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Case Report

A case of hypertrophic obstructive cardiomyopathy in which left ventricular remodeling and reverse remodeling were seen with pacing on and off

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ABSTRACT

A 77-year-old woman with hypertrophic obstructive cardiomyopathy was admitted to the hospital in March 2005 with a chief complaint of chest discomfort, and a left ventricular outflow tract (LVOT) gradient was seen. After starting apical dual chamber (DDD) pacing and oral cibenzoline 300 mg/day to relieve the stenosis, the pressure gradient and subjective symptoms disappeared. The patient was then followed as an outpatient. In January 2008, cibenzoline was discontinued because the patient experienced a hypoglycemic attack. Plain chest radiographs showed an increased cardiothoracic ratio from May 2009 and ventricular remodeling was suspected, although there were no changes in chest symptoms. Therefore, pacing off was considered. Acute changes in the LVOT gradient were evaluated with echocardiography before and after pacing on–off, but no changes were seen, so the course was observed in the pacing-off state. The LVOT gradient gradually increased again from 7 months after pacing off, and the pressure gradient decreased again after pacing was restarted. On the electrocardiogram, a deep negative T wave was seen in V_{4-6} immediately after pacing off, but with time, it became positive, similar to before an implantable cardioverter defibrillator was inserted. Reverse remodeling was judged to occur after pacing off, and pacing therapy was restarted. The patient is currently under observation.

1. Introduction

Right ventricular pacing (apical) therapy is reported to be effective in relieving stenosis of the left ventricular outflow tract in hypertrophic obstructive cardiomyopathy, but there are no reported cases in which pacing off was carried out and the subsequent course observed in a long-term follow up. Here we report a case of hypertrophic obstructive cardiomyopathy in which reverse remodeling occurred after pacing off, the outflow tract gradient increased again, a new stenosis occurred in the center of the left ventricle, and apical ventricular aneurysm-like changes were seen.

2. Case history

History of current illness: the patient was a 77-year-old woman. She was treated for hypertension and dyslipidemia. She had no siblings with heart disease or who had experienced sudden death. She began to feel exertional chest discomfort beginning in Octobar 2000. From about March 2005, the chest discomfort started to occur frequently, and she was examined in our department. Asymmetric septal hypertrophy (ASH), systolic anterior motion (SAM), and left ventricular outflow tract (LVOT) stenosis (PG max = 124 mm Hg) were seen on echocardiography (Fig. 1), and the patient was hospitalized with a diagnosis of suspected hypertrophic obstructive cardiomyopathy.

No significant stenosis was seen on coronary angiography, and the lumen was banana shaped in systole on left ventriculography. The maximum LV-Ao gradient was 130 mmHg with pullback pressure. On histopathological examination of a myocardial biopsy specimen obtained at the same time, myocardial disarray was seen, and hypertrophic obstructive cardiomyopathy was diagnosed.

The outflow tract gradient was judged to be the cause of the chest discomfort. With the aim of relieving the pressure gradient, oral cibenzoline 300 mg/day and apical DDD pacing were started. The patient had no episode of syncope, but considering the proarrhythmic effect of cibenzoline (there are some case reports regarding class Ic antiarrhythmic drug-induced ventricular tachycardia and fibrillation in patients with hypertrophic cardiomyopathy) [1,2], an implantable cardioverter defibrillator (ICD) (PRIZM2, Boston scientific, St. Paul, MN) was implanted. The atrial pacing lead was located in the right atrial appendage, and the

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defibrillation lead was located in the right ventricular apex, with a set-rate of 60 bpm and a paced AV delay of 70 ms. With these findings, both the gradient (124 mm Hg→24.6 mm Hg) and subjective symptoms were resolved. The patient's symptoms improved from the New York Heart Association (NYHA) class III to class I after which she was discharged from the hospital.

