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J C Steele's reflections on progressive supranuclear palsy

The well known tetrad of supranuclear ophthalmoplegia, axial rigidity, pseudobulbar palsy, and subcortical dementia is the hallmark of progressive supranuclear palsy (PSP), a disease of late middle life, terminating in five to 10 years. Dr Steele's recent observations may be of interest.

"Progressive supranuclear palsy is the name Dr J Clifford Richardson chose to designate an unusual clinical syndrome he first identified in the 1950s. Neurofibrillary degeneration is the hallmark of this fatal brain disease, and during our study of Richardson's patients, Professor Jerzy Olszewski and I also observed granulovascular degeneration, and widespread nerve cell loss and gliosis in subcortical and brain stem nuclei. The histopathological features bear a striking resemblance to those seen in postencephalitic parkinsonism after von Economo's epidemic encephalitis, and in the parkinsonism-dementia complex of Guam (PDC).

During the past 30 years, neurologists confirm that progressive supranuclear palsy is a universal, sporadic and not uncommon neurodegeneration of middle and late life. Many fine studies...have advanced our understanding of PSP but its cause, and thereby its cure, is still to be revealed. These historical notes tell of our observations from 1955 to 1975. We are pleased that colleagues remember these early descriptions and honor us by calling this disease, the Steele-Richardson-Olszewski (SRO) syndrome."22

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