PATTERNS OF LATE GADOLINIUM ENHANCEMENT PREDICT SURVIVAL IN CARDIAC AMYLOIDOSIS: A SYSTEMATIC REVIEW OF 95 CASES WITH AL OR TTR TYPE

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Background: Cardiac MRI (CMR) is increasingly used to investigate patients with suspected amyloidosis. Circumferential subendocardial late gadolinium enhancement (LGE) has been reported to be typical in AL amyloidosis, whereas, transmural LGE has been associated with transthyretin amyloidosis (ATTR).

Methods: We studied the CMR LGE pattern in 95 patients who had been referred to the UK National Amyloidosis Centre (NAC) between 2007 and 2011.

Results: The mean age was 69.2 ± 11.3 years with male predominance (73%). Amyloid type was AL in 39 patients (41%) and ATTR in 56 patients (59%). LGE was evident in all ATTR patients and 36 AL patients (92%, p<0.01). Fifty-three ATTR patients (95%) demonstrated ≥ 1 transmural segment, compared to 12 AL patients (31%, p<0.01). Circumferential transmural LGE was specific to ATTR (6 patients (10.7%), p<0.01). Right ventricular (RV) LGE was present in all ATTR patients but only 26 AL patients (67%, p <0.01). Circumferential subendocardial LGE was present in only 8 AL patients (21%) and 2 ATTR patients (3%, p <0.01). Median survival was significantly reduced in AL patients (24 vs 36 months, p<0.01). However, the presence of transmural LGE in any segment was associated with improved survival in AL amyloid (p=0.04).

Conclusion: A transmural pattern of LGE is more suggestive of ATTR but only the absence of RV LGE in one-third of AL patients reliably distinguished the two types of amyloid. Transmural LGE in any segment was associated with prolonged survival in patients with AL amyloid.