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Predaval bo

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PATHOLOGY AND CURRENT MOLECULAR CLASSIFICATION OF ALS/FTD

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POVZETEK PREDAVANJA

Amyotrophic lateral sclerosis (ALS) is the major motor neurone disorder. The hallmark features are progressive, irreversible motor neuron loss leading to denervation atrophy of muscles and death usually within 5 years of disease onset. The hallmark proteins of the pathognomonic inclusions are either SOD-1, TDP-43 or FUS; rarely the disease is caused by mutation of the respective genes. Frontotemporal lobar degenerations (FTLDs) are genetically, neuropathologically and clinically heterogeneous dementias presenting with three major clinical syndromes dominated by behavioural, language and motor disorders, respectively. The characteristic aggregate-forming proteins in non-tau FTLDs are TDP-43 and FUS. It has been known for long that frontotemporal dementia (or less severe forms of cognitive impairment) may coexist with ALS. Recent discoveries in genetics (e.g. *C9orf72* mutation) and the subsequent neuropathological characterisation have revealed remarkable overlap between ALS and non-tau FTLDs also on molecular level indicating common molecular pathways in pathogenesis. After an historical overview we demonstrate and compare the macroscopic and microscopic appearances and molecular characteristics with emphasis on genetic background, neuroanatomical distribution and morphology of abnormal protein aggregates (and their possible association with specific mutations). The clinico-pathological classifications and correlations are also discussed.

Gostitelj je prof. dr. Boris Rogelj.
Predavanje bo v angleškem jeziku.