Miocardiopatia Hipertrófica Apical: Um Caso de Imagem

Apical Hypertrophic Cardiomyopathy: An Image Case

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An eighty-six-year-old female patient with a previous history of myocardial infarction, uncharacterized heart failure and permanent atrial fibrillation was admitted in an Internal Medicine Department due to acute decompensated heart failure. Electrocardiogram (ECG) was notable for the presence of left ventricle hypertrophy (LVH) voltage criteria and biphasic T waves / T wave inversion in leads V3 to V6 (Fig. 1). After instituting diuretic therapy and achieving euvolemia, a transthoracic echocardiogram was performed, which revealed isolated apical hypertrophy of the left ventricle (LV), with a wall thickness of 16 mm (Fig. 2); anomalous apical displacement of papillary muscles, an "ace-of-spades" configuration of the LV cavity (Fig. 3), elevated filling pressures (mean E/e' 18) with grade III diastolic dysfunction and an LV ejection fraction of 55%. These data allowed us to establish a diagnosis of apical hypertrophic cardiomyopathy (HCM).

Apical HCM is a morphological subtype of HCM, which was first described in Japan, in 1976.[1] Although its prevalence has been difficult to estimate, some studies report that apical HCM accounts for 25% of all HCM cases in Asian populations and around 1%-10% in non-Asian populations.[2] Mortality appears to be like classical HCM, with an annual cardiac death rate of up to 4%.[2,3] Although there are no specific criteria for diagnosing apical HCM, it can be defined as the presence of apical LVH (≥15 mm) and a ratio of maximal apical to posterior wall thickness ≥1.5, based on echocardiography or cardiac magnetic resonance.[4] Apical HCM may coexist with septal hypertrophy – mixed form.[4] A relative apical HCM form may be diagnosed, when LVH criteria are not met but there is a loss of apical wall thickness tapering, due to apical wall exceeding basal wall thickness – this is thought to be an early disease phase.[4]

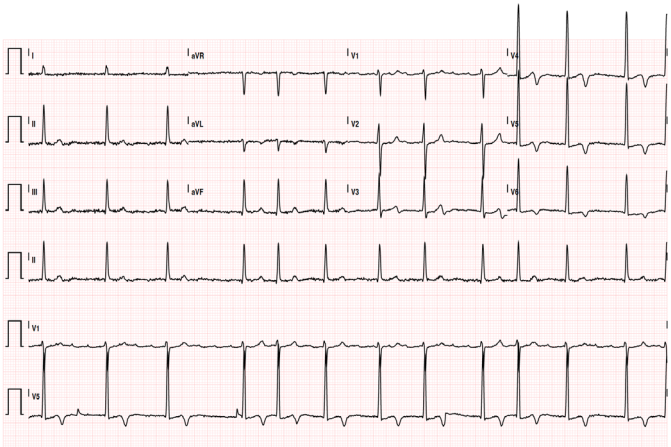

Figure 1. ECG showing LVH criteria (R in V5 + S in V1 >35mm) and biphasic T waves / T wave inversion in leads V3-V6 (red circles)

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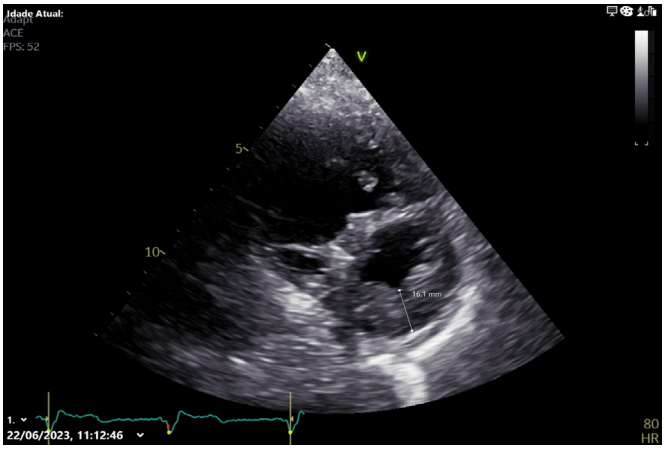

Figure 2. Short axis view displaying apical hypertrophy (16mm)

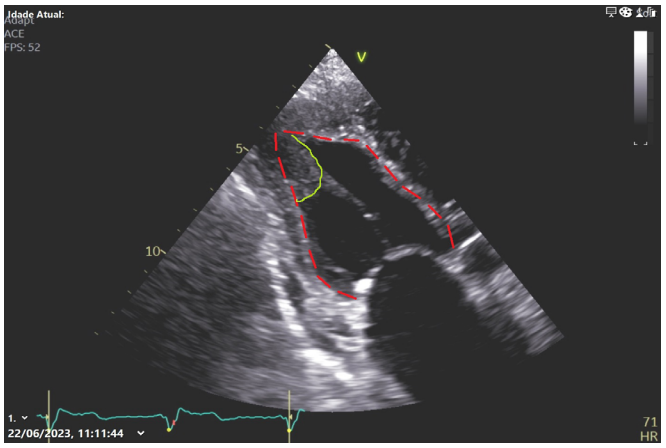

Figure 3. Three-chamber view depicting "ace-of-spades"-like configuration of LV cavity (red dashed line) and apical displacement of papillary muscle (green line)

One of the most typical findings of apical HCM on ECG is giant T wave inversion (≥10 mm); however, it is not mandatory for establishing the diagnosis, as they are not always present and are found in other diseases.[4] ECGs of most of the patients with apical HCM exhibit LVH voltage criteria and/or T wave inversion (<10 mm) on ECG.[4] Additional features that may be found on imaging include an "ace-of-spades" configuration of the LV cavity and apical displacement of the papillary muscles, the latter oftentimes preceding the development of hypertrophy.[5] Other findings such as diastolic dysfunction, midventricular obstruction and cavity obliteration, or apical aneurysms may be present and indicate worse prognosis[4].

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