

CHORDAL RUPTURE RESOLVES HEART FAILURE AND LEFT VENTRICULAR OUTFLOW OBSTRUCTION

Complete Resolution of Left Ventricular Outflow Tract Obstruction After Spontaneous Mitral Valve Chordal Rupture in a Patient With Hypertrophic Cardiomyopathy



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INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is characterized by left ventricular hypertrophy with variable clinical manifestations such as left ventricular outflow tract (LVOT) obstruction and mitral valve regurgitation (MR). The presence of LVOT obstruction is related to poor clinical outcome in HCM.¹ In hypertrophic obstructive cardiomyopathy (HOCM), rupture of mitral chordae tendineae is a rare but severe complication that causes acute MR resulting in acute exacerbation of clinical symptoms. Here we describe a case of HOCM in which symptoms improved after spontaneous mitral valve chordal rupture with the resolution of LVOT obstruction.

CASE PRESENTATION

A 78-year-old woman presented to the hospital with shortness of breath of New York Heart Association (NYHA) class II and fatigue. Four years before, she was diagnosed with HOCM by echocardiography, which revealed a peak pressure gradient of 40 mm Hg at the LVOT, and she started to take 200 mg of cibenzoline daily. However, her symptoms gradually worsened to NYHA class III. She was referred to our hospital for further evaluation and treatment. She had no history of hypertension and was prescribed statin for dyslipidemia. She had a family history of HCM (brother and elder sister). Chest auscultation revealed a systolic murmur (Levine IV/VI) at the left upper sternal border. Laboratory tests showed elevation of plasma N-terminal-pro B-type natriuretic peptide (NT-pro BNP) level (2,752 pg/mL), normal blood cell counts, and normal renal and hepatic function. Electrocardiogram demonstrated normal sinus rhythm with narrow QRS width and increased QRS voltage in precordial leads, suggesting left ventricular hypertrophy (Figure 1A). Cardiac magnetic resonance imaging (MRI) revealed systolic anterior motion (SAM) of the mitral leaflet on cine image (Figure 1B, Video 1) and regions of late

gadolinium enhancement in the lateral and anterolateral wall of the left ventricle (Figure 1C).

Although she began to take 2.5 mg of bisoprolol daily, her symptoms did not improve during 2 months of treatment. Transthoracic echocardiography revealed asymmetric septal hypertrophy with normal left ventricular ejection fraction (69%), SAM of the mitral leaflet, severe dynamic LVOT obstruction (peak systolic pressure gradient = 102 mm Hg), and severe MR due to SAM (Figure 2A–C, Videos 2 and 3). NT-pro BNP remained high (2,249 pg/mL) even after introduction of bisoprolol. Surgical or catheter septal reduction therapy was considered for heart failure (HF) refractory to medical therapy, and she was scheduled to be admitted to our hospital for presurgical evaluation. However, her condition improved markedly before the admission. Even though she had not taken any additional treatment, her symptoms improved to NYHA class I. Repeated transthoracic echocardiography showed severe MR, which was the same grade as the previous time. However, it revealed new mitral valve prolapse with complete resolution of SAM, while peak LVOT pressure gradient decreased from 102 to 15 mm Hg (Figure 2D–F, Videos 4 and 5). For further evaluation of this new mitral valve prolapse, transesophageal echocardiography was performed, and it revealed flail motion of the medial scallop of posterior mitral leaflet (P3) and ruptured chordae tendineae (Figure 3A–C, Videos 6–8). Four-dimensional CT also showed mitral valve prolapse and ruptured chordae tendineae but no subvalvular apparatus abnormalities (Figure 3D, Video 9). Coronary angiography was performed and proved to be normal. Her HF symptoms improved dramatically, and the NT-pro BNP level also decreased to 310 pg/mL. Her remaining severe MR has been medically treated because of lack of symptoms and preserved LV function. Mitral valve repair for HOCM has a high incidence of relapse of SAM and LVOT obstruction. Her NT-pro BNP level has remained low, and she has been clinically stable for 4 years since this event.

DISCUSSION

Chordal rupture is an infrequent complication in HOCM patients.^{2,3} A literature search found 14 cases of HOCM with ruptured chordae tendineae (Table 1).^{2,4–12} In the present case, ruptured chordae tendineae were confirmed by transesophageal echocardiography. We were not able to identify any specific causes of chordal rupture, such as myocardial infarction, infective endocarditis, or trauma. Chordae tendineae of the posterior mitral leaflet were involved in the present case, which were also reported to be primarily involved in HOCM and HCM cases.^{2,3} Chordal rupture in HOCM has been reported to cause two major hemodynamic changes. One is the negative effect of acute MR,

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VIDEO HIGHLIGHTS

Video 1: Cardiac cine MRI image before chordal rupture. SAM of mitral leaflet and acceleration jet are observed.

