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Case Report

Apical Hypertrophic Cardiomyopathy Presenting as Peripheral Cyanosis on Exertion and Paroxysmal Nocturnal Dyspnea: An Atypical Case Report

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ABSTRACT

Background: Hypertrophic cardiomyopathy (HCM) is defined by the presence of significant left ventricular hypertrophy (LVH) in the absence of secondary factors like systemic hypertension, aortic stenosis, and athlete's heart syndrome.

Case presentation: A 67-year-old woman, with a complaint of severe fatigue, peripheral cyanosis on normal daily activity life, and paroxysmal nocturnal dyspnea, was admitted to Cardiac Care Unit, Razavi Hospital, Mashhad, Iran. In the primary physical examination, cardiac auscultation revealed pathologic S4 sound. Clinical investigations such as electrocardiography, chest X-ray, and echocardiography approved Apical Hypertrophic Cardiomyopathy (AHCM). Only administration of Metoprolol succinate with a short-term follow-up showed completely relieved pathologic presentation of this case.

Conclusion: In this case report, the management of a patient with peripheral cyanosis on normal activity, paroxysmal nocturnal dyspnea, and AHCM was emphasized. This case showed that early diagnosis followed by medication and supportive care, can control the patient's symptoms and postpone the progression of heart failure symptoms.

Keywords: Apical Hypertrophic Cardiomyopathy, Dyspnea, Echocardiography

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Introduction

Hypertrophic cardiomyopathy (HCM) is an autosomal dominant familial disease due to mutations of the myocardial sarcomeric contractile proteins that occurs in all races worldwide (1). In many series, annual death rates are as high as 3-6%, but recent studies from centers in Europe and the United States have reported substantially better prognosis with overall mortality rates of 1% or less (2). It is defined by the presence of significant left ventricular hypertrophy (LVH) in the absence of secondary factors such as systemic hypertension, aortic stenosis, and athlete's heart (3). Apical hypertrophic cardiomyopathy (AHCM) is a rare variant of HCM that it is more frequent in Japan (4). We reported a case of AHCM presenting as severe dyspnea on exertion and peripheral cyanosis.

Case Report

A 67-year-old woman was admitted to the Cardiac Care Unit, Razavi Hospital, Mashhad, Iran. with a complaint of fatigue, peripheral cyanosis on normal daily activity life, and paroxysmal nocturnal dyspnea. She had no history of hospitalization for these complaints, but she had a history of numbness and tingling in fingers and severe fatigue that was neglected by the patient. There was also no reported family history of cardiac disease. She reported worsening of symptoms in the week before admission. On admission, she was in stable condition with a blood pressure of 145/84 mmHg, regular heart rate at 102 beats per minute, respiratory rate of 26 breaths per minute, and was afebrile. Oxygen saturation, at rest, with finger pulse oximeter on the index finger was 82% and the patient had a cyanotic appearance in the upper and lower extremities. In the initial physical examination, a pathologic S4 on cardiac auscultation was found. The lung sounds were clear.

In the electrocardiogram (ECG), it was found an evidence of narrow complex, regular and normal sinus rhythm with normal axis deviation. Moreover, the ECG showed LVH, T-wave inversions in the precordial leads (V2-V6), and ST-segment depression in I and aVL (Figure 1).

