Abnormal Mitral Valve Apparatus in a Case of Hypertrophic Obstructive Cardiomyopathy: Intraoperative Transesophageal Echocardiography

Abstract

Hypertrophic obstructive cardiomyopathy is a relatively common disorder that signifies asymmetric hypertrophy of interventricular septum causing obstruction of the left ventricular outflow tract (LVOT). However, more recent studies have shown that during ventricular systole, flow against an abnormal mitral valve apparatus results in drag forces on the part of the leaflets. The mitral leaflet is pushed into the LVOT to obstruct it. We present a case where intraoperative transesophageal echocardiography played a crucial role in defining the etiology of LVOT obstruction that subsequently helped in deciding the surgical plan.

Keywords: *Abnormal mitral valve apparatus, Asymmetric septal hypertrophy, transesophageal echocardiography*

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Introduction

Hypertrophic obstructive cardiomyopathy (HOCM) is defined as asymmetrical LV thickening without chamber dilation that develops in the absence of an identifiable cause (aortic stenosis, hypertension). 1 Disorganized myocyte pattern, increased wall/lumen ratio of coronaries, and remodeling changes lead to impaired coronary reserve, diastolic dysfunction, ventricular dysrhythmias and sudden death in HOCM patients.

In addition to the septal hypertrophy, the mitral valve (MV) apparatus is intrinsically abnormal in HOCM patients contributing to LVOT obstruction. 1,2 A number of surgical techniques are available but the technique should be individualized on a case-to-case basis. 3,4 Intraoperative transesophageal echocardiography (TEE) has a vital role to play in describing the accurate etiology of LVOT obstruction and therefore, it helps in selection of proper surgical technique. The present case report precisely discusses the importance of intraoperative TEE in evaluating a patient of LVOT obstruction and surgical decision-making.

Case Report

A 32-year-old male patient presented with a 1-year history of chest pain and dyspnea on exertion. Preoperative transthoracic echocardiography revealed a hypertrophic septum obstructing the flow in the left ventricular outflow tract (LVOT) with a peak gradient of 120 mmHg, an ejection fraction of 60% and severe eccentric mitral regurgitation due to systolic anterior motion (SAM). He was planned for septal reduction surgery along with the detailed intraoperative transesophageal echocardiography (TEE) evaluation of the mechanism of SAM. After uneventful standard anesthesia induction, the heart was inspected with a TEE probe and an ultrasound system (Epiq 7, Philips Ultrasound, Andover, MA, USA). The TEE examination detected a catenoid type hypertrophy of mid-ventricular septum (diastolic septal thickness of 19.8 mm) with turbulent flow across the LVOT. During midsystole, the anterior mitral leaflet (AML) was seen dragging into the LVOT to obstruct the flow, whereas a regurgitant jet was directed toward the posterior mitral leaflet (PML) as shown in Figure 1. Other interesting findings were the elongated mitral leaflets resulting in anterior displacement of the coaptation point, small LV cavity, anterior, and medially displaced hypertrophied anterolateral papillary muscles (AL-PM) with bifurcated heads. The echocardiographic parameters described in the study by

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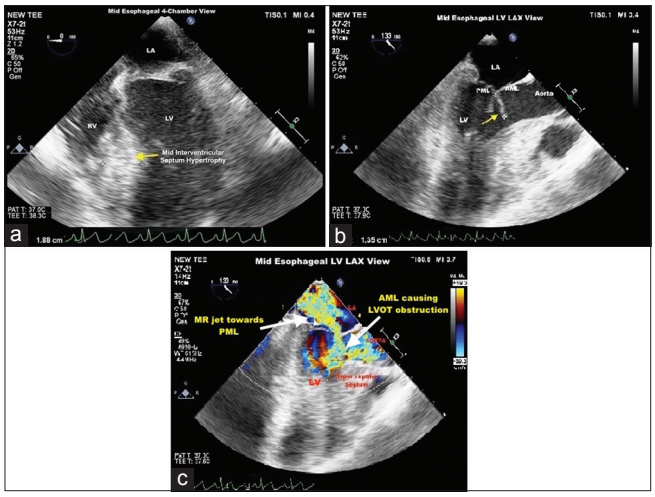

Figure 1: Echocardiographic details showing left ventricular outflow tract obstruction. (a) Mid-septum hypertrophy (Catenoid septum). (b) Residual anterior mitral leaflet beyond the point of coaptation shown by yellow arrow dragged into left ventricular outflow tract causing obstruction. (c) Color Doppler shows flow turbulence in left ventricular outflow tract and eccentric mitral regurgitation