Video 2: Parasternal long-axis view of transthoracic echocardiography showing SAM of mitral leaflet.

Video 3: Color Doppler of parasternal long-axis view showing an LVOT acceleration jet.

Video 4: Parasternal long-axis view of transthoracic echocardiography after mitral chordal rupture. SAM is not observed.

Video 5: Color Doppler of parasternal long-axis view.

Video 6: Transesophageal echocardiography midesophageal long-axis view showing posterior mitral leaflet prolapse.

Video 7: Color Doppler of midesophageal long-axis view showing the severe MR.

Video 8: Three-dimensional full-volume image of mitral valve acquired from 6 beats by transesophageal echocardiography. Medial scallop of posterior leaflet prolapse and ruptured chordae tendineae are observed.

Video 9: Four-dimensional computed tomography movie of heart after chordal rupture. No LVOT obstruction is observed.

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and the other is the positive effect of alleviation of LVOT obstruction.^{4,7-11} Most of the previously reported cases had acute deterioration of HF due to acute MR, requiring emergent or urgent surgical treatment. However, one case was reported to be clinically stable, and left ventricular wall thickness decreased after chordal rupture.⁸ In that case, it was suggested that the negative effect of acute MR was counterbalanced by amelioration of LVOT obstruction. Two cases have reported that mitral chordal rupture caused acute deterioration in HF due to acute MR, despite the resolution of LVOT obstruction after chordal rupture (Table 1).^{9,11} In the present case, severe MR due to SAM was already present before the chordal rupture. We consider that her HF symptoms improved due to resolution of LVOT obstruction

alone, while the severity of MR did not change. In other words, the pathophysiology of severe MR was changed from SAM to prolapse by spontaneous chordal rupture. The findings of previous cases and of the present case indicate that clinical manifestations of chordal rupture in HOCM vary depending on the degree of alleviation of LVOT obstruction and the degree of emerging acute MR. Chordal rupture in HCM is infrequent, but a previous study reported that chordal rupture in HCM occurred more often in aged patients with obstructive disease and was more frequently associated with posterior mitral leaflet than with anterior leaflet.³ In the present case, our patient was also elderly with markedly elevated LVOT gradient (peak LVOT pressure gradient of 102 mm Hg), which is consistent with the prior report. Our literature search found that 12 cases out of previously reported 14 cases involved posterior mitral leaflet (Table 1). Mitral leaflet elongation, abnormal structure of papillary muscle, and chordae tendineae are often seen in HOCM patients.¹³ Four-dimensional computed tomography is useful for depicting the distribution of myocardial hypertrophy and the position or size of the papillary muscle^{14,15} and is used to evaluate the precise structure in left ventricular myocardium and to detect the abnormalities in mitral subvalvular apparatus, including hypertrophied or anteriorly displaced papillary muscle or apical-basal muscle bundles.¹⁶ These abnormalities in the mitral valve apparatus displace the mitral valve coaptation point anteriorly closer to the septum. The thickened septum makes the flow drag, which pushes the mitral valve, more anteriorly, resulting in SAM and LVOT obstruction.¹⁷ In the present case, we speculate that rupture of chordae tendineae caused a lack of support of the anterior mitral leaflet, shifting the coaptation more posteriorly with resolution of SAM and LVOT obstruction.

A recent study suggests that cutting the secondary mitral valve chordae tendineae in addition to septal myectomy improves LVOT pressure gradient compared with myectomy alone in HOCM patients with mild septal hypertrophy.¹⁸ The reason is that the mitral valve coaptation point contributes mainly to LVOT obstruction in such patients. The present case, in which rupture of chordae tendineae improved LVOT obstruction, supports the hypothesis that the mitral valve plays a principal role in the mechanism of LVOT obstruction and SAM. It is possible that intervention with the mitral valve alone may be a treatment option for HOCM.

