

A rare case of hypertrophic obstructive cardiomyopathy with mitral valve stenosis

Anil Özen¹, Aytaç Çalışkan², Utku Ünal¹, Emre Kubat³, Levent Birincioglu¹

Received: February 08, 2015 Accepted: December 25, 2015 Published online: March 03, 2016

ABSTRACT

A 50-year-old male was referred with a 10 day history of tachypnea and dyspnea. Transthoracic echocardiography showed serious fibrotic mitral stenosis and a mean gradient of 18 mmHg. The interventricular septum was hypertrophic with a width of 2.1 cm. A standard septal myectomy and mitral valve replacement were performed. Hypertrophic obstructive cardiomyopathy is mostly associated with mitral insufficiency rather than mitral stenosis. Surgery can be life-saving in acute deterioration in patients with coexisting hypertrophic obstructive cardiomyopathy and mitral valve pathology.

Keywords: Hypertrophic obstructive cardiomyopathy; mitral stenosis; mitral valve replacement; septal myectomy.

Hypertrophic obstructive cardiomyopathy (HOCM) is a cause of sudden cardiac death in young patients.^[1] Intraventricular anatomical and contractile features are the determinants of the clinical course and management modality in most of the cases. Herein, we discuss an unusual case of HOCM and mitral stenosis (MS).

CASE REPORT

A 50-year-old male was referred to our hospital with a 10-day history of tachypnea and dyspnea. Physical examination revealed widespread crepitations at the base of the lungs, particularly. His blood pressure was 100/70 mmHg. Electrocardiography revealed atrial flutter with a rate of 150 bpm. Transthoracic echocardiography (TTE) demonstrated serious fibrotic MS with a valve area of 1.2 cm² and a mean gradient of 18 mmHg. The interventricular septum was hypertrophic with a width of 2.1 cm (Figure 1). The pressure gradient of the left ventricular outflow tract (LVOT) was 76 mmHg and the ejection fraction was 60%. The preoperative TTE did not reveal an abnormal anterior motion of the anterior leaflet. Coronary angiography showed clinically insignificant coronary plaques. Physical examination and a chest X-ray revealed signs of pulmonary edema and increased cardiothoracic ratio. A decision was made to operate on an emergency basis. A written informed consent was obtained from the patient.

Transesophageal echocardiography probe was placed. Sternotomy was followed by bicaval

cannulation. Cardiopulmonary bypass (CPB) was initiated. After cross-clamping, an oblique aortotomy was made towards the non-coronary sinus. The first incision was made to the subannular region at the level of the right and the left leaflet commissures. The second incision was made to the subannular region at the level of the right coronary ostium. These two incisions were adjoined with the third incision which was made 5 mm below the annulus. The myectomy was performed with a thickness of 1 cm and a length of 3-4 cm towards the apex. The myectomy was decided to be sufficient, when the trunk and the base of the papillary muscles became visible. As the mitral valve leaflets were severely thickened and fibrosed with a narrow valve orifice, the mitral valve replacement was performed. Transesophageal echocardiography revealed the pressure gradient of the LVOT as 15 mmHg and no systolic anterior motion (SAM) of the mitral valve was detected. The postoperative course was uneventful. He was followed in the intensive care unit (ICU) for two days and discharged in the eight postoperative day.

¹Department of Cardiovascular Surgery, Türkiye Yüksek İhtisas Training and Research Hospital, Ankara, Turkey

²Department of Cardiovascular Surgery, Karşıyaka State Hospital, İzmir, Turkey

³Department of Cardiovascular Surgery, Karabük University Karabük Training and Research Hospital, Karabük, Turkey

Corresponding author: Aytaç Çalışkan, MD. Türkiye Yüksek İhtisas Eğitim ve Araştırma Hastanesi Kalp ve Damar Cerrahisi Kliniği, 06230 Altındağ, Ankara, Turkey.

Tel: +90 312 - 306 12 34 e-mail: aytac.caliskan@gmail.com



Figure 1. A preoperative transthoracic echocardiographic image showing an interventricular septum thickness of 2.1 cm.

