Mitral stenosis and hypertrophic obstructive cardiomyopathy: An unusual combination

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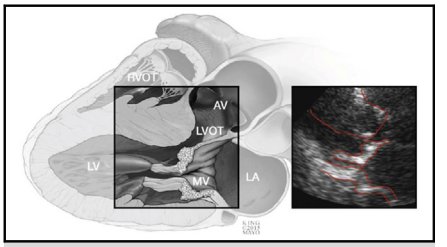
ABSTRACT

Objective: Systolic anterior motion of mitral valve (MV) leaflets is a main pathophysiologic feature of left ventricular outflow tract (LVOT) obstruction in hypertrophic obstructive cardiomyopathy. Thus, restricted leaflet motion that occurs with MV stenosis might be expected to minimize outflow tract obstruction related to systolic anterior motion.

Methods: From January 1993 through February 2015, we performed MV replacement and septal myectomy in 12 patients with mitral stenosis and hypertrophic obstructive cardiomyopathy at Mayo Clinic Hospital in Rochester, Minn. Preoperative data, echocardiographic images, operative records, and postoperative outcomes were reviewed.

Results: Mean (standard deviation) age was 70 (7.6) years. Preoperative mean (standard deviation) maximal LVOT pressure gradient was 75.0 (35.0) mm Hg; MV gradient was 13.7 (2.8) mm Hg. From echocardiographic images, 4 mechanisms of outflow tract obstruction were identified: systolic anterior motion without severe limitation in MV leaflet excursion, severe limitation in MV leaflet mobility with systolic anterior motion at the tip of the MV anterior leaflet, septal encroachment toward the LVOT, and MV displacement toward the LVOT by calcification. Mitral valve replacement and extended septal myectomy relieved outflow gradients in all patients, with no death or serious morbidity.

Conclusions: Patients with mitral stenosis and hypertrophic obstructive cardiomyopathy have multiple LVOT obstruction mechanisms, and MV replacement may not be adequate treatment. We favor septal myectomy and MV replacement in this complex subset of hypertrophic obstructive cardiomyopathy. (J Thorac Cardiovasc Surg 2016;151:1044-8)



Displacement of the MV toward the LVOT by calcification. (Used with permission of Mayo Foundation for Medical Education and Research.)

Central Message

Mitral valve stenosis can co-occur with obstructive hypertrophic cardiomyopathy and is best managed with myectomy and mitral valve replacement.

Perspective

Systolic anterior motion of the mitral valve has a central role in obstructive physiology of hypertrophic cardiomyopathy. However, we have encountered patients who have both obstructive hypertrophic cardiomyopathy and limited mobility of mitral leaflets due to mitral valve stenosis. Multiple mechanisms can lead to obstruction, and optimal treatment is septal myectomy and mitral valve replacement.

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Left ventricular outflow tract (LVOT) obstruction in hypertrophic cardiomyopathy is related to systolic anterior motion of the mitral valve (MV) leaflets in contact with the subaortic septum.[1,2] In patients who have hypertrophic cardiomyopathy and MV stenosis, the valve leaflets usually have limited mobility, which might be expected to minimize systolic anterior motion and dynamic LVOT obstruction. Some studies have reported isolated cases of the combination of MV stenosis and hypertrophic obstructive cardiomyopathy,[3-8] but none have identified or proposed mechanisms of LVOT obstruction.

The aim of the present study was to evaluate the pathophysiologic characteristics and surgical outcome of patients who underwent MV replacement and septal myectomy for mitral stenosis and hypertrophic obstructive cardiomyopathy at our institution.

METHODS

Patients

From January 1993 through February 2015, a total of 12 patients with the preoperative diagnosis of MV stenosis and obstructive hypertrophic cardiomyopathy underwent MV replacement and septal myectomy at Mayo Clinic Hospital in Rochester, Minn. During that interval, a total of 2162 adults (age ≥18 years) had an operation for hypertrophic

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Abbreviations and Acronyms
LVOT = left ventricular outflow tract
MV = mitral valve

cardiomyopathy. The study protocol was approved by the Mayo Clinic Institutional Review Board. Preoperative data, operative records, and postoperative outcomes in the electronic health records and our prospective surgical database were reviewed. Detailed analysis of preoperative transthoracic and intraoperative transesophageal echocardiographic images was performed to characterize the anatomic relationship between the MV and the subaortic septum, to identify possible mechanisms of outflow tract obstruction.

