

Latent Obstructive Hypertrophic Cardiomyopathy: Is Echocardiography Enough?

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Case Report

We present a clinical case of a 51 year-old Caucasian male patient referred for exercise stress test because of non-progressive exertional chest pain. His past medical history was unremarkable. Physical examination was negative for specific cardiovascular abnormalities. His resting 12-lead electrocardiogram (ECG), made in a lying position, had terminal T-wave inversion in the precordial leads (Figure 1, A). M-mode and two-dimensional echocardiographic exam was reported as normal. An exercise ECG stress test was then performed. By this time, repolarization abnormalities were no longer present in standing resting ECG and exercise test was positive for ischemia, as the patient referred similar exertional chest pain with simultaneous near-3mm ST segment depression (Figure 1, B). A single photon emission computed tomography was requested attending to late onset of symptoms and ECG changes (4th stage at Bruce protocol), good exercise workload and immediate ECG normalization at recovery. Stress perfusion suspected of an inferior defect (Figure 1, C) and a coronary angiography was attempted, negative for major vessel disease.

In attempting to achieve diagnosis, a complete transthoracic echocardiogram was performed. It showed an asymmetric mild left ventricle (LV) hypertrophy: LV mass index 110g/m² by cubed formula, with a ratio of septal/posterior wall thickness of 1.2. Nevertheless, in parasternal short axis view, a 19mm maximum wall thickness was measured at end-diastole in the inferior interventricular septum at the level of the papillary muscles and LV mass by area length was determined at 123g/m². Both papillary muscles were mildly anteriorly displaced and there was a trace of systolic anterior motion of the anterior mitral valve leaflet with a mild protosystolic mitral regurgitation. There was no intraventricular or outflow tract obstruction, even after Valsalva maneuver and in standing position. Global LV systolic ejection fraction was preserved with normal filling pressures albeit reduced mitral annular velocities at tissue Doppler examination. Right ventricle did not seem to be involved. (Figure 1, D-I, Clip 1, 2, 3).

Keywords

Cardiomyopathy, hypertrophic; echocardiography; cardiac magnetic resonance.

Deformation analysis under speckle tracking algorithm (60 and 82fps) was performed to evaluate both two-dimensional longitudinal and radial strain (at apical and parasternal views, respectively). LV torsion and twisting velocity was also assessed (peak torsion, peak torsion rate and time of onset untwisting velocity). Average global longitudinal strain was assessed as normal (-19.9%) but there were some asymmetry in its distribution. In fact, the basal lateral wall had a positive pre-systolic longitudinal strain and both mid septal and mid lateral wall had reduced systolic longitudinal deformation (Figure 1, J, Clip 4). At mid-LV short axis level, radial deformation (thickening) was equally reduced at inferior septal and inferior wall mid-segments, matching hypertrophic areas (Clip 5). Peak torsion was determined at 18.05° (Figure 1, K).

The patient also underwent an exercise stress echocardiography to assess the development of dynamic intraventricular gradient, which could possibly explain the symptoms. A significant (>30mmHg) peak intraventricular gradient at the 3rd stage of Bruce protocol was detected, apparently not involving the LV outflow tract, with no worsening of mitral regurgitation (Figure 1, L). These findings were simultaneous to chest pain development. The patient was placed under beta-blockers, with complete resolution of complaints. The 24-hour ECG Holter was negative for ventricular arrhythmias.

In order to confirm the diagnosis and to assess the presence of fibrosis, a Cardiac Magnetic Resonance (CMR) was performed. Steady state free precession (SSFP) cine images revealed asymmetric LV hypertrophy, mainly involving the interventricular septum and inferior wall (Indexed LV mass: 175g/m²; maximum LV wall thickness: 17mm at mid inferior septum). The right ventricle was not hypertrophic and biventricular ejection fraction was normal (Clip 6, 7). From tagging, qualitative analysis showed abnormal deformation at the inferior septum and inferior wall (Figure 2). There was no delayed enhancement after gadodiamide injection.

Discussion

We presented the case of an obstructive form of hypertrophic cardiomyopathy in which the diagnosis could only be brought along after a careful and detailed echocardiographic study: 1. LV mass was increased on two-dimensional evaluation, as confirmed by CMR. 2. Regional LV thicknesses were high above normal on two-dimensional echo and CMR measurements¹. 3. LV papillary muscles were abnormally displaced. 4. Tissue Doppler-derived velocities were abnormally low in the inferior septum. Besides this, two-dimensional strain evaluation

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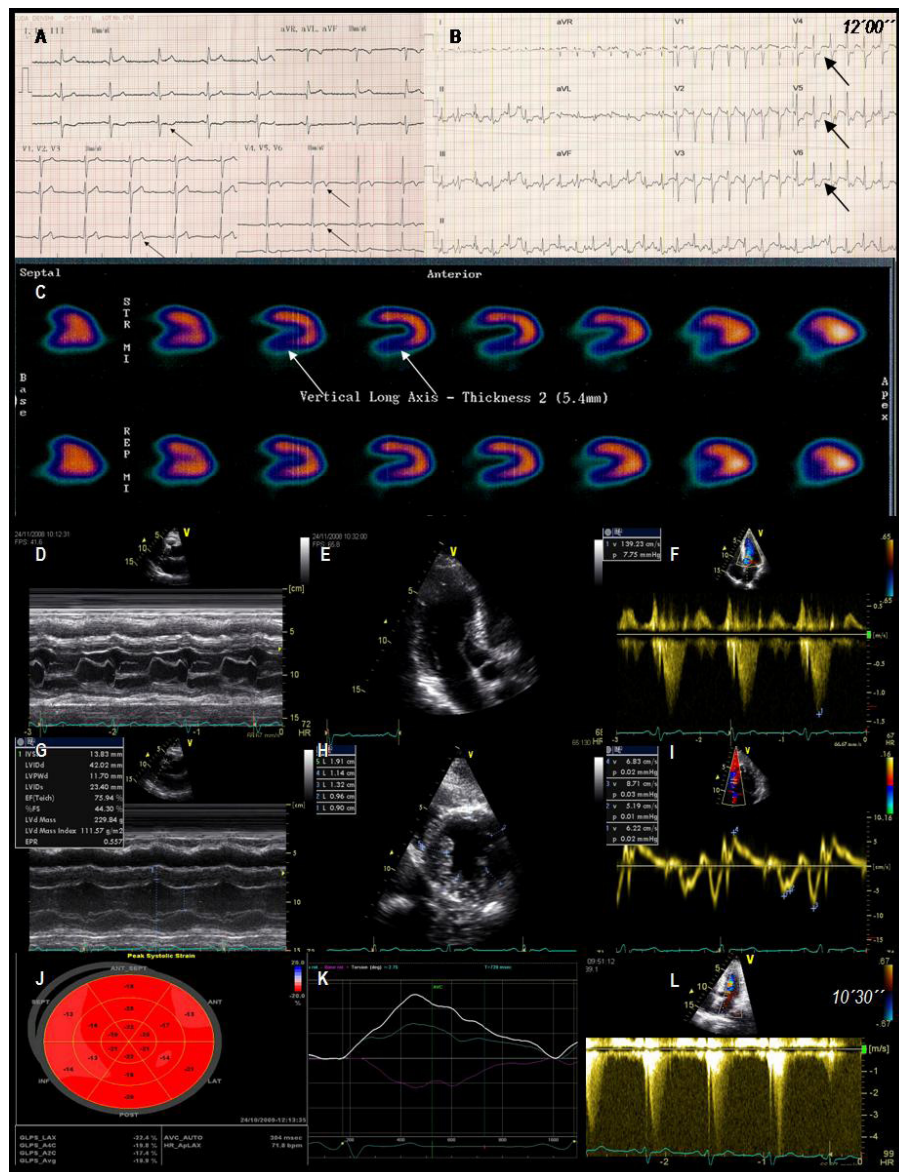


Figure 1 - (A) Decubitus twelve lead electrocardiogram with abnormal repolarization in DIII and terminal T-wave inversion in the precordial leads (arrows). (B) Standard Bruce treadmill protocol showing an almost 0.30 mV ST segment displacement (arrows). (C) ^{99}Tc -labelled scintigraphy showing a mild, reversible, inferior wall defect (arrows). (D-L) Transthoracic echocardiography; (D) M-mode image with an incomplete systolic anterior motion of the anterior mitral valve leaflet (G) LV mass index (Teichholz). (E, H) Two-dimensional images showing chordal elongation and mid ventricle end-diastolic thicknesses. (F) Spectral pulsed wave Doppler with no significant LV outflow tract gradient. (I) Tissue Doppler evaluation in mitral annulus, showing a reduced S' wave velocity (7cm/s) and an E'/A' ratio below 1. (J) "Bull's eye" global longitudinal strain assessed as normal with preserved basal to apical gradient. (K) Torsion. Time to peak torsion: 462ms; apical rotation rate: 21.5%/s, basal rotation rate:- 17.7%/s. (L) Treadmill exercise echocardiography with significant intraventricular gradient development (almost 80mmHg).

Clip 1 - Two-dimensional apical four chamber view.

Clip 2 - Two-dimensional mid-ventricle short axis view.

Clip 3 - Modified apical four chamber view with colour flow Doppler.

Clip 4 - Longitudinal strain at apical four chamber view.

Clip 5 - Radial strain at mid ventricle level.

Clip 6 - Short axis SSFP cine sequences.

Clip 7 - Long axis SSFP cine sequences.

Case Report

by speckle tracking allowed us to identify regional systolic dysfunction, as already described, mainly involving hypertrophic segments². Furthermore, as recently reported, both hypertrophic cardiomyopathy and hypertensive heart disease patients have enhanced peak torsion and this could be suggested in this case³.

In spite of this, we needed multiple imaging modalities for diagnostic confirmation and symptomatic correlation. Actually, we had to exclude coronary artery disease and we performed CMR to confirm the diagnosis and obtain prognostic information⁴. Nevertheless, we should emphasize that echocardiography should be fully explored in its different modalities (regional function, flow and myocardial velocities, deformation) when this diagnosis is in suspicion, as this technique may provide key structural and functional features. Indeed, we should perform exercise echocardiography when in doubt for symptomatic correlation in the absence of significant outflow tract and intracavitary gradient at rest, even after load changing maneuvers.