A hypoglycemic attack occurred in January 2008, and cibenzoline was discontinued. Afterward, no changes in the LVOT gradient were seen on echocardiography, but enlargement of the cardiothoracic ratio was seen on plain chest radiographs from May 2009. No changes occurred in chest symptoms (NYHA class I), but an effect from left ventricular remodeling was suspected, and pacing off was considered. Acute changes in the LVOT gradient were evaluated with echocardiography before and after pacing on–off. No changes were seen before and after pacing on–off (6.8 mm Hg and 8.1 mm Hg, respectively), and so the patient's course was observed in a pacing-off state after March 2009. New functional stenosis (PG max=36 mm Hg) in the center of the left ventricle (Fig. 2), which had not been seen previously, was seen on echocardiography in May 2009, but outflow tract stenosis did not recur. The outflow tract gradient gradually increased (9–16 mm Hg) from October 2009. A small cavity was seen in the left ventricular apex in December 2009. The gradient decreased again with the restart of pacing, and reverse remodeling was judged to be the cause of the repeat increase in the outflow tract gradient in this case. The course of the outflow tract gradient is shown in Fig. 3. A deep negative T wave was seen immediately after pacing off on electrocardiogram in March 2009, but the negative T wave improved with partial induction on electrocardiogram in October 2009 (Fig. 4). The reverse remodeling after pacing off was judged to be due to this improvement in the T wave, and DDD pacing therapy is currently under way.

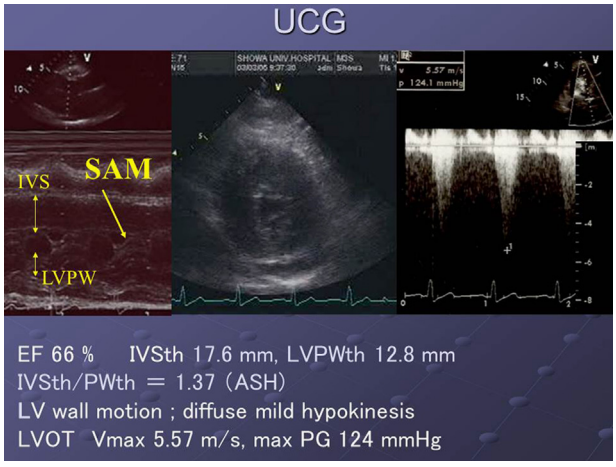

Fig. 1. Asymmetric septal hypertrophy (ASH), systolic anterior motion (SAM), and left ventricular outflow tract (LVOT) stenosis (PG max=124 mm Hg) were seen on echocardiography. EF=ejection fraction, IVSth=interventricular septum thickness, LVPWth=left ventricular posterior wall thickness, ASH=asymmetrical septal hypertrophy, LV=left ventricule, LVOT=left ventricular outflow tract.

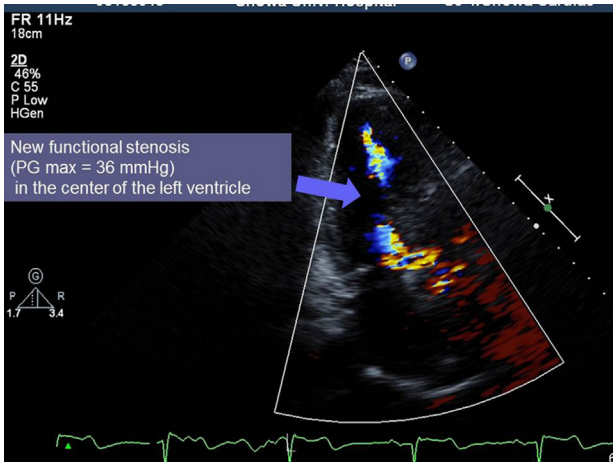

Fig. 2. New functional stenosis (PG max=36 mm Hg) in the center of the left ventricle, which had not been seen previously, was seen on echocardiography in May 2009, but outflow tract stenosis did not recur.

3. Discussion

This case is the first report of hypertrophic obstructive cardiomyopathy in which electrical and functional reverse remodeling occurred after pacing off, and a new stenosis in the center of the left ventricle and apical ventricular aneurysm-like changes were seen when the outflow tract gradient increased again.

There are many reports on the effectiveness of pacing therapy for hypertrophic obstructive cardiomyopathy. Fananapazir et al. reported that long-term right ventricular apical pacing improved both the left ventricular outflow tract gradient and subjective symptoms [3]. Maron et al. reported improvement in quality of life scores with long-term DDD pacing, but no relationship with gradient decrease was seen, and the improvements in subjective symptoms were considered a placebo effect from pacemaker implantation [4].