DISCUSSION

Asymmetrical septal hypertrophy is the most common cause of HOCM which results in SAM of the anterior leaflet of the mitral valve.^[2] This abnormal movement of the mitral apparatus may also cause mitral insufficiency and contributes to the worsening of the left ventricular function.^[3] Patients may be asymptomatic, until the hypertrophy becomes prominent and causes geometrical impairment of the left ventricle.^[1] Sudden cardiac death may be even the presenting symptom in some of the cases.^[1]

The unusual presentation of our case is what makes this case interesting. Hypertrophic obstructive cardiomyopathy is mostly associated with mitral insufficiency rather than MS.^[3] The preoperative TTE did not reveal an abnormal anterior motion of the anterior leaflet nor did we observe any regurgitation intraoperatively. There was a severe pressure gradient caused by the MS. The MS may have decreased the already low left ventricular filling even further. The clinical deterioration and pulmonary edema seen in our case, despite medical treatment, was due to the sum of the pathophysiological effects of HOCM and MS; HOCM increased the left ventricular end-diastolic pressure with outflow obstruction resulting in back pressure on the lungs, while MS caused dyspnea due to the restricted left ventricular inflow, thereby, resulting in left atrial pressure and back pressure on the pulmonary circulation.

Pharmacological therapy with beta-blockers, calcium channel blockers, and disopyramide is often beneficial for patients with mild symptoms.^[4] However, for patients refractory to the pharmacological therapy, more invasive therapeutic procedures can be life-saving.

In addition, alcohol-induced septal ablation (ASA) is an interventional treatment modality which aims to decrease the septal muscle mass.^[5] Long-term follow-up results are comparable with surgical myectomy.^[5] Alcohol-induced septal ablation can also achieve significant reduction of the LVOT obstruction gradient and symptomatic relief.^[5] However, complications such as complete heart block are more common after ASA than surgical myectomy.^[5] In daily practice, it is mostly performed in older patients with high surgical risk and in those without any intracardiac disease and not requiring open surgery.^[5]

Furthermore, surgery yields the best long-term results with good mortality and morbidity rates.^[6] Septal myectomy is the conventional procedure in most of the cases.^[7] Almost always, sufficient myectomy is achieved, if the papillary muscles are easily visible. Nevertheless, in some patients, myectomy may be insufficient in creating a wide outflow tract. In such cases, mitral valve replacement is the treatment of choice to relieve the LVOT obstruction.^[7] In this case, mitral valve replacement was performed, as the mitral valve was not appropriate for repair. We believe that replacement of the valve also contributed to the abolition of the LVOT gradient.^[8]

In conclusion, in most cases, hypertrophic obstructive cardiomyopathy does not consist of a single anatomical pathology and it can manifest itself with an unusual clinical presentation, as in our case. Therefore, the approach to each case should be individualized and treatment decisions should be made by expert surgeons. Surgery is a more favorable approach for hypertrophic obstructive cardiomyopathy and can be life-saving in acute deterioration in patients with coexisting hypertrophic obstructive cardiomyopathy and mitral valve pathology.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

REFERENCES

1. Maron BJ, Olivetto I, Spirito P, Casey SA, Bellone P, Gohman TE, et al. Epidemiology of hypertrophic cardiomyopathy-related death: revisited in a large non-referral-based patient population. *Circulation* 2000;102:858-64.
2. Wigle ED, Sasson Z, Henderson MA, Ruddy TD, Fulop J, Rakowski H, et al. Hypertrophic cardiomyopathy. The importance of the site and the extent of hypertrophy. A review. *Prog Cardiovasc Dis* 1985;28:1-83.
3. Cavalcante JL, Barboza JS, Lever HM. Diversity of mitral valve abnormalities in obstructive hypertrophic cardiomyopathy. *Prog Cardiovasc Dis* 2012;54:517-22.
4. Nishimura RA, Holmes DR. Hypertrophic obstructive cardiomyopathy. *N Engl J Med* 2004;350:1320-7.
5. Yavuzgil O, Tülüce K, Şimşek E, Gürgün C, Kültürsay H, Payzın S ve ark. Hipertrofik obstrüktif kardiyomyopatide septal alkol ablasyonu: Retrospektif bir inceleme. *Turk Gogus Kalp Dama* 2012;20:427-33.
6. Ünal EU, Özen A, İşcan HZ, Birincioğlu CL. Obstrüktif hipertrofik kardiyomyopatide cerrahi tedavinin erken ve orta dönem sonuçları. *Turk Gogus Kalp Dama* 2012;20:421-6.
7. Mataracı İ, Polat A, Songur ÇM, Aydın C, Yanartaş M, Erentuğ V ve ark. Hipertrofik obstrüktif kardiyomyopatide cerrahi tedavi ve sonuçları. *Turk Gogus Kalp Dama* 2009;17:243-8.
8. McIntosh CL, Maron BJ. Current operative treatment of obstructive hypertrophic cardiomyopathy. *Circulation* 1988;78:487-95.