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CASE REPORT

INTERMEDIATE

CLINICAL CASE

Pacemaker Treatment for Apical Hypertrophic Cardiomyopathy in the Setting of an Apical Ventricular Aneurysm

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ABSTRACT

We describe the case of a patient with apical hypertrophic cardiomyopathy with concomitant apical aneurysm. We measured the aneurysmal cavity pressure using the pressure guidewire system. The patient underwent implantable cardioverter-defibrillator treatment successfully to reduce the pressure gradient between the aneurysmal cavity and the true left ventricle. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:1150–5) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 78-year-old man, who was given a diagnosis of apical hypertrophic cardiomyopathy (ApHCM) 17 years earlier, was admitted to Dokkyo Medical University Hospital (Mibu, Tochigi, Japan) for detailed examination. Ten years ago, cardiac computed tomography showed a small amount of contrast medium pooling in a hypertrophic apical area (Figure 1A). Follow-up computed tomography 1 year before this admission revealed a larger amount of pooling contrast material, consistent with an aneurysm (Figure 1B). Cardiac magnetic resonance at that time revealed late gadolinium enhancement (Figures 2A to 2D). Holter electrocardiogram monitoring documented 198 beats/d of premature ventricular beats with a single 4-beat short run. He reported exertional dyspnea of 6 months' duration. On admission, his plasma B-type natriuretic peptide level was 185 pg/dL.

LEARNING OBJECTIVES

- To understand that apical aneurysms increase the risk of adverse cardiac events in HCM.
- To understand that the pressure guidewire system for fractional flow reserve measurement in coronary artery stenosis is useful to measure intracavitary pressure in the apical aneurysm.
- To understand that implantable cardiac defibrillator in the AV sequential pacing mode is a promising treatment strategy for a case such as in this patient.

PAST MEDICAL HISTORY

Fifteen years earlier, the patient received a diagnosis of coronary spastic angina. He was subsequently treated with oral benidipine hydrochloride (8 mg/d) and nicorandil (15 mg/d).

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DIFFERENTIAL DIAGNOSIS

The other causes of left ventricular aneurysm were excluded.

INVESTIGATIONS

The patient underwent cardiac catheterization using a conventional fluid-filled catheter-transducer system, with previous home medications withheld. Left ventricular systolic and end-diastolic pressures were 115 and 10 mm Hg, respectively. Next, we tried to measure the pressure of the aneurysmal cavity. Because measurement using the conventional system was difficult, we measured intracavitary pressure by using a pressure guidewire system for fractional flow reserve measurement in coronary artery stenosis (COMET, Boston Scientific). The systolic intracavitary pressure was

