A COMPARATIVE ANALYSIS OF SPINAL CORD PATHOLOGY IN 4R-TAUOPATHIES: PPND, PSP AND CBD

K. Markopoulou1,2, J. Slowinski1,3, D.W. Dickson4, Z.K. Wszolek1

1Neurology, Mayo Clinic Jacksonville, Jacksonville, FL, USA, 2Neurology, University of Thessaly Medical School, Larissa, Greece, 3Neurochirurgia, Specjalistyczny Szpital im. Dr Alfreda Sokolowskiego, Walbrzych, Poland, 4Neuroscience, Mayo Clinic Jacksonville, Jacksonville, FL, USA

Introduction: Pallido-ponto-nigral degeneration (PPND), progressive supranuclear palsy (PSP) and corticobasal ganglionic degeneration (CBD) are 4R-tauopathies. PPND is a familial disease associated with the N279K mutation in the MAPT gene, belonging to the group of frontotemporal dementias and parkinsonism linked to chromosome 17 (FTDP-17). In contrast, PSP and CBD are mostly sporadic and are not associated with a MAPT mutation.

Aims: The aim of this study is to compare the neuropathology in spinal cord tissue from PPND, PSP and CBD patients.

Methods: Spinal cords from 4 PPND, 4 PSP, 2 CBD and 5 control cases were analyzed using H&E staining and immunostaining with monoclonal antibodies specific for phospho-tau. Pairwise multiple comparisons were performed.

Results: The mean age at autopsy was: PPND, 51 ± 5; PSP, 69 ± 9; CBD, 56 ± 13; and controls, 63 ± 7 years. Tau burden was greater in PPND compared to CBD and controls (p< 0.05), but the difference between PPND and PSP did not reach statistical significance. Both CBD and PSP have more spinal cord tau burden than controls.

Conclusions: The comparison of spinal cord pathology in 4R-tauopathies revealed a statistically significant greater tau burden in PPND, PSP and CBD than in controls, with PPND having the greatest tau burden. These findings provide further support for the involvement of the spinal cord in 4R-tauopathies and suggest that familial forms of 4R-tauopathies have a more severe phenotype compared to sporadic forms.