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Acute Mitral Regurgitation Due to Ruptured Chordae Tendineae in a Patient With Hypertrophic Obstructive Cardiomyopathy: A Case Report

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Abstract

A 63-year-old woman had been followed up for hypertrophic obstructive cardiomyopathy with 85 mmHg of left ventricular outflow tract pressure gradient over 7 years. She was hospitalized because of acute dyspnea and syncope. On admission, echocardiography revealed severe mitral regurgitation with ruptured chordae tendineae at the medial scallop of the posterior mitral leaflet. Mitral valve replacement was successfully performed and her symptoms improved to 28 mmHg of left ventricular outflow tract pressure gradient. In patients with hypertrophic obstructive cardiomyopathy, elevated left ventricular systolic pressure and systolic anterior motion of the mitral leaflets may lead to mucoid degeneration in the chordae tendineae. Rupture of the mitral chordae tendineae should be considered in the differential diagnosis of acutely deteriorated mitral regurgitation in patients with hypertrophic obstructive cardiomyopathy, because this is a rare but critical complication.

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Key Words

■**Cardiomyopathies, hypertrophic** ■**Mitral regurgitation (ruptured chordae tendineae)**
■**Echocardiography, transesophageal**

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is a genetic disease, characterized by hypertrophy of the left ventricle, with markedly variable clinical manifestations and morphological and hemodynamic abnormalities. The clinical course is relatively benign for the majority of patients, although disease-related complications can develop. However, the site and extent of cardiac hypertrophy and systolic anterior motion of the mitral valve leaflets which contact with the ventricular septum result in obstruction to left ventricular outflow tract (LVOT) in a subset of patients. The presence of LVOT stenosis is an independent predictor of poor prognosis in patients with HCM[1]. Ruptured chordae tendineae represents a rare cause of mitral regurgitation in hypertrophic obstructive cardiomyopathy (HOCM)[2–6].

We describe a case of acute deterioration of mitral regurgitation due to ruptured chordae tendineae identified preoperatively by trans-

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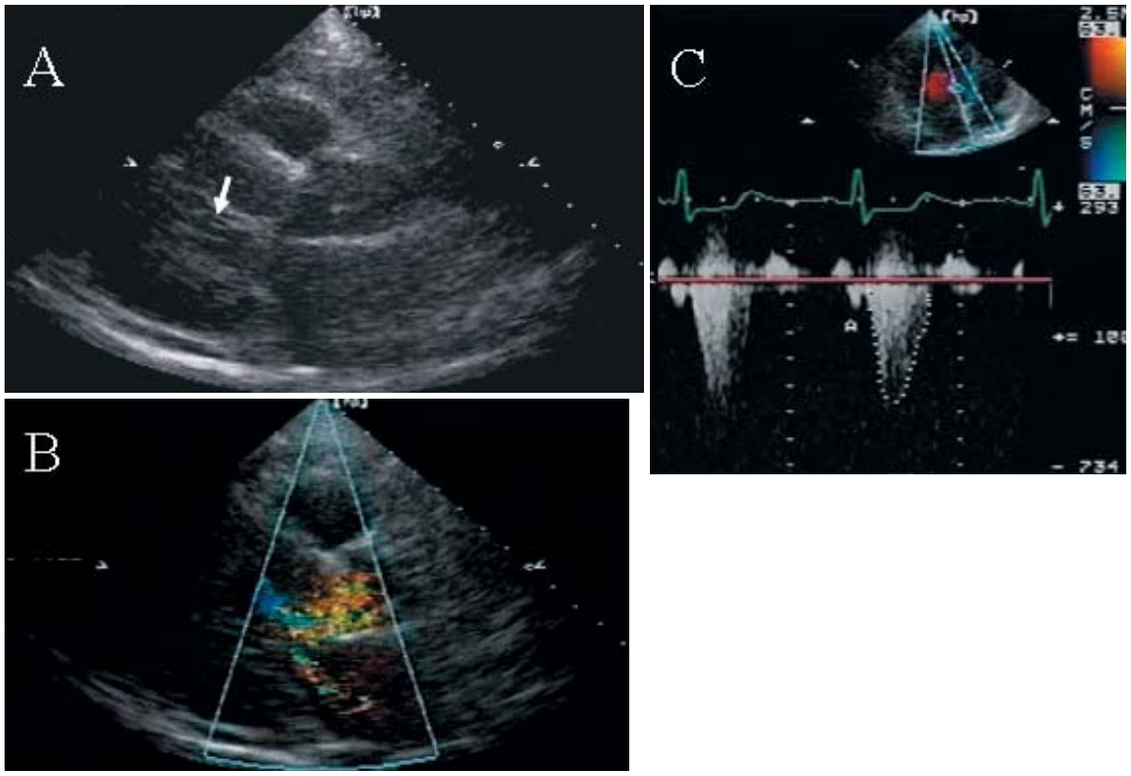

Fig. 1 Two-dimensional echocardiograms taken in 1996, showing the apical three chambers
A: Systolic anterior motion of the mitral valve and asymmetric septal hypertrophy is apparent.
B: Mitral regurgitation jet on color Doppler imaging.
C: Left ventricular outflow tract pressure gradient was 85mmHg.

esophageal echocardiography.

