

## Apical Hypertrophic Cardiomyopathy: The Ace-of-Spades as the Disease Card

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Apical cardiac hypertrophy (Yamaguchi syndrome) is a relatively rare form of hypertrophic cardiomyopathy. A 67-year-old woman presented with exertional angina and moderate dyspnea. Her electrocardiogram showed sinus rhythm, flattened T waves in the limb leads and a strain pattern in the V<sub>2</sub>-V<sub>6</sub> leads. Echocardiographic examination showed that apical wall thickness of the left ventricle (LV) was 19 mm. Coronary angiography was normal, but left ventriculography revealed a spade-like shaped LV cavity, typical for apical cardiac hypertrophy. The diagnostic criteria for apical cardiac hypertrophy are: 1) asymmetric LV hypertrophy - predominantly at the apex of the ventricle; 2) LV wall thickness of 15 mm or more during diastole; and 3) apical to posterior wall thickness ratio of 1.5 or more determined by 2-dimensional echocardiography or cardiac magnetic resonance imaging. Although relatively rare, Yamaguchi syndrome must enter into the differential diagnosis of patients presenting with exertional angina pectoris.

**Key Words:** Angina pectoris • Hypertrophic cardiomyopathy

### INTRODUCTION

Yamaguchi syndrome (apical cardiac hypertrophy) is a relatively rare form of hypertrophic cardiomyopathy (HCM). In Japan, where it was first described, it constitutes 15-25% of all cases of hypertrophic cardiomyopathies.<sup>1</sup>

### CASE REPORT

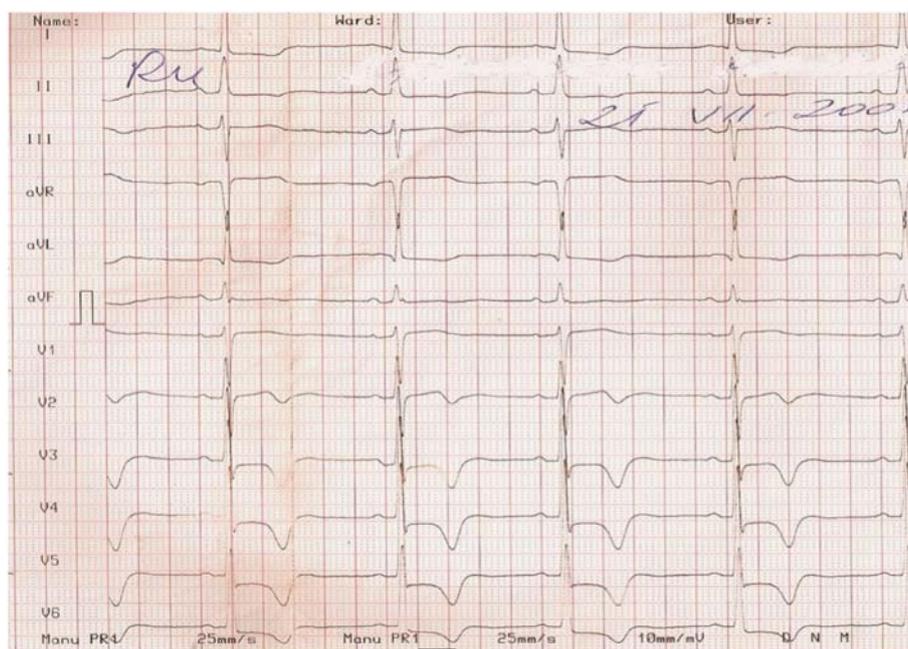
A 67-year-old non-smoking woman presented to the emergency department with worsening exertional an-

gina and moderate to severe exertional dyspnea, palpitations and fatigue. The cardiovascular risk factors included hyperlipidemia, hypertension, documented paroxysmal atrial tachycardia and a family history of cardiac disease and stroke, without any family history of HCM. Clinical examination showed blood pressure of 140/70 mmHg, with a regular heart rate of 63 beats per minute, no superimposed heart sounds or murmurs and no resting dyspnea. The 12-lead electrocardiogram showed sinus rhythm with a rate of 56 beats per minute, with flattened T waves in the limb leads and a strain pattern (ST segment depression of 1.5-2 mm and deep inverted T waves) in leads V<sub>2</sub> to V<sub>6</sub>, along with voltage criteria of LV hypertrophy and a Romhilt-Estes score of 9 points (Figure 1). The apical third of the left ventricle (LV) was hypokinetic on echocardiography and appeared as hypertrophic (the LV apical wall thickness was 19 mm), with an estimated LV mass of 400 g, normal systolic function and an enlarged left atrium of 53 mm. Blood tests showed no anemia, no leukocytosis and no elevation of myocardial enzymes or cardiac troponin. On coronary angiography, the coronary arteries were normal,

Received: November 22, 2013 Accepted: March 10, 2014

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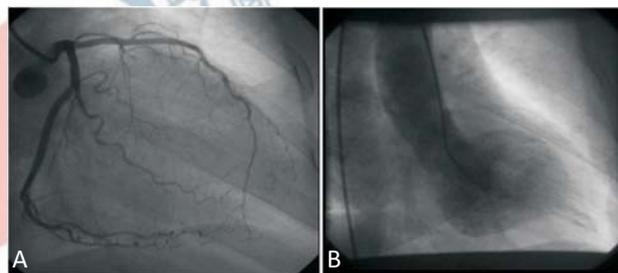
**Figure 1.** Electrocardiogram – sinus rhythm with a rate of 56 beats per minute, flattened T waves in the limb leads and a strain pattern (ST segment depression of 1.5-2 mm and deep inverted T waves) in leads  $V_2$  to  $V_6$  along with voltage criteria of left ventricular hypertrophy.

but the left ventriculography showed a spade-like shaped LV cavity in systole - a typical aspect for apical HCM (Figure 2).

