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Recurrent Cardiogenic Shock Caused by Dynamic Mitral Regurgitation in a Patient With Hypertrophic Obstructive Cardiomyopathy: A Case Report

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Abstract

A 68-year-old man with hypertrophic obstructive cardiomyopathy developed recurrent cardiogenic shock due to dynamic mitral regurgitation. The pressure gradient in the left ventricular outflow tract under medication was 30 mmHg, and he complained of no symptom. He developed shock with dyspnea suddenly after bathing and defecation. Echocardiography and left ventriculography revealed massive mitral regurgitation and mild increment of pressure gradient in the left ventricular outflow tract (50 mmHg). He underwent successful mitral valve replacement following treatment with beta-blocker under intraaortic balloon pumping support. This case illustrates that exacerbation of the systolic anterior motion of the mitral anterior leaflet can cause dynamic severe mitral regurgitation with ' mild ' increment of pressure gradient in the left ventricular outflow tract, resulting in cardiogenic shock with severe lung edema.

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Key Words

■Cardiomyopathies, hypertrophic **■Mitral regurgitation** **■Shock**

INTRODUCTION

Hypertrophic cardiomyopathy has a diverse clinical course characterized by both sudden death and disabling symptoms related to heart failure[1,2]. Left ventricular outflow tract (LVOT) obstruction at rest is recognized as an independent predictor of heart failure and sudden death[3]. We report a case of recurrent cardiogenic shock caused by transient severe mitral regurgitation in a patient with obstructive hypertrophic cardiomyopathy, who was rescued by intraaortic balloon pumping (IABP) and beta-blocker administration, and successfully underwent mitral valve replacement.

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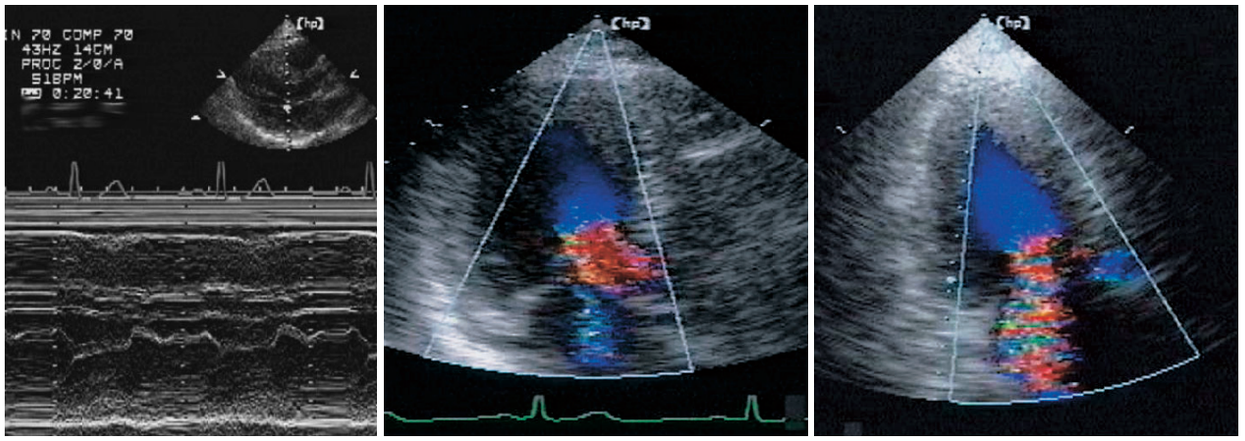

Fig. 1 M-mode echocardiograms
Mild left ventricular hypertrophy and incomplete systolic anterior motion of the mitral valve are present (*left*). Mild mitral regurgitation is present in the stable condition (*middle*), and is exacerbated during shock (*right*).

