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Case Report

A case of hypertrophic obstructive cardiomyopathy with aortic stenosis

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ABSTRACT

A 78-year-old woman complained of experiencing dyspnea (New York Heart Association II) and faintness. Echocardiography revealed she had asymmetric left ventricular hypertrophy, and a dynamic left ventricular outflow tract (LVOT) obstruction due to systolic anterior motion of the mitral valve. It also revealed calcification of the noncoronary cusp and a high-flow velocity in the LVOT (6.3 m/s). The planimetry measurement with transesophageal echocardiography was 0.89 cm^2 (aortic valve area/body surface area: 0.69 cm^2/m^2). Later, she was diagnosed with hypertrophic obstructive cardiomyopathy (HOCM) and aortic stenosis (AS). However, during the catheterization, the transvalvular pressure gradient (PG) was only 25 mmHg. In order to solve this, we performed a percutaneous transluminal septal myocardial ablation. As a result, the PG of the LVOT decreased from 152 mmHg to 25 mmHg.

We first thought that the LVOT obstruction had reduced the flow passing through the aortic valve, and restricted the motion of the aortic valve leaflets. We also considered the possibility that the aortic valve area had been underestimated. The hemodynamic study played an important role in the decision for the treatment plan. The present case was a combination of HOCM and "mild" AS.

<**Learning objective**: We know that we can distinguish between a left ventricular outflow tract obstruction and aortic stenosis using continuous-wave Doppler according to the phase of the peak gradient. However, if both are present, it is uncertain whether we can distinguish between them. It is necessary to measure the subaortic pressure and flow passing through the aortic valve accurately by catheterization in order to know which is the chief pathology.>

Introduction

Hypertrophic cardiomyopathy (HCM) is a primary disorder of the myocardium caused by a missense mutation in the cardiac sarcomere [1,2]. This phenotypic expression occurs in 1 out of every 500 adults in the general population [3,4]. Hypertrophic obstructive cardiomyopathy (HOCM) is characterized by asymmetric septal hypertrophy (ASH), systolic anterior motion (SAM) of the mitral valve, and a left ventricular outflow tract (LVOT) obstruction [4]. These abnormalities reduce the left ventricular (LV) diastolic performance and cause a high LV diastolic pressure, mitral regurgitation (MR), and low cardiac output, which lead to dyspnea, chest pain, and syncope [5,6]. The diagnosis can be confirmed by abnormalities on the echocardiogram, which show not only an abnormal morphology, but also a high jet velocity in the LVOT. We here report a case of HOCM with aortic stenosis (AS). Further, we will discuss the pathology and treatment plan.

Case report

A 78-year-old woman diagnosed with HOCM was admitted for an evaluation of the severity of her AS. Six years previously, she initially experienced chest discomfort. She was diagnosed with HOCM by echocardiography because it revealed ASH, SAM, and a systolic pressure gradient (PG) in the LVOT (64 mmHg). We chose conservative drug therapy as the first-line approach, and she began to take beta-blockers (carvedilol 10 mg). However, after 6 years, the symptoms became worse and she also had dyspnea [New York Heart Association (NYHA) II] and faintness when she was admitted. She did not have a family history of sudden death. A physical examination revealed a midsystolic murmur (Levine III/IV) which significantly increased during the Valsalva maneuver

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and when standing up. Her blood pressure was 118/50 mmHg and heart rate was 78 beats/min. A laboratory test revealed mild anemia (hemoglobin 11 g/dl) and a high N-terminal-pro B-type natriuretic peptide level (4822 pg/ml). The electrocardiogram at rest exhibited normal sinus rhythm. There was ST-segment depression and terminal T-wave negativity in leads II, III, aVF, V5, and V6. The chest X-ray did not reveal any cardiomegaly. Transthoracic echocardiography (TTE) revealed ASH (the septal wall thickness was 17 mm and posterior wall thickness 13 mm). The LV diameters were 37 mm in the end-diastolic phase and 23 mm in the end-systolic phase. The ejection fraction was 60%. The mitral valve exhibited a significant SAM phenomenon, with complete systolic septal contact. The MR was mild. There was a calcification change in the noncoronary cusp of the aortic valve. The aortic valve area (AVA) was unclear due to the calcification change. Continuous-wave Doppler echocardiography revealed that the PG in the LVOT was 159 mmHg at rest (Fig. 1). Magnetic resonance imaging also depicted an obstructive LVOT, ASH, and SAM, but we could not confirm any delayed contrast enhancement of the hypertrophic cardiac muscle. We also performed transesophageal echocardiography (TEE) to measure an accurate AVA. The planimetry measurement of the TEE showed that the AVA was 0.89 cm^2 (AVA/body surface area: 0.69 cm^2/m^2). It also revealed that there was a calcification change in the noncoronary cusp and the leaflet motion was slightly decreased. We could confirm a mosaic flow in the LVOT (Fig. 2). The Holter electrocardiography showed that the total heartbeats were 93,252/day. The minimum heart rate was 56 bpm and maximum heart rate 86 bpm. The Holter electrocardiography did not reveal any ventricular tachycardia.

