

personal events experienced prior to the time of his injury (episodic memory), with relative preservation of memory for personal and world facts (semantic memory). This pattern of spared and impaired memory extended to spatial memory for large-scale environments and beyond memory to future imagining and decision-making. MRI showed widespread changes including diffuse atrophy, left PCA infarct, and left anterior frontal encephalomalacia. Notably, there was severe atrophy of the bilateral hippocampi, parahippocampal gyri, left amygdala, mammillary body, and anterior thalamus (after adjusting for generalized atrophy). We present neuropathological autopsy findings and clinicopathologic correlations in a case that has contributed greatly to our understanding of mechanisms of memory, especially the distinction between episodic and semantic memory.

ABSTRACT A9

Neuropathological correlates of Corticobasal Syndrome

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Corticobasal syndrome (CBS) is characterized by asymmetric parkinsonism, apraxia, cortical sensory deficits, dystonia, myoclonus, and cognitive dysfunction. Although it was originally described as the clinical manifestation of corticobasal degeneration (CBD), it is now recognized that CBS may be the clinical presentation of a variety of neurodegenerative pathologic processes including, but not limited to, CBD, Alzheimer disease (AD) and frontotemporal lobar degeneration (FTLD). In order to evaluate the neuropathological correlates of CBS in our center, 37 retrospectively identified cases with clinical CBS and post-mortem examination were analyzed. CBD was the primary pathological diagnosis in only eight cases (22% of the total cohort), whereas the most common underlying pathology was AD with or without Lewy body disease (LBD) in 16 cases (43%). The remaining 35% of cases were found to have progressive supranuclear palsy (N=5), FTLD-TDP (N=3), LBD (N=2), unclassifiable tauopathy (N=1), mild developmental abnormality (N=1) and neuronal intranuclear inclusion body disease (N=1). Moreover, 27% of the CBS cases had other pathological findings in addition to the main neurodegenerative process, most often cerebrovascular disease and/or mild AD-related pathology. These findings confirm and illustrate the heterogeneous pathological substrate for CBS.

ABSTRACT A10

Adult-onset progressive dementia and myoclonic epilepsy with polyglucosan bodies

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This 65 year-old left hand dominant male was referred for progressive cognitive decline with a working diagnosis of cortical

basal degeneration versus Alzheimer's disease. The patient also had a 10-12 year history of spontaneous myoclonic jerks partially controlled with Valproic Acid. There were no reported sensory or bladder changes and no episodes of status epilepticus. Neuropsychological assessment was consistent with generalized cognitive impairment that suggested a widespread dementing illness with a MoCA of 8/30 which had deteriorated from 14/30 in the year prior. Other exam findings demonstrated difficulty with upward gaze, apraxia and a wide-based and unsteady gait. Electroencephalographic studies revealed dysrhythmia Grade IV, generalized spikes, polyspike and wave discharges, several of which were associated with myoclonic jerks, consistent with generalized epilepsy. MRI revealed generalized cerebral and cerebellar atrophy with ventriculomegaly.

Post-mortem examination failed to demonstrate significant neurofibrillary degenerative changes. Of note however, there were abundