Natural history of sporadic Inclusion Body Myositis – an observational longitudinal study

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Background/objective: There have been few studies prospectively assessing the clinical characteristics of sporadic inclusion body myositis (IBM) and its progression. Our aim was to assess prospectively the clinical features and functional impact of IBM.

Methods: Patients were diagnosed with either probable or definite IBM, according to the Griggs' criteria (Griggs RC, et al. Ann Neurol 1995;38:705-713) or with clinically defined IBM, according to the Medical Research Council criteria (Hilton-Jones D, et al. Neuromuscul Disord 2010;20:142-147). Clinical data, manual muscle testing (MMT), quantitative muscle testing (QMT) of quadriceps extensors with HUMAC Norm CSMi™ dynamometer and IBM functional rating scale (IBM-FRS) were collected according to a standardised protocol (IBM-Net) at baseline (n=51) and one-year follow-up (n=23, QMT performed in a subgroup of 13 patients). The responsiveness to change of MMT, QMT and IBM-FRS was assessed by calculating the standardized response mean (SRM). Cox-regression analysis was performed to estimate the effect of sex, age at disease onset and previous or current treatment with steroids or immunosuppressants on the time to using a walking stick. Time to using a walking stick was modelled using a Kaplan-Meier curve.

Results: Mean age at disease onset was 58 years. After a median time of 7 years of disease, 63% of patients had lost independent walking ability and needed assistive devices. MMT, IBM-FRS and quadriceps QMT significantly declined after one year (by 5.2%, 13.8% and 27.9%, respectively). QMT of the quadriceps muscle (SRM=1.8) and IBM-FRS (SRM=1.3) were the most sensitive measures of disease progression. Disease onset after 55 years of age (HR=4.1; 95% CI=1.7, 9.8; p=0.001), but not sex or treatment, was predictive of a shorter time to requirement of a walking stick. We detected no differences in disease presentation/progression between clinically and pathologically defined IBM patients.

Conclusions: IBM is a disabling myopathy with prominent involvement of muscle groups essential for activities of daily living. Targeted markers of disease progression such as quadriceps QMA and IBM-FRS could prove helpful as outcome measures in future therapeutic trials in IBM. Onset of the disease after 55 years of age was predictive of a shorter time to using a walking stick.

Conflicts of interest: None declared.

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