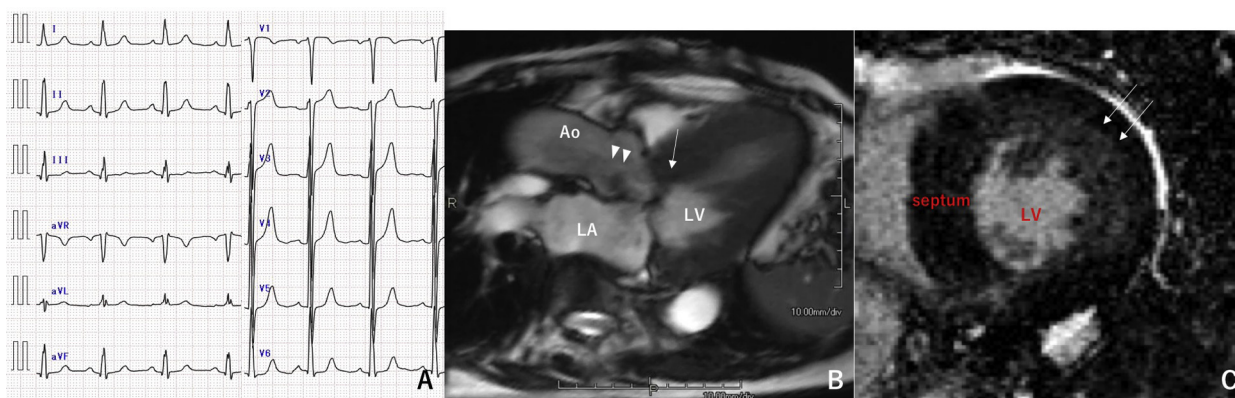


Figure 1 Electrocardiogram and MRI at first visit. **(A)** Electrocardiogram at presentation. Sinus rhythm of 77 bpm with high voltage in precordial lead, suggesting left ventricular hypertrophy. **(B)** Cardiac cine MRI. SAM of mitral leaflet (arrow) and acceleration jet (arrowheads). **(C)** MRI short-axis view of left ventricle. Late gadolinium enhancement in the anterolateral wall (arrows). Ao, Aorta; LA, left atrium; LV, left ventricle.

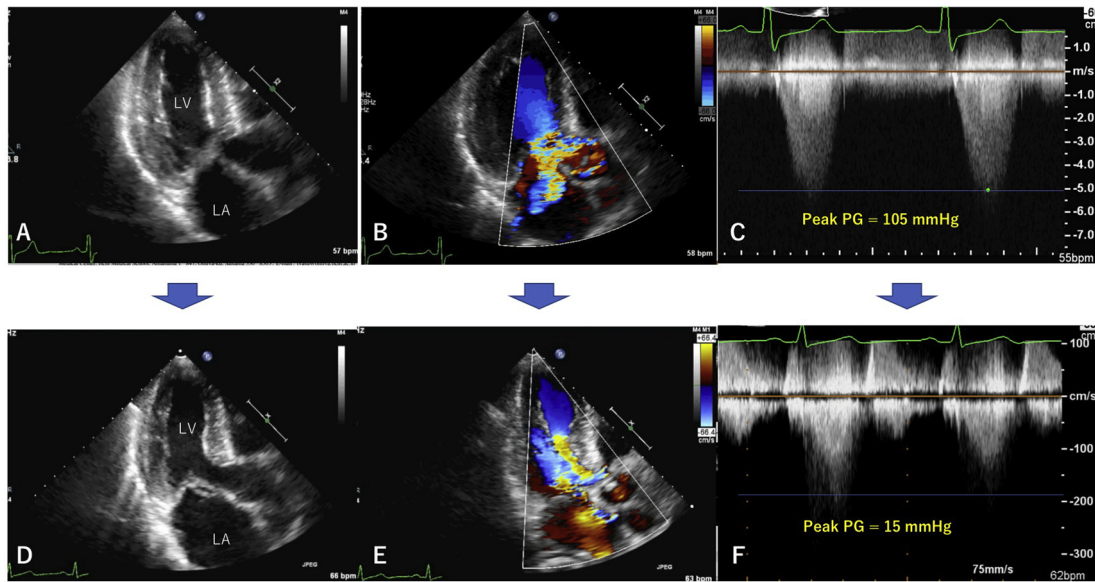


Figure 2 Transthoracic echocardiography before (A, B, C) and after (D, E, F) mitral valve chordal rupture. (A) Apical three-chamber view shows SAM of mitral valve with mitral-septal contact. (B) Color Doppler image shows severe MR and acceleration jet in LVOT. (C) LVOT peak pressure gradient is 105 mm Hg. (D) Apical three-chamber view. SAM is not observed. (E) Color Doppler image. Although severe MR remains, left ventricular outflow acceleration jet is not observed. (F) LVOT peak pressure gradient is 15 mm Hg. LA, Left atrium; LV, left ventricle.

