Hypertrophic obstructive cardiomyopathy causing severe right and left ventricular outflow tract obstruction

Sağ ve sol ventrikül çıkış yolunda ileri derecede tıkanıklığa neden olan hipertrofik tıkayıcı kardiyomiyopati

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An 18-year-old male patient presented with a 3-year history of exertional dyspnea, dizziness, and angina. Echocardiography showed advanced hypertrophy of the left ventricle (LV), right ventricle (RV) free wall, and interventricular septum. There were apparent muscular bundles especially at the level of the right ventricular outflow tract (RVOT). Maximal pressure gradients across the RVOT and left ventricular outflow tract (RVOT) were 141 mmHg and 66 mmHg, respectively. There was also grade 2 aortic regurgitation. Transesophageal echocardiography and cardiac magnetic resonance imaging confirmed these findings. Despite treatment with propranolol and cibenzoline, the patient remained symptomatic with unchanged pressure gradients. Corrective surgery including an extensive muscular resection of the RVOT, minimal resection of the LVOT, and interposition of a graft patch in the RVOT resulted in complete disappearance of the RVOT gradient and a significant decrease to 28 mmHg in the LVOT gradient. During a year follow-up, aortic valvular insufficiency remained clinically stable and the patient was asymptomatic. This is the first case of hypertrophic obstructive cardiomyopathy with predominant RVOT obstruction treated by myectomy and patch graft interpositioning.

Key words: Cardiomyopathy, hypertrophic/surgery; hemodynamics; ventricular outflow obstruction/surgery.

On sekiz yaşında erkek hasta, üç yıldır var olan egzersiz dispnesi, baş dönmesi ve angina yakınmalarıyla başvurdu. Ekokardiyografide sol ventrikülde (SolV), sağ ventrikül (SağV) serbest duvarında ve interventriküler septumda ileri derecede hipertrofi görüldü. Özellikle sağ ventrikül çıkış yolu (SağVÇY) seviyesinde belirgin kas demetleri vardı. Sağ ve sol ventrikül çıkış yolunda (SolVÇY) ölçülen en yüksek basınç gradiyentleri sırasıyla 141 mmHg ve 66 mmHg idi. Hastada ayrıca derece 2 aort yetersizliği saptandı. Bu bulgular transözofageal ekokardiyografi ve kardiyak manyetik resonans görüntüleme ile de doğrulandı. Propranolol ve cibenzolin ile tedaviye rağmen hastanın semptomlarında düzelme ve basınç gradiyentlerinde değişiklik olmadı. Cerrahi tedaviye karar verilerek, SağVÇY için geniş kas rezeksiyonu, SolVÇY için minimal rezeksiyon uygulandı ve SağVÇY seviyesine yama yerleştirildi. Bu tedaviyle SağVÇY'deki gradiyent tamamen kaybolurken, SolVÇY'deki gradiyent 28 mmHG'ye düştü. Hastanın bir yıllık takibi sırasında semptomlar tekrarlamadı ve aort yetersizliğinde klinik kötüleşme olmadı. Sunulan olgu, baskın olarak SağVÇY tıkanıklığı ile kendini gösteren hipertrofik tıkayıcı kardiyomiyopatinin miyektomi ve yama grefti ile tedavi edildiği ilk olgudur.

Anahtar sözcükler: Kardiyomiyopati, hipertrofik/cerrahi; hemodinami; ventrikül çıkış yolu tıkanıklığı/cerrahi.

Right ventricular outflow tract (RVOT) obstruction is seen in 15% of patients with hypertrophic cardiomyopathy.[1,2] Although different therapeutic modalities including surgical myectomy and alcohol septal ablation in addition to medical therapy exist for left ventricular outflow tract (LVOT) obstruction, treatment of RVOT obstruction is not well established. We present a case of hypertrophic obstructive cardiomyopathy (HOCM) with left and right ventricular outflow tract obstruction treated with surgical myectomy and graft patch interposition.

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CASE REPORT

An 18-year-old male was admitted to the emergency department with a 3-year history of exertional dyspnea, dizziness, and angina. He had no other history of medical or surgical disease or family history. On physical examination, he was in good condition with a blood pressure of 90/60 mmHg, heart rate of 66 beats/min, respiratory rate of 21/min, and body temperature of 36 °C. Auscultation showed a thrill and a grade 5/6 harsh, systolic ejection murmur over the apex and a grade 2-3/6 early diastolic murmur over the mesocardiac area.