CASE REPORT

A 68-year-old man was referred to Omiya Medical Center, Jichi Medical School because of chest oppression during exercise in March 2002. The diagnosis was hypertrophic cardiomyopathy with mild obstruction in the LVOT. Echocardiography (**Figs. 1−left**, **middle**) revealed mild left ventricular hypertrophy of both the septum (interventricular septal thickness: 15 mm) and the posterior wall (posterior wall thickness: 11 mm), the narrow region of the LVOT caused by incomplete systoloic anterior motion (SAM) of the mitral valve, and mild mitral regurgitation. Therefore, he had been treated with metoprolol 30 mg, disopyramide 300 mg and diltiazem 300 mg per day. These medications relieved him from chest oppression, and maintained the pressure gradient in the LVOT at around 30 mmHg.

He developed shock suddenly after bathing, and was transferred to our hospital by ambulance in January 2003. On admission, systolic blood pressure was 40 mmHg, heart rate was 45 beats/min, and SpO_2 under 6 *l* with O_2 inhalation of 86%. Coarse crackle was audible in all lung fields, and grade Ⅳ/Ⅵ pan-systolic murmur was audible at the apex. Electrocardiography showed sinus bradycardia and depression of the ST segment in leads Ⅰ, Ⅱ, Ⅲ, aV_F, and V_3−V_6 (**Fig. 2−left**). Chest radiography showed cardiomegaly and butterfly shadow (**Fig. 2−right**). Echocardiography revealed that the anterior ' deviation ' of the mitral anterior leaflet had caused severe mitral regurgitation and LVOT narrowing (**Fig. 1−right**). Regional wall motion abnormalities and papillary muscle rupture were not detected, and ejection fraction of the left ventricle was approximately 70%. Emergent coronary angiography showed no fixed stenosis. The mitral regurgitation jet reached the pulmonary veins, and was recognized as Seller's grade Ⅳ/Ⅳ by left ventriculography (**Fig. 3−left**). The pressure gradient between left ventricle and the ascending aorta was approximately 50 mmHg (**Fig. 4−left**). Left ventricular wall motion revealed hyperkinesis without asynergy (left ventricular end-diastolic volume index: 91 m*l*/m^2, left ventricular end-systolic volume index: 11 m*l*/m^2, left ventricular ejection fraction: 88%).

He was treated using a respirator and a temporary VVI mode pacemaker and sufficient fluid therapy. However, his hemodynamic state was not improved. Therefore, an IABP was inserted through the right femoral artery. Blood pressure was slightly increased, although severe mitral regurgitation remained. Therefore, propranolol (2 mg) was administered intravenously to reduce SAM. Catecholamine was not administered during the entire course of treatment. After these procedures, the degree of mitral regurgitation was decreased significantly by left ventriculography (**Fig. 3−right**), and the pressure gradient between the left ventricle and ascending aorta almost vanished (**Fig. 4−right**). Consequently, his vital signs and general condition were dramatically improved.

Metoprolol, 180 mg and disopyramide, 300 mg per day orally were administered after recovery.

