Current Research in MRI

Case Report

Apical Hypertrophic Cardiomyopathy: A Case Report

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Abstract

In this case report, a patient with apical hypertrophic cardiomyopathy (AHCM) who presented to the emergency department with chest pain is presented. The symptoms, clinical findings, diagnostic methods, differential diagnosis, and treatment options for apical hypertrophic cardiomyopathy are briefly described. An uncommon type of hypertrophic cardiomyopathy, AHCM typically affects the left ventricle's apex.
Keywords: Cardiomyopathy, hypertrophic cardiomyopathy, magnetic resonance imaging

INTRODUCTION

Apical hypertrophic cardiomyopathy usually affects the apical region of the left ventricle and is morphologically divided into 3 categories: diffuse, focal, and mixed type with interventricular septum hypertrophy. However, this classification is not widely used in clinical practice.

CASE PRESENTATION

A 46-year-old man with no previously known chronic disease presented with chills, tremor in the hands, and chest pain. There was no familial history of sudden cardiac death. Vital findings and physical examination were unremarkable. The electrocardiogram showed deep T negativity in the anterior precordial leads. Laboratory values showed a troponin value of 12 ng/L. On transthoracic echocardiogram, left ventricular ejection fraction was 30%, apex was akinetic, and left ventricular hypertrophy was present. The patient was evaluated with digital subtraction angiography, and no stenosis was detected. The patient was then evaluated with cardiac contrast MRG. Myocardial hypertrophy was observed in the apical septum and left ventricular free wall adjacent to the apex. The apex was relatively thin, and delayed intraventricular contrast evacuation in the apical region was detected in dynamic series. Mid-wall contrast enhancement was present in the apical septum and free wall adjacent to the apex of the left ventricle in late contrast series. The findings were evaluated as AHCMP. The patient was followed up in the cardiology service, and saneloc (50 mg) and panto (40 mg) were started as treatment. An implanted cardiac defibrillator (ICD) was planned for the patient whose risk ratio was above 0.6 in cardiomyopathy risk calculation. Since this case report is retrospective, the need for a consent form was waived.

DISCUSSION

Apical hypertrophic cardiomyopathy is most commonly diagnosed in the middle-age group and is more frequently observed in people of Asian ethnicity.[1] The prevalence of hypertrophic cardiomyopathies in the general population ranges from 0.02% to 0.2%. The incidence of apical hypertrophy is at 1%-2% of all hypertrophied cardiomyopathies compared to 25% in Japan. It is known to affect men more than women.[2] Although autosomal dominant (OD) inheritance has been observed in certain cases of AHCM, the condition is mostly sporadic.[3] In genetically inherited cases, it is known that the alpha cardiac actin gene has a sarcoma mutation called E101K.[4] Patients with AHCM may present with chest pain, dyspnea, palpitations, syncope, atrial fibrillation, myocardial infarction, ventricular rhythm disorders, and heart failure, or may be asymptomatic. The primary diagnostic method for AHCM is a transthoracic echocardiogram. In addition to ECG, multislice spiral CT, left ventriculography, and cardiac magnetic resonance imaging are alternative diagnostic methods.[4] Although transthoracic echocardiogram is the first diagnostic test, the gold standard in diagnosis is cardiac MRI, as stated in various case reports. The cardiac MRI images of apical hypertrophic cardiomyopathy are similar to those of hypertrophied cardiomyopathies. Apical hypertrophic cardiomyopathy holds only the apex. The wall thickness is diagnosed by an intervention septum of 15 mm, a free wall of 10 mm, and mid-wall contrast in late-phase gadolinium images. The traditional appearance of the left ventricle is a configuration resembling a spade or ace-of-spades symbol. A patient with chest pain considered to have apical hypertrophic cardiomyopathy does not need a routine coronary CT scan to rule out other diagnoses. Because of its high spatial resolution, multi-detector computed tomography has recently emerged as a new technique for assessing the morphology and function of the heart. Multi-layered reconstructions allow measurement of heart function as well as myocardial thickness. Cardiac CT can be used to evaluate anatomy in the presence of suboptimal