CASE REPORT

A 63-year-old woman with HOCM was referred to our hospital with acute severe dyspnea and syncope. She had noted mild dyspnea on exertion [New York Heart Association (NYHA) class II] at age 56 years. The diagnosis was HOCM based on the results of Doppler echocardiography, showing asymmetric septal hypertrophy, systolic anterior motion of mitral valve and a pressure gradient of 103 mmHg at the LVOT. Treatment with disopyramide 150 mg/day, atenolol 50 mg/day and verapamil 120 mg/day resulted in improvement of symptoms. Two-dimensional echocardiography showed the left ventricular end-diastolic dimension was 44 mm and the left ventricular end-systolic dimension was 26 mm, and the pressure gradient at LVOT had decreased to 85 mmHg (**Fig. 1**). Moderate mitral regurgitation blowing back to the posterior of the left atrium with systolic anterior motion of the mitral valve persisted. Sixteen days prior to admission, the patient suffered acute onset of severe dyspnea and paliptation when she took a short walk in the morning. She had syncope for a few seconds just after standing up after a few minutes of rest.

On admission, blood pressure was 136/68 mmHg with a regular heart rate of 92 beats/min. Pan-systolic murmur grade III/IV and the third heart sound were audible in the apex. Crackle was audible in the lower lungs. No edema was found in the legs. Electrocardiography revealed sinus rhythm, complete right bundle branch block and right ventricular overload. Chest radiography indicated lung congestion and cardiomegaly. Two-dimensional echocardiography (**Fig. 2−A**) showed severe prolapse of the posterior mitral valve leaflet (medial scallop). End-diastolic and end-systolic dimensions of the left ventricle were 49 and 20 mm, respectively. Interventricular septum thickness and posterior wall thickness at end diastolic phase were 16 and 10 mm, respectively.