Basic laboratory tests and cardiac enzyme levels were normal. The chest roentgenogram showed an increased cardiothoracic ratio and diminished pulmonary vascular markings indicating pulmonic obstruction. Electrocardiography showed sinus rhythm with right-axis deviation and right ventricular strain pattern (Fig. 1). Echocardiography revealed advanced hypertrophy of the left ventricle (LV), right ventricle (RV) free wall, and interventricular septum (IVS). There were apparent muscular bundles at the level of the RVOT (Fig 2). Maximal pressure gradients at the RVOT and LVOT were 141 mmHg and 66 mmHg, respectively (Fig. 3). There was grade 2 aortic regurgitation. Transesophageal echocardiography showed severe hypertrophy of the LV, RV, and IVS, and dynamic obstruction of the LVOT and RVOT. Cardiac magnetic resonance imaging confirmed these findings (Fig. 4). The diameters of the main, right and left pulmonary arteries were normal. Cardiac catheterization revealed a peak-to-peak gradient of 100 mmHg at the level of the RVOT and a gradient of 50 mmHg at the level of the LVOT.

Despite treatment with a beta-blocker (propranolol 40 mg) and class Ia antiarrhythmic agent (cibenzoline 300 mg), the patient remained symptomatic with unchanged gradients necessitating surgical treatment. Under cardiopulmonary bypass, deep hypothermia, and complete circulatory arrest, an extensive muscular resection of the RV was performed. However,

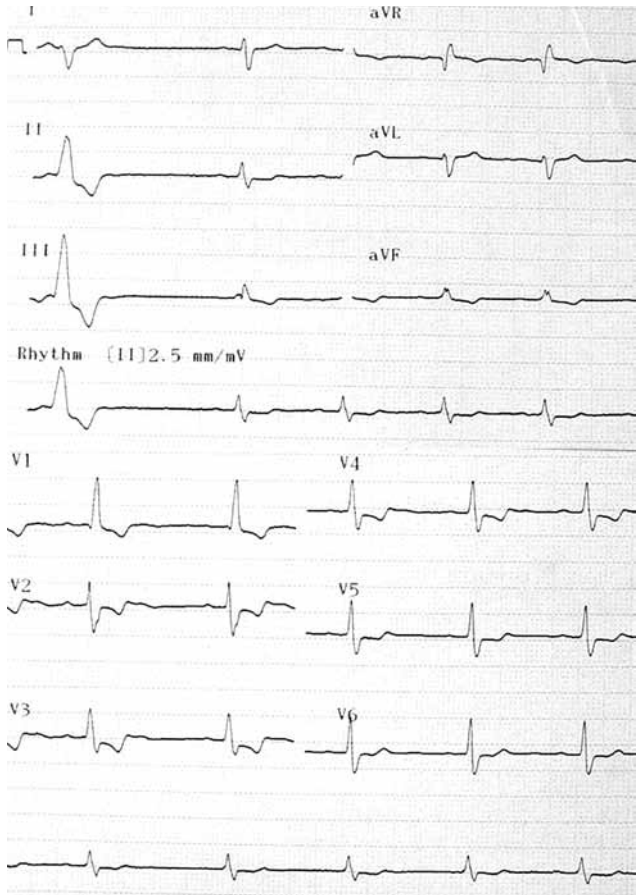

Figure 1. The 12-lead electrocardiogram on admission shows sinus rhythm, bradycardia and inverted T waves in D3, aVf, and V1-V3, and hypertrophy signs in the right ventricle.

