第24回 IBS セミナー

（共催）名古屋市立大学医学会

Institute of Brain Science
Graduate School of Medical Sciences
Nagoya City University

Exploring functional similarities between neurodegeneration in progressive myoclonus epilepsies and the aging brain

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The term "progressive myoclonus epilepsies" (PME) refers to a category of conditions characterized by epileptic seizures that begin in adolescence. In addition to seizures, PME patients exhibit a progressive deterioration of neurological features such as ataxia, muscular atrophy, dementia, psychosis, and hallucinations. Interestingly, these neurological disorders are also widespread in older people, which raises the possibility of a shared underlying mechanism for neurological deficiencies. Most PMEs are loss-of-function mutations, and the gene products are known to participate in pathways that mediate stress responses. According to our hypothesis, the PME-related genes are neuroprotective, their level of expression in the brain declines with aging, and their gradual loss may result in neurodegeneration and neurological impairments. On the other hand, we also proposed that some of the typical neurological deficiencies found in PMEs are brought on by aging. Using mice models, we investigated this theory and showed that the PME gene might contribute to brain aging.

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