Iranian Heart Journal; 2020; 21 (2)

Case Report

Familial Apical Hypertrophic Cardiomyopathy With Atrioventricular Block

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ABSTRACT

A 53-year-old woman was referred to us with frequent episodes of palpitation, near-syncope, and dizziness. The patient had a history of apical hypertrophic cardiomyopathy in her sister and aborted sudden cardiac death in her aunt. Because of first-degree atrioventricular block and high atrioventricular Wenckebach point, cardiac magnetic resonance imaging was done, and apical hypertrophic cardiomyopathy was diagnosed. ***(Iranian Heart Journal 2020; 21(2): 90-93)***

KEYWORDS: Hypertrophic cardiomyopathy, Atrioventricular block, Sudden cardiac death

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Received: May 6, 2019 **Accepted:** June 20, 2019

In hypertrophic cardiomyopathy (HCM), a slow rhythm is an uncommon finding. However, isolated cases of HCM have been reported to have cardiac conduction disorders (CCDs) and PRKAG1 (protein kinase AMP-activated non-catalytic subunit gamma 1) mutations associated with CCDs. [1-3]

Most cases of pacemaker implantations in patients with HCM are aimed at reducing the left ventricular outflow tract gradient or treating iatrogenic atrioventricular (AV) block due to myectomy or alcohol septal ablation. [2]

Apical HCM is a specific variant first described in Japan by Yamaguchi. It is characterized by apical hypertrophy and a spade-like configuration in left ventriculography. First-degree AV block is the most commonly seen block type in ECG. The percentage of apical HCM in Japan is about 15% of all HCM cases, while in the United States is about 3% of the population with HCM. [4-7]

The diagnostic criteria for apical HCM are apical wall thickness of 15 mm or greater and a ratio of maximal apical to posterior wall thickness of 1.5:8 or greater.

Deep negative T-waves in the precordial leads are found in more than 90% of patients with apical HCM; nonetheless, in about half of them, the depth of T-wave is not more than 10 mm. [8]

Cardiac magnetic resonance imaging (MRI) is a valuable tool for the diagnosis of these patients with inconclusive echocardiography findings.

Case Report

A 53-year-old woman presented with episodes of palpitation and presyncope without chest pain and dyspnea. ECG illustrated first-degree AV block with PR of 320 ms and sinus tachycardia. An electrophysiology study revealed no inducible ventricular or supraventricular arrhythmias. Because of high AV Wenckebach point, cardiac MRI was recommended to rule out sarcoidosis. Cardiac MRI showed apical wall thickening, a small left ventricular apical aneurysm, and late gadolinium enhancement in the mid-to-apex of the left ventricle and the junction between the left and right ventricles. A diagnosis of apical HCM was considered for the patient (Fig. 1–3).

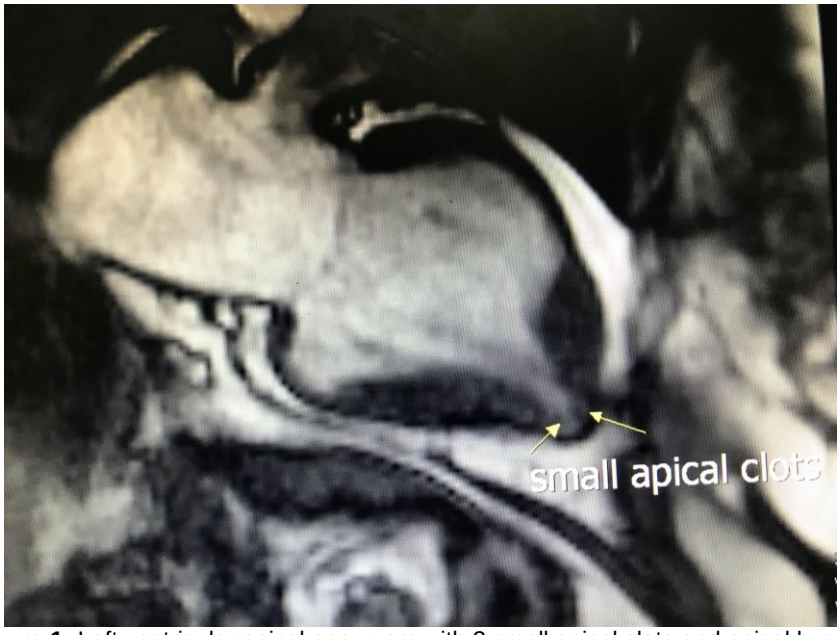

Figure 1: Left ventricular apical aneurysm with 2 small apical clots and apical hypertrophy