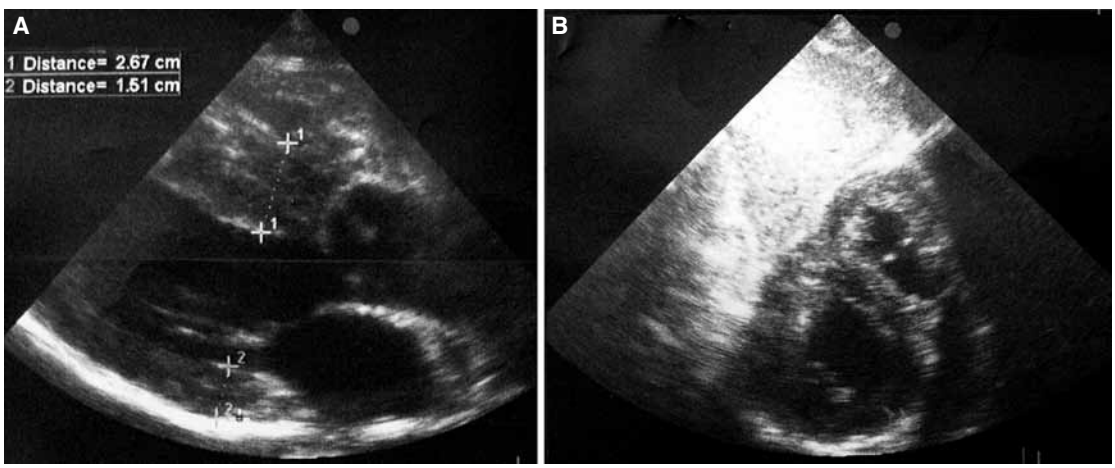

Figure 2. Parasternal **(A)** long-axis and **(B)** short-axis echocardiograms showing biventricular hypertrophy and right ventricle outflow tract obstruction with muscular bundles.

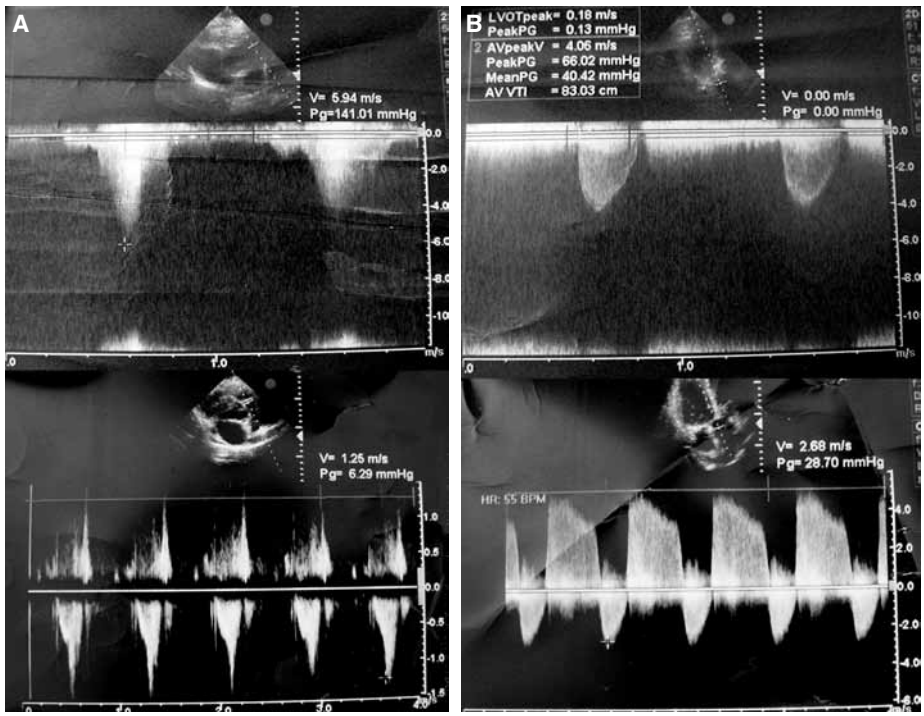

Figure 3. Doppler flow velocity profiles before and after surgery at the levels of the **(A)** right and **(B)** left ventricle outflow tracts.

this did not provide a satisfactory relief of the RVOT obstruction and a 3.5 cm Dacron graft patch was interpositioned (Fig 5). A minimal resection of the LVOT was also performed. The postoperative course was uneventful. Control echocardiograms demonstrated no residual gradient across the RVOT and a gradient of only 28 mmHg across the LVOT. During the follow-up, the diameters of the LV, RV, and IVS decreased progressively. Mild-to-moderate aortic valvular insufficiency remained clinically stable. The patient was asymptomatic at the six-month and one-year follow-ups.

DISCUSSION

Hypertrophic obstructive cardiomyopathy is a complex cardiac disease presenting with distinct pathophysiological characteristics and a wide spectrum of morphologic, functional, and clinical features. The existence of biventricular pressure gradients, as seen in our case, is a severe clinical presentation of HOCM.[3]

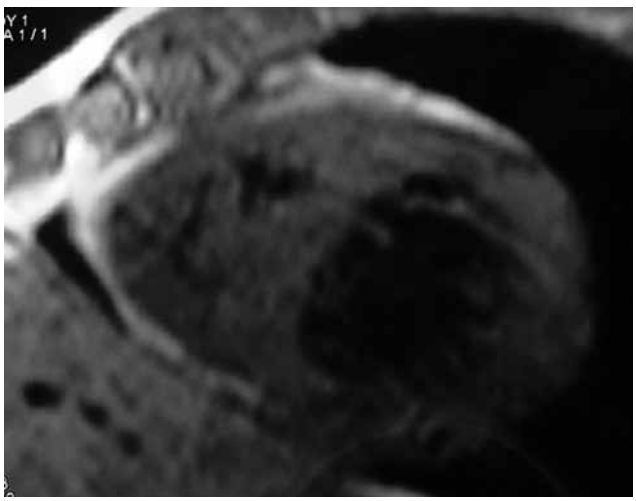

Figure 4. Magnetic resonance scan showing hypertrophy of the biventricular free wall and interventricular septum and apparent muscular bundles occluding the right ventricle outflow tract.

