

Apical Hypertrophic Myocardopathy Mimicking Acute Coronary Syndrome. Contribution of Three-Dimensional Echocardiography

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ABSTRACT

The apical hypertrophic myocardopathy is characterized by the presence of predominant hypertrophy localized to the left ventricular apex. This type of hypertrophic myocardopathy usually presents favorable clinical outcomes and a benign prognosis. Nevertheless, the typical presence of significant alterations in the electrocardiogram may mimic the existence of coronary artery disease. In this sense, image tests have great clinical implications to arrive at a definite diagnosis.

We present a case report of apical hypertrophic myocardopathy in a context of chest pain and difficulties to perform a diagnosis; the advantages of three-dimensional echocardiography for the assessment of this disorder are described.

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Key words > Apical hypertrophic myocardopathy – Three dimensional echocardiogram

Abbreviations > ECG Electrocardiogram | AHM Apical hypertrophic myocardopathy

CASE REPORT

Male patient, aged 29, with controlled hypertension (stage I of JNC 7) and a history of recent hospitalization in another facility for oppressive chest pain associated to significant alterations in ECG, which was diagnosed as unstable angina. The transthoracic echocardiography reported anteroseptal hypokinesia in apical segments, and the myocardial perfusion test result was normal. The patient was therefore discharged, medicated with aspirin, beta blockers, angiotensin converting enzyme inhibitors, and lipid lowering drugs.

Fifteen days later, the patient consults in our center for a new event of chest pain, resulting in his hospitalization.

The ECG (Figure 1) showed ST elevation of 3 mm in DII, DIII, aVF and negative asymmetric T in V2 to V4, DI and aVL (no changes regarding previous ECG), with cardiac enzymes CK and CK MB and negative troponin T.

A new transthoracic echocardiogram performed during his hospitalization showed a suspected image at left ventricular apex level that suggested apical hypertrophy (Figure 2A). In addition, a 3D echocardiography was performed (Figure 2B), which clarified the diagnosis and made it possible to better characterize hypertrophy morphology by the segment measurement of the ventricular walls. The myocardial hypertrophy was located in the apical anterior and lateral segments, with a maximum thickness of 27 mm (Figure 3). The left ventricular mass was estimated in 162 grams.

The 24-hour ECG Holter did not evidence atrial or ventricular arrhythmias. The patient was treated with atenolol, and he was discharged.

DISCUSSION

The AHM is a morphologic type of hypertrophic myocardopathy, characterized by the presence of predominant hypertrophy localized at cardiac apex level. Since the prevalence of hypertrophic myocardopathy (HM) is between 1/500 and 1/1000, this variable stands for about 20% in Japanese series, and 2% to 3% in the rest of the population. (1)

It is an autosomal dominant disorder caused by the mutation of one of the 13 possible genes present in chromosomes 7, 11, 14, and 15, which codify different sarcomeric proteins, resulting in its characteristic heterogeneity regarding the apical hypertrophy degree and distribution. (2)

The pathognomonic aspect presents deep negative T waves in the ECG precordial derivations; this finding is one of the main causes of suspected diagnosis in asymptomatic patients. Clinically, it can occur with angina (16%), atypical pain chest (14%), palpitations (10%), dyspnea (6%), and presyncope/syncope (6%). (3, 4)

An accurate diagnosis is clinically relevant, since the AHM prognosis is generally benign, with a published annual mortality lower than 0.1%. (3) The demonstration of the asymmetric ventricular hypertrophy at apex level, with an apical thickness of ≥ 15 mm

and a relation between the maximum apical thickness and the posterior wall of ≥ 1.5 mm, are the necessary elements that image tests provide to be able to diagnose. (4)

The bi-dimensional echocardiography with harmonic image is the first diagnostic tool, which must be supplemented with a contrast echocardiography when there is high clinical suspicion with non-diagnostic echocardiogram or bad acoustic window. (5)

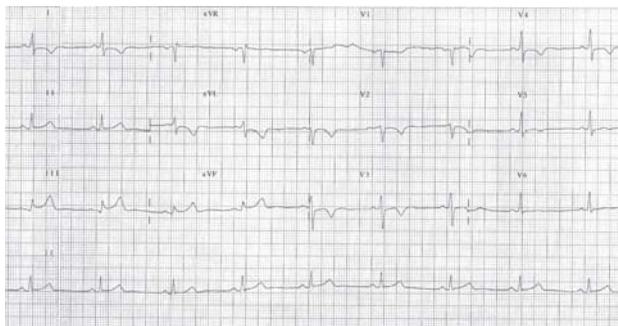


Fig. 1. Surface electrocardiogram that shows negative T in anterolateral wall (DI-aVL, VI-V4).

The magnetic resonance imaging (MRI) is the most sensible and specific test to diagnose that entity due to its high tissue definition. It measures thicknesses more precisely, with a correct grading and hypertrophy extension assessment. (6)

In this case report, the apical hypertrophy had not been first observed through bi-dimensional echocardiography, probably because of the difficulties caused by the assessment of the cardiac segments. Then, on a second chest pain event, a new 2D echocardiography was performed showing a suspicious apical image that suggested a diagnosis of AHM. It was the tri-dimensional echocardiography that determined the right diagnosis, since it was possible to perform the exact section in the heart through the real cardiac apex, usually difficult through conventional apical echocardiographic images. On the other hand, it was possible to perform planes of section in the apex on different space points, resulting in a better morphologic characterization of the hypertrophy distribution and measurement of maximum thicknesses. (7) This technique is particularly useful in morphologic forms with hypertrophy localized in an area of the left ventricle, as in the case of our patient, where we clearly detected that the apical anterior and lateral segments were the ones affected, with no involvement of the rest of the walls. Finally, the tri-dimensional echocardiography also

Fig. 2. A. 2D echocardiography with suspected image in apex. **B.** 3D echocardiographical image with multiplane sections that show the asymmetric distribution of hypertrophy.

