Contrast Echocardiography for Diagnosis of Apical Hypertrophic Cardiomyopathy

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Abstract

Most patients with hypertrophic cardiomyopathy (HCM) have asymmetric septal hypertrophy; and among them, 25% present dynamic left ventricular outflow tract obstruction. Apical HCM is unusual. Echocardiography has been the first imaging method for patients with suspected HCM, but its shortcomings in evaluating the apex are well known. We present a 56-year-old woman with a history of left hemithorax pain for five years and myocardial perfusion defect in the inferoseptal region, who was a candidate for selective coronary angiography. On echocardiography, a mildly thickened apex was mistaken for apical foreshortening on planar imaging, but contrast echocardiography revealed apical HCM ***(Iranian Heart Journal 2010; 11 (3): 37-39).***

Key words: contrast echocardiography ■ hypertrophic cardiomyopathy

When patients present with unexplained ECG repolarization abnormalities and the history, examination, and echocardiography are not illuminating, the diagnosis poses a significant challenge. There is a wide differential diagnosis, including various cardiomyopathies. The patients may undergo lengthy investigation, which may ultimately be fruitless, with implications for lifestyle and life insurance.

Hypertrophic cardiomyopathy (HCM) is a primary disease of cardiac muscles that is characterized by a hypertrophied, non-dilated left ventricle (LV) unassociated with other cardiac diseases. Left ventricular hypertrophy (LVH), particularly asymmetric septal hypertrophy, is the most characteristic feature of HCM. The apical form of HCM, often associated with an abnormal ECG, in which left ventricular wall thickening is confined to the region of LV apex, was originally reported from Japan as a subset of nonobstructive HCM in which hypertrophy is limited to the apical region of the left ventricle.

This morphologic variety of HCM is characterized by a striking electrocardiographic pattern of giant negative T waves, defined as ≥10 mm deep, in addition to tall R waves in the left precordial leads, associated with an angiographic spade-shape deformation of the LV cavity at end diastole that indicates hypertrophy localized to the LV apex. Apical HCM is a clinically benign and non-familial disease that is identified predominantly in older men and is commonly associated with mild systemic hypertension. Most of these patients are asymptomatic and usually have a good long-term prognosis. In cases reported from centers outside of Japan, characteristic electrocardiographic and angiographic features are not often observed.[1] Apical HCM appears to be more benign in western countries than in Japan, with atrial fibrillation and myocardial infarction being the most frequent complication occurring in up to a third of patients during long-term follow-up, rather than sudden death.[2,3]

In patients with apical HCM, a transthoracic echocardiogram may be inadequate, and the hypertrophy localized to the apex may be missed.[4] In these patients, techniques such as transesophageal echocardiography have been utilized to better visualize the apex. In some cases, apical hypertrophy may be confused with apical thrombus. Differentiation between these conditions can be done by utilizing myocardial contrast echocardiography. In this technique, a contrast agent is injected intravenously to highlight hypertrophied myocardium by echocardiography.

Case presentation

The patient was a 56-year-old woman with a history of prolonged hypertension, without any other coronary risk factor or family history of HCM. She was admitted to the coronary care unit for left hemithorax pain with radiation to the left arm.

Her physical examination revealed only a fourth heart sound. Her ECG on admission showed sinus rhythm, LVH, and ST depression and deep negative T waves in the inferolateral leads (Fig. 1).

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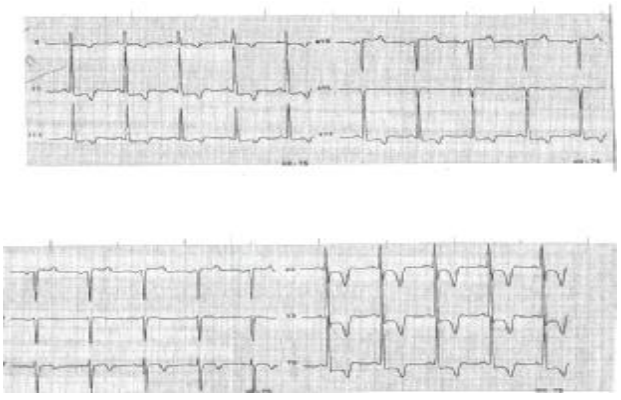


Fig. 1. 12-lead electrocardiogram shows diffuse ST-T changes in inferior and left precordial leads.