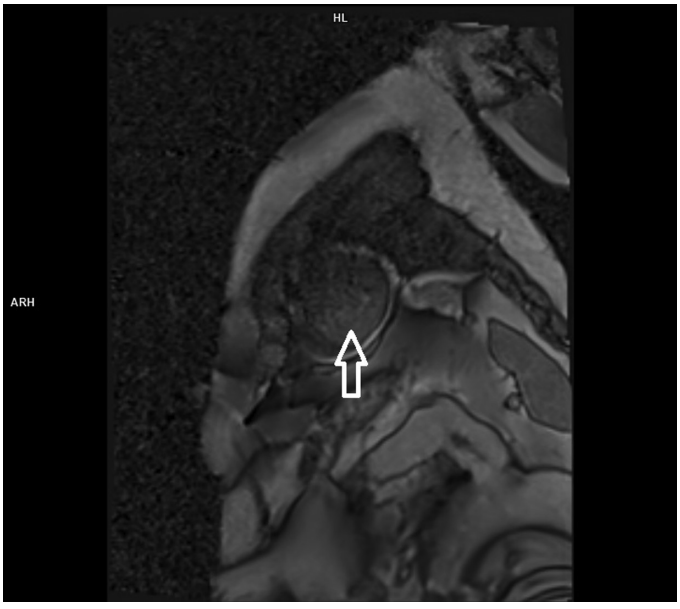

Figure 2. Late-phase gadolinium contrast-enhanced images showed gadolinium uptake in areas of the myocardium with hypertrophy, confirming a diagnosis of focal hypertrophic cardiomyopathy.

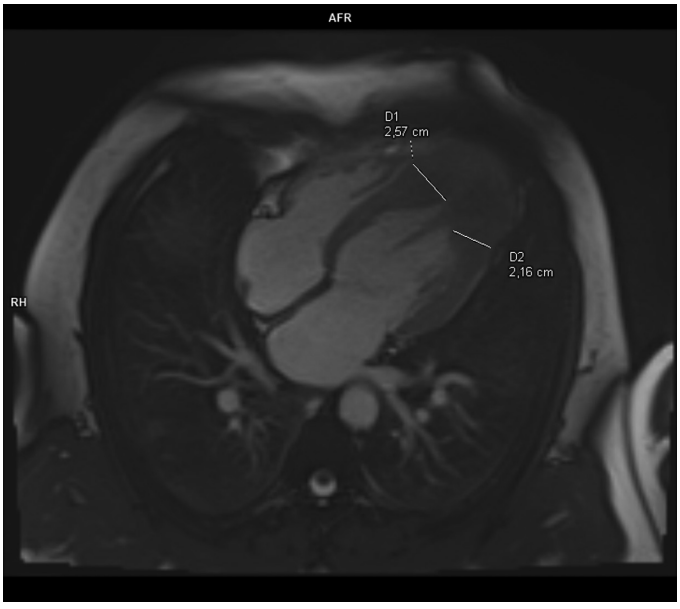

Figure 1. Cine images showed thickening of the apex and interventricular septum surrounding the apex, along with a focal area in the left ventricular free wall. The end-diastolic interventricular septum diameter was 25 mm, and the left ventricular free wall diameter was 21 mm and increased.

echocardiographic images and contraindications of cardium MRI. In this case, the definitive diagnosis was made by cardiac MRI (Figures 1 and 2). As in this case, the most common ECG finding of AHCM is negative T waves in the precordial leads, followed by left ventricular hypertrophy (Figure 3). Differential diagnoses may include left ventricular neoplasms, coronary artery disease, endomyocardial fibrosis, and left ventricular apical thrombus. Antiarrhythmic agents such as