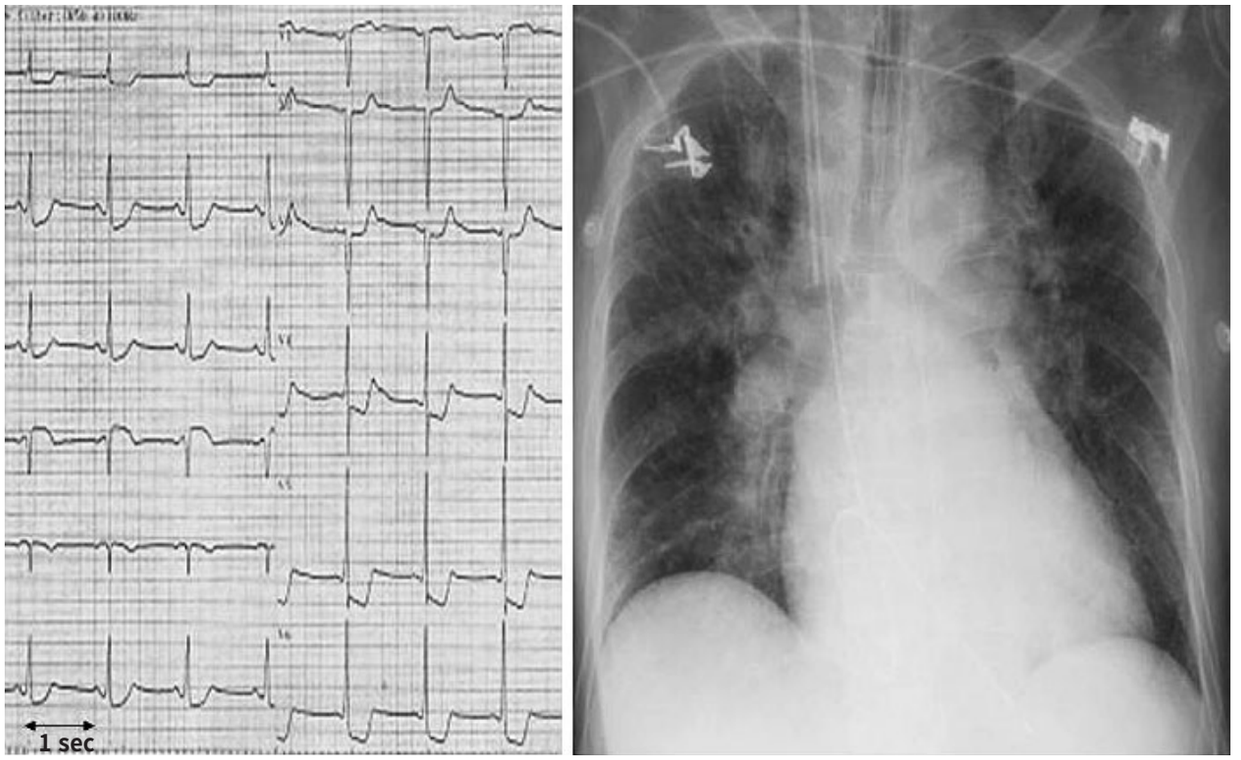

Fig. 2 Electrocardiogram and chest radiograph on admission
Left: Electrocardiogram shows sinus bradycardia and depression of the ST segment in the Ⅰ, Ⅱ, Ⅲ, aVF, and V_3-V_6 leads.
Right: Chest radiograph shows the butterfly shadow and the cardiothoracic ratio is 62%.