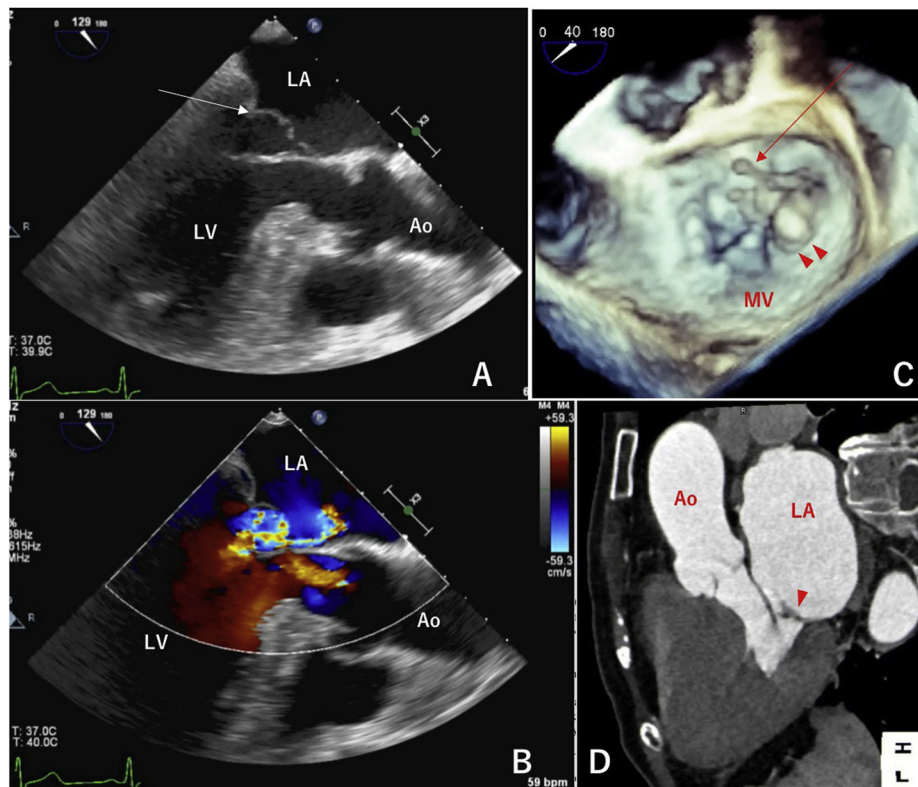


Figure 3 Transesophageal echocardiography and four-dimensional computed tomography (CT) after spontaneous mitral chordal rupture. (A) Midesophageal long-axis view shows flail motion of posterior mitral leaflet (arrow) in systolic phase. LVOT obstruction is not observed. (B) Color Doppler image shows MR. (C) Three-dimensional image of mitral valve shows medial mitral valve leaflet prolapse (arrowheads) and ruptured chordae tendineae (arrow). (D) Four-dimensional cardiac CT images (systolic phase). Posterior mitral leaflet prolapses into left atrium (arrowhead). LVOT obstruction is not observed. Ao, Aorta; AV, aortic valve; LA, left atrium; LV, left ventricle; MV, mitral valve.

Table 1 Summary of reported cases of HOCM with concomitant ruptured chordae tendineae of the mitral valve

Reference, first author	Age (years), sex	Prolapsed leaflet	Clinical manifestations	LVOT peak PG, mm Hg	LVOT obstruction after chordal rupture	Treatment
Zhu (1992) ²	68, F	P3	ADHF	100	Yes	MVr + Morrow
Zhu (1992) ²	77, F	P3	ADHF	158	Yes	MVr + Morrow + CABG
Zhu (1992) ²	39, M	P2	ADHF	64	Yes	MVr + Morrow
Zhu (1992) ²	76, F	PML	ADHF	0	No	MVr + Morrow
Zhu (1992) ²	26, M	A2	ADHF	N/A	N/A	Morrow
Shibata (1996) ⁴	61, M	PML + AML	ADHF	N/A	N/A	MVR
Kioka (1997) ⁵	70, F	PML	ADHF	125	Yes	MVR + Morrow
Yano (2000) ⁶	59, M	PML	ADHF	N/A	N/A	MVR
Wakeyama (2004) ⁷	61, M	A3	ADHF	120	Yes	MVR + Morrow
Araujo (2005) ⁸	38, M	A3, P3	Stable	105	No	Medication
Koshino (2006) ⁹	63, F	P3	ADHF	85	No	MVR + TAP
Morris (2009) ¹⁰	63, F	P2	ADHF	109	N/A	MVR
Yang (2009) ¹¹	69, M	A3, P3	ADHF	85	No	MVR + Morrow + Maze
Suehiro (2015) ¹²	69, F	P3	ADHF	71	Yes	MVr + Morrow

A2, Middle portion of the AML; A3, medial portion of the AML; *ADHF*, acute decompensated heart failure; *CABG*, coronary artery bypass graft; *AML*, anterior mitral leaflet; *F*, female; *M*, male; *Morrow*, ventricular septal myectomy; *MVr*, mitral valve repair; *MVR*, mitral valve replacement; *P2*, middle scallop of PML; *P3*, medial scallop of PML; *PML*, posterior mitral leaflet; *TAP*, tricuspid valve annulus plasty.

CONCLUSION

Chordal rupture is a rare but serious complication that could change the hemodynamics by generating acute MR and alleviating LVOT obstruction in HOCM. In the present case, symptoms of HF and LVOT obstruction were completely resolved after rupture of mitral valve chordae tendineae, suggesting that the mitral valve plays a pivotal role in the pathophysiology of LVOT obstruction.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2019.03.001>.

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