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Case Report

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Apical Hypertrophic Cardiomyopathy: A Case Report

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Abstract

Apical hypertrophic cardiomyopathy (ApHCM) is a rare variant of hypertrophic cardiomyopathy, characterized by a spade-like left ventricular cavity. A 58-year-old African-American female with past medical history of hypertension presented for evaluation of recurrent exertional chest tightness, palpitations and headache. Prior workup including multiple stress tests and angiogram was non-conclusive. Electrocardiogram (EKG) showed characteristic marked T-waves inversions in inferior leads and left ventriculography revealed left ventricle apical hypertrophy with spade like left ventricular cavity that was typical of Yamaguchi syndrome. This case highlights the rare incidence of the disease among African American as well as the challenging diagnostic and presentation features of the disease.

Keywords: Apical hypertrophic cardiomyopathy; Yamaguchi syndrome; African American; Coronary angiography; Ventriculography

Introduction

Apical hypertrophic cardiomyopathy (ApHCM) is a rare variant of hypertrophic cardiomyopathy, first described in Japan in 1976, characterized by a spade-like left ventricular cavity. ApHCM is more commonly seen in the Asian population; however, it has been well documented among many other population groups worldwide. Few cases were reported among African Americans. This case highlights the rare incidence of the disease among African Americans and the associated diagnostic challenge.

Case Report

A 58-year-old African-American female with past medical history of hypertension presented for evaluation of exertional chest tightness, palpitations and headache. Over preceding 3 years, patient had multiple hospital admissions with similar complaints, and she underwent extensive cardiac workup, including multiple stress tests as well as an angiogram, but a correct diagnosis was not reached. She had no family history of premature coronary artery disease or sudden cardiac death, and no history of tobacco use. Physical examination was within normal limits. Electrocardiogram (EKG) (Fig. 1) showed normal sinus rhythm, left ventricle hypertrophy and marked T waves inversions in leads II, III and aVF. Troponin was minimally elevated but stable at 0.008 ng/mL, and other labs were unremarkable including normal thyroid-stimulating hormone and hemoglobin A1c. In lights of persistence of the symptoms, decision was made to take patient to cath lab. Coronary angiography (Fig. 2) showed preserved ejection fraction as well as angiographically normal coronary arteries. Left ventriculography revealed left ventricle apical hypertrophy with spade-like left ventricular cavity that was typical of the Japanese variant of asymmetrical apical hypertrophy known as "Japanese heart disease", or Yamaguchi syndrome.

Discussion

We report a case of a 58-year-old African American with ApHCM. ApHCM is a subtype of non-obstructive hypertrophic cardiomyopathy characterized by a spade-like left ventricular cavity. The disease is common in Japan where it was first reported [1]. In addition, ApHCM has been well documented among many other population groups worldwide [2-6]. Few cases were reported among African Americans [7, 8].

ApHCM has familial inheritance in an autosomal dominant pattern, sporadic forms of the disease caused by acquired genetic mutations of the sarcomere protein gene also exist [9]. Characteristic histological findings include edema, fibrosis, disorganization and bizarre nuclei in the myocardium [10]. The disease commonly presents on middle age and is most commonly seen in males [11] though in few cases, it was reported in both adolescents [12] and advanced age [13].

Common presenting symptoms include chest pain mimicking acute coronary syndrome [14], exertional dyspnea, palpitations, dizziness and fatigue [1]. Other reported features include syncope [15] and symptoms of heart failure [16].

ApHCM commonly has a benign course and is not associated with sudden cardiac death. Unfavorable outcomes include apical fibrosis, apical aneurysm formation, and heart failure. Life-threatening complications, such as myocardial infarction, arrhythmias, and stroke develop in around one-third of the patients, thus close follow-up is needed [17, 18]. Predictors of poor prognosis include advanced age, presence of hyperten-

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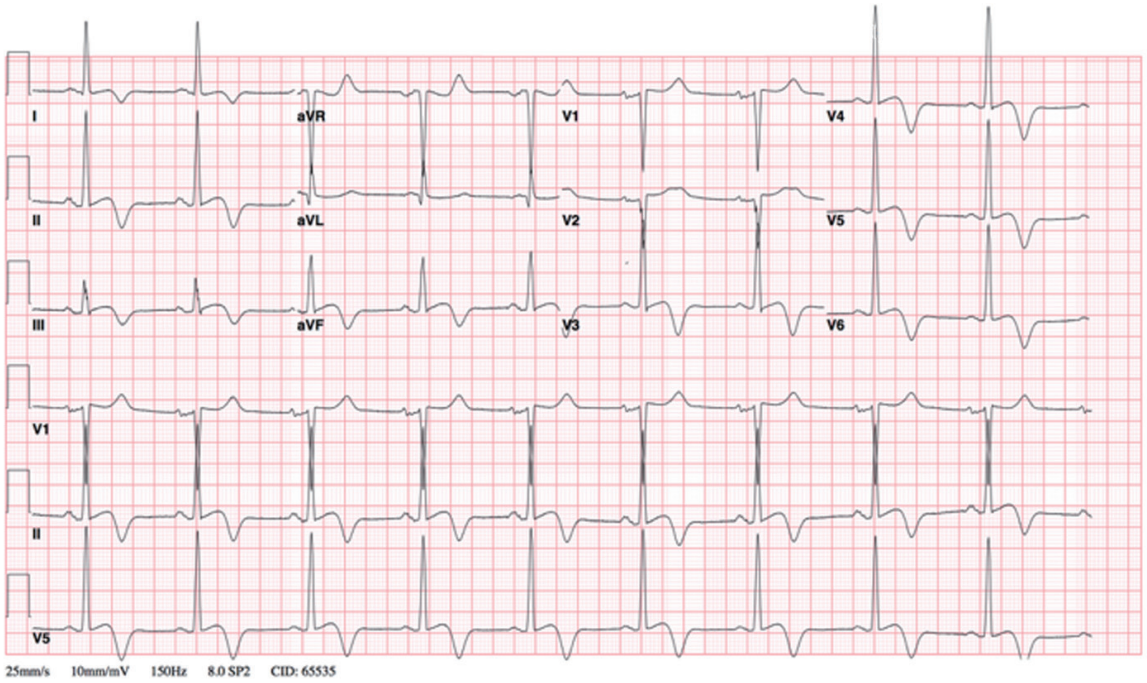

