A 57-year-old man presented with exertional dyspnea. Cardiac examination was normal. Transthoracic echocardiogram showed asymmetrical septal hypertrophy with maximum thickness of 22 mm at the basal septum (Figure 1). There was no significant left ventricular outflow tract obstruction at rest. Parasternal short axis (PSAX) view at the level of the mitral valve (MV) leaflets demonstrated a double orifice mitral valve (DOMV), separated by an incomplete bridge with two asymmetrical orifices (Figure 2A, Movie 1). PSAX view at the level of the MV annulus showed a normal MV opening with an indentation on the anterior
leaflet characterizing the “lovely heart” sign (Figure 2B). Apical four-chamber (A4-C) view demonstrated a separate subvalvular apparatus for each orifice (Figure 3). A4-C view with color flow Doppler demonstrated two regurgitant jets through the two asymmetrical orifices of the DOMV amounting to mild mitral regurgitation (Figure 4). There was no significant gradient across the MV.

Various anomalies of the MV apparatus have been described with hypertrophic cardiomyopathy (HCM). However, DOMV has not been described in a case of HCM previously. DOMV is a rare congenital anomaly which can occur either isolated or in association with other congenital heart defects with varied presentations, ranging from symptomatic severe stenosis or regurgitation to asymptomatic ones. Herein, we report the first case of HCM in combination with DOMV.

Figure 3. Apical four-chamber view showing separate subvalvular apparatus (yellow double arrow) for each mitral valve orifice. LA: left atrium, LV: left ventricle.

Figure 4. Apical four-chamber view with color Doppler showing two jets of mitral regurgitation. LA: left atrium, LV: left ventricle, MR: mitral regurgitation.
SUPPLEMENTARY MATERIAL

Movie 1
Parasternal short axis view at the level of the mitral leaflets showing two asymmetrical orifices.

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REFERENCES

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