Echocardiography revealed normal LV systolic function (LVEF= 60%) and mild apical hypertrophy (Fig. 2).

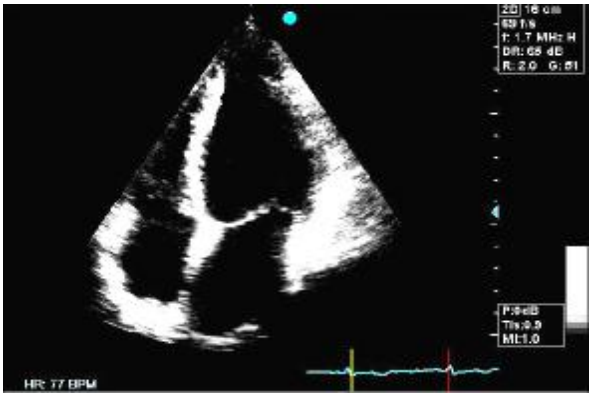


Fig. 2. Apical four-chamber view shows mild hypertrophy of the apical portion.

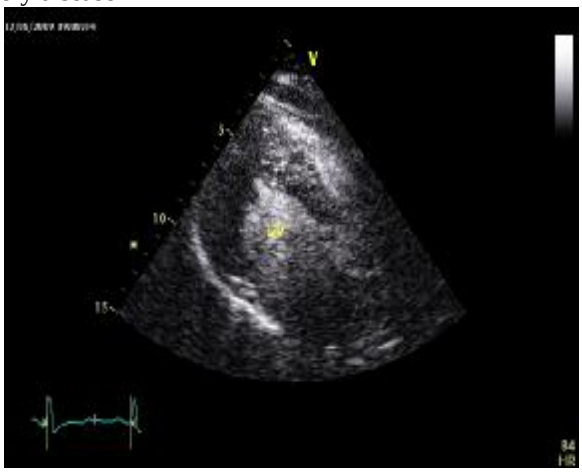
Contrast echocardiography, which was performed for a better evaluation of LVH and regional wall motion abnormality (RWMA), showed significant apical hypertrophy without RWMA or LV obstruction (Fig. 3).

Delayed images showed normal perfusion throughout the myocardium.

Considering the result of a previous myocardial perfusion scan which had shown ischemia in the inferoseptal segment, the patient was scheduled for coronary angiography, which revealed normal coronary arteries. The left ventricle was hyperdynamic, with systolic cavity obliteration of the distal third and apical portions. Simultaneous hemodynamic measurements in the apical area and the base of the left ventricle did not reveal a pressure gradient within the LV cavity.

Conclusion

Contrast echocardiography is a good alternative method for the diagnosis of apical HCM and coronary artery disease



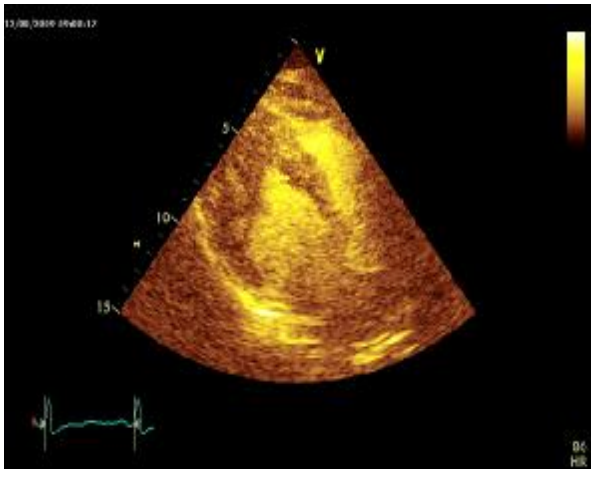


Fig. 3. Contrast echocardiography: apical two-chamber view sh and systole (right panel) along with severe hypertrophy of apical LV: left ventricle.

Conflict of Interest

No conflicts of interest have been claimed by the authors.

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