

The Value of MRI in Diagnosing Cardiac Amyloidosis

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Background: Extracellular space infiltration and direct toxicity of amyloid proteins mediate heart wall stiffening, conduction disturbances, contractile dysfunction and arrhythmia in cardiac amyloidosis. Recent studies suggest improved outcome in this lethal disease with therapies suppressing production of the abnormal protein. Diagnosis of cardiac amyloidosis is complicated by high variability in clinical presentation and inadequate performance of various diagnostic measures. A characteristic diffuse subendocardial pattern of late gadolinium enhancement (LGE) using MRI was reported to characterize cardiac amyloidosis.

Methods and Results: Our cardiac amyloidosis registry comprises 29 patients, 90% of light chains (AL) type. We assessed the sensitivity and specificity of the characteristic LGE for diagnosing cardiac amyloidosis within cardiomyopathy patients undergoing MRI in Sheba Medical Center. Cardiac amyloidosis was diagnosed by endomyocardial biopsy or the presence of typical echo-doppler pattern and a positive Congo-Red in another tissue. Between 2007 and 2009, 13 of our patients with cardiac amyloidosis underwent MRI. The sensitivity of MRI for the diagnosis of cardiac amyloidosis was 85%. Among patients with other cardiomyopathies, such an MRI pattern was encountered only in 1 patient with familial dilated cardiomyopathy caused by LMNA mutation, giving a specificity of 99%.

Conclusion: MRI may facilitate the diagnosis of cardiac amyloidosis. While a negative study cannot rule out cardiac involvement, a positive result is highly suggestive of the disease. We suggest that in a suitable patient, a combination of monoclonal gammopathy and characteristic MRI findings are sufficient to establish the diagnosis and helps initiate early chemotherapy.