

Images in Cardiovascular Disease



A Rare Cause of Syncope: Apical Biventricular Hypertrophic Cardiomyopathy Complicated by Atrial Flutter

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Conflict of Interest

The authors have no financial conflicts of interest.

A 62-year-old male patient was admitted to our emergency service with symptom of palpitation and syncope attack. The patient's palpitation developed about 6 hours prior, and he had a brief period of syncope attack following palpitation. On medical history, the patient had only mild chronic obstructive pulmonary disease. The patient did not have any family history of cardiomyopathy. The patient had tachycardia, irregular pulse, and blood pressure of 130/80 mmHg on physical examination. No pathological findings were present on neurological examination. Electrocardiography revealed an atrial flutter (AFL), left axis deviation, and ST segment depression along with T wave negativity on the leads I, aVL, and V4-V6. Two-dimensional transthoracic echocardiography examination showed an isolated hypertrophy located at the left (LV) and right ventricular (RV) apex with sparing the interventricular and posterior septum (**Figure 1, Movie 1, 2**). Despite the treatment with IV amiodarone, normal sinus rhythm was not restored. Therefore, the patient was scheduled for electrical cardioversion. After two failed attempts of 125J and 200J of direct cardioversion, sinus rhythm was obtained by 275J biphasic cardioversion. Coronary angiography revealed normal coronary arteries. A cardiac magnetic resonance imaging (MRI) study was performed to confirm diagnosis. On cardiac MRI examination, the four-chamber long-axis view showed a clear illustration of apical hypertrophy both on the LV and RV apex with sparing the interventricular septum (**Figure 2, Movie 3**). Also, there was no thrombus formation and

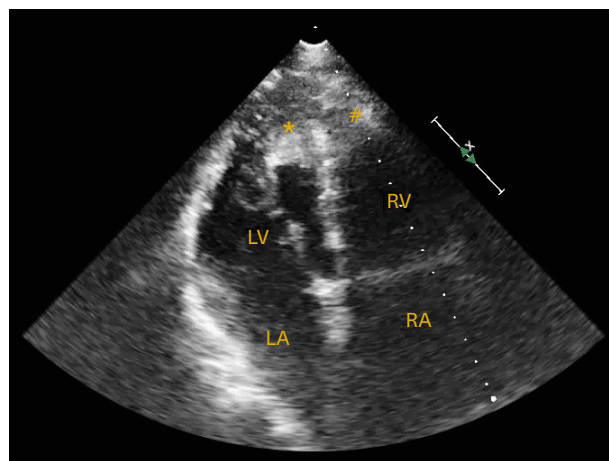


Figure 1. Two dimensional transthoracic echocardiography image showing a hypertrophy located at the apex of the LV (*) and RV (#). LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle.

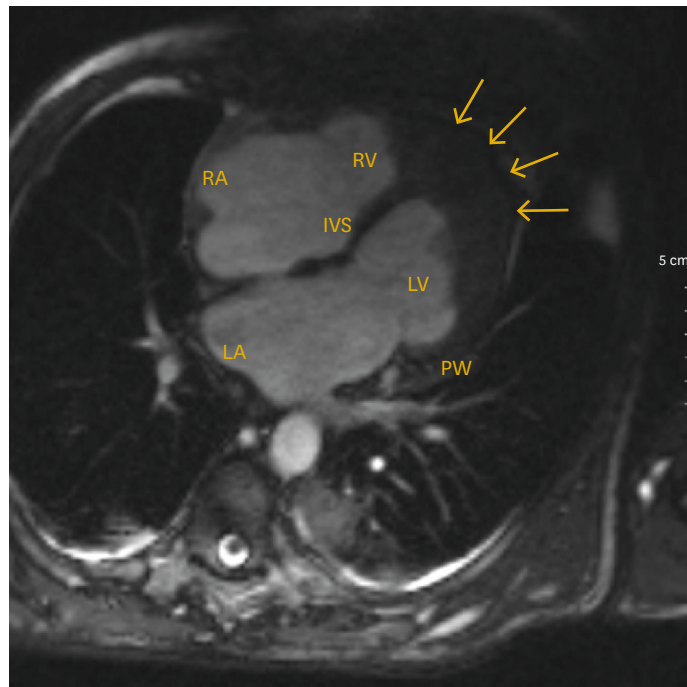


Figure 2. A clear illustration of isolated hypertrophy (long arrows) located at the apex of both ventricles on four-chamber cardiac MRI. IVS: interventricular septum, LA: left atrium, LV: left ventricle, MRI: magnetic resonance imaging, PW: posterior wall, RA: right atrium, RV: right ventricle.

