Cardiac Magnetic Resonance Imaging Demonstrates the Coexistence of Hypertrophic Cardiomyopathy and Ischemic Cardiomyopathy due to Coronary Vasospasm

Junghwa Choi¹, Whal Lee², Eun Ah Park²

¹Department of Radiology, Uijeongbu St. Mary's Hospital, College of Medicine, The Catholic University of Korea, Uijeongbu, Korea

²Department of Radiology, Seoul National University Hospital, Seoul, Korea

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Corresponding author

Junghwa Choi, MD
 Department of Radiology,
 Uijeongbu St. Mary's Hospital,
 College of Medicine,
 The Catholic University of Korea,
 271 Cheonbo-ro, Uijeongbu 11765, Korea
 Tel: 82-31-820-4215
 Fax: 82-2-960-4568
 E-mail: cjh@catholic.ac.kr

Whal Lee, MD
 Department of Radiology,
 Seoul National University Hospital,
 101 Daehak-ro, Jongno-gu,
 Seoul 03080, Korea
 Tel: 82-2-2072-0898
 Fax: 82-2-743-6385
 E-mail: whal.lee@gmail.com

We present the case of a 46-year-old male who visited our clinic for the evaluation of ventricular fibrillation and cardiac arrest that occurred one week prior to presentation. He had a family history of hypertrophic cardiomyopathy (HCM). Echocardiography and cardiac magnetic resonance (CMR) imaging revealed myocardial hypertrophy, and he was diagnosed with HCM. Coronary angiography with the ergonovine provocative test showed vasospasm in the proximal left anterior descending artery (LAD), and CMR demonstrated a late-gadolinium enhancement pattern of ischemic cardiomyopathy in the LAD territory and septal HCM. There have been several reports suggesting that coronary spastic angina is not uncommon in Japanese HCM patients, and indicating that calcium channel blockers should be the first-line therapy for variant angina in HCM patients instead of beta-blockers. CMR images displaying the coexistence of HCM and ischemic cardiomyopathy caused by coronary vasospasm have not been previously reported.

Key words Coronary vasospasm · Cardiomyopathy, Hypertrophic · Magnetic resonance imaging.

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is the most common genetic heart disease and the most common cause of sudden cardiac death (SCD) in the young [1]. Cardiac magnetic resonance (CMR) is one of the imaging modalities of choice in most HCM patients because it is useful for both morphologic and functional assessments, and observation of the late gadolinium enhancement (LGE) pattern of ischemic cardiomyopathy due to vasospasm in HCM might help predict the prognosis and change the first-line management of chest pain in these patients.

CASE REPORT

A 46-year-old male was referred to our hospital for evaluation of ventricular fibrillation resulting in cardiac arrest that occurred one week prior to presentation.

The patient had an unremarkable medical history. However, both his father and brother had been diagnosed with HCM, and his uncle died from sudden cardiac arrest in his mid-forties.

The patient underwent electrocardiography, and the findings suggested anterior myocardial infarction. Two-dimensional transthoracic echocardiography showed hypertrophy of the myocardium at the basal anteroseptum and the anterior and anterolateral segment of the left ventricle (LV). The maximal thickness of the myocardium was 19 mm at the basal anteroseptum in the end-diastolic phase.

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Coronary angiography (CAG) revealed normal caliber coronary arteries (Fig. 1A). A provocative test with ergonovine induced focal vasospasm of the proximal segment of the left anterior descending artery (LAD), causing a 90% stenosis which was relieved by nitroglycerin injection (Fig. 1B).

CMR was performed one week after discharge and revealed myocardial hypertrophy (measured as 16.5 mm at the basal

septum, end-diastolic phase) (Fig. 2) and multifocal, fuzzy and patchy delayed enhancement of the septal wall in the basal and mid LV (Fig. 3A and B). Subendocardial delayed enhancement and hypokinesia of the basal, mid anteroseptal wall, and apical septal wall of the LV were also noted (Fig. 3C-E). The systolic function of the LV was preserved, and cavity size was within normal range (ejection fraction 68.9%, end-diastolic volume

Fig. 1. A 46-year-old male with hypertrophic cardiomyopathy and coronary vasospasm. Initial coronary angiography revealed normal caliber coronary arteries (A). Administration of ergonovine induced focal vasospasm of the proximal segment of the left anterior descending artery, causing a 90% stenosis which was relieved by nitroglycerin injection (B).

