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Multimodality Imaging in Hypertrophic Cardiomyopathy Associated With Anomalous Hypertrophied Papillary Muscles: A Case Report

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Introduction: Multimodality imaging can help rule in/out the diagnosis of hypertrophic cardiomyopathy (HCM) in patients with significant left ventricular (LV) hypertrophy.

Case Presentation: We describe a 73-year-old woman referred to us for consultation because of a giant negative T wave on her electrocardiography. Echocardiography revealed diffuse severe hypertrophy associated with hypertrophied anterolateral papillary muscles with a bifid head and with extensive wall insertion into the apicolateral segment. Three-dimensional echocardiography and cardiac magnetic resonance confirmed these data. Importantly, automated function imaging determined the global longitudinal strain at -10.2%.

Conclusions: According to our multimodality imaging approach, hypertrophic cardiomyopathy was the most probable diagnosis.

Keywords: Multimodality Imaging; Hypertrophy; Left Ventricular; Anomalous Papillary Muscle; Global Longitudinal Strain

1. Introduction

Multimodality imaging can help rule in/out the diagnosis of hypertrophic cardiomyopathy (HCM) in patients with significant left ventricular (LV) hypertrophy.

HCM is a primary disease of the cardiac muscle characterized generally by the typical asymmetrical hypertrophy of the LV, in the absence of another cardiac or systemic disease to account for the observed abnormality (1). Non-invasive cardiovascular imaging, especially echocardiography, has advanced our knowledge about the morphological aspects and also abnormalities of the mitral valve in HCM. The most reported morphological abnormalities of the papillary muscles are their anomalous insertions and hypertrophy (2). In the presence of a giant negative T wave in the precordial leads on 12-lead electrocardiograms (ECG), a diagnosis of apical HCM has often been made in patients with apical hypertrophy on two-dimensional (2D) echocardiography. However, based on literature review, an extensive insertion of the papillary muscles into the apical segment, a situation referred to as apically displaced papillary muscles, may also be characterized by a negative T wave on 12-lead ECG, rendering the differentiation between these two conditions difficult. Apicoseptal hypertrophy would be helpful in this regard (3).

Recently, 2D strain or speckle tracking imaging has conferred a robust and rapidly evolving technology that has enhanced our understanding of regional myocardial mechanics in HCM.

We herein report a case of HCM associated with the anterolateral papillary muscle hypertrophy with a large extensive wall insertion into the apicolateral segment, increasing the possibility of an error in the measurement of the LV thickness.

2. Case Presentation

We describe a 73-year-old woman with a history of controlled diabetes type II and hypertension grade I treated with captopril. She had chest tightness and palpitations of 5 months' duration. Her family history showed no sudden cardiac death. She was referred to our hospital for an echocardiography checkup because of her ECG abnormalities.

On admission, her blood pressure was 120/70 millimeter Hg and heart rate 86 beats/minutes with regular rhythm. ECG found sinus rhythm with LV hypertrophy (Cornell index = 34 millimeter), left auricular hypertrophy, giant negative T wave in V4 - V5, I, aVL, II, and aVF.

2D transthoracic echocardiography demonstrated a preserved biventricular systolic function (LV ejection fraction = 63% and right ventricular tricuspid annular plane systolic excursion = 20 millimeter) and symmetric hypertrophy of all the segments except the apical segment of the septal wall. The maximum wall thickness was 21 millimeter.

In the parasternal short-axis view, the anterolateral papillary muscle were severely hypertrophied (Figure 1). It had a bifid head and an extensive wall insertion into the apical and mid segments of the lateral region (Figure 2). No abnormality regarding the insertion of the chordae tendineae was noted, and nor were there any LV outflow tract obstruction and mitral regurgitation. There was, however, an abnormal relaxation of the LV. The atrial volume was 44 milliliter/square meter. The mitral annulus S' and e' velocity on tissue Doppler imaging was low (4.5 centimeter/second). The LV global longitudinal strain, according to automated function imaging, was -10.2%, which significantly contributed to the final diagnosis. The border between the free wall of the LV and the abnormal papillary muscle insertion was delicately visible and was clear on three-dimensional (3D) echocardiography as well (Figure 3).

