SATELLITE SYMPOSIUM: TAUOPATIES OVERLAPPING SYNDROMES

SOFT OVERLAPS BETWEEN PROGRESSIVE SUPRANUCLEAR PALSY AND CIRTICOBASAL DEGENERATION

Vladimir S. Kostić

Institute of Neurology, Belgrade, Serbia

Progressive supranuclear palsy (PSP) is the second most common neurodegenerative extrapyramidal disorder after idiopathic Parkinson's disease (PD) Globose neurofibrillary tangles (NFTs), tau positive astrocytes, and occasional ballooned argyrophilic neuronal degeneration involving brainstem, basal ganglia, and frontal lobe represent the pathological hallmarks of PSP. The cardinal clinical features of PSP are an insidious early onset of a symmetric akinetic-rigid syndrome with vertical supranuclear gaze palsy, early backwards falls, and frontal dysfunction. Magnetic resonance imaging (MRI) usually shows third ventricle dilatation and significant midbrain atrophy especially of the anteroposterior diameter. The classic clinical description of PSP, however, does not adequately describe one-third of cases in pathologically confirmed series. Patients with normal eye movements, or PD-like presentation with asymmetrical onset and good response to levodopa (L-dopa), have been described. Despite the publication of consensus operational criteria, an accurate diagnosis of PSP remains indeed a challenge for each neurologist. Particular clinical and pathological overlap exist between PSP and cirtocobasal degeneration.

In an attempt to unravel these diagnostic difficulties, PSP has been recently classified into two major clinical entities. The most common form is the classic clinical picture originally described by Richardson (Richardson's syndrome or PSP-RS). The second clinical phenotype associated with PSP is the PSP-parkinsonism (PSP-P), in which parkinsonism dominates the early clinical picture with initial moderate response to L-dopa, falls are delayed, and if gaze palsy and dementia develop, they occur late in the course of the disease. Disease duration in PSP-RS is significantly shorter and age at death earlier than in PSP-P. Pathological and genetic heterogeneity of PSP syndromes has also been reported. Tau pathology is more severe and the effect of the H1/H1 PSP susceptibility genotype appears stronger in clinically defined PSP-RS than in PSP-P.