Doppler echocardiography revealed severe mital

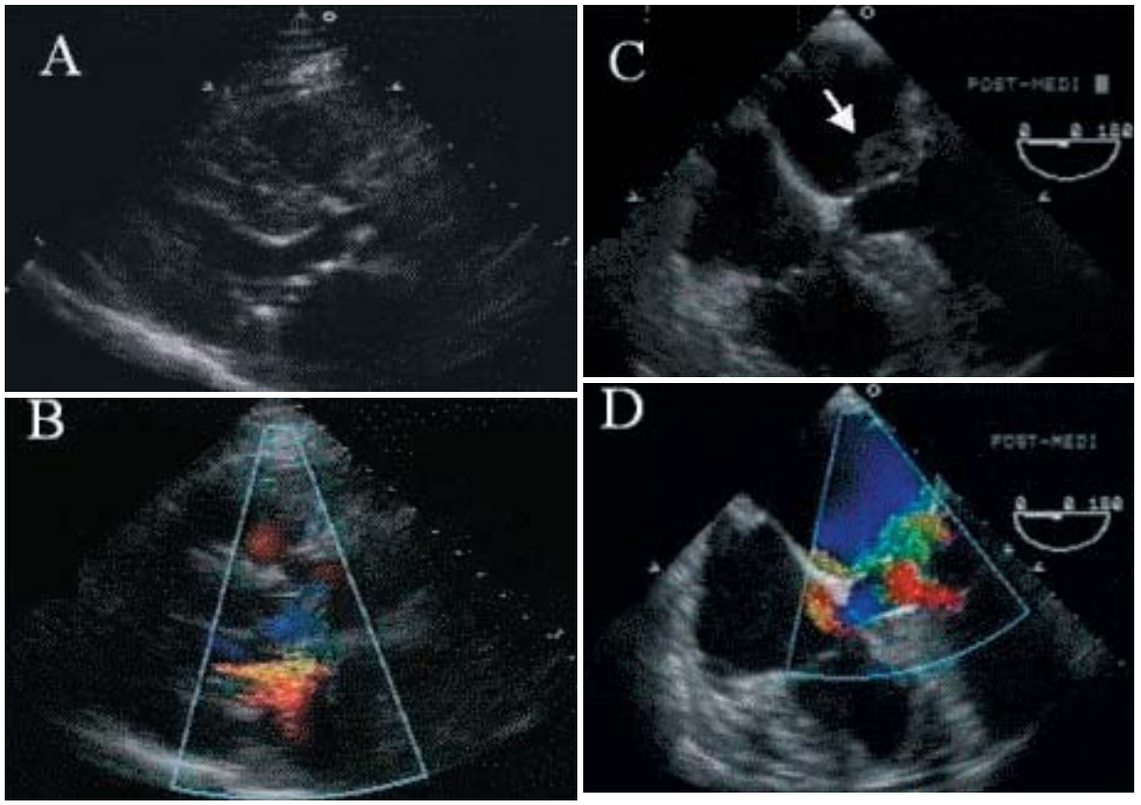

Fig. 2 Echocardiograms taken on admission in 2003
A: Two-dimensional imaging shows left ventricular outflow tract narrowing has disappeared due to left ventricular overload. Worsening of mitral regurgitation was counterbalanced by relief of the left ventricular outflow obstruction.
B: Color Doppler imaging shows severe mitral regurgitation.
C: Transesophageal imaging shows ruptured chordae tendineae at the posterior leaflet of the mitral valve.
D: Color Doppler transesophageal imaging shows mitral regurgitation jet.

regurgitation blowing back to the left auricular appendage of the left atrium. LVOT was wider, possibly due to left vertricular volume overload resulting from mitral regurgitation(**Fig. 2−B**). Systolic anterior motion had disappeared and the LVOT pressure gradient was less than 30 mmHg. Severe tricuspid regurgitation was present with a systolic pressure gradient of 60 mmHg. Transesophageal echocardiography confirmed ruptured chordae tendineae in the posteromedial scallop of the posterior leaflet without any sign of vegetation(**Figs. 2−C, D**). Right heart catheterization revealed the cardiac index was as low as 2.1 *l*/min/m^2 and pulmonary capillary wedge pressure as high as 37 mmHg. Treatment with furosemide improved the hemodynamic condition of the patient. Twelve days after admission, cardiac index and pulmonary capillary wedge pressure improved to 3.2 *l*/min/m^2 and 18 mmHg. Coronary arteriography showed no stenosis. Left ventriculography showed severe mitral regurgitation of grade Ⅳ according to the Sellers ’classification.

Mitral valve replacement and tricuspid annuloplasty were performed on the 27th day of hospitalization. Intraoperatively, a flail segment of the posterior leaflet was noted secondary to ruptured commissural chordae tendineae at the medial portion of the posterior leaflet. No calcification or thickening of the mitral valve was apparent. Histological examination of the lesion revealed mucoid degeneration without inflammation. Doppler echocardiography on postoperative day 16 showed no residual mitral regurgitation, with left ventricular dimensions of 36 mm at enddiastole and 27 mm at endsystole. The reduction in left ventricular dimensions induced mid-ventricular stenosis with a pressure gradient of 28 mmHg on Doppler echocardiography. Pharmacotherapy with β-blockers was resumed. The postoperative course was uneventful and the patient was discharged 4 weeks postopera-

Table 1 Case reports of ruptured chordae tendineae with hypertrophic obstructive cardiomyopathy

Report	Patient (age, sex)	RCT	PA pressure (mmHg)	LVOT (mmHg)		MVR	Myotomy myectomy
				Pre-ope	Post-ope		
Kioka (1997)	70 F	+ (PML)	80	125	0	+	+
Shibata (1996)	61 M	+ (PML, AML)			0	+	−
Zhu (1992)	68 F	+ (PML)	70	100	19	+	+
	77 F	+ (PML)	90	158	15	+	+
	39 M	+ (PML)		64	0	+	+
	76 F	+ (PML)	44	20	0	+	+
	26 M	+ (AML)				−	+

RCT = ruptured chordae tendineae; PA = pulmonary artery; LVOT = left ventricular outflow tract; MVR = mitral valve replacement; F = female; M = male; PML = posterior mitral leaflet; AML = anterior mitral leaflet.

tively.

DISCUSSION

Regardless of the presence of symptoms, significant outflow tract obstruction represents an independent predictor of poor prognosis in patients with HCM. A multicenter study of 1,101 patients with HCM revealed that 25% had outflow tract obstruction at rest with a peak gradient >30 mmHg[1]. At a mean follow-up of 6 years, 12% had died, and another 20% had undergone progression to NYHA class III or IV heart failure. After adjusting for age, sex, heart failure at entry, presence of atrial fibrillation, and left ventricular wall thickness ≥ 30 mm, patients with obstruction displayed a higher rate of HCM-related mortality.

Mitral regurgitation in most patients with HOCM is caused by systolic anterior motion of the anterior and posterior mitral leaflets. The present patient with HOCM displayed acute severe mitral regurgitation due to ruptured chordae tendineae. Preoperative identification of ruptured chordae tendineae as the cause of mitral regurgitation was established using transesophageal echocardiography. The patient underwent successful mitral valve replacement for relief of LVOT obstruction and mitral valve competence was restored by valve replacement rather than by prosthetic valve repair.