Cardiac magnetic resonance (CMR) underscored the diagnosis of HCM with diffuse hypertrophy, involving 16 of the 17 segments, as well as the anterolateral papillary muscle hypertrophy with a large insertion into the apicolateral segment (Figure 4). In addition, no systolic signal void jet or delayed hyper-enhancement was observed. Ambulatory monitoring via ECG found no arrhythmia risk at this period. Finally, we decided to devise a medical follow-up program for our patient based on medical treatment (beta blockers) and periodic arrhythmic-risk-assessment monitoring.

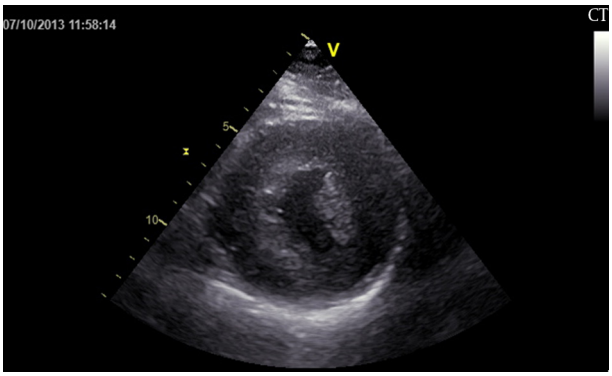

Figure 1. Hypertrophy and the Abnormal Position of the Lateral Papillary Muscles in the Parasternal Short-Axis View

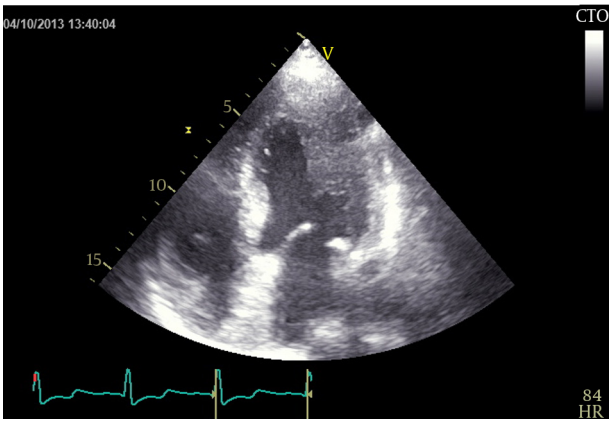

Figure 2. Four-Chamber Apical View Shows the Large Insertion of the Anterolateral Papillary Muscles Into the Mid and Apical Segments of the Lateral Wall

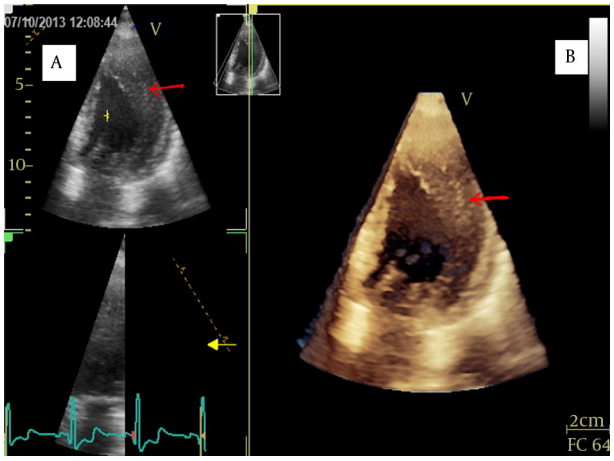

Figure 3. Three-Dimensional Echocardiography Clearly Distinguishes the Border Between the Left Ventricular Free Wall and the Base of the Anterolateral Papillary Muscles (Arrow)