Figure 1. EKG shows normal sinus rhythm, left ventricle hypertrophy and marked T-wave inversions in leads II, III and aVF.

sion or diabetes, baseline atrial fibrillation, presence of echocardiographic findings including left atrial volume index, Sa velocity, and E/Ea ratio [19, 20] as well as cardiac magnetic resonance imaging finding including presence of apical aneurysm and extent of late gadolinium enhancement [21].

Typical EKG findings for hypertrophic cardiomyopathy include repolarization changes and giant (> 10 mm), inverted T waves in the anterolateral leads [22]. The giant negative T wave can be used as an index of severity of ApHCM [23]. Diagnostic imaging modalities include contrast-enhanced echocardiography [24], ventriculography and cardiac magnetic resonance imaging [25]. Transthoracic echocardiography (TTE) is a helpful non-invasive tool in diagnosis of ApHCM; however, it is operator-dependent, and due to low familiarity and degree of suspicion, ApHCM is frequently missed by TTE [26]. In our patient, given high risk profile and high index of suspicion for non-ST elevation myocardial infarction, echocardiography was not performed and a decision was made to take the patient immediately to the cath lab. Surprisingly, the patient had normal coronaries and based on ventriculography a

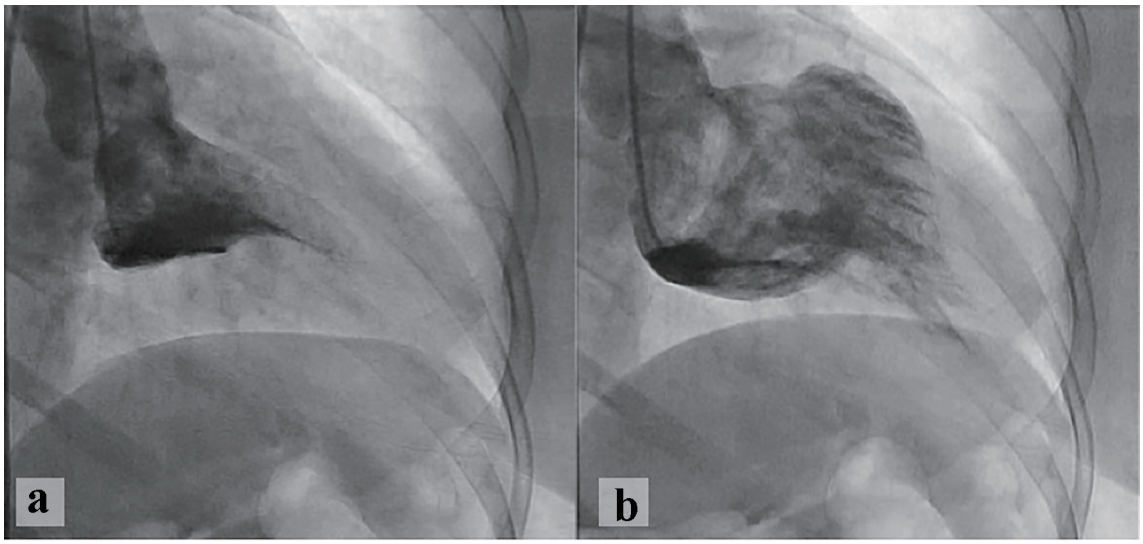

Figure 2. Left ventriculography revealed left ventricle apical hypertrophy with spade-like left ventricular cavity during systole (a) and diastole (b).

diagnosis of ApHCM was made.

Treatment options for patients with symptomatic ApHCM are aimed towards decreasing heart rate and reduced left ventricular afterload through use of β-blockers, calcium-channel blockers, and angiotensin-converting enzyme inhibitors [27]. For patients with severe heart failure symptoms refractory to medical therapy, available options include heart transplant and apical myectomy [28].

Conclusion

With the increasing numbers of cases with ApHCM reported among non-Asian populations, physicians need to consider ApHCM in the evaluation of typical chest pain. Diagnosis of ApHCM is clinically challenging. Despite recurrent hospitalizations of the patient and repeated cardiac workup, the correct diagnosis remained elusive. Given its diverse presentation, understanding the unique EKG features with giant T-wave inversions provides the initial clues to making the diagnosis.

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