The clinical courses of the reported cases illustrate important management considerations in addition to the utility of transesophageal echocardiography for diagnosis. Chordal rupture should be considered in the differential diagnosis of mitral regurgitation in patients with HOCM, particularly in those with acute hemodynamic deterioration. Prominent septal hypertrophy and possible abnormal location of the mitral valve reduce LVOT. This mechanical impedance to outflow occurs in the presence of high velocity ejection in which a variable proportion of the forward blood flow may be ejected early in systole. Systolic anterior motion is probably attributable to a drag effect and/or possibly a Venturi effect, and is responsible not only for subaortic obstruction but also the concomitant mitral regurgitation due to incomplete leaflet apposition, which is typically directed posteriorly into the left atrium[7].

In the present case, increases in preload by acute mitral regurgitation abolished the pressure gradient at LVOT, although septal thickness was present. Previous reports of the ruptured chordae tendineae associated with HOCM (**Table 1**) showed reduced pressure gradients at LVOT with improvement in symptoms after mitral valve replacement or mitral valvuloplasty with septal myectomy in all 10 patients[2–6]. Histological examination of the mitral chordae lesion revealed mucoid degeneration without inflammation. Possible explanations for the mechanism underlying chordae tendon rupture include elevated left ventricular systolic pressure and mechanical movement of the chordae tendon anteriorly. The residual portion of the posterior leaflet may be pulled anteriorly toward the ventricular septum by the high-velocity jet of blood ejected through the anatomically narrow LVOT[8]. This may lead to mucoid degeneration in the chordae tendineae of the mitral valve[9]. Rupture of the chordae tendineae occurs more frequently at the posterior mitral leaflet than the anterior leaflet, as the chordae tendineae of the posterior leaflet are thinner than those of the anterior leaflet[2,10]. In addition, HOCM displays typical features including abnormal location of the mitral valve. Some patients with HOCM had a marked segmental elongation that

was confined to the middle scallop of the posterior mitral leaflet[8]. The free caudal margins of such an abnormal portion of the posterior leaflet are probably more affected by stress from the elevated left ventricular pressure. Chordae tendineae of the posterior leaflet may thus rupture more easily than those of the anterior leaflet in HOCM.

Mitral valve replacement produced a substantial and consistent reduction in LVOT pressure gradient. Left ventricular end-diastolic pressure was predicted to fall after mitral valve replacement because of the decrease in tricuspid regurgitation gradient. Although myectomy reportedly results in generally satisfactory outcomes with regard to symptoms, myectomy is associated with higher frequencies of complications, such as arrhythmias and the rare but critical complication of ventricular septal perforation[3]. As a result, the American College of Cardiology and the European Society of Cardiology suggest that myotomy-myectomy should be employed in patients with HOCM if the septal thickness is over 18 mm and that mitral valve replacement should be employed if it is under 18 mm[7,11]. Mitral valve replacement or repair has been employed in selected patients judged to have severe mitral regurgitation due to intrinsic abnormalities of the valve apparatus（such as myxomatous mitral valve）.

The present case with ruptured chordae tendinae of the posterior mitral leaflet was considered to be of sufficient severity to preclude repair. We successfully utilized a simpler procedure, using mitral valve replacement without myectomy or myotomy because of 17 mm of left ventricular septal wall thickness. The subjective symptoms were improved along with optimal improvement of the LVOT obstruction, and the patient is now able to maintain an active social life.

Rupture of the mitral chordae tendineae should be considered in the differential diagnosis of acutely deteriorated mitral regurgitation in patients with HOCM, because it is a rare but critical complication.

要　　約

閉塞性肥大型心筋症の経過中に腱索断裂による急性僧帽弁閉鎖不全症をきたした1例

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症例は63歳，女性で，7年前に心エコー図法で閉塞性肥大型心筋症と診断された．内服薬を開始し，左室流出路圧較差は85 mmHgで，胸部症状もなく近医で経過観察されていたが，突然，呼吸困難が出現し，意識消失したため入院となった．入院時の心エコー図検査で僧帽弁後尖medial scallopの腱索断裂に伴う重度の僧帽弁逆流が認められた．僧帽弁置換術により症状は改善し，左室流出路圧較差は28 mmHgであった．左室流出路圧較差を伴う肥大型心筋症では，僧帽弁の収縮期前方運動と高い左室内圧のため僧帽弁ならびに腱索に退行性変性を生じることが報告されており，本症例は感染性心内膜炎の合併はなく，腱索の退行性変性に起因することが推測された．閉塞性肥大型心筋症における僧帽弁後尖の断裂はまれであるが，難治性の心不全をきたすため，早期の診断が肝要である．

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