At last, we also found this case peculiar for the data provided by different imaging techniques.

These findings were not only important for supporting and confirming the diagnosis but they also matched correctly (Figure 2), particularly concerning morphological two-dimensional echo and CMR data. Furthermore and in spite of its lower spatial resolution, stress perfusion by SPECT seemed to be impaired in the segments subsequently identified as hypertrophic. This could even support the concept of abnormal perfusion and impaired coronary flow reserve in HCM⁴.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

Sources of Funding

There were no external funding sources for this study.

Study Association

This study is not associated with any post-graduation program.

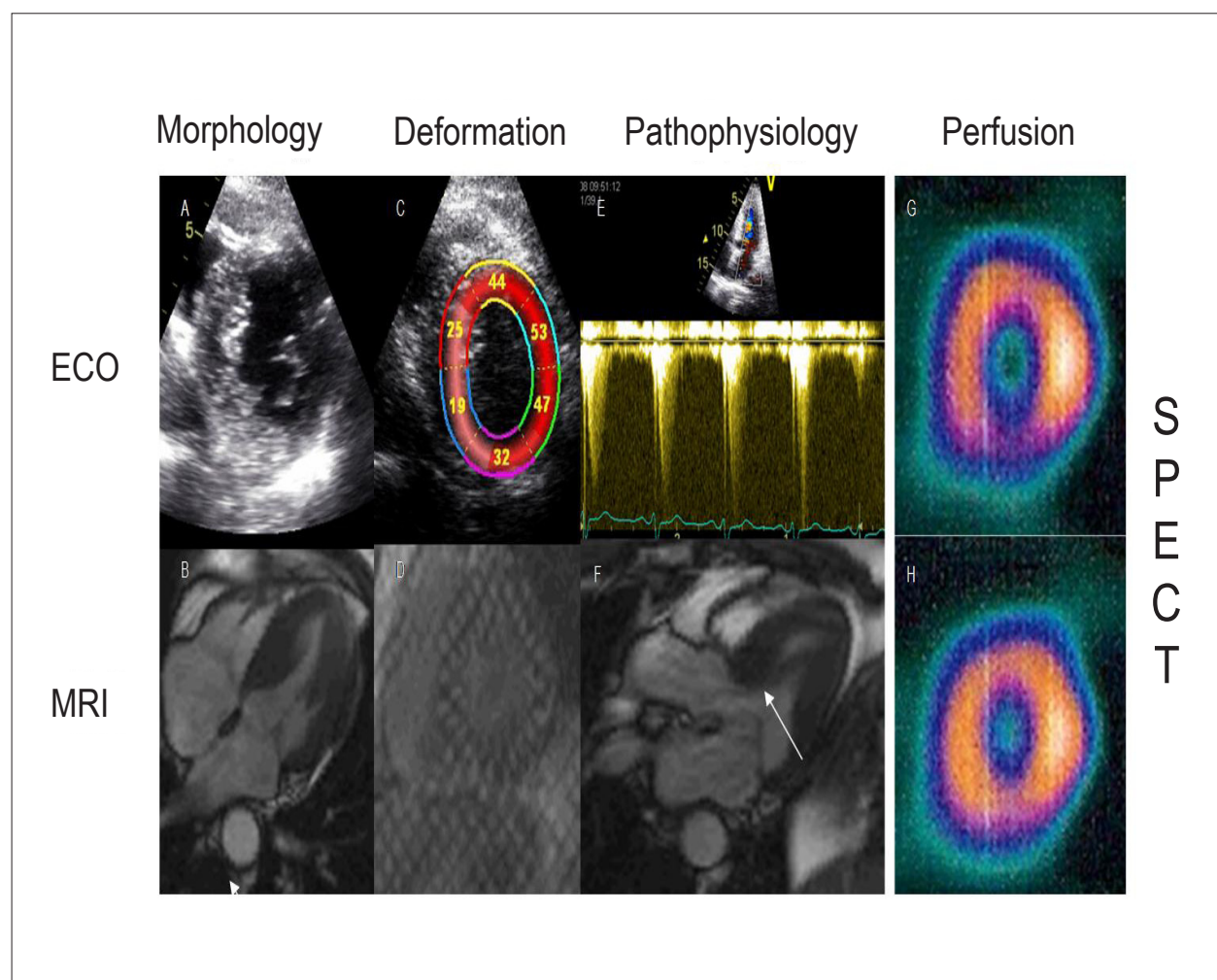


Figure 2 - Echocardiography (A-two-dimensional echo, C-radial strain, E- spectral Doppler) and MRI (B-SSFP, D-tagging, F-SSFP) derived frames showing similar findings from these two different imaging techniques, matching morphology, tissue behaviour and pathophysiology (arrow - slight intraventricular signal void meaning turbulent flow). Scintigraphy: stress (G) and rest (H) perfusion.

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