Hypertrophic cardiomyopathy associated with mid-ventricular obstruction and apical aneurysm

A 31-year-old woman was referred to our hospital because of dyspnea and palpitation upon effort. On physical examination, there was a grade 3/6 systolic ejection murmur most prominent in the 4th intercostal space, left sternal border. Chest X-ray revealed a normal cardiothoracic ratio and no pulmonary congestion. Her electrocardiogram was consistent with atrial fibrillation rhythm and left ventricular hypertrophy with high voltage on precordial leads and ST-segment depression on V4–V6. Echocardiography disclosed asymmetric septal hypertrophy (the interventricular septal thickness was 22 mm and the posterior wall thickness was 13 mm) (Figs. A, B) and mid-ventricular obstruction during systole and an apical aneurysm. Continuous wave Doppler recording revealed a peak velocity of 4.6 m/s equivalent to the pressure gradient of 84 mmHg between the distal and proximal portions within the left ventricle (Figs. C, D, Video 1 and Video 2*). Cardiac catheterization showed angiographically normal coronary arteries. A left ventriculogram revealed severe left ventricular hypertrophy with mid ventricular total obstruction-like hour-glass appearance at systole and a large apical aneurysm (Figs. E, F; Video 3 and Video 4*). A peak-to-peak intraventricular pressure gradient of 75 mmHg was documented during pullback from the apical high pressure chamber (185 mmHg) to the basal low pressure chamber in the left ventricle (110 mmHg). Because of severe symptoms of progressive heart failure despite medical treatment, the patient underwent surgical myectomy on the operative day, the patient suffered sudden cardiac death that could not be resolved with resuscitation.

Figures—Transthoracic echocardiograms in the parasternal (A) long and (B) short axis views showing left ventricular hypertrophy, (C) intraventricular pressure gradients by Continuous-wave Doppler recordings and (D) apical aneurysm. Left ventriculography showing (E) mid ventricular obstruction and (F) large apical aneurysm. *Supplementary video files associated with this case can be found in the online version.