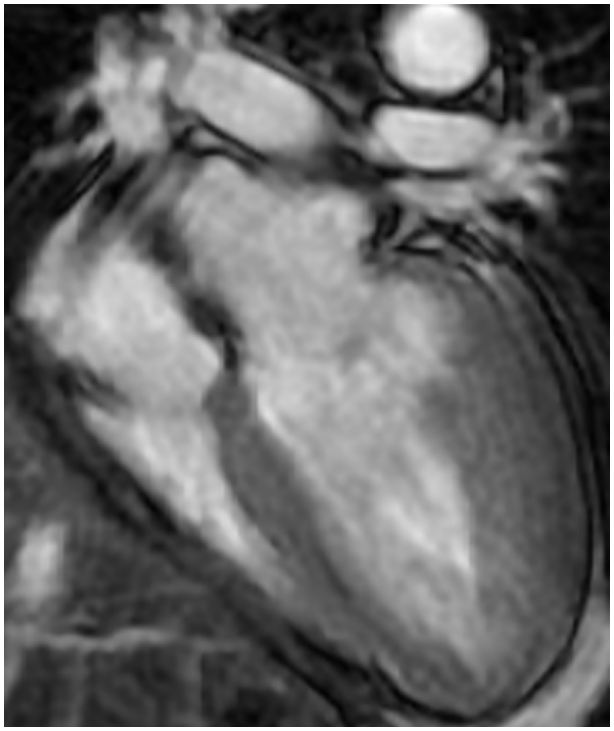

Figure 4. Cardiac Magnetic Resonance in the Four-Chamber View Shows the Hypertrophy and Large Insertion of the Anterolateral Papillary Muscles

3. Discussion

The diagnosis of HCM is sometimes challenging and requires a multimodality imaging approach. Our patient had hypertension and diffuse hypertrophy, which could have led to a diagnosis of hypertension-induced LV hypertrophy. However, her grade-I hypertension and echocardiographic findings (i.e. the absence of the concomitant hypertrophy of the apical segment of the septal region, low S' wave velocity on tissue Doppler imaging (4), low global longitudinal strain, and mitral abnormalities) further hinted at a diagnosis of HCM. Indeed, 2D strain or speckle tracking imaging offers a robust and rapidly evolving technology that has advanced our understanding of regional myocardial mechanics in HCM.

Regarding our patient, we had concordant data between the three imaging modalities, demonstrating the LV wall hypertrophy, papillary muscle hypertrophy, and anomalous insertion of the papillary muscles.

The morphological abnormalities of the mitral valve and the subvalvular apparatus are often observed in HCM and are accused in the physiopathology of subaortic obstruction (2).

In our patient, the anterolateral papillary muscles were hypertrophic (horizontal and vertical diameters were 15/27 milliliter, respectively). The hypertrophy of the papillary muscles is confirmed if the horizontal or vertical diameter or both are more than 1.1 centimeters in the parasternal short-axis view (5). Moreover, our patient's anterolateral papillary muscles had a morphological abnormality characterized by a bifid head, increasing the risk of confusion with supernumerary papillary muscles when seen in the parasternal short-axis view. Furthermore, the base of the anterolateral papillary muscles had an anomalous insertion, which doubled the thickness of the LV and became a source of error in the estimation of the degree of hypertrophy. On echocardiography, the border between the LV endocardium and the base of the anterolateral papillary muscles should be meticulously distinguished in HCM. Overestimation of the LV thickness could result in excessive treatment decisions such as implantation of an implantable automatic defibrillator if the thickness is ≥ 30 millimeter (6). A giant negative T wave is not specific for apical HCM and can be found in other cardiac diseases such as apically displaced papillary muscles (3). Our case was unique in as much as apical HCM was associated with apically displaced papillary muscles.

3D echocardiography and CMR can be very useful to confirm anomalous papillary muscles. This abnormality in the papillary muscles may have clinical implications such as a dynamic LV outflow tract obstruction even in the absence of hypertrophy (7). No data are available on the prevalence of this anomaly, however.

Abnormalities in the mitral apparatus such as those in the papillary muscles should be carefully analyzed in HCM. Our case report highlights the risk of error in the echocardiographic measurement of the thickness of the LV wall adjacent to hypertrophied papillary muscles. In our patient, the LV global longitudinal strain played a role in the final diagnosis, and 3D echocardiography and CMR proved instrumental in the confirmation of the morphological abnormalities.

Authors' Contributions

Ikram Kammoun performed the echocardiographic exam, wrote the paper and, supervised a multicentric study on hypertrophic cardiomyopathy, Lemone Houchinne participated in writing the paper, Sonia Marrakchi and Zied Ibn Elhaj reviewed the relevant literature, Wael Ben Amara and Souha Mokrani are members of the group who supervised a multicentric study on hypertrophic cardiomyopathy, Salem Kachboura provided critical input into the manuscript.

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