Incremental value of cardiovascular magnetic resonance imaging in the differential diagnosis of hypertrophic cardiomyopathy

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A 71-year-old male was admitted to our tertiary care centre for further differential diagnosis due to hypertrophic cardiomyopathy of unknown origin. Clinically, the patient showed signs of heart failure with cachexia, progressive dyspnoea (NYHA class III), peripheral oedema, and bilateral pleural effusion. The electrocardiogram revealed atrial fibrillation, right bundle branch block, and repolarization changes. Echocardiography demonstrated concentric left ventricular (LV) thickening with a granular ‘sparkling’ wall appearance without obstruction of the LV outflow tract and normal LV ejection fraction (Panels A and B). Initial working diagnosis were infiltrative cardiac diseases (glycogen storage diseases, sarcoidosis, amyloidosis, Morbus Fabry, haemochromatosis) and the differential diagnosis hypertrophic non-obstructive cardiomyopathy.

For further tissue characterization, late gadolinium-enhancement (LGE) cardiovascular magnetic resonance (CMR) imaging was performed. CMR confirmed biventricular thickening, a small pericardial effusion, significant bilateral pleural effusion, and normal ejection fraction (Panel C). LGE imaging revealed a characteristic global sub-endocardial gadolinium uptake throughout the myocardium of both ventricles (Panels D and E). Therefore, the diagnosis of cardiac amyloidosis was presumed and LV biopsy was performed. Anatomic pathology showed a diffuse Congo-red positivity (Panel F) with apple-green birefringence under polarized light, confirming the diagnosis of cardiac amyloidosis. Additional staining revealed immunoglobulin-light-chain amyloidosis with subsequent haematologic treatment including chemotherapy with melphalan plus dexamethasone. One month after initiation of the chemotherapy, the condition of the patient stabilized with current dyspnoea (NYHA class II).

This case underlines the incremental value of CMR for further differential diagnosis in hypertrophic cardiomyopathy. Accumulation of amyloid in the myocardial interstitium results in LGE, often with a predominant diffuse, global and sub-endocardial distribution that matches the distribution of amyloid on histology. This is associated with substantial alterations in gadolinium-chelate kinetics, with a higher affinity of the contrast medium to amyloid-loaden myocardium, and hence a relative increase in washout kinetics from normal myocardium. In addition, CMR provides direct information on the spatial distribution of cardiac amyloid in the myocardium and thus might be useful to monitor progression or regression of cardiac amyloid depositions.

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