiapine, lioresal, paroxetine, alprazolam, rotigotine, bisoprolol. The objective examination recorded a holosystolic murmur at centrum cordis irradiated to the aortic region. The ECG observed signs of left ventricular hypertrophy. Computed tomography of the head and the chest radiograph showed a cardiac silhouette at the upper limits of normal. Blood exams highlighted a slight increase of troponin T (25 ng/L). The echocardiographam instead demonstrated a pronounced left ventricular hypertrophy with a 240 mmHg dynamic gradient, a moderate mitral regurgitation secondary to SAM (Figures 1-3).

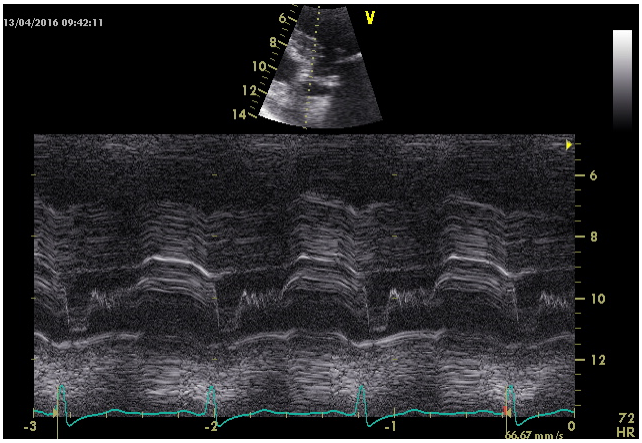

Figure 3: M-Mode profile of the left ventricular outflow with protosystolic opening of the aortic valve cusps and meso/telesystolic fluttering.

As a consequence, the patient was admitted to the hospital at the Department of Cardiology for an in depth-analysis with a symptomatic obstructive hypertrophic cardiomyopathy diagnosis.

In the ward, the echocardiographic pattern was repeated and confirmed the clinical picture of a pronounced ventricular hypertrophy at the expense of the IVS (VTD 45 ml), a diastolic dysfunction Grade I with anterior systolic movement of the mitral flap valves with a dynamic obstruction to the left ventricular outflow (DP max 290 mmHg) (Figures 4 and 5). Moreover, a moderate-severe mitral insufficiency was reported. Vital signs were stable [14].

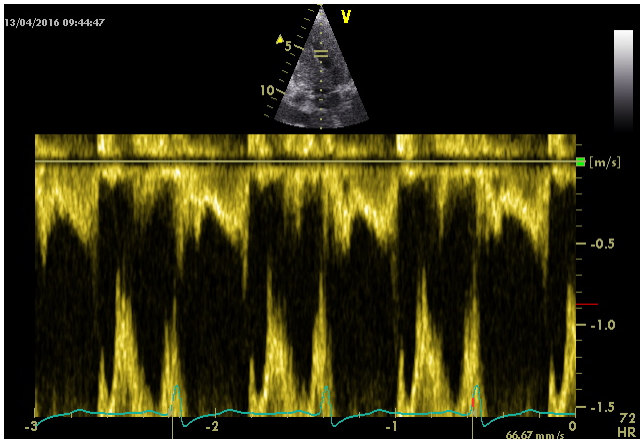

Figure 4: Pulsed Doppler in the middle of the left ventricular cavity.

Lastly, ECG Holter excluded significant arrhythmias (1 couple of ventricular contractions).

The antihypertensive therapy was enhanced with added ACE inhibitors, loop diuretics, beta blockers and the disopyramide administration started. The patient was discharged from the hospital and was scheduled a cardiologic echocardiographic follow up in order to evaluate the effectiveness of the therapy [15-17].

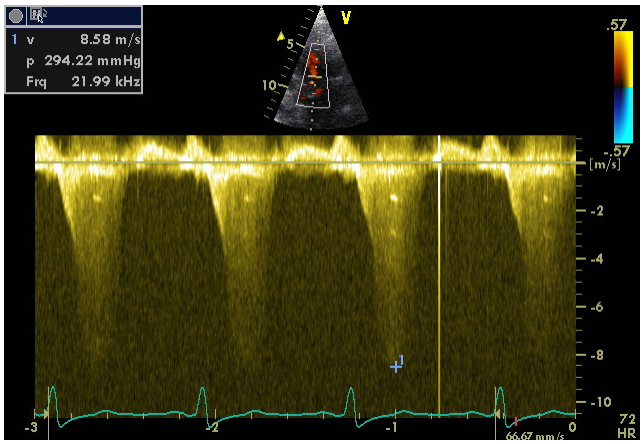

Figure 5: Continuous wave Doppler.

Conclusions

This study has demonstrated the association between the clinical symptom, the clinical objectivity and the echocardiographic anomalies typical of the obstructive hypertrophic cardiomyopathy. Severe blood pressure values of dynamic obstruction to the left ventricular outflow have been recorded and which are not currently known by the scientific literature. Our purpose will be to evaluate the clinical and echocardiographic follow up and the potential response to the pharmacological therapy prescribed.

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