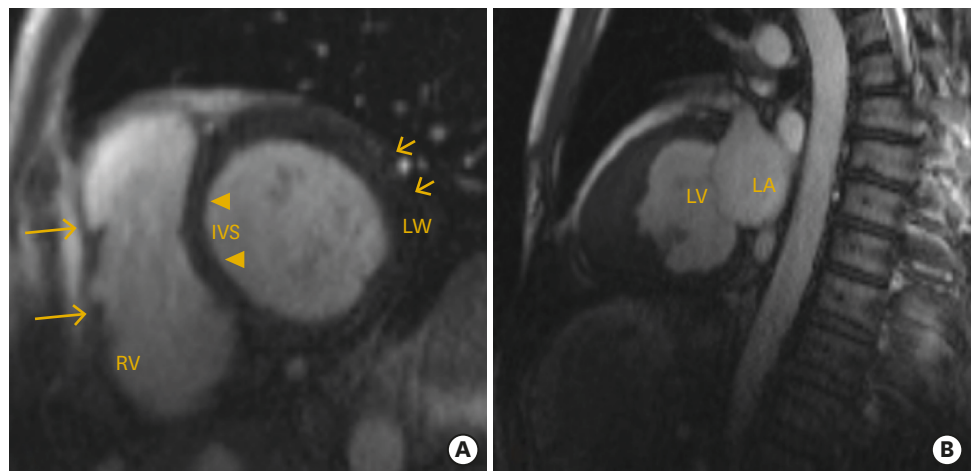


Figure 3. (A) Two-chamber short-axis cardiac MRI showing no involvement of the interventricular septum (arrowheads) or lateral wall of the LV (short arrows) or the free wall of the RV (long arrows). **(B)** Still frame from parasagittal two-chamber cardiac MRI showing no thrombus formation on the LV apex. IVS: interventricular septum, LA: left atrium, LV: left ventricle, LW: lateral wall, MRI: magnetic resonance image, RV: right ventricle.

involvement of interventricular septum, lateral wall of the LV, and free wall of the RV in a two-chamber short- and long axis view in cardiac MRI (**Figure 3, Movie 4, 5**).

Among hypertrophic cardiomyopathy (HCM) cases, the RV involvement is usually in diffuse character and, it is found approximately 30% of such cases.¹⁾ However, as shown in our case, both the LV and the RV showed a typical apical hypertrophy pattern and sparing the interventricular septum, which is an extremely rare clinical entity. In addition, to the

best of our knowledge, this might be the first case of adult patient who had a very clear demonstration of apical biventricular HCMP using cardiac MRI.

Prior studies have demonstrated that arrhythmic events are more frequent in patients with apical HCMP than normal subjects.^{2,3)} In particular, atrial fibrillation is the most commonly seen arrhythmia followed by ventricular tachycardia and AFL. Although the exact relationship between AFL and HCMP is uncertain, those with AFL were older, male, and had higher burden of comorbidities.

In terms of clinical perspective, apical biventricular HCMP should be kept in mind when assessing patients presenting with syncope and AFL even though it is very rare.

SUPPLEMENTARY MATERIALS

Movie 1

Cine image showing hypertrophy at the apex of the LV (*) along with apical thickness in the RV (#) on four-two-chamber transthoracic echocardiography. LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle.

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Movie 2

Cine image showing a hypertrophy located at the apex of the LV (*) on a two-chamber transthoracic echocardiography. LA: left atrium, LV: left ventricle.

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Movie 3

Cine image showing a clear illustration of hypertrophy located at the apex of both ventricles (long arrows). LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle.

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Movie 4

Cine image from short-axis cardiac MRI showing no involvement of the interventricular septum (arrowheads) or lateral wall of the LV (short arrows) or free wall of the RV (long arrows). IVS: interventricular septum, LW: lateral wall, MRI: magnetic resonance imaging, RV: right ventricle.

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Movie 5

Cine image from parasagittal two-chamber cardiac MRI showing no thrombus formation in the LV apex region. LA: left atrium, LV: left ventricle, MRI: magnetic resonance imaging.

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