

# Muscular Mitral Chord Contribution to Left Ventricular Outflow Tract Obstruction in HOCM

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## Abstract

Papillary muscle anomaly with a muscular chord directly attached to the anterior mitral leaflet is a rare mitral valve disease. A 62-year-old man with systolic anterior motion of the anterior mitral leaflet and hypertrophic obstructive cardiomyopathy presented to surgical intervention after unsuccessful transcatheter ablation of septal hypertrophy with alcohol. Intraoperative findings revealed a primarily not detected anomalous muscular mitral chord (0.8 × 2.2 cm) connecting the base of the A1 segment to the anterolateral papillary muscle. Resection of this chord and additional septal myectomy treated systolic anterior motion and obstruction of the outflow tract. In spite of the infrequent occurrence, anomalies of the subvalvular apparatus, such as muscular chords, should be ruled out by thorough transesophageal echocardiography imaging before decision on the therapeutical strategy.

## Keywords

- ▶ hypertrophic obstructive cardiomyopathy
- ▶ chordae anomaly
- ▶ systolic anterior motion
- ▶ mitral valve disease

## Introduction

In patients with hypertrophic cardiomyopathy, anomalies of the mitral valve structures can contribute to left ventricular outflow tract (LVOT) obstruction and mitral valve insufficiency by systolic anterior motion (SAM). Common pathologies comprise leaflet elongation and, less frequently, papillary muscle anomalies (increased papillary muscle mass while overall left ventricle mass is normal, anterior displacement of the papillary muscle and anomalous insertion of the papillary muscle).<sup>1</sup> Papillary muscle anomalies may be underdiagnosed due to their rare occurrence. In this article, we present the case and surgical treatment of a patient with hypertrophic cardiomyopathy who additionally exhibited a primarily not detected anomalous muscular chord directly attached to the anterior mitral valve leaflet as underlying pathology for SAM and subsequent LVOT obstruction.

## Case Presentation

A 62-year-old man diagnosed with hypertrophic obstructive cardiomyopathy (HOCM) 18 months ago now suffered from

progressing dyspnea (New York Heart Association [NYHA] functional classification class III). The LVOT gradient at rest was 115 mm Hg, and up to 200 mm Hg during exercise. The patient initially underwent transcatheter ablation of septal hypertrophy (TASH) by means of alcohol, which proved ineffective with continuous symptoms postinterventionally. Thorough transesophageal echocardiography (TEE) control revealed an unaltered pathological LVOT gradient, SAM with mild mitral regurgitation and de novo detection of a subvalvular structure attached to the anterior mitral leaflet (AML) that seemed to contribute to LVOT obstruction (▶ Fig. 1A). Therefore, the patient was admitted to our center for surgical treatment.

The intraoperative TEE confirmed the finding of an echogenic structure connected to the AML in area A1 as well as to the anterolateral papillary muscle. Applying a transaortic surgical approach, a muscular chord with dimension of 0.8 × 2.2 cm was detected in the position described above. The muscular chord was completely resected without necessity of mitral valve reconstruction (▶ Fig. 1B). Furthermore, a septal myectomy was performed resecting a segment of 1.0 cm depth, 1.5 cm width, and 2 cm length. The

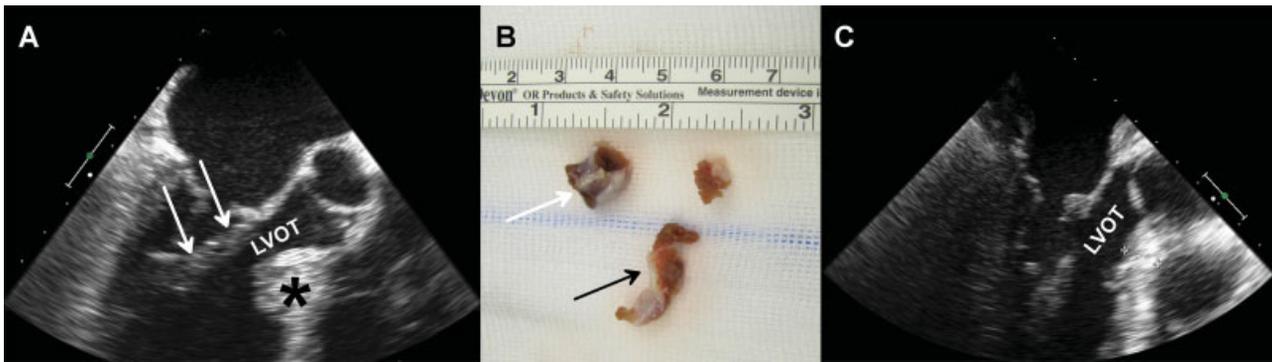
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**Fig. 1** Transesophageal echocardiography B-mode views showing (A) preoperative septal hypertrophy (black asterisk) and a thick chord (white arrows) attached to the anterior mitral leaflet and the anterolateral papillary muscle, and (C) the postoperative result without left ventricular outflow tract (LVOT) obstruction. Measured septal wall thickness in (C) = 12 mm. (B) Image of the resected muscular chord (black arrow) and the resected segment of the hypertrophic septum (white arrow). Top scale on the bar measures centimeters.

postoperative TEE showed a wide LVOT and the absence of SAM (→Fig. 1C). The LVOT gradient amounted to 10 mm Hg. A pacemaker had to be implanted due to postoperative atrioventricular block III.

## Comments

Muscular chords are rare findings in the group of pathological mitral valve structures leading to LVOT obstruction. Only a few reports on abnormal chordae tendineae with muscular chords connecting the leaflets to the ventricular septum or free wall have been published.<sup>1,2</sup> A modified classification of chords was proposed by Gunnal et al, counting muscular chords to the structural groups of chordae, such as tendinous, muscular and membranous chordae.<sup>3</sup>

While for single hypertrophic obstructive cardiomyopathy, interventional therapy provides a good alternative to surgical myectomy,<sup>4</sup> there is no evidence on the optimal treatment in case of additional muscular mitral chords. In the presented case, we think that the surgical approach should have been the first choice instead of TASH, which actually did not remove the LVOT obstruction, but it predisposed the patient to a postoperative atrioventricular block. In a previous study on 31 patients undergoing septal myectomy following failed TASH, the incidence of complete AV block was significantly increased as compared with patients with septal myectomy only (19.4 vs. 1.6%;  $p < 0.01$ ).<sup>5</sup>

In spite of the infrequent occurrence, anomalies of the subvalvular apparatus, such as muscular chords, should be ruled out by TEE imaging before decision on the therapeutic strategy.

## Conflict of Interest

None.

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