## DISCUSSION

The apical nonobstructive type of HCM, also known as Yamaguchi syndrome, is an uncommon type of HCM in the European population. Sakamoto et al. first described its electrocardiographic pattern and echocardiographic findings in 1976<sup>2</sup> in Japanese patients, but it was Yamaguchi that described the syndrome and its ventriculographic features in 1979.<sup>3</sup> In the Asian population, this type of HCM seems to be more frequently diagnosed in men (23.8% vs. 8.9%,  $p = 0.03$ ) and at a younger age ( $57.2 \pm 12.9$  vs.  $64.8 \pm 11.3$ ,  $p < 0.001$ ) compared to women.<sup>4</sup> Also, the prevalence of the LV outflow obstruction is reported as being higher in Asian elderly women compared to men.<sup>4</sup>

Although patients with apical HCM may develop symptoms and signs that appear in other forms of HCM, its course is usually benign (with a 74% percent probability of survival at 15 years without morbid events) in the absence of predictors for poor prognosis, such as a



**Figure 2.** (A) Coronary angiography: patent coronary arteries. (B) Ventriculography: ace of spades-like shape of the left ventricular cavity in systole.

family history of sudden death, younger age at diagnosis, males, the appearance of syncope and effort induced hypotension.<sup>5</sup>

There are studies that have tried to determine the outcome of patients with apical HCM.<sup>4-6</sup> One study determined the overall survival at 15 years being 95% and identified three predictors of cardiovascular morbidity: age at presentation  $< 41$  years, left atrial enlargement and New York Heart Association functional class II or more at baseline.<sup>5</sup> The annual cardiovascular mortality in these patients was of 0.1%.<sup>5</sup> One other study (on an Asian population) found that LV outflow obstruction, atrial fibrillation, and female gender were predictors of mortality.<sup>4</sup> The patients with apical HCM are at risk of

developing arrhythmias such as atrial fibrillation (12%), paroxysmal atrial tachycardia, ventricular arrhythmias (ventricular tachycardia or ventricular fibrillation) or myocardial infarction with a secondary apical LV aneurysm (10%), that might determine the disappearance of the giant T waves.<sup>5</sup>

Typical features of apical HCM include an audible fourth heart sound, giant T wave negativity on the electrocardiogram, especially in the left precordial leads, a “spade-like” configuration of the left ventricular cavity at end-diastole on left ventriculography. Patients with apical HCM can have associated apical wall motion abnormalities like hypokinesis and aneurysm formation.<sup>5</sup> The imaging diagnostic criteria for apical HCM are: asymmetric LV hypertrophy – predominantly at the apex of the ventricle, LV wall thickness of 15 mm or more during diastole (17 mm in our case), apical to posterior wall thickness ratio of 1.5 or more determined by 2-dimensional echocardiography (ratio of 1.81 in our case) or cardiac magnetic resonance imaging (MRI)/multi-slice computed tomography (MSCT). It is well known that contrast-enhanced echocardiography can provide better images. 2-dimensional speckle tracking echocardiography allows the assessment of LV apical deformation in apical HCM.<sup>7</sup> There are studies that support the importance of cardiac MRI as being the gold standard in differentiating between hypertensive LV hypertrophy and the LV hypertrophy determined by a sarcomeric mutation (as in apical HCM).<sup>8</sup> In patients with apical HCM, first-pass gadolinium delayed-enhanced cardiac MRI may show local delayed enhancement in the subendocardium in the hypertrophic lesion.<sup>8</sup> A more recent method of evaluating cardiomyopathy is cardiac MSCT. Cardiac MSCT has the advantage of evaluating the coronary artery when trying to rule out coronary artery disease. Cardiac MSCT has the advantages of evaluating both morphology of the LV wall, the shape of the LV cavity and the morphology of the coronary arteries. It has the disadvantage of involving radiation and contrast exposure and not providing information about the degree of tissue fibrosis that MRI does.

We found a similar case of apical HCM masquerading as acute coronary syndrome cited in the literature.<sup>9</sup> The exclusion diagnosis has been made by the same invasive and noninvasive procedures. One study found significant coronary stenosis (> 50% of one or more coro-

nary arteries) in 35.5% of patients with apical HCM.<sup>10</sup> Additionally, reports about the coexistence of multiple coronary artery to LV microfistulae in patients with apical HCM are increasing. These microfistulae can cause LV hypertrophy through induction of ischemia by means of “coronary steal phenomenon” or through chronic volume overload.

Ruling out coronary artery disease is very important in these patients because the symptoms usually occur in middle-aged or elderly patients that might as well have this associated pathology. It is also important to establish the type of HCM, because apical HCM has a good long-term prognosis, with only a small percent of patients being affected by the complications that usually appear in other forms of HCM.

## ACKNOWLEDGMENTS

None.

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