RESULTS

Preoperative Characteristics

Mean (standard deviation) age was 70 (7.6) years; 11 patients were women. Mean (standard deviation) New York Heart Association functional class was 3 (0.6); 10 patients (83%) were in New York Heart Association functional class III or IV. On preoperative echocardiography, the mean (standard deviation) was 75.0 (35.0) mm Hg (range: 29-125 mm Hg) for maximal LVOT pressure gradient, and for MV gradient, 13.7 (2.8) mm Hg (Table 1).

Pathophysiologic Mechanisms of LVOT Obstruction

Four main mechanisms of LVOT obstruction were identified in the patients with mitral stenosis and hypertrophic obstructive cardiomyopathy (Figure 1). Eleven of our 12 patients had combinations of pathophysiologic mechanisms of LVOT obstruction (Table 2). The first mechanism observed was systolic anterior motion of MV leaflets without severe limitation in leaflet motion. In patients with this mechanism, MV leaflet motion was not severely limited, even with significant MV stenosis; thus, the mechanism of LVOT was similar to that in hypertrophic obstructive cardiomyopathy without mitral stenosis.

The second mechanism identified was systolic anterior motion at the tip of the MV leaflet, with severe limitation in MV motion. With this mechanism, only the body of the MV was calcified with limited motion, leaving the tip mobile, and the tip of the MV leaflet moved toward the LVOT, causing systolic anterior motion and LVOT obstruction. The third mechanism was focal septal hypertrophy encroaching on the LVOT. With this mechanism, septal hypertrophy is prominent at the base of the septum, forming a fixed obstruction of the LVOT. With the fourth mechanism, marked calcification and displacement of the anterior leaflet of the MV toward the LVOT resulted in LVOT obstruction. A separate level of obstruction was present in the midventricle in 3 patients.

Operations and Outcomes

All 12 patients underwent MV replacement (6 mechanical valves and 6 bioprostheses) and septal myectomy. Mean (standard deviation) aortic crossclamp time and cardiopulmonary bypass time were 89.8 (30.7) minutes and 107.7 (32.1) minutes, respectively. In 11 patients, LVOT pressure gradients were relieved completely after the surgery; the sole patient with residual gradient had concomitant aortic valve replacement with a 21-mm aortic bioprosthesis in 1999. No operative deaths occurred, and the median length of hospital stay was 8 days (range: 5-27 days; Table 1).

DISCUSSION

The relationship between MV regurgitation and obstructive hypertrophic cardiomyopathy is well known, and experience has shown that in most patients, systolic anterior motion of the MV and related MV regurgitation resolve after successful septal myectomy[9,10] (JH Hong, HV Schaff, RA Nishimura, MD Abel, JA Dearani, Z Li, and SR Ommen, unpublished data, 2015). However, little is known about the mechanisms of outflow tract obstruction in patients who have hypertrophic cardiomyopathy and restricted motion of the MV leaflets.

Systolic anterior motion of the MV leaflets is caused by complex anatomic and physiologic interactions between the left ventricular outflow and the MV apparatus that produce systolic anterior motion of the MV.[11,12] However, none of the previous explanations of systolic anterior motion has accounted for the possible mechanism of LVOT obstruction in patients who have mitral stenosis and hypertrophic cardiomyopathy.

In the present review, we found that mechanisms of the LVOT obstruction in patients with mitral stenosis and hypertrophic cardiomyopathy were multiple and complex. In our case patients, the various pathophysiologic mechanisms included: systolic anterior motion with or without severe limitation of the MV leaflets; septal encroachment toward the LVOT; and distortion of the base of the MV toward the LVOT with calcification. Thus, although systolic anterior motion producing outflow tract obstruction may occur in patients who have restrictive MV disease, the latter 2 mechanisms explain how LVOT obstruction can occur without systolic anterior motion.

In addition to these mechanisms, a separate level of obstruction at midventricle may cause symptoms.[13] Categorizing 4 mechanisms of LVOT obstruction on the basis of echocardiography data of a limited number of patients may be an oversimplification, and more mechanisms of LVOT obstruction could be present in other patient groups. But, even if other mechanisms are present that might not be found in our patient groups, we assume that they are few and are concurrent with the 4 mechanisms we found.

Concomitant MV surgery is recommended at the time of myectomy for patients who are known to have significant MV stenosis,[14] because relief of outflow tract obstruction alone would not be expected to correct symptoms related