Chapter 2
A Case of Mid-ventricular Obstructive Hypertrophic Cardiomyopathy

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Abstract Mid-ventricular hypertrophic obstructive cardiomyopathy is a rare type of hypertrophic cardiomyopathy that can be accompanied by an apical aneurysm.

Left heart catheterization and continuous-wave Doppler echocardiography revealed a pressure gradient between the apical and basal chambers of the left ventricle in concomitant with other important and prognostic echocardiographic findings.

Hypertrophic cardiomyopathy (HCM) is a genetic cardiac disorder characterized by marked variability in morphological appearance and natural history. The hypertrophic myocardium is frequently confined to the septum or lateral wall of the left ventricle (LV), however, it can also be encountered in the middle or apical regions of the LV myocardium [1–3].

Case Presentation

A 34-year-old woman presented with a history of chest pain and also shortness of breath producing important limitations on her daily activities. She had no history of hypertension, diabetes, and coronary artery disease. Her physical exam was unremarkable.

Transthoracic echocardiography (TTE) revealed normal systolic function and significant concentric LV hypertrophy that was greater in the mid-LV cavity region. There was high LV end-diastolic pressure; normal right ventricle function and normal pulmonary artery pressure (Fig. 2.1).

Our patient had persistent symptoms despite receiving optimized medical management, and a surgical approach was indicated for her. Coronary CT-angiography

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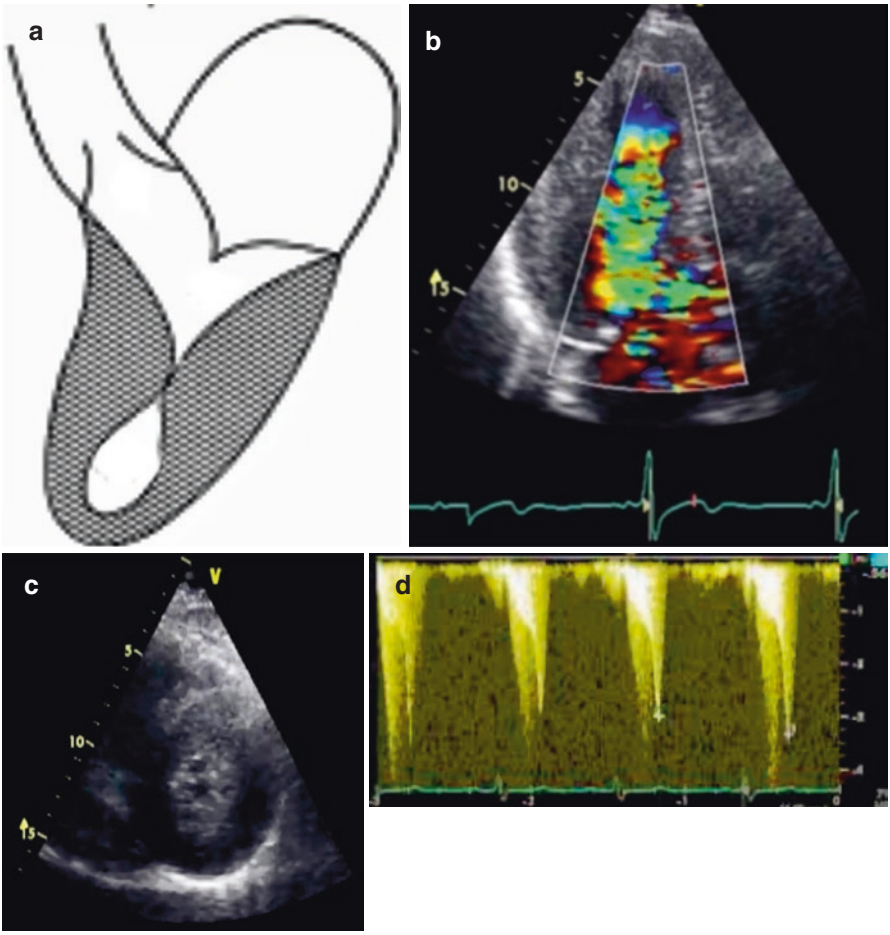

Fig. 2.1 (**a**) Mid-LV cavity hypertrophy, (**b**) Turbulent flow in mid-LV cavity in apical 3-chamber view, (**c**) LV short-axis view with significant hypertrophy, (**d**) High Doppler Flow gradient in mid-LV cavity

was normal; Precise myectomy surgery was done with preservation of papillary muscles and fortunately, no complications occurred after surgery, and her symptoms resolved completely.

In the follow-up beyond 1 month, the patient's NYHA class improved from III to I. Additionally, the medication being used was meaningfully reduced postoperatively. New TTE was performed after the surgery, and it confirmed the disappearance of the mid-ventricular pressure gradient.

Discussion

It might involve principally the proximal septum, or there can be diffuse LV hypertrophy. However, there are other forms, such as mid-ventricular and also apical hypertrophy. In the mid-LV cavity HCM pattern, there may be an intraventricular pressure gradient that creates an obstruction at the level of the papillary muscles, which may lead to apical myocardial infarction and an apical aneurysm. Mid-ventricular obstruction is defined as a ventricular gradient ≥30 mm Hg [1, 4] (Fig. 2.2).

Patients with mid-ventricular HCM are more symptomatic than HCM patients without obstruction or LV outflow tract obstruction. Moreover, the incidence of sudden death and arrhythmic events is relatively high in this pattern of HCM [3, 4]. Minami and colleagues, by using multivariate models, demonstrated that mid-ventricular obstruction is an independent determinant of HCM-related cardiac death [1, 5].

Fig. 2.2 Mid-LV cavity obstruction, apical four-chamber view, (white arrow)

The pharmacologic management of HCM, specially in obstructive variants, is based on β blockers, calcium-channel blockers, and disopyramide. This treatment is effective in many patients, however, those with mid-ventricular hypertrophy have worse results in terms of symptom relief. In these patients, the standard treatment is surgical resection of the hypertrophied portion of the ventricle or even cardiac transplantation [3–5].

Conclusion

Our patient had a mid-ventricular HCM with symptoms refractory to a optimize medical therapy. In addition, the patient's hypertrophy obstructed the cavity and created a significant intraventricular gradient. After consultation with the heart team, the surgical approach was chosen for her.

The procedure was performed without complications, and no damage in the mitral valve apparatus or disturbance in the left bundle branch was found too.

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