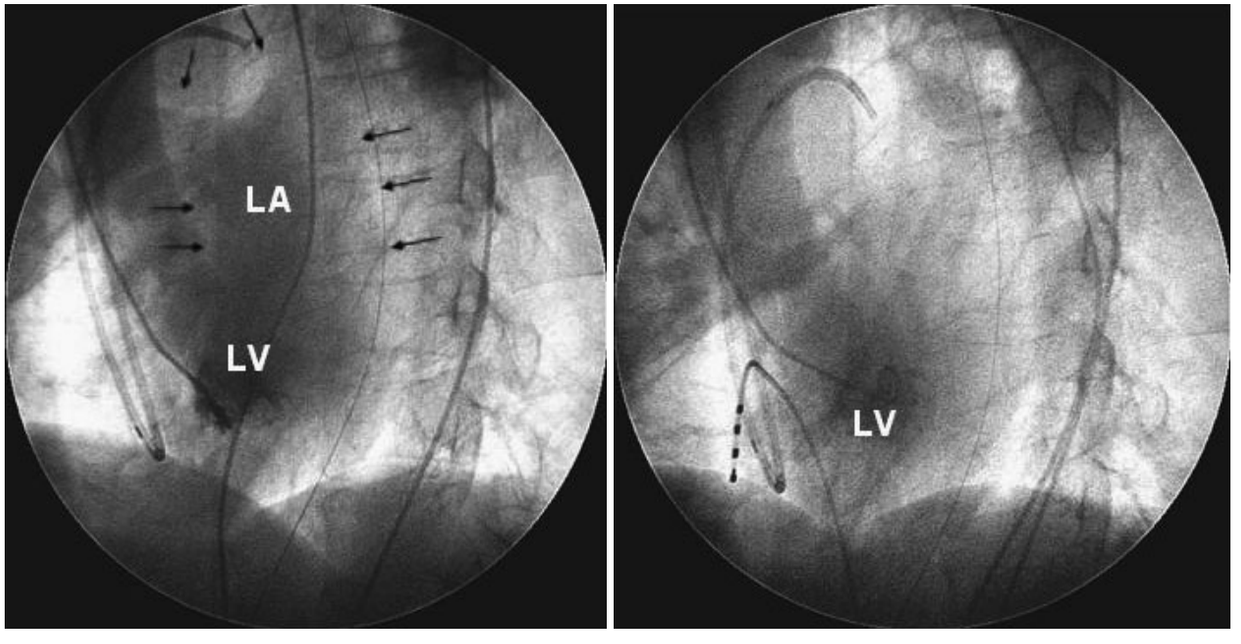

Fig. 3 Left ventriculograms showing the changes in mitral regurgitation
Left: Before treatment, severe mitral regurgitation reached the pulmonary vein.
Right: After propranolol(2 mg)injection during intraaortic balloon pumping support, the mitral regurgitation was decreased significantly. *Arrows* indicate the left atrium and pulmonary vein.
LA = left atrium; LV = left ventricle.

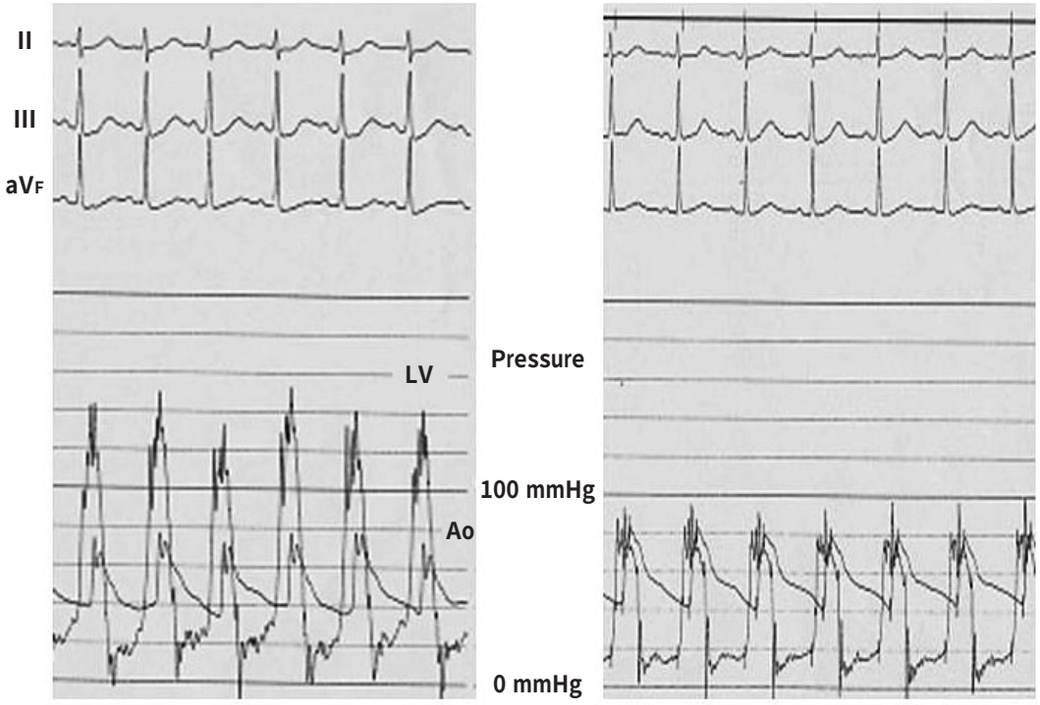

Fig. 4 Changes in the pressure gradient between left ventricle and ascending aorta
Left: On admission, the pressure gradient was approximately 50 mmHg, and the systolic pressure of the ascending aorta was approximately 60 mmHg.
Right: After beta-blocker administration, the pressure gradient decreased to 0 mmHg. Measurement of these pressures was performed without intraaortic balloon pumping support.
Ao = ascending aorta. Other abbreviation as in Fig. 3.