We performed a cardiac catheterization and hemodynamic study (we used a specific catheter similar to a pig-tail catheter). Firstly, we measured the right atrial pressure (RAP), right ventricular pressure (RVP), pulmonary artery pressure (PAP), and pulmonary capillary wedge (PCWP) pressures and cardiac output (CO) using a Swan-Ganz catheter (RAP: 9 mmHg, RVP: 72/5–11 mmHg, PAP: 70/26(48) mmHg, PCWP: 42 mmHg, CO: 5.17 L/min) The hemodynamic study revealed a high RAP, evidence of pulmonary hypertension, high left atrium pressure (the mean PCWP was 42 mmHg even though the end-diastolic pulmonary artery pressure was 26 mmHg) and adequate CO. We then placed one catheter in the ascending aorta, and another into the LV cavity. We measured the intra-ventricular pressure and ascending aortic pressure at the same time. When we placed the catheter in the LV apex, the peak to peak PG was 132 mmHg. However, when we pulled it back to the subaortic valve, the PG decreased to 25 mmHg (Fig. 3a and b). We thought that the true transvalvular PG was 25 mmHg. Coronary angiograms revealed no arteriosclerotic changes or narrowing of the extramural coronary arteries. Left ventriculography, as well as the echocardiogram, revealed SAM and an LVOT obstruction. It also revealed grade II MR due to the SAM.

As a result, we determined that the AS was not so severe. Through the informed consent process, we decided to perform a percutaneous transluminal septal myocardial ablation (PTSMA).

We performed an elective PTSMA with myocardial contrast echocardiography (MCE). First, we inserted a temporary pacemaker into the right ventricle. Following that, we introduced a balloon catheter into the first septal branch of the left anterior descending coronary artery. After the balloon was inflated, the distribution of the first septal branch was verified by contrast two-dimensional echocardiography after an injection of an echo contrast agent. After confirming which territory of the basal septum contributed to the LVOT obstruction, we infused 1.7 ml of alcohol selectively through an over-the-wire catheter with the balloon inflated. We injected a little morphine hydrochloride because the patient complained of crushing pressure in her precordial chest. We confirmed the complete ablation of the basal septum with the MCE.

Immediately after the PTSMA, we measured each intra-cardiac pressure and the PG between the LV cavity and ascending

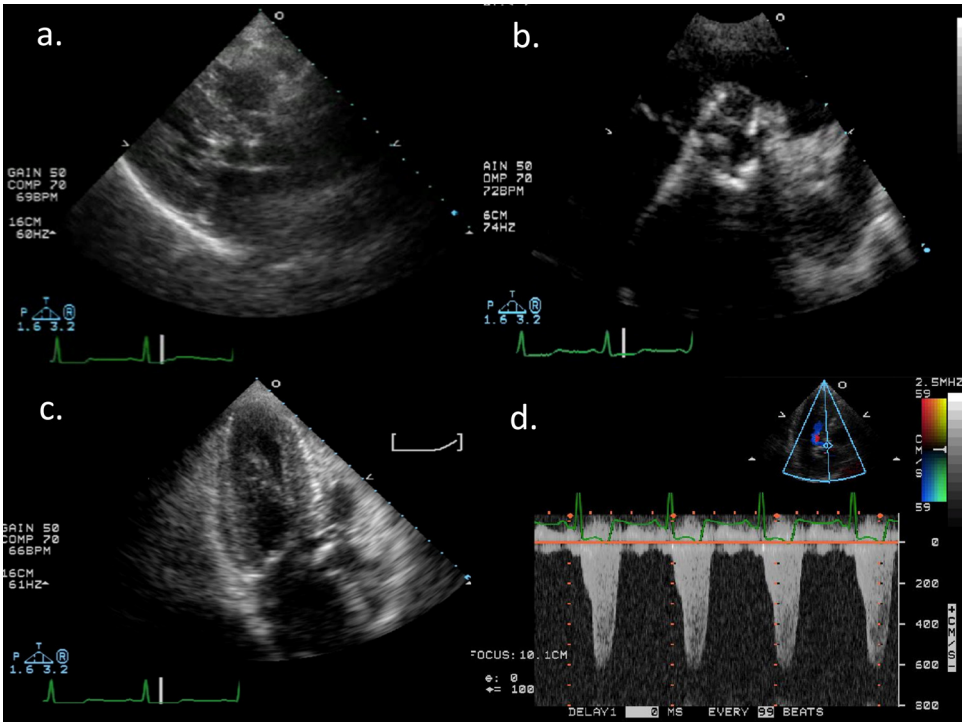

Fig. 1. The transthoracic echocardiograms. (a) The transthoracic echocardiogram in the parasternal long-axis view during the mid-systolic period reveals asymmetric septal hypertrophy, systolic anterior motion (SAM) of the mitral valve, and a left ventricular outflow tract (LVOT) obstruction. (b) The short-axis view of the aortic valve reveals a calcification of the noncoronary cusp. (c) This three-chamber view also shows SAM and an LVOT obstruction. There were calcific changes in the aortic valve. (d) Continuous-wave Doppler echocardiography revealed that the pressure gradient was 159 mmHg (6.3 m/s) at rest.