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IMAGING VIGNETTE

INTERMEDIATE

CLINICAL VIGNETTE

Hypertrophic Midventricular Obstructive Cardiomyopathy Revealed by a Stroke

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ABSTRACT

This case report describes the contributions of multimodality imaging to the diagnosis and management of midventricular hypertrophic cardiomyopathy revealed by a transient thromboembolic stroke. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2019;1:62–3) © 2019 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 39-year-old nonhypertensive man without a past medical history was admitted for a transient left hemiplegia.

Electrocardiography (ECG) depicted sinus rhythm and inverted ischemic T waves in the anterior leads (**Figure 1A**). Echocardiography showed myocardial hypertrophy (30 mm) with a mean midventricular systolic gradient (21 mm Hg) (**Figure 1B**) and an apical aneurysm (**Figure 1C**). Multislice dual-source computed tomography showed nonobstructive coronary atherosclerosis and severe focal midventricular hypertrophy (**Figure 1D**) with systolic obliteration of the left ventricle (**Figure 1E**, asterisks) leading to an apical aneurysm containing 2 small clots (**Figure 1E**, arrows). In this patient, 24-h ECG monitoring, carotid Doppler imaging, and transesophageal echocardiography did not show an alternative cause for the stroke. Cerebral embolism of cardiac origin was diagnosed. The apical pouch filled with the 2 small clots showed transmural delayed enhancement on cardiac magnetic resonance (CMR) imaging (**Figure 1F**), suggesting myocardial fibrosis.

The patient was treated with a beta blocker and a vitamin K antagonist. One year later he survived a witnessed sudden cardiac death. He subsequently underwent implantable cardioverter-defibrillator (ICD) insertion. Repeated 24-h ECG monitoring showed nonsustained ventricular tachycardia.

Hypertrophic cardiomyopathy (HCM) is a genetic cardiac disease sometimes related to sarcomeric protein mutation (1) with variable localization of myocardial hypertrophy. Dynamic ventricular obstruction can be found in the outflow tract (22.4%) (2) or the midventricle ("hourglass" shape). The latter comprised 9.4%, 12.9%, and 10.9% of HCM cases in Japan, the United States, and Italy (2). Apical aneurysm has been found in 4.8% of all HCM cases (3), in 28% and 1.8% of those with midventricular obstruction (MVO) and outflow tract obstruction, respectively (1). Aneurysm can result from chronic myocardial ischemia in patients without significant epicardial coronary artery disease (1). Delayed enhancement of the apical myocardium on CMR can precede the development of an aneurysm. Fibrosis can also be located in the aneurysm-contiguous areas, the left ventricular free wall, or the proximal ventricular septum (1). Apical aneurysm predicted a 6.4%/year rate of adverse events (sudden death, appropriate ICD use, nonfatal thromboembolic stroke, heart failure, and death) over 4.4 ± 3.2 years of follow-up (3). Although HCM-related mortality does not differ between patients with and without apical aneurysm, the former had a 5-fold greater rate of arrhythmic events (5%/year) and a 3-fold

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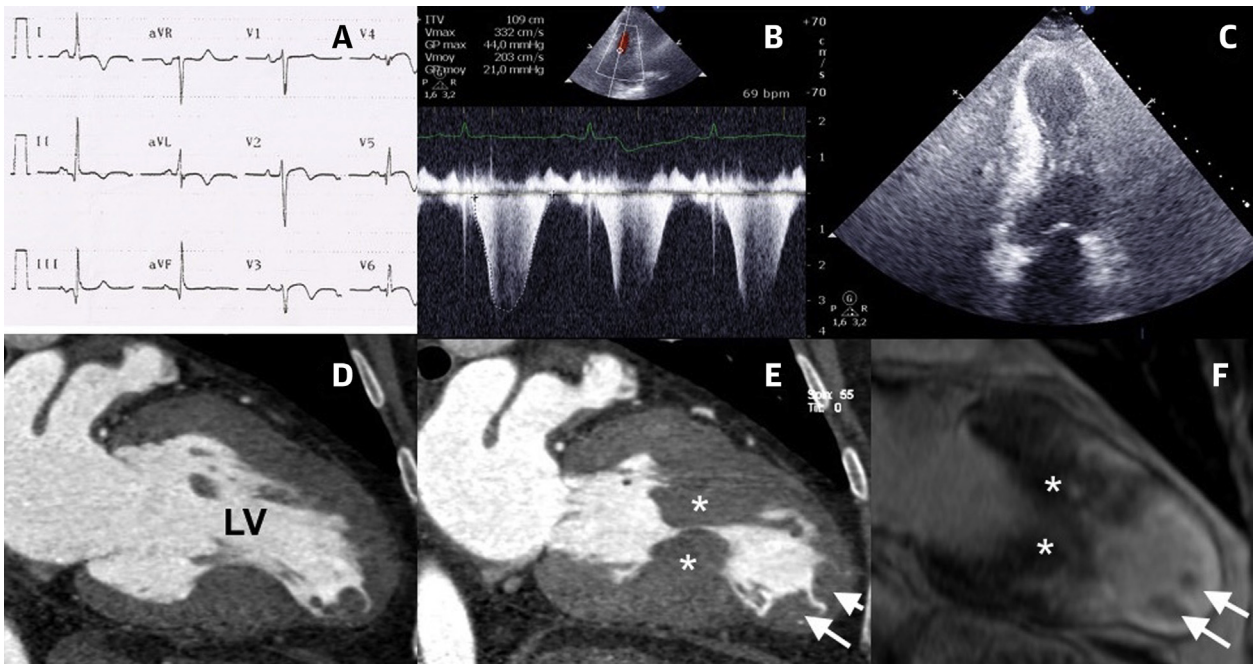
FIGURE 1 Multimodality Imaging of a 39-Year-Old Patient With Midventricular HCM Revealed by a Stroke



(A) Inverted apical T waves on electrocardiogram. **(B)** Transthoracic echocardiography showing hypertrophic cardiomyopathy (HCM) with its mean systolic obstructive gradient leading to **(C)** an apical aneurysm. **(D)** Dual-source computed tomography showing midventricular myocardial hypertrophy in diastole and **(E)** its hourglass shape **(asterisks)** in systole with an apical aneurysm filled by 2 clots **(arrows)** missed by transthoracic echocardiography. **(F)** Cardiac magnetic resonance imaging showing systolic mid-ventricular obliteration of the LV **(asterisks)** and transmural fibrosis of the apical aneurysm containing the clots **(arrows)**. LV = left ventricle.

greater rate of combined mortality and nonfatal adverse disease-related events (3). Relying only on the European Society of Cardiology risk score for ICD insertion could have missed 19 of 93 (20%) patients with HCM and apical aneurysm who had arrhythmic events. Therefore, apical aneurysm per se should be included in this risk score (3).

Transthoracic echocardiography is less reliable than computed tomography and CMR for visualizing apical aneurysms (1,3). CMR is also useful for depicting myocardial fibrosis. In 93 patients presenting with an apical aneurysm (34 patients with MVO) and followed up for 4.4 ± 3.2 years, 5 patients (5%; 1.1%/year) had a nonfatal embolic event while in sinus rhythm, and 13 others had an aneurysm filled with an uncomplicated thrombus (3).

As shown in this case, multimodality imaging is very useful for accurate diagnosis and management of a patient with HCM and MVO.

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