

A rare manifestation of a known disease: Severe non-obstructive hypertrophic cardiomyopathy

Bilinen bir hastalığın nadir bir bulgusu: Ağır nonobstrüktif hipertrofik kardiyomiopati

Yalçın Velibey

Ahmet Ekmekçi#

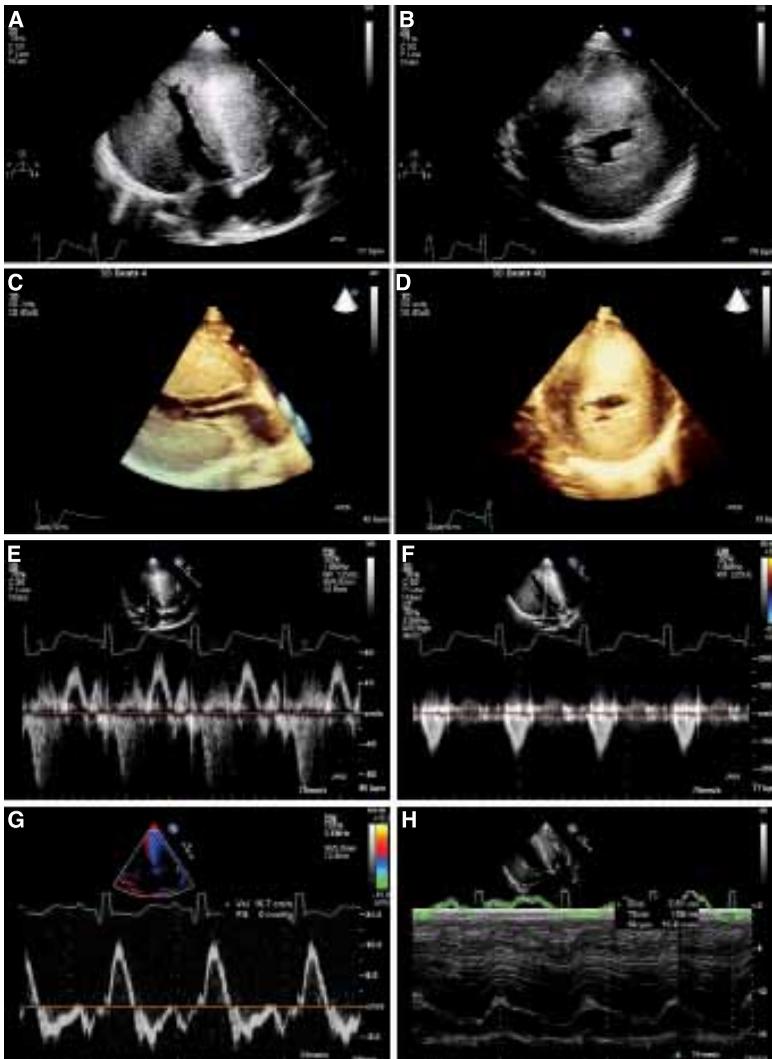
Eyüp Tusun#

Mehmet Eren#

Department of Cardiology,
Uskudar State Hospital, Istanbul;

Department of Cardiology,
Siyami Ersek Thoracic and
Cardiovascular Surgery
Center, Training and
Research Hospital, Istanbul

A 17-year-old man was admitted to the cardiology department complaining of chest pain and shortness of breath. At age 15, he had been admitted to hospital for the first time and was at that time diagnosed with hypertrophic cardiomyopathy. Due to increased septal thickness (>30 mm) and syncope episodes, an implantable cardioverter defibrillator was implanted at age 16. He had been taking 50 mg of metoprolol succinate once daily since age 15. At admission, arterial blood pressure and heart rate were 110/70 mmHg and 80 bpm respectively. Clinical examination of the respiratory system was normal. A chest X-ray revealed cardiomegaly, and an electrocardiography (ECG) demonstrated left ventricular hypertrophy by voltage. A two- and three-dimensional transthoracic echocardiography revealed severe hypertrophic cardiomyopathy (interventricular septum, 54 mm; posterior wall, 44 mm; and LV end-diastolic diameter, 29 mm) with an ejection fraction of 60% (Figure 1A-D, Video 1*). Right ventricular hypertrophy (9 mm) with normal right ventricle systolic functions were also noted. There was no left or right ventricular outflow tract gradient at the mid-ventricular level at rest or after the Valsalva maneuver (Figure E-H). A forty-eight-hour ambulatory ECG recording revealed frequent premature ventricular contractions. Echocardiographic screening of family members was performed, but no evidence of hypertrophic cardiomyopathy was found. The patient was conducted for the transplantation program and discharged with optimal medications (100 mg of metoprolol succinate once daily).



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Figures— Two-dimensional transthoracic echocardiographic views of severe hypertrophic cardiomyopathy. (A) Apical four chambers view; (B) parasternal short axis view. Three-dimensional transthoracic echocardiographic views of the severe hypertrophic cardiomyopathy. (C) Parasternal long axis view; (D) parasternal short axis view. (E) CW Doppler echocardiography showed normal mitral valve inflow (E/A ratio of 1.2) with a normal deceleration time (DT=170 ms). (F) CW Doppler echocardiography showed no left ventricular outflow tract gradient at rest or after the Valsalva maneuver. (G, H) Normal right ventricle systolic functions of the lateral tricuspid annulus as demonstrated through pulsed tissue Doppler. *Supplementary video files associated with this presentation can be found in the online version of the journal.

