

Extended Myectomy for Apical Hypertrophic Cardiomyopathy: A Case Report

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Case report

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Abstract

Background

Apical hypertrophic cardiomyopathy is a variant of hypertrophic cardiomyopathy that predominantly affects the left ventricle apex and rarely involves the right ventricular apex or both apices. Traditional treatment for apical hypertrophic cardiomyopathy is heart transplantation. Although surgical myectomy approaching the apex has been available for decades, its safety and accuracy greatly depend on the surgeon's skills and experience.

Case presentation

The first case is of a 63-year-old man with apical hypertrophic cardiomyopathy in whom a preoperative contrast computed tomography revealed apical hypertrophy and complete apical cavity obliteration. The patient underwent extended myectomy. The inside of the apex cavity was filled with abnormal muscles. Using the transaortic approach, the location of the bilateral papillary muscle was confirmed, which provided the required orientation. The abnormal muscle mass was successfully resected, and postoperative end-diastolic volume was extremely increased. The second case is of a 43-year-old man with an apical left ventricular aneurysm and mid-hypertrophic cardiomyopathy obstruction. The thin-walled apical aneurysm contained a large apical-basal band. After detecting the bilateral papillary muscle, mid-ventricular myectomy was performed from the apex. During postoperative catheterization, there was no pressure gradient between the left ventricle and aorta.

Conclusions

We reviewed two cases of apical hypertrophic cardiomyopathy, efficiently treated using extended apical myectomy. Although apical myectomy is an uncommon procedure, the cases presented herein show that it can successfully manage apical hypertrophic cardiomyopathy. However, it is important to secure the postoperative left ventricular end-diastolic volume.

Background

Apical hypertrophic cardiomyopathy (aHCM) is a variant of HCM that predominantly affects the left ventricle apex and rarely involves the right ventricular apex or both apices. aHCM was first described in 1976 and presents unique echocardiographic findings of asymmetrical apical hypertrophy in an ace-of-spades configuration [1, 2]. Approximately 40% of HCM cases are aHCM. The condition is relatively uncommon in western countries and occurs more frequently in Asian populations [3–7]. aHCM is traditionally treated using heart transplantation. Surgical myectomy approaching the apex (apical myectomy) has been available for decades [8]; however, the safety and accuracy of apical myectomy greatly depend on the surgeon's skills and experience [9].

Herein, we report two cases of patients with aHCM who underwent extended myectomy.

Case Presentation

Case 1

A 63-year-old man with an 8-year history of aHCM had been healthy until 7 years after his aHCM diagnosis when he experienced exertional dyspnea. He was referred to our hospital for surgical aHCM treatment. Preoperative contrast computed tomography (CT) revealed apical hypertrophy and complete apical cavity obliteration. Preoperative transesophageal echocardiography (TEE) revealed no systolic anterior motion (SAM) and an abnormal apical-basal band.

The patient underwent extended myectomy. The inside of the apex cavity was filled with abnormal muscles (Figure 1). Therefore, we could not decide on the orientation for making an incision on the myocardium. Using the transaortic approach, we confirmed the location of the bilateral papillary muscle, which provided us with the knowledge of required orientation. We first used a bidirectional tunnel approach between the bilateral papillary muscle and resected area to avoid the apex. The hypertrophic myocardium appeared whitish. We used 2-0 monofilament sutures for left ventricular (LV) closure. The cardiopulmonary bypass time was 148 min, and the aortic clamp was on for 93 min.

Postoperative contrast CT showed a small LV aneurysm, which was surgically resected 3 weeks after initial myectomy. On postoperative day 3, the patient developed septic shock because of urinary tract infection and mediastinitis. After antibiotic administration and omentopexy, he was transferred to his previous hospital for 90 days of rehabilitation. Postoperative TEE demonstrated an LV end-diastolic volume (LVEDV)/end-systolic volume of 105.3/56.5 mL (preoperative, 67.2/35.9 mL). The postoperative left ventriculogram and angiography are shown in Figure 2. The patient's exertional dyspnea improved. He was discharged 30 days after changing the hospital.