ABBREVIATIONS AND ACRONYMS

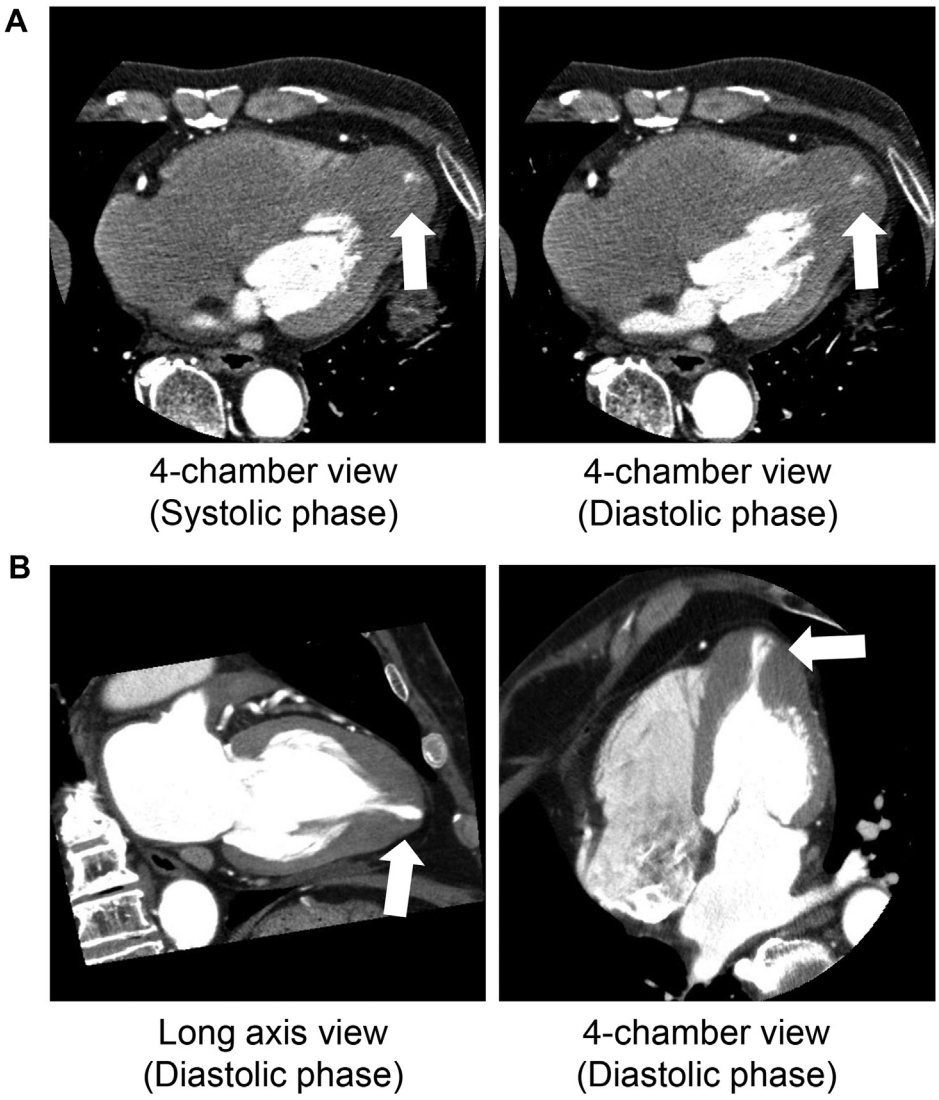
ApHCM = apical hypertrophic cardiomyopathy

AV = atrioventricular

HCM = hypertrophic cardiomyopathy

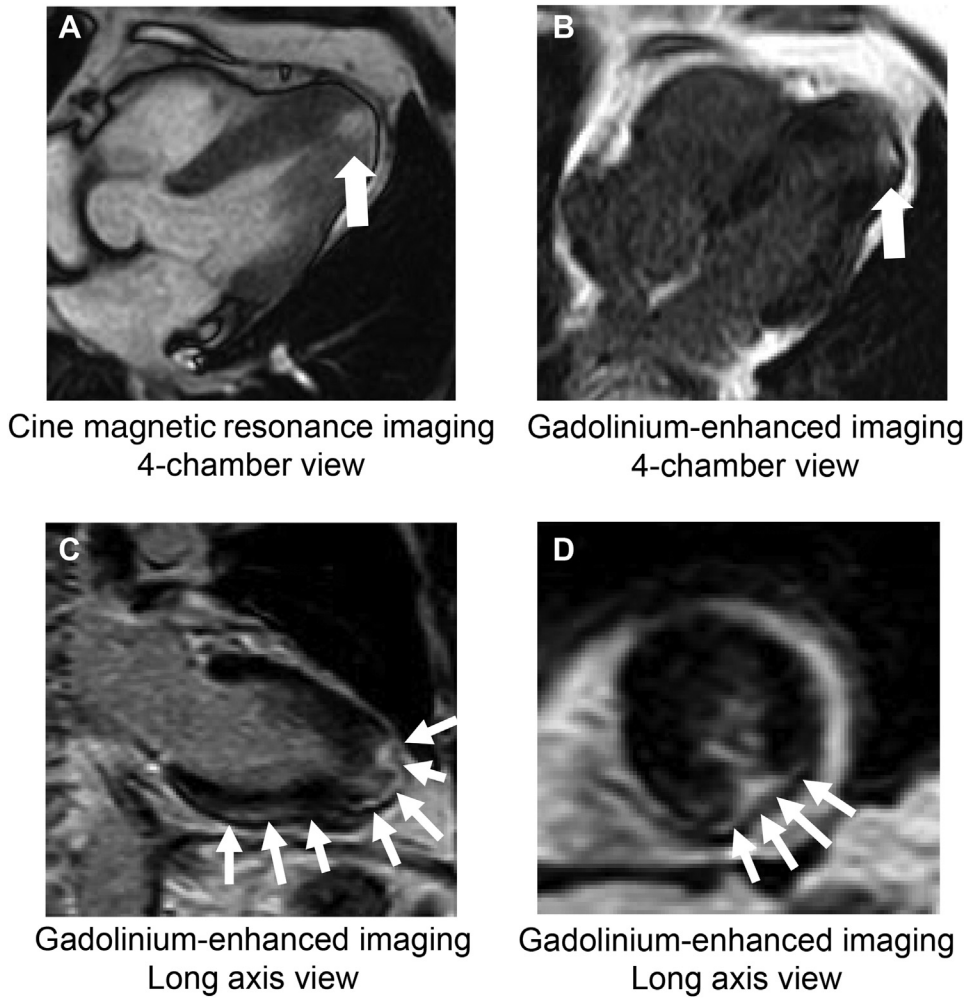
ICD = implantable cardioverter-defibrillator