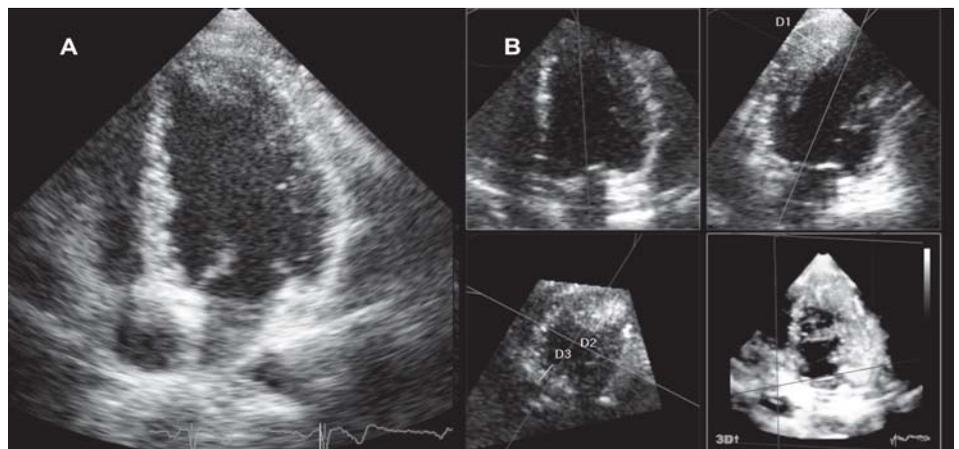
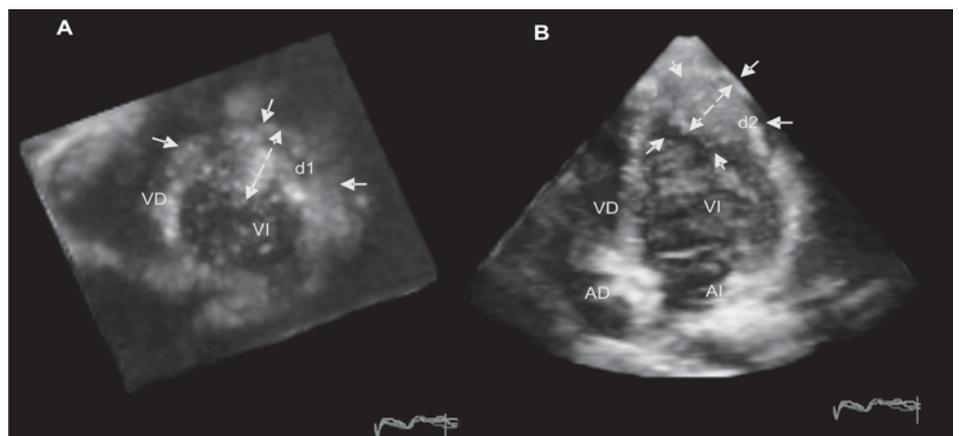


Fig. 3. See the segment distribution of the apical hypertrophy. **A.** Section of the VI from the apex, where the maximum hypertrophy of the anterior and anteriolateral segment is evidenced. $d1 = 26$ mm. **B.** Image of four chambers with elimination of the inferior area of the heart. Note the disproportionate hypertrophy of the apical segment on the lateral wall. $d2 = 27$ mm.



provides interesting data regarding the accurate calculation of the left ventricular mass. (8)

CONCLUSION

The AHM should be suspected in patients with pathological ECG associated to deep negative T waves in anterior precordials. These alterations may mimic the presence of coronary artery disease, so the definite diagnosis by image tests is clinically relevant. The bi-dimensional echocardiography is the first diagnostic tool; the three-dimensional echocardiography provides additional information in order to determine the segmental apical hypertrophy localization, and an accurate quantification of the left ventricular mass.

RESUMEN

Apical Hypertrophic Myocardopathy Mimicking Acute Coronary Syndrome. Contribution of Three-Dimensional Echocardiography

La miocardiopatía hipertrófica apical es una entidad que se caracteriza por la presencia de hipertrofia predominante de los segmentos apicales del ventrículo izquierdo. Esta forma de miocardiopatía hipertrófica en general presenta una evolución clínica y un pronóstico benignos. Sin embargo, la típica presencia de alteraciones significativas del electrocardiograma puede simular la existencia de enfermedad coronaria, motivo por el cual el diagnóstico definitivo por técnicas de imagen tiene una gran implicación clínica. En esta presentación se describe un caso de miocardiopatía hipertrófica apical en el contexto de dolor precordial y difi-

cultades diagnósticas, en el que se demuestran las ventajas de la ecocardiografía tridimensional para su evaluación.

Palabras clave > Miocardiopatía hipertrófica, apical - Ecocardiograma tridimensional

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