Abstract 65

MACULAR INNER NEURODEGENERATION MAY PREDICT THE RESPONSE TO IDEBENONE IN PATIENTS WITH LEBER'S HEREDITARY OPTIC NEUROPATHY

Oral

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Purpose:

To assess the relationship of demographics, clinical characteristics and structural optical coherence tomography (OCT) findings to long-term visual outcomes in patients with Leber's hereditary optic neuropathy (LHON) treated with idebenone. Design: Retrospective cohort study.

Methods:

A total of 17 participants (34 eyes) with LHON treated with idebenone within 1 year after disease onset and 24 months of regular follow-ups were retrospectively enrolled. At baseline, structural OCT volume scans of the macula and optic nerve were reviewed to measure metrics reflecting neuronal loss (macular ganglion cell complex (GCC) and peripapillary retinal nerve fiber layer (RNFL) thicknesses). Stepwise multiple regression analyses were computed to assess associations between final best-corrected visual acuity (BCVA) at 2 years and change in BCVA from baseline at 2 years as dependent variables with demographics, clinical characteristics and structural OCT metrics at baseline.

Results:

The BCVA was 1.6±0.8 LogMAR (Snellen VA of ~ 20/800) at baseline (visit before the initiation of treatment) and 1.0±0.7 LogMAR (Snellen VA of 20/200) at the 2- year follow-up visit ($p<0.0001$). Mean±SD change in BCVA from baseline at 2 years was -51.9±35.9%. In multivariable analysis, the strongest associations with final BCVA were with baseline BCVA ($p=0.012$), superior macular GCC thickness ($p=0.044$), superotemporal macular GCC thickness ($p=0.010$), and inferotemporal macular GCC thickness ($p=0.015$). Similarly, the strongest associations with delta BCVA were with superior macular GCC thickness ($p=0.045$), superotemporal macular GCC thickness ($p=0.047$), and inferotemporal macular GCC thickness ($p=0.030$).

Conclusions:

We identified OCT biomarkers associated with long-term (i.e. 2-year) visual outcomes in patients with LHON treated with idebenone therapy in the first year after disease onset. Thinning of the GCC in the superior and temporal parafoveal regions was associated with worse long-term visual outcomes in these patients