FIGURE 1 Cardiac Computed Tomography Findings



(A) At 10 years before admission, a small amount of contrast material was observed pooling in the hypertrophic apical area during both systole and diastole in the 4-chamber view **(arrows)**. **(B)** In the follow-up study 1 year previously, the amount of contrast material pooling was larger, consistent with diagnosis of an aneurysm **(arrows)**.

FIGURE 2 Cardiac Magnetic Resonance Findings



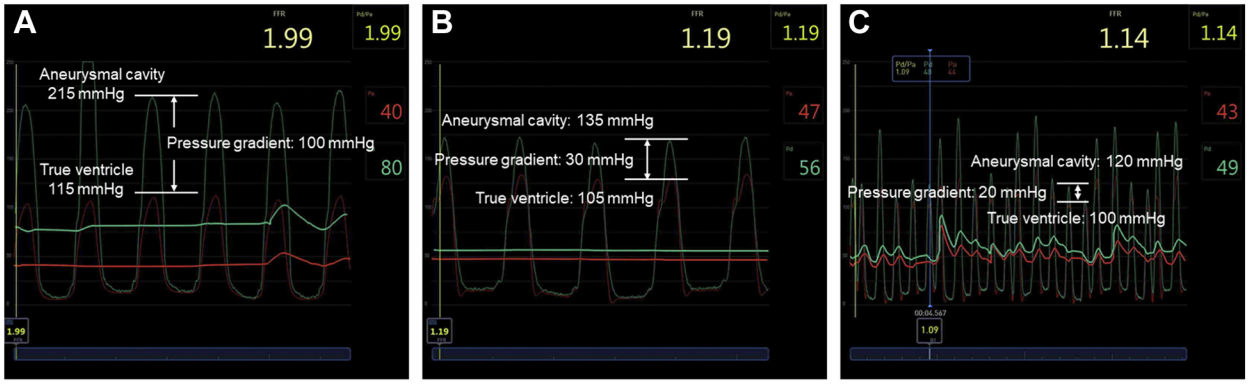
(A) Plain cine cardiac magnetic resonance shows the apical aneurysm **(arrows)**. Gadolinium-enhanced imaging demonstrated late gadolinium enhancement in the **(B)** 4-chamber view **(arrows)**, **(C)** long-axis view **(arrows)**, and **(D)** short-axis view **(arrows)**.