Hosoda et al. reported a case of hypertrophic obstructive cardiomyopathy treated with DDD pacing therapy. In that report, cardiac catheterization study was performed 7 years after ICD implantation, and LVOT gradient was significantly low (10 mm Hg) and this state continued more than 60 min after pacing was turned off [5]. In the current case, no changes in the left ventricular outflow tract gradient were seen for 7 months after pacing off. Pacing therapy is reported to not only reduce myocardial constriction around the pacing site, but also produce constriction from mechanical dyssynchrony, with remodeling in the long term [6–10]. It has also been reported that an abnormal excitation pattern in the left ventricle continues for some time after pacing off [6]. In the current case, long-term right ventricular apical pacing brought about left ventricular myocardial remodeling, and it is thought that the gradient did not increase because

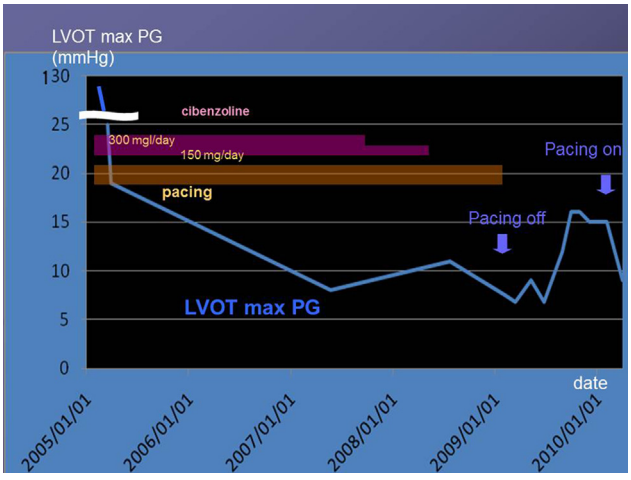

Fig. 3. The patient's course was observed in a pacing-off state after March 2009. The outflow tract gradient gradually increased (9–16 mm Hg) from October 2009. The gradient decreased again with the restart of pacing. LVOT=left ventricular outflow tract.

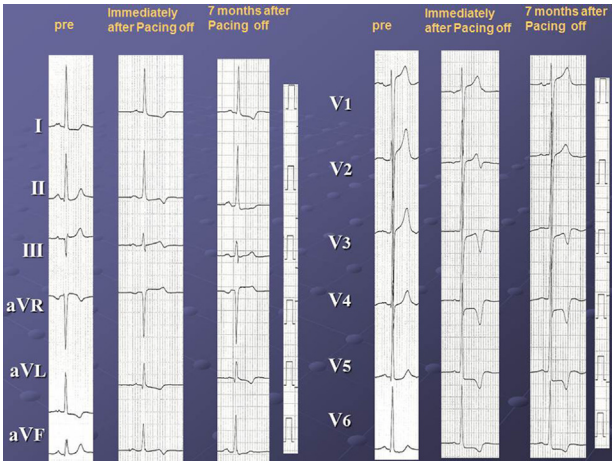

Fig. 4. A deep, negative T wave was seen immediately after pacing off on electrocardiogram, but the negative T wave improved with partial induction over time after pacing off.

this state continued for some time even after pacing off. Over time, after pacing off, the LVOT tract gradient increased again. It was therefore judged that reverse remodeling had occurred.

A new stenosis in the center of the left ventricle and apical ventricular aneurysm-like changes were seen at this time. There have been no other similar reports. Stenosis in the center of the left ventricle is a relatively rare condition even in hypertrophic cardiomyopathy, and it is reported to often be accompanied by apical ventricular aneurysm from the frequently increased pressure load in the apex [11,12]. Although there are no reports of stenosis in the center of the left ventricle during the course of hypertrophic cardiomyopathy accompanied by LVOT stenosis, this may be the natural course of hypertrophic cardiomyopathy.

In this case, functional stenosis occurred in the center of the left ventricle after pacing was stopped. Afterward, a cavity was seen, so that development of a ventricular aneurysm in the future is also a possibility. Therefore, restarting right ventricular apical pacing is thought to have been very important for the outcome of this case.

In the current patient, right ventricular apical pacing was performed for hypertrophic obstructive cardiomyopathy, and later stopped. There was no increase in the gradient even in a state without pacing for a certain time; thus, there would seem to be some room for thought with regard to future pacing therapy. In addition, the location of the hypertrophy shifted from the LVOT to mid-ventricular-apical, and careful observation will be needed in the future.

Conflict of interest

None declared.

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