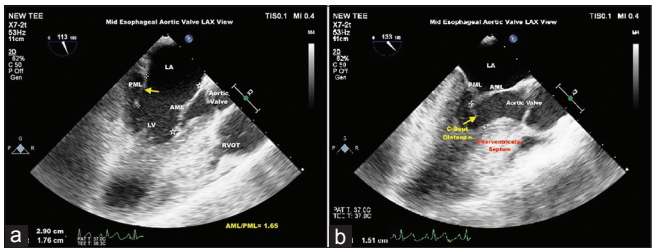

Figure 2: Predictors of residual systolic anterior motion following mitral valve repair. (a) Measurement of mitral valve leaflets in end diastole; elongated posterior mitral leaflet is shown by yellow arrow. (b) Measurement of coaptation-septum distance in the end systole

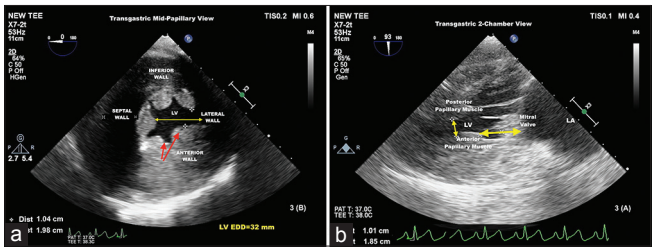

Figure 3: Abnormal Mitral valve apparatus. (a) Septal hypertrophy (19.8 mm); reduced left ventricular end diastolic dimension; hypertrophied and bifurcated anteriorly displaced. (b) Superior displacement of anterolateral papillary muscle and reduced interpapillary muscle distance (10.1 mm) produce chordal slack, sweep the anterior mitral leaflet into the left ventricular ejection flow stream

Varghese *et al*. such as coaptation-septal distance, ratio of length of mitral leaflets (AML/PML), anterior location of the PMs, inter-PM distance, and LV end-diastole dimension predicted the postoperative SAM potential,[1] and therefore, the likelihood of mitral valve (MV) repair failure [Figures 2 and 3]. Subsequently, the surgeon was informed about the obstructive pathophysiology due to intrinsically abnormal MV apparatus in addition to the septal hypertrophy. A collaborative decision was taken to replace the abnormal MV along with septal reduction surgery to prevent high residual postoperative LVOT gradient. After sternotomy and systemic heparinization, standard normothermic cardiopulmonary bypass was established utilizing aortobicaval cannulation. A shelf of hypertrophied muscle was removed from the basal septum to enlarge the LVOT, and a 29 mm sized mechanical prosthesis was used to replace the MV. Adrenaline (0.05 μg/kg/min) was initiated in the immediate postbypass period in view of low heart rate to maintain the systemic pressure. He developed atrioventricular (AV) conduction block and required temporary pacing. A detailed TEE examination was performed in the postoperative period to rule out the presence of any residual high LVOT gradient (peak gradient of 15 mmHg), aortic insufficiency, ventricular septal defect, and prosthetic valve dysfunction. Table 1 outlines the echocardiographic parameters that should be assessed in all the hypertrophic obstructive cardiomyopathy (HOCM) patients undergoing repair surgery. The patient was shifted to ICU with sequential AV pacing and no inotropic support.

The literature mentions different phenotypes of HOCM patients based on the location of septal wall thickness and has found a striking association between one particular subtype and certain characteristics.[2] Patients with the catenoid morphology, predominant thickening of the mid-septum, have the youngest age at the time of diagnosis, larger LV mass with restrictive diastolic dysfunction and a higher prevalence of MV abnormalities. The most frequent symptoms are exercise intolerance, angina, dyspnea, dizziness, syncope, dysrhythmias, and sudden death.

The LV cavity has an inflow and an outflow compartment. The normal LV inflow occurs posteriorly during diastole, while systolic outflow from LV occurs anteriorly after the MV closure. The separation of these two functional compartments and the normal coaptation of the MV leaflets posteriorly away from the LVOT are determined by anatomically interrelated variables such as size of ventricular cavity in relation to the size and location of components of the mitral apparatus.[2] Anatomic abnormalities of the MV are well recognized among patients with HOCM [Table 1] that contribute to subvalvular LVOT obstruction. Flow-drag-flow forces occur when anteriorly displaced PMs cause diastolic inflow to be directed toward the septum opposed to the normal flow pattern, and outflow is directed posteriorly, causing drag forces on the MV leaflets into the LVOT. Moreover, elongated leaflets reach into the LV cavity well above the plane of mitral annulus and displace the coaptation point anteriorly. The residual portion of AML beyond the point of coaptation moves with the LV flow and contacts the septum contributing to the flow