215 mm Hg, and 100 mm Hg of the peak-to-peak pressure gradient was evident between the aneurysmal cavity and the true left ventricle (Figure 3A). We attempted to intervene in the pressure gradient by using pharmacologic and mechanical approaches. First, we confirmed that intravenous injection of cibenzoline (1.4 mg/kg) for 3 minutes substantially decreased the pressure gradient to 30 mm Hg (Figure 3B). Next, right ventricular pacing revealed beat-by-beat variation of the pressure gradient, depending on the atrioventricular (AV) interval, and the 60 beats/min pacing for 1 minute showed a decrease in the gradient minimally to 8 to 20 mm Hg (Figure 3C). Concomitant coronary angiography showed no organic stenotic lesions in the left and right coronary arteries, but an acetylcholine provocation test of the left coronary artery (100 μg) resulted in 99% narrowing of the midportion of the left circumflex artery.

MANAGEMENT

After the cardiac catheterization, the patient received 300 mg/d of oral cibenzoline for reduction of his intraventricular pressure gradient. His home benidipine and nicorandil were continued. Four months later, according to his wish, he underwent placement of an implantable cardioverter-defibrillator (ICD)

FIGURE 3 LV Pressure Wave in Cardiac Catheterization Study



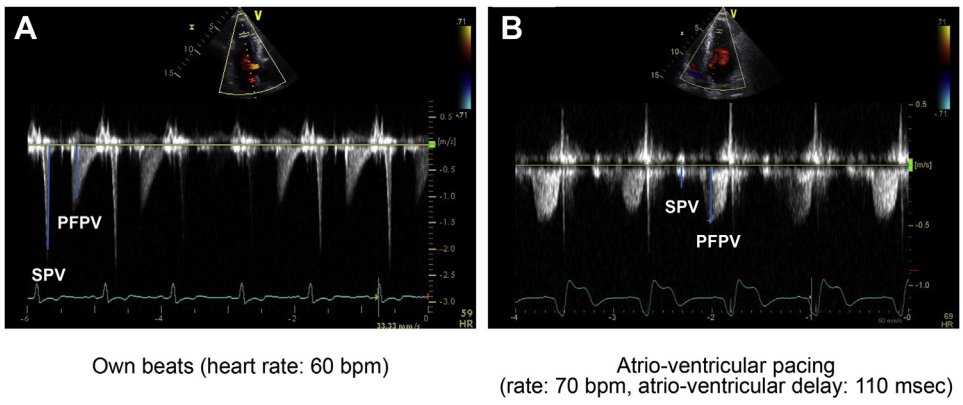
For the ventricular waveform, the **red wave** indicates pressure in the true left ventricle, and the **green wave** indicates pressure in the aneurysmal cavity. **(A)** The peak-to-peak pressure gradient was 100 mm Hg between the aneurysmal cavity and the true left ventricle at rest. **(B)** Intravenous injection of cibenzoline subsequently decreased the pressure gradient to 30 mm Hg. **(C)** Right ventricular pacing represented beat-by-beat variation of the pressure gradient, depending on the atrioventricular interval. The 60 beats/min pacing for 1 minute produced 5 cycles of pressure gradient variation. Minimal pressure gradients for each cycle were 8 to 20 mm Hg. LV = left ventricular.

(Evera MRI XT DR SureScan, Medtronic) for AV sequential pacing under the setting of DDD mode with a basic rate of 70 beats/min and an AV interval of 110 ms. As a result, the high-pulse repetition frequency pulse Doppler echocardiogram showed that the systolic peak flow velocity was reduced from 2.4 to 0.1 m/s and the diastolic paradoxical flow peak velocity was reduced from 1.0 to 0.5 m/s at the ostium of the true ventricle to aneurysmal cavity and that the estimated pressure gradient between the aneurysmal cavity and the true left ventricle was reduced from 24 mm Hg to 0 mm Hg (Figures 2B and 4A).