However, on day 20, he developed shock suddenly after defecation, and again needed treatment with beta-blocker and IABP. Echocardiography showed massive mitral regurgitation with 50 mmHg pressure gradient in the LVOT, similar to the first attack. Finally, he underwent mitral valve replacement successfully at Shin-Tokyo Hospital in June 2003. There were no primary changes in the mitral complex apparatus such as papillary muscle displacement and leaflet elongation. Histological examination showed focal fibrosis and myxoid degeneration of the mitral valve complex.

DISCUSSION

The present case demonstrated that a hypertrophic cardiomyopathy patient with mild stenosis of the LVOT who developed cardiogenic shock due to dynamic mitral regurgitation could be successfully treated with beta-blocker and IABP. Dynamic mitral regurgitation is often induced by papillary muscle ischemia, but coronary stenosis was not detected in this patient. In patients with hypertrophic cardiomyopathy, mitral regurgitation depends on the SAM of the anterior mitral leaflet, and the severity of mitral regurgitation is directly related to the magnitude of the LVOT gradient[4]. In this patient, it is important to note that the SAM affected the severity of mitral regurgitation more than the LVOT gradient. We think that degeneration of mitral leaflets affects the development of mitral regurgitation. SAM was easily provoked by bathing and defecation, which resulted in reduction in preload and afterload.

Beta-blocker and IABP support were effective in this case. IABP during acute mitral regurgitation may lower aortic impedance, resulting in less mitral regurgitation and more output toward the aorta[5]. However, in patients with obstructive hypertrophic cardiomyopathy, IABP use is considered to be contraindicated because reducing afterload results in exacerbation of SAM and stenosis at the LVOT. Moreover, to reduce the obstruction at the LVOT, beta-blocker and fluid therapy are recommended because SAM is mainly caused by the Venturi effect. The combination of beta-blocker and IABP was effective to rescue our patient with dynamic

severe mitral regurgitation due to SAM.

Major therapeutic interventions such as ventricular septal myetomy, percutaneous alcohol septal ablation, mitral valve replacement, and dual-chamber pacing have been introduced to improve the disabling symptoms associated with LVOT obstruction[6–8]. In this case, mitral valve replacement was selected as the radical treatment because residual mitral regurgitation was recognized after recovery, the hypertrophy of interventricular septal was not so severe, so percutaneous alcohol septal ablation might not be so effective, and mitral valve degeneration was believed to be the partial cause of mitral regurgitation.

This case suggests that beta-blocker administration during IABP support can be used to successfully treat a patient with cardiogenic shock due to dynamic mitral regurgitation and mild increment of the LVOT gradient.

要　　約

可逆性僧帽弁閉鎖不全により心原性ショックを繰り返した閉塞性肥大型心筋症の1例

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閉塞性肥大型心筋症を基礎疾患とし，可逆性の僧帽弁逆流による心原性ショックと肺水腫を繰り返し引き起こした症例を報告する．症例は68歳，男性で，2002年3月より中等度の僧帽弁逆流を伴う閉塞性肥大型心筋症の診断で外来通院した．薬物治療下での流出路圧較差30 mmHg程度であり，症状もなく経過していた．2003年1月，入浴後突然ショック状態となり搬送された．心臓超音波，心臓カテーテル検査では重度の僧帽弁逆流と圧較差50 mmHgが認められた．大動脈バルーンパンピングによる補助の下，プロプラノロール投与を行ったところ，圧較差は10 mmHgまで改善し，軽度の僧帽弁逆流を呈するのみとなり，最終的に僧帽弁置換術を施行し独歩で退院となった．入浴，排便を契機に僧帽弁前尖の収縮期前方運動が増強したことで，左室流出路圧較差の軽度上昇と僧帽弁逆流の急激な増悪を引き起こし，心原性ショックと肺水腫を呈したと考えられた．また治療として，大動脈バルーンパンピングによる補助の下でのβ遮断薬投与が有効であった．

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