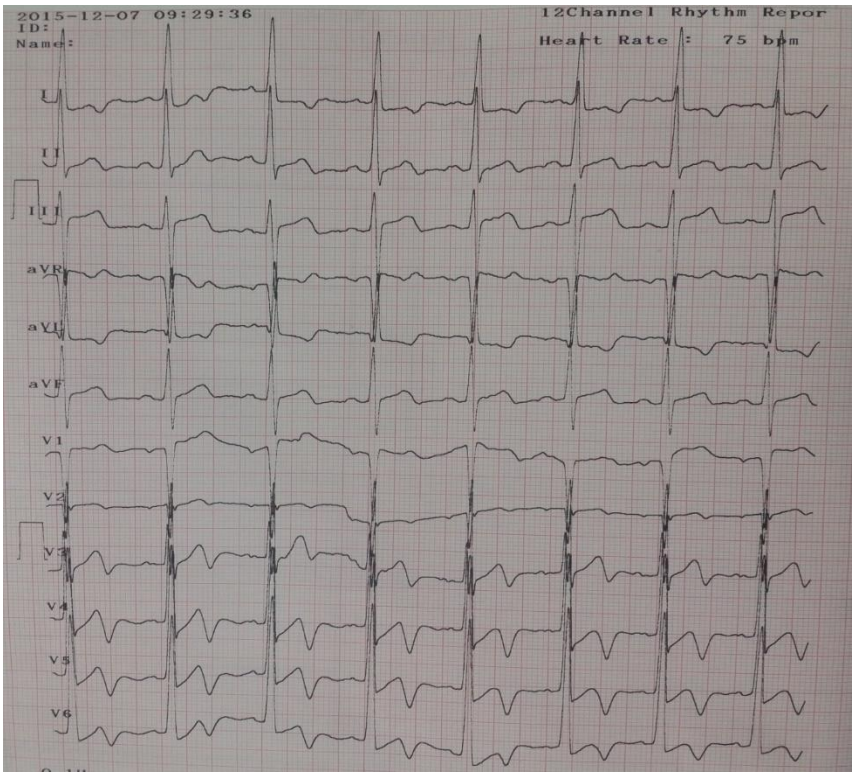

Figure 1. The 12-lead electrocardiograms on admission showing LVH, T-wave inversion in the precordial leads; V2-V6, and ST segment depression in I, aVL.

For other diagnostic investigations, the patient was referred to the echocardiography unit for transthoracic echocardiography. It was performed in the Cardiac Care Unit and showed a left atrium size of 45 mm and the apex of 21 mm. Transthoracic echocardiography in the parasternal long-axis view in end diastole revealed that the inter-ventricular septum and posterior wall thickness was 14 mm. There was also an evidence of asymmetric septal hypertrophy (ASH) in short axis and parasternal long-axis views. The left ventricular end diameter was 52 mm at the end of diastole in the parasternal long-axis view, with preserved systolic function (ejection fraction of 55%) and Left ventricular diastolic dysfunction. The right ventricle was normal with a good function. The patient did not have any evidence of left ventricular outflow tract obstruction (LVOTO), systolic anterior motion (SAM), or significant mitral regurgitation. An apical four-chamber view of the patient's transthoracic echocardiography is shown in Figure 2. Chest X-ray (in PA view) showed an increased cardiothoracic ratio. The aortic arch was dilated, the main pulmonary artery (MPA) and the left atrial appendage (LAA) were normal. Considering these findings, the patient was scheduled for a 30-day follow-up with the prescription of Metohexal (Metoprolol succinate) with a dose of 23.75 mg twice a day, as a β-blocker. After this period, her symptoms were relieved and she had no complaint of dyspnea on exertion. Also, she had no evidence of cyanosis in the upper and lower extremities. Finally, the patient was suggested to return to the clinic for adjustment of her medications, and screening of first-degree family members, if symptoms recurred.

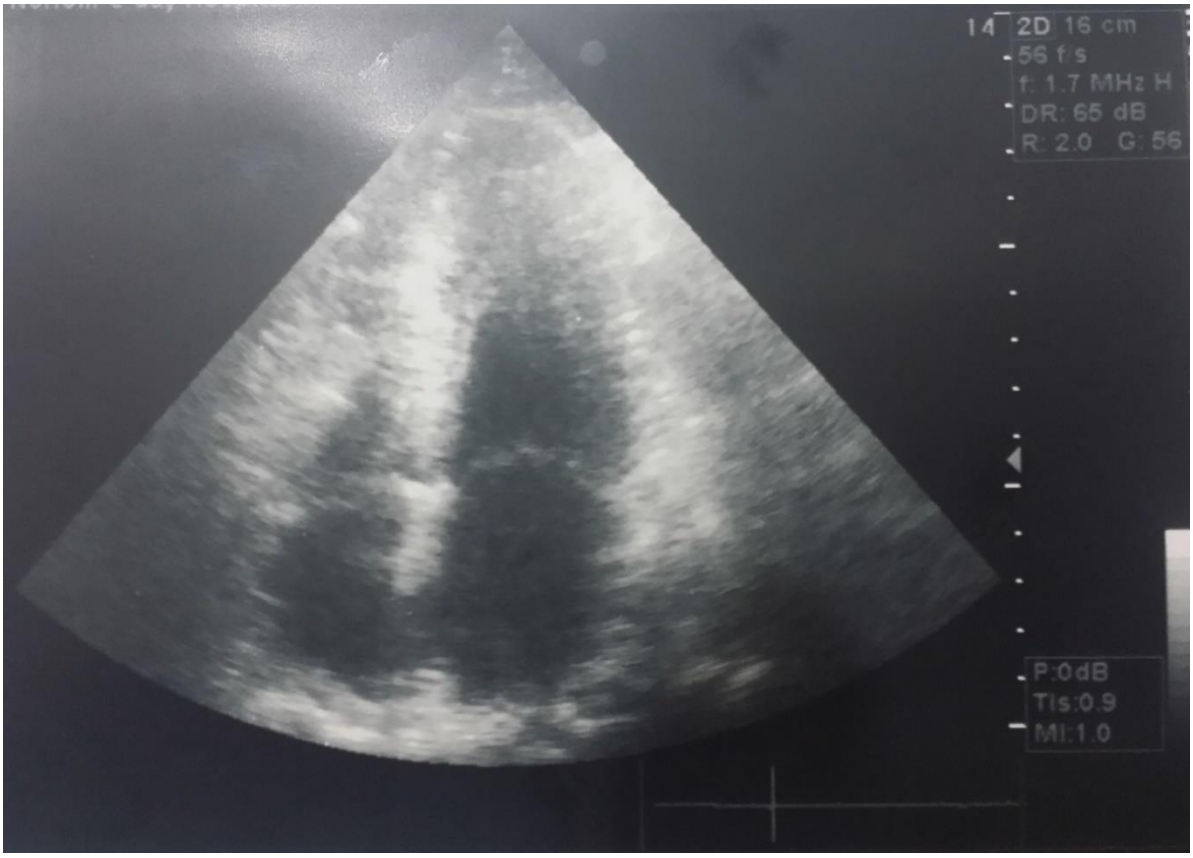

Figure 2. Transthoracic echocardiographic four-chamber view of patient demonstrated apical hypertrophic cardiomyopathy.

Discussion

Cardiac diseases are significant since they are the most common chronic diseases of the 21st century and are regarded as the main causes of death (5). Hypertrophic cardiomyopathy (HCM) is a genetic cardiac disease characterized by marked variability in morphological expression and natural history (6). It may involve mainly the proximal septum, or there may be diffuse LVH. However, there are other patterns, such as apical hypertrophy (7).

Apical Hypertrophic Cardiomyopathy (AHCM) can be asymptomatic or present with syncope, chest pain (symptoms similar to those of acute coronary syndrome), palpitations, and dyspnea (4). Another study by Ahmed et al. showed that the most common presenting symptoms were dyspnea with or without chest pain (1). The main complaint and unique presentation of our patient was a cyanotic appearance in the upper limbs extremities that was not a common presentation. This presentation may be misdiagnosed with severe respiratory diseases at the first time. Also, paroxysmal nocturnal dyspnea in our patient is a relatively uncommon presentation of that seen in the progressive HCM. But our patient had no other symptoms of advanced features of AHCM such as syncope or pre-syncope, arrhythmia, angina, and dizziness.

AHCM is predominantly a hereditary disease, although it can also be present in patients with no family history (8). Our patient had no evidence of familial history.

In Western patients, there seems to be a varied presentation as far as clinical and ECG features are concerned, compared to the classic AHC as defined in the Asian population. Hence, in the presence of AHCM situations, understanding the unique ECG features of AHCM can be of assistance in the diagnostic process of this uncommon condition (4,8). The presence of ECG findings indicative of LVH with giant T-wave inversions (especially in precordial leads) and loss of septal Q-waves should raise strong suspicions of AHCM. These are considered pathogenic for this disorder (4, 9, 10). Our patient's ECG demonstrated the same features, and based on these characteristics, additional investigations such as transthoracic echocardiography were performed.

Numerous studies have investigated the effect of β-blockers on HCM patients and reported symptoms improvement (11,12). A study by Kasirye et al. also mentioned that β-blockers were the first drugs used for the treatment of symptomatic HCM (4). Hence, our patient mentioned relief of her symptoms after 1-month use of this drug agent and it can be a selective drug in the first step of treatment of AHCM. On the other hand, in some studies (10-12), it was stated that β-blockers can relieve the symptoms in the two to five months after administration, but our patient's symptoms improved after only one month and in a short-term use of metoprolol succinate. This is a rare case that was treated with only one agent β-blocker in a short time. So, in other cases of AHCM, metoprolol succinate is suggested to be considered in the first step of treatment.

Symptoms such as severe dyspnea should be controlled in patients diagnosed with AHCM. The treatment process can include β-blockers, ACEi, and calcium channel blockers. If this approach is not successful, patients with this condition should be referred for changes in the medication protocol to postpone heart failure progression due to AHCM complications.

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Conflict of interests

The authors declare that there is no conflict of interests.

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