DISCUSSION

Left ventricular apical aneurysm was thought to be a rare complication of hypertrophic cardiomyopathy (HCM), but it is increasingly identified with the

FIGURE 4 High-Pulse Repetition Frequency Pulse Doppler Recording



(A) At baseline heart rate and rhythm, the systolic peak flow velocity (SPV) was 2.4 m/s and the diastolic paradoxical flow peak velocity (PFPV) was 1.0 m/s at the ostium of the true ventricle to aneurysmal cavity, and the estimated pressure gradient between the aneurysmal cavity and the true left ventricle was 24 mm Hg, with the patient taking oral cibenzoline 300 mg/d. **(B)** At atrioventricular sequential pacing, the systolic peak flow velocity was 0.1 m/s, the paradoxical flow peak velocity was 0.5 m/s, and the estimated pressure gradient was almost 0 mm Hg.

advancement of imaging modalities. In 2017, Rowin et al (1) retrospectively analyzed 1,940 consecutive patients with HCM and reported that 93 patients (4.8%) had a concomitant apical aneurysm; 3 of the 93 patients with apical aneurysm (3%) died either suddenly or of heart failure. The life-threating complications include fatal arrhythmias, such as ventricular tachycardia or ventricular fibrillation, and thromboembolic events. Regarding aggressive treatment strategies, ICD, radiofrequency catheter ablation, alcohol septal myocardial ablation, surgical myectomy, and aneurysmectomy have been proposed (1–4).

ApHCM is an uncommon phenotype of HCM, with an incidence of only 1% to 2% of HCM cases in Western countries, but it accounts for up to 25% of cases in Japan (5,6). In addition, the percentage of concomitant apical aneurysms in patients with ApHCM is 2% in Western countries, but 24% in Japan (7,8). The prognosis of ApHCM seems to be benign, but when it is accompanied by apical aneurysm, it can lead to adverse cardiac events (8). Nevertheless, there are no evidence-based established treatments for ApHCM in combination with apical aneurysm. Although the mechanisms for the development of apical aneurysm remain unknown, high intracavitary pressure may promote myocardial degeneration and fibrosis, which potentially lead to adverse cardiac events, including fatal arrhythmias.

In the presented case of ApHCM, the patient's apical aneurysm had been developing over at least 10 years. Cardiac magnetic resonance revealed the presence of late gadolinium enhancement, which is considered a significant predictor of adverse clinical events in HCM (9). During cardiac catheterization, we observed a high intracavitary pressure and a high pressure gradient between the aneurysmal cavity and the true left ventricle, by using a pressure guidewire system for fractional flow reserve measurement. To our knowledge, this method of measuring the pressure of the aneurysmal cavity has not been reported elsewhere, and we believe that this method was crucial as a diagnostic tool. On the basis of these results, we proceeded to pressure gradient reduction as a first-line treatment strategy. We could not treat the patient with β-blockers and could not withhold use of vasodilating agents such as benidipine and nicorandil because of his underlying coronary spastic angina, but we found that oral cibenzoline was effective. In addition to oral cibenzoline, we inserted the ICD for the purpose of further pressure gradient reduction by AV sequential pacing function and also as the countermeasure for fatal arrhythmias, which could happen in the future. More than anything, the device treatment was what the patient wanted. Consequently, the systolic peak flow velocity and diastolic paradoxical flow peak velocity at the ostium into the aneurysmal cavity were substantially reduced, and the estimated pressure gradient was substantially reduced, as shown by Doppler echocardiographic findings. To our knowledge, this is the first case of AV sequential pacing treatment used for the purpose of pressure gradient reduction in a patient with ApHCM and an apical aneurysm. Because high intracavitary pressure may be associated with adverse cardiac events, we believe that the pacemaker treatment would be promising in patients with ApHCM who have apical aneurysms.

FOLLOW-UP

After the treatment with AV sequential pacing, the patient's exertional dyspnea improved, and his B-type natriuretic peptide level 2 months later was 48 pg/dL.

CONCLUSIONS

Given that high intracavitary pressure may be related to adverse cardiac events, we believe that AV sequential pacing treatment would be promising in patients with ApHCM and a concomitant apical aneurysm.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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