Case 2

A 43-year-old man with an apical LV aneurysm and mid-HCM obstruction was referred to our hospital for further evaluation. Eight years earlier, he was fitted with an implantable cardioverter-defibrillator (ICD) for non-sustained ventricular tachycardia. A mid-HCM was detected using CT and transthoracic echocardiography (TTE), and he was followed up as an outpatient for 5 years. One month before presenting at our hospital, he developed syncope and palpitations.

Preoperative contrast CT revealed an apical aneurysm and mid-ventricular obstruction. Preoperative TEE revealed mid-acceleration but no SAM. He underwent an extended myectomy for his palpitations and arrhythmia. Inside the thin-walled apical aneurysm, there was a large apical-basal band. Mid-ventricular myectomy was performed from the apex after detecting the bilateral papillary muscle. The resected myocardium weighed 9 g. The aneurysm was removed, and the incision was closed. The cardiopulmonary bypass time was 85 min, and the aorta was clamped for 50 min. Postoperative TEE revealed no SAM or ventricular septal perforation. During postoperative catheterization, there was no pressure gradient between the left ventricle and aorta.

On postoperative day 8, the patient had atrial flutter, and defibrillation was performed via an ICD. His sinus rhythm recovered, and he was discharged on postoperative day 10.

Discussion And Conclusions

Left ventricular septal myectomy is the gold standard surgical treatment for LV outflow tract obstruction (LVOTO) in HCM patients [10]. However, the safety and accuracy of apical myectomy depend on the surgeon's skills and experience [9] as it is an uncommon procedure. Apical myectomy can be used for aHCM repair, but the postoperative LVEDV should be secured. Postoperatively, the cardiac output likely improves because of both increased outflow area and reduced drag forces on the anterior mitral leaflet (AML) by changing the flow vector in the outflow tract (the flow is more parallel to the AML).

The patient in case 1 had extensive hypertrophy, especially in the apex; therefore, we could not perform myectomy using only the transaortic approach. However, using both apex and transaortic approaches, we could determine the positions of the bilateral papillary muscles, chordates, and mitral valve. This was our first case of treating aHCM using a bidirectional approach. In case 2, extended myectomy was preferred because we thought that removal of both the thin apex aneurysm and part of the septum was necessary, and we could secure the patient's postoperative LVEDV. The comparison of pre- and post-CT scans in both cases, especially the first one, showed increased LVEDV.

In the first case, poor visibility caused considerable difficulty (Table 1). Therefore, we could not determine the correct direction or depth from the apex for the incision. Although this depends on the surgeon's skills and experience, adding a transaortic incision and using a finger as a guide from the LVOT can be helpful. Intraoperative TEE is also beneficial.

Table 1. Comparison of the two aHCM and LVOTO cases

	aHCM	LVOTO
Cause of decreasing SV	LVDEV↓	LVOTS, SAM
Difficulties of procedure	Invisible	More visible
Surgical complications	Mitral valve injury, ventricular septal perforation, left coronary injury	Aortic valve injury, ventricular septal perforation

aHCM, apical hypertrophic cardiomyopathy; LVEDV, left ventricular end-diastolic volume; LVOTO, left ventricular outflow tract obstruction; LVOTS, left ventricular outflow tract stenosis; SAM, systolic anterior motion; SV, stroke volume

After a discussion with our team of cardiovascular surgeons and cardiologists, we reached a consensus to use these procedures rather than medical treatment for these two cases. We hope that the positive outcomes from these two cases will lead to improvements in extended myectomy.

Apical myectomies are performed rarely; however, there is a need to standardize this method. In Japan, heart transplantation for aHCM is unrealistic because there are very few transplantation facilities. Heart transplantation is a longer procedure than extended myectomy. Extended myectomy can be challenging, but a detailed surgical procedure has been recently reported [3, 11-13]. Almost half of the global aHCM cases occur in Asian countries; therefore, clinicians should gather sufficient surgical experience in extended myectomy.

