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Prevalence, risk factors and correlation with cardiac involvement of carpal tunnel syndrome in amyloidosis

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Background

Carpal tunnel syndrome (CTS) is one of the most common clinical manifestations of TTR-related amyloidosis, both hereditary (ATTR), and wild type (senile systemic amyloidosis, SSA) and often precedes cardiac symptoms. The exact prevalence of CTS in light-chain amyloidosis (AL), ATTR and SSA is not known. We therefore aimed to establish prevalence, risk factors and possible association with cardiac involvement in patients with TTR-related and AL amyloidosis.

Methods

We retrospectively analyzed clinical and instrumental (ECG and echocardographic) findings of 260 patients with TTR-related, and 175 with AL amyloidosis evaluated at our Centre between 1990 and September 2013.

Results

Prevalence was 35% in TTR-related amyloidosis (35% in ATTR and 32% in SSA) and 8% in patients with AL ($p < 0.001$). Among TTR patients, CTS was more frequently associated with cardiac involvement (76% vs. 42%; $p<0.0001$) as reflected by the presence of pathological ECG and echocardiogram. Moreover, CTS manifested 9 years before the onset of cardiac symptoms. Among patients with cardiomyopathy with/without CTS there were no significant clinical/instrumental differences. At univariate analysis male gender and genotype were not associated with CTS.

Conclusion

CTS is specifically associated with TTR-related (but not AL) amyloidosis independently from patient gender. In TTR-related amyloidosis, CTS is more frequently associated with cardiac involvement, even though patients with cardiomyopathy with/without CTS have a comparable clinical/instrumental profile. CTS precedes cardiac symptoms onset by 9 years and this awareness is important for an early diagnosis of amyloidotic cardiomyopathy.

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