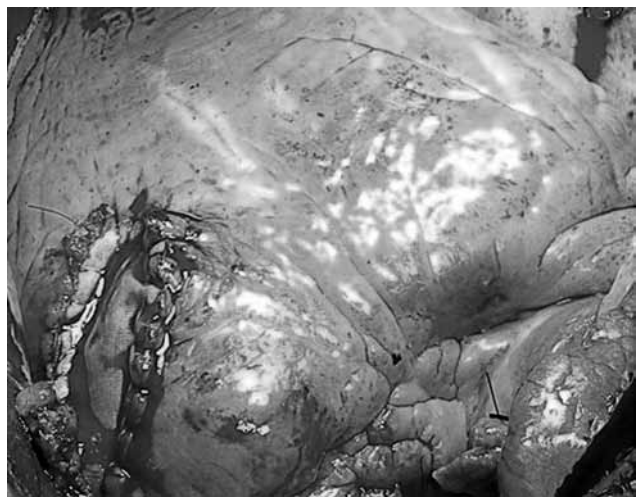

Figure 5. Operative view showing interposition of a Dacron graft patch in the right ventricle outflow tract after muscular bundle resection.

Management options for HOCM accompanied by infundibular stenosis of the RV include surgical removal of hypertrophied muscle bundles, resulting in a decreased gradient and improvement in the symptoms. Surgical relief of infundibular pulmonary stenosis is associated with a very low risk of mortality and morbidity. Reoperation is rarely required and surgery remains the treatment of choice in patients with isolated RVOT obstruction.[4-6]

As an alternative to palliative surgical relief of infundibular pulmonary stenosis, Gibbs et al.[7] treated four patients with right ventricular outflow stent implantation, which resulted in significant improvement in three cases during a mean follow-up of 9.7 months, whereas one patient had mild-to-moderate residual stenosis in the early postprocedure period and required stent enlargement at 17 months because of neoendothelial proliferation within the stent.

Percutaneous transluminal septal myocardial ablation, on the other hand, as an alternative procedure to reduce the LVOT gradient in HOCM may have limited effectiveness due to the anatomic variability of the vascularization of the obstructing septal bulge.[8] Alcohol ablation therapy was also used in the treatment of infundibular hypertrophy in patients with infundibular pulmonary stenosis.[9] Alcohol ablation was applied to the conus branch of the right coronary artery supplying the hypertrophied myocardium of the RVOT which was demonstrated by myocardial contrast echocardiography. Akinesia of the anterior wall of the RVOT was thought as the main mechanism in the relief of obstruction.

Cibenzoline is a class Ia antiarrhythmic drug with remarkably weak anticholinergic effect and is effective in attenuating LVOT obstruction and LV diastolic dysfunction in HOCM.[10,11] Administration of a single oral dose of 200 mg cibenzoline to a patient with biventricular HOCM decreased the pressure gradients in both ventricles and 300 mg daily cibenzoline was effective in maintaining low pressure gradients.[12]

The presence of varying clinical approaches indicates how difficult it is to define precise guidelines for management of patients with HOCM. Thus, it is often necessary to individualize the therapy. Although medical treatment is usually accepted as the first-line therapy, surgical septal myectomy offers substantial long-term hemodynamic and symptomatic benefits in severely symptomatic patients with HOCM. The efficacy of alcohol septal ablation, on the other hand, remains unclear in the treatment of HOCM.

Considering that all these modalities may have different clinical and hemodynamic improvements or complications, the merits and demerits of each therapeutic approach must be balanced when deciding on treatment for LV and RV obstructions. Reduction in the pressure gradients may probably be more satisfactory and long-standing after surgery. In our case, we did not see any clinical and hemodynamic improvement after medical therapy and performed surgical myectomy to reduce hypertrophied muscle bundles especially at the level of the RVOT. This operation was combined with a patch interposition at the level of the RVOT to obtain a satisfactory relief of obstruction. There were no perioperative or postoperative complications and the patient was asymptomatic during a year follow-up.

In conclusion, medically refractory HOCM is still challenging and requires increasing experience with individual patients to define the best effective therapeutic approach.

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