Abbreviations

AML anterior mitral leaflet

CT computed tomography

aHCM apical hypertrophic cardiomyopathy

ICD implantable cardioverter-defibrillator

LV left ventricular

LVEDV left ventricular end-diastolic volume

LVOTO LV outflow tract obstruction

LVOT LV outflow tract

LVOTS left ventricular outflow tract stenosis

SAM systolic anterior motion

SV stroke volume

TEE transesophageal echocardiography

TTE transthoracic echocardiography

Declarations

Ethics approval and consent to participate: This was a case report and a requirement for an approval from the Institutional Review Board of Sakakibara Heart Institute was waived.

Consent for publication: The patient provided informed consent for the publication of images

Availability of data and materials: All data generated or analyzed during this study are included in this published article

Competing interests: None

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Authors' contributions: DS performed data collection and wrote the manuscript. MS supported the writing of the manuscript. ST participated in the design of the study.

All authors read and approved the final manuscript.

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Figures

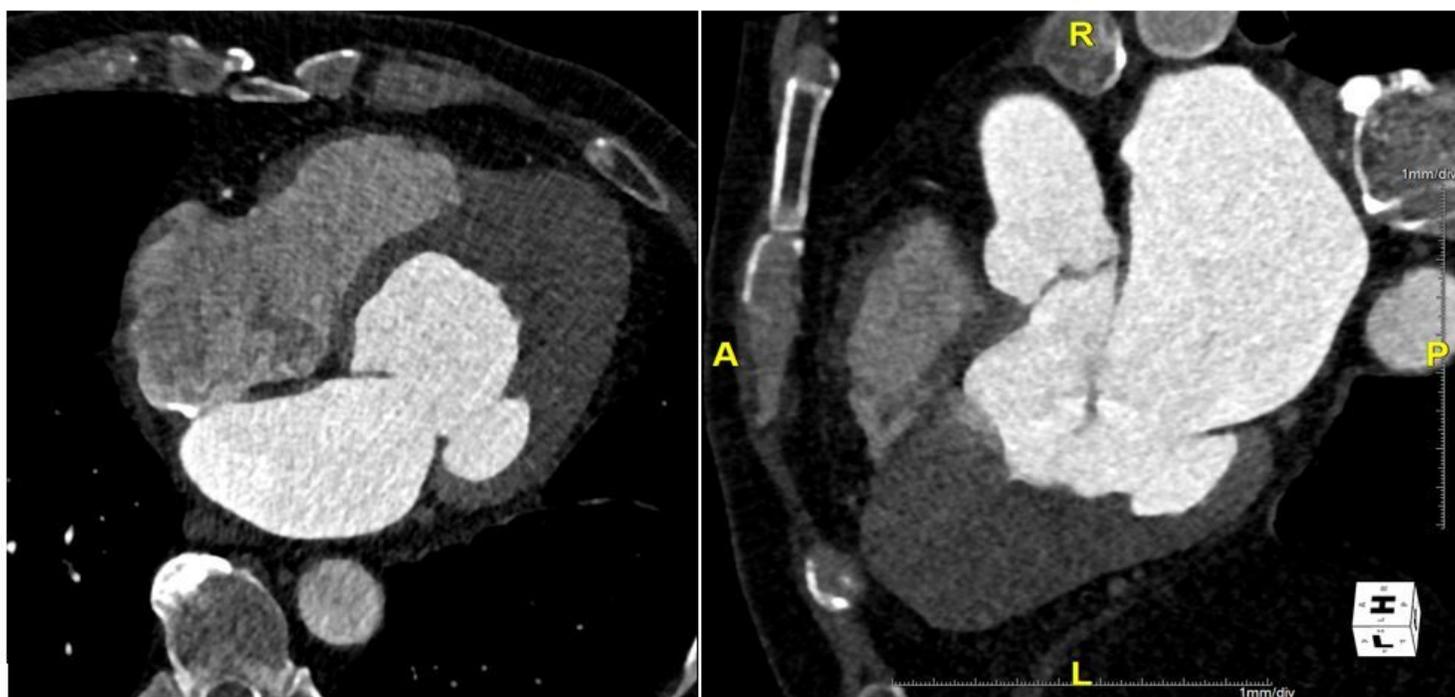


Figure 1

Preoperative CT images from case 1

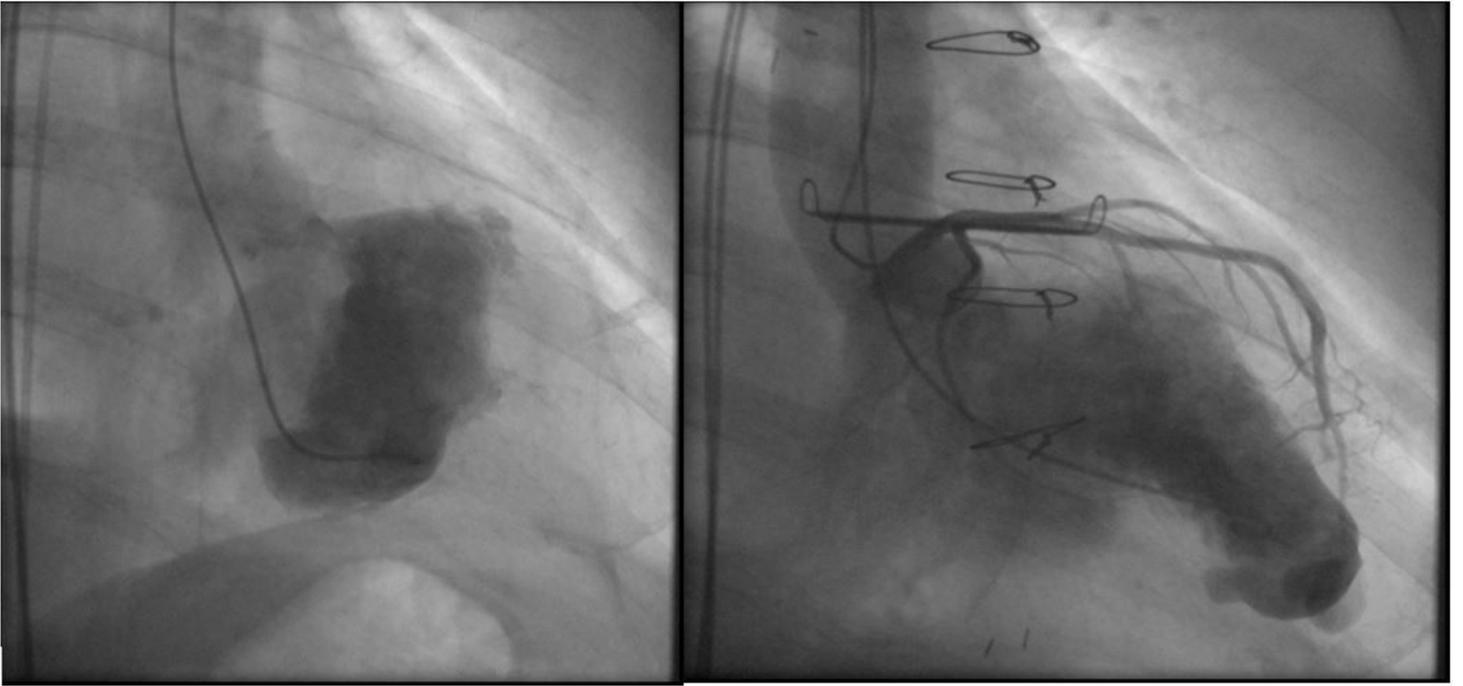


Figure 2

Pre- and postoperative cardiography images from case 1

Supplementary Files

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