Table 1: Echocardiographic parameters to be assessed in hypertrophic cardiomyopathy

Prebypass parameters	Postbypass parameters
Hypertrophic septum	Color jet in LVOT: Rule out
The location and distribution of maximal septal thickness (basal sigmoid, catenoid, neutral and apical)	Iatrogenic ventricular septal defect
	Iatrogenic aortic insufficiency
The ratio of septal/posterior wall thickness to report asymmetry (asymmetrical hypertrophy if ratio >1.3)	Severed septal perforator
Exact site of obstruction and peak left ventricular outflow gradient	Any residual obstruction; site and gradient
Abnormal MV[2,3]	Residual SAM
Details of MR (mechanism, severity)	Residual MR
MV leaflet length (AML>33 mm/ >17 mm/m2, PML >15mm)	Prosthetic MV function (normal seating, leaflet motion, paravalvular leak, mean gradient, DVI and EOAI)
Papillary muscle hypertrophy, location of papillary muscles (anteriorly displaced) and their anomalous insertion onto AML	Iatrogenic mitral stenosis after MV repair
Reduced interpapillary distance (normal value: 20mm-25 mm)	
Mitral annular calcification	
Anterior mitral tenting due to fibrotic and retracted secondary chordae	
Prediction of failure of MV repair	Successful MV repair
Prediction of SAM potential after MV repair2 (LV EDD <45 mm, aorto-mitral angle <120°, c-sept distance <25 mm, PML height >15 mm and basal septal diameter ≥15 mm)	Coaptation zone (6-9 mm)
	Residual MR <1+
	Mean transmitral gradient ≤6 mmHg
MV leaflet morphology	MV area ≥1.8 cm2
LV systolic and diastolic function	LV function assessment
LV ejection fraction, TDI and strain imaging of wall motion (LVEF <50% and systolic strain of ≤−10.6% signify poor LV systolic function)	
LV diastology (TDI velocities of mitral annulus, LA volume index >34 ml/m2)	
LV dimensions (small LV cavity)	
RV function and pulmonary artery systolic pressure	RV function assessment

LVOT: Left ventricular outflow tract, MV: Mitral valve, MR: Mitral regurgitation, SAM: Systolic anterior motion, c-sept: Coaptation-septum, AML: Anterior mitral leaflet, PML: Posterior mitral leaflet, LV: Left ventricle, EDD: End-diastolic dimension, TDI: Tissue Doppler imaging, LA: Left atrium, RV: Right ventricle, DVI: Dimensionless velocity index, EOAI: Effective orifice area index, LVEF: Left ventricular ejection fraction

obstruction.[3] Reduced interpapillary distance due to muscle hypertrophy and medial displacement produces slack in the chords attached to the center (A2 segment) of the AML, allowing it to be swept anteriorly toward the septum.

Although adequate septal myectomy relieves outflow tract gradients in many patients, some may have persistent obstruction due to SAM of mitral leaflets. Therefore, several reports have described adjunctive techniques of mitral valvuloplasty, and the controversy regarding the optimal surgical strategy for HOCM patients still continues.[3,4] A number of surgical techniques are available, given the diversity of mitral abnormalities; the proper technique should be selected for the individual patient. The techniques include plication of AML when it is > 17 mm/m^2 (vertical or horizontal plication), excision of excessive leaflet, release of the AL-PM by extending the resection laterally into the free wall above its base along with thinning of the hypertrophied heads. PM release or their surgical reorientation brings the plane of the mitral annulus and aortic valve into a more normal parallel orientation. Accessory PM heads and anomalous chordae should be removed. Nevertheless, repair of subvalvular apparatus is technically demanding and introduce additional complexity and potential complications. The decision of MV repair depends on surgeon's expertize and speculation. In addition to the surgery, HOCM patients may require implantable cardioverter-defibrillator in following conditions: (1) positive family history of premature sudden cardiac death as a result of HOCM; (2) documented nonsustained ventricular tachycardia; (3) syncope at rest or during exercise; and (4) an abnormal arterial blood pressure response to exercise (increase in systolic blood pressure of <20 mm Hg from the baseline value).

In our case, we detected the need to repair the abnormally elongated mitral leaflets. However, abnormal morphology and location of PMs made the repair more challenging. The authors analyzed the postoperative SAM potential after MV reconstruction. Therefore, the MV was replaced along with resection of hypertrophied septum.

Intraoperative TEE has a definite role to play in HOCM patients to detect the abnormal MV geometry, predict the success of repair, plan the surgical repair and confirm the successful repair without any complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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