Fig. 2. A 46-year-old male with hypertrophic cardiomyopathy and coronary vasospasm. Cardiovascular magnetic resonance imaging. (A and B) Short-axis cine sequence of the patient demonstrated myocardial hypertrophy at the basal LV anteroseptal, anterior walls and the mid LV anteroseptal, inferoseptal walls (maximal wall thickness measured as 16.5 mm at the basal septum, end-diastolic phase). LV: left ventricular.

Fig. 3. A 46-year-old male with hypertrophic cardiomyopathy and coronary vasospasm. Cardiovascular magnetic resonance imaging. (A and B) Short-axis late-gadolinium enhanced sequence of the patient demonstrated myocardial hypertrophy and multifocal, fuzzy and patchy late-gadolinium enhancement pattern of the septal wall in the basal and mid LV. (C-E) In the mid to apical LV, the late-gadolinium enhanced sequence pattern was restricted to the subendocardial layer of the septal wall, corresponding to the vascular territory of the left anterior descending artery. LV: left ventricle.

index 67.5 mL/m^2).

The patient was asymptomatic, and blood analysis, including cardiac enzymes, were all within normal range on admission and throughout the study.

The patient was treated with vasodilators with a presumed diagnosis of variant angina which might have caused the ventricular fibrillation in addition to the ischemic cardiomyopathy observed on CMR. The patient was also diagnosed with HCM given the family history of HCM and the myocardial hypertrophy demonstrated on CMR. An implantable cardioverter defibrillator was considered to manage the HCM and prevent SCD. Informed consent was obtained.

DISCUSSION

HCM is the most common genetic heart disease with a prevalence of 1:500 in the general population. HCM is also the most common cause of SCD in the young [1]. CMR imaging helps establish a diagnosis and predict the risk of SCD in patients with HCM, providing tissue characterization as well as morphologic

and hemodynamic information. LGE of the hypertrophied myocardium is observed in 60–80% of HCM patients undergoing CMR [1-3]. Post-mortem studies of HCM patients with SCD displayed vessel wall thickening of the intramural coronary arterioles, and this finding explains the hypoperfusion of the myocardium during stress and the myocardial fibrosis revealed as LGE on CMR in HCM patients [1].

Our patient was diagnosed with HCM based on a family history of HCM and myocardial hypertrophy seen on echocardiography and CMR images. The multifocal and patchy LGE in the basal and mid LV septum on CMR corresponded to the typical finding of HCM. However, a linear LGE pattern in the mid and apical LV septum was confined to the vascular territory of the LAD and the subendocardial layer. This imaging feature was not explained by HCM and was highly suggestive of ischemic cardiomyopathy.

The presence of LGE has been associated with the development of heart failure symptoms and decreased systolic function of the LV in HCM patients [1,3]. However, our patient's admission evaluation revealed preserved contractile function one

week after the cardiac arrest, indicating that the etiology of his arrhythmia and chest pain may have been coronary vasospasm rather than HCM.

There have been several reports suggesting that HCM can coexist with coronary spastic angina in Japanese patients, and one retrospective study showed the mutation of the delta-sarcoglycan gene was associated with coronary spasm in Japanese patients with HCM [4]. These reports suggested that coronary vasospasm in patients with HCM might play a potential role for the conversion of HCM to dilated cardiomyopathy, thus affecting their prognosis. Also in the clinical setting, calcium channel blockers should be the first-line therapy recommended for coronary spastic angina in HCM patients in order to induce coronary vasodilatation and to prevent the aggravation of coronary spasm by beta-blockers which are commonly administered to HCM patients to relieve chest pain [5].

Despite the recognized clinical significance of the coexistence of HCM and coronary vasospasm, this is the first report of variant angina in an HCM patient which presented with the LGE pattern of HCM and also ischemic cardiomyopathy on CMR images.

In summary, we have presented an interesting case of concurrent HCM and ischemic cardiomyopathy due to coronary vasospasm on CMR images in South Korea. The utilization of CMR to evaluate cardiomyopathy is increasing due to its ob-

jective assessments and development in spatial and temporal resolution. The recognition of the LGE pattern of ischemic cardiomyopathy in HCM patients is expected to provide the information necessary for the proper recommendations in the evaluation of chest pain in patients with HCM, which includes using cardiac catheterization and the ergonovine or acetylcholine provocation test and proper management with calcium channel blockers as a first-line therapy.

Conflicts of Interest

The authors declare that they have no conflict of interest.

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