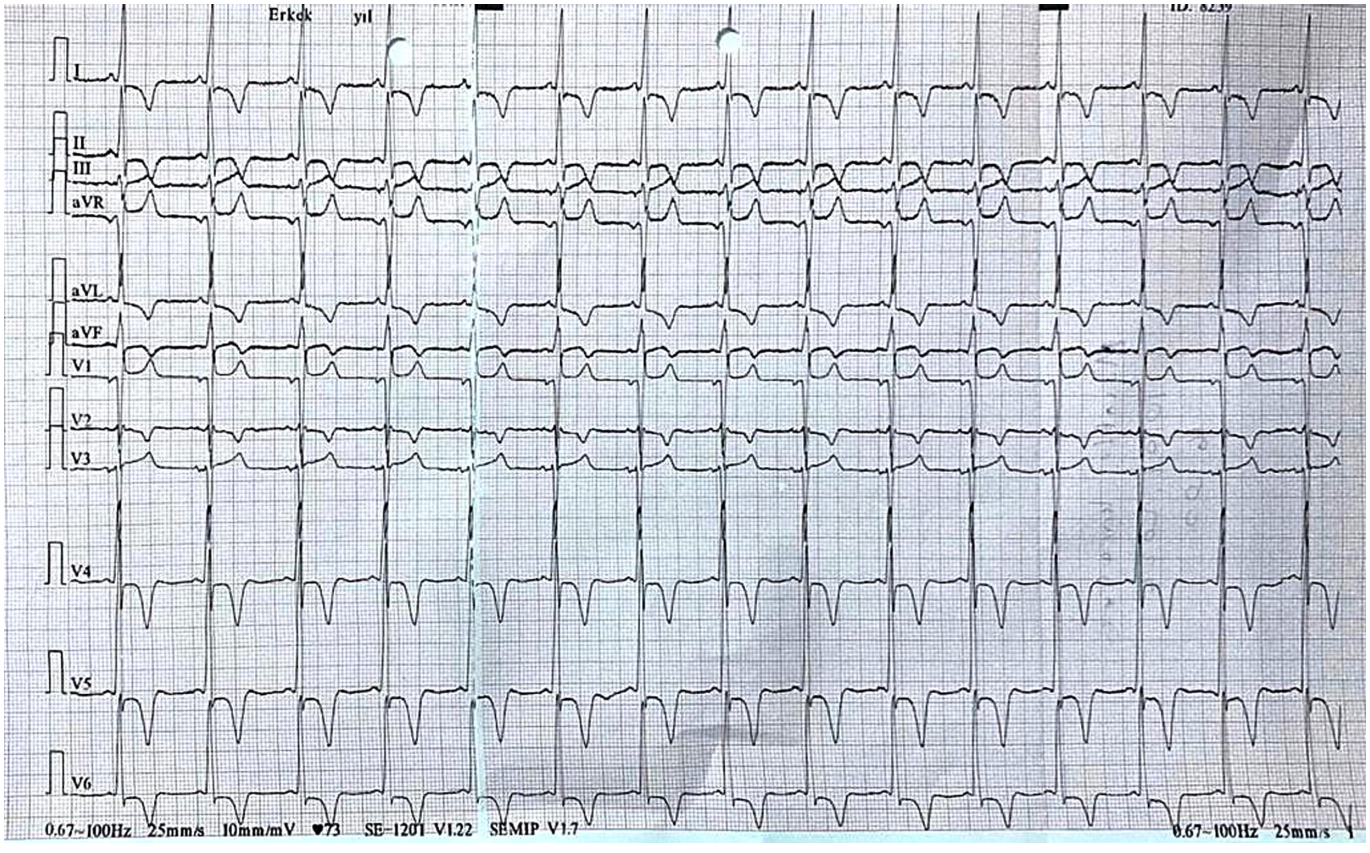

Figure 3. Common T negativity and left ventricular hypertrophy on ECG.

verapamil, beta-blockers, procainamide, and amiodarone are among the medications used to treat AHCM, which usually has a benign course. Implanted cardiac defibrillators, alcohol septal ablation, and apical myomectomy may be treatment options in high-risk patients.

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