CASE REPORT

Hypertrophic obstructive cardiomyopathy complicated by ruptured chordae tendineae to the posterior mitral leaflet and severe congestive heart failure

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A 63-year-old woman with hypertrophic obstructive cardiomyopathy developed rapidly progressive fatigue, shortness of breath and congestive heart failure. A transesophageal echocardiogram demonstrated ruptured chordae to the posterior mitral valve leaflet with severe mitral regurgitation. Mitral valve replacement eliminated the outflow gradient. Acute or subacute hemodynamic deterioration in a patient with hypertrophic obstructive cardiomyopathy should lead to a search for associated lesions.

Key Words: *Hypertrophic cardiomyopathy; Mitral valve prolapse; Ruptured chordae tendineae*

Une myocardiopathie obstructive hypertrophique compliquée par une rupture des cordons tendineux du feuillet valvulaire mitral postérieur et une insuffisance cardiaque congestive grave

Une femme de 63 ans ayant une myocardiopathie obstructive hypertrophique s'est mise à présenter une lassitude, un essoufflement et une insuffisance cardiaque congestive à évolution rapide. Un échocardiogramme transoesophagien a démontré une rupture des cordons tendineux du feuillet valvulaire mitral postérieur accompagnée d'une grave régurgitation mitrale. Le remplacement de la valvule mitrale a éliminé le gradient d'éjection. La détérioration hémodynamique aiguë ou subaiguë des patients ayant une myocardiopathie obstructive hypertrophique devrait susciter la recherche de lésions connexes.

Hypertrophic obstructive cardiomyopathy (HOCM) is frequently associated with mitral valve regurgitation (MR). In most instances, the latter is related to functional left ventricular outflow tract (LVOT) obstruction rather than intrinsic disorders of the mitral valve. The latter, however, need to be considered in presentations that are inconsistent with the rather stable natural history of HOCM. Mitral valve prolapse (MVP), with or without chordal rupture, can complicate HOCM by adding to the burden of MR and thereby aggravating and/or precipitating congestive heart failure.

CASE PRESENTATION

A 63-year-old woman presented to the emergency department because of progressive, painless shortness of breath. She had suffered from fatigue for the past two weeks, shortness of breath on a home treadmill for three days, and dyspnea at rest for one day. Previous dyspnea, chest discomfort, ischemic heart disease, palpitations, presyncope or syncope were denied. She was hypertensive and had had 'asthma' for approximately 10 years. Three years before admission, a transthoracic echocardiogram revealed symmetrical left ventricular hypertrophy with hyperdynamic left ventricular systolic function and systolic anterior motion (SAM) of the mitral valve. Eccentric MR of uncertain severity was presumed to be secondary to HOCM.

The patient was taking 2.5 mg of ramipril, 25 mg of hydrochlorothiazide and 240 mg of diltiazem daily.

She was short of breath at rest. Her blood pressure was 162/79 mmHg and her pulse was 123 beats/min and regular. Her respiratory rate was 30 breaths/min. A grade 4/6 systolic crescendo-decrescendo murmur was noted. The jugular venous pressure was normal. The chest was clear. There was no edema.

The electrocardiogram revealed sinus tachycardia at 114 beats/min, borderline left atrial abnormality and voltage criteria for left ventricular hypertrophy. The chest x-ray revealed congestive heart failure with bilateral pleural effusions. The laboratory data were unremarkable.

The patient was admitted and treated with oxygen and diuretics. Her dyspnea and tachycardia persisted. Low-dose metoprolol was added but shortness of breath continued. Her clinical response was believed to be inconsistent with hypertrophic cardiomyopathy (HCM) alone. An urgent transthoracic echocardiogram revealed MR with probable prolapse of the posterior leaflet of the mitral valve. There was marked SAM of the mitral valve with a peak instantaneous LVOT gradient of 148 mmHg. A transesophageal echocardiogram examination revealed asymmetrical septal hypertrophy (septal thickness 17 mm), SAM of the mitral valve, ruptured chordae to the posterior leaflet of the mitral valve and severe MR. There was no evidence of bacterial endocarditis.

The coronary angiogram was normal. Surgery revealed ruptured chordae to the P2 segment of the posterior leaflet, with mild thickening of the anterior leaflet. The mitral valve was replaced using a St Jude's prosthesis (#29; St Jude Medical Inc, USA). A septal myomectomy was not performed. The intraoperative LVOT gradient fell from 109 mmHg to 6 mmHg. Gross pathological examination revealed 'myxoid degeneration'. The postoperative course was uneventful.

DISCUSSION

MR is an intrinsic component of the functional LVOT obstruction seen in patients with HOCM. Such MR, however, is usually associated with an intrinsically normal mitral valve and the severity of the MR is directly related to the LVOT gradient (1). The association of MVP, with or without chordal rupture, has been reported to complicate HCM (1-5).

A report from the National Institutes of Health (United States) (2) described prospective M-mode and two-dimensional echocardiographic findings in 528 patients with HCM. Sixteen patients, or 3% of the study population, met their criteria for MVP. Ten of these sixteen patients had small or absent LVOT gradients. Three patients had ruptured chordae. The authors suggested that the association of MVP and HCM "may reflect the coexistence of two separate but primary disease entities."

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Yu et al (3), from the Toronto General Hospital (Toronto, Ontario), reviewed their operative experience with HOCM and reported that 11 of 104 patients (11%) had independent mitral valve disease. Of these, five had more than moderate MVP and one patient had ruptured chordae.

The transesophageal echocardiographic operative findings in HOCM patients were reported by Ommen et al (4) from the Mayo Clinic (Rochester, Minnesota, USA). In a series of 256 surgeries, seven patients had MVP, four had flail leaflets and three had severe MR.

Zhu et al (5) summarized the Mayo Clinic experience in five patients with HOCM and ruptured chordae tendineae. New or progressive congestive symptoms led to the discovery of ruptured chordae tendineae with severe MR. Mitral valve repair and septal myectomy were successful in all instances, and were associated with an excellent clinical response.

Sporadic reports of the association of these two disorders continue to appear and emphasize the need to consider other cardiac disorders in the differential diagnosis of patients with HCM whose congestive symptoms become slowly and/or acutely progressive. In such individuals, echocardiographic studies should be directed toward the identification of intrinsic abnormalities of the mitral valve producing degrees of MR that are inconsistent with the presence of HCM alone. Surgical

treatment may then result in resolution of the MR and elimination of the LVOT gradient.

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