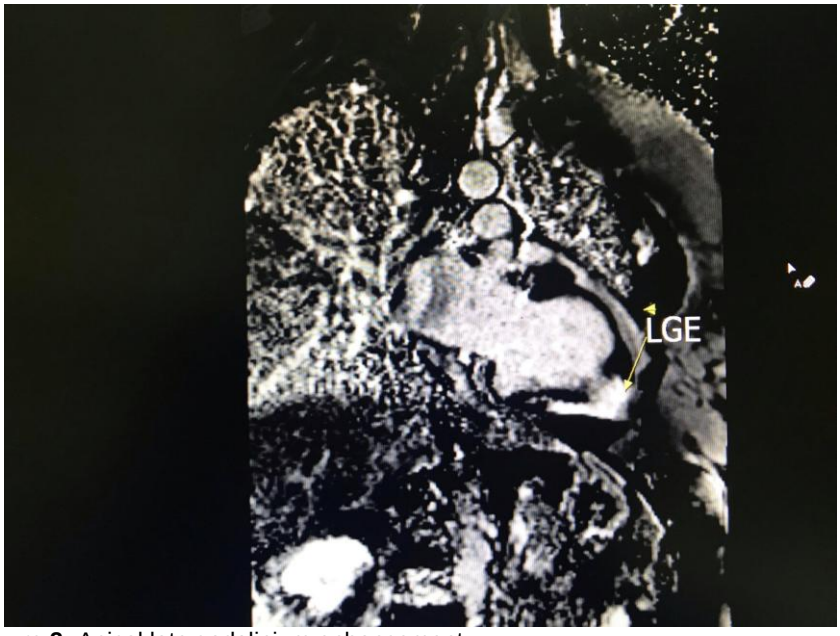

Figure 2: Apical late gadolinium enhancement

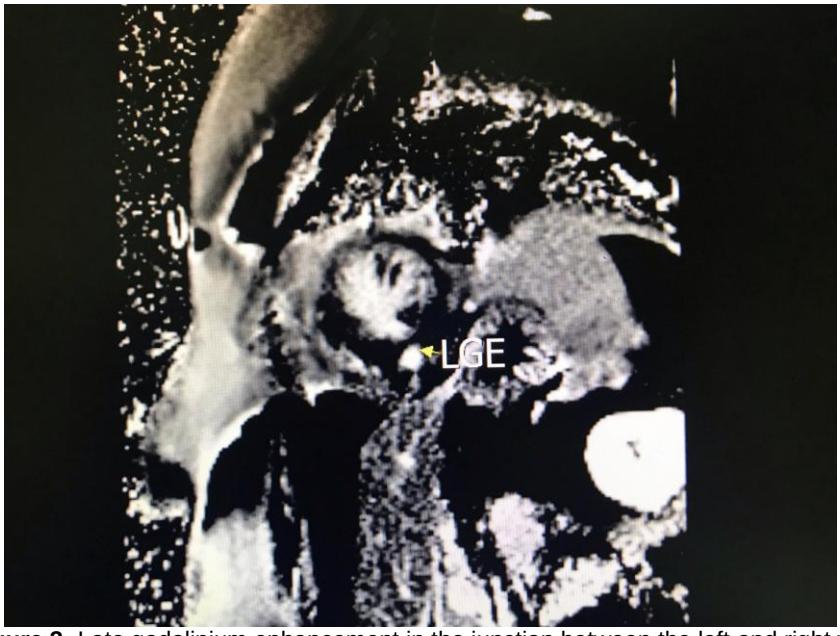

Figure 3: Late gadolinium enhancement in the junction between the left and right ventricles

A meticulous retrospective history taking of the patient revealed that her sister also had apical HCM, for which she had received an implantable cardioverter-defibrillator (ICD). There was also a history of aborted sudden cardiac death in her aunt. The female predominance in this familial involvement was intriguing.

A cardiac genetic study was recommended for the patient; however, financial constraints precluded it.

DISCUSSION

The incidence of arrhythmias in HCM is well documented, with most of such arrhythmias being premature ventricular complexes, ventricular tachycardias, and supraventricular premature depolarizations. Although the association between AV block and HCM has been reported in adults, it is very rare. [9]

Sudden cardiac death in patients with HCM is caused by ventricular arrhythmias, but it can also be caused by CCDs. [10-12]

In an evaluation of the anatomical basis of complete heart block in cats with HCM, extensive fibrosis of the central fibrous body and endocardial and myocardial fibrosis in the upper part of the septum were reported. [3]

In a case series in 2010 of about 450 patients, Barrialous et al [1] reported that about 8% of their study patients experienced CCDs, which required permanent pacemaker implantation. Additionally, their study showed the existence of the family history of CCDs in their patients with HCM.[1]

Our patient was a rare case of familial apical HCM presenting with first-degree AV block. Because of high AV Wenckebach point, extensive late gadolinium enhancement, and apical aneurysm, we decided to implant an ICD for this patient.

What is remarkable in this case is the involvement of females only. Had we been able to conduct a genetic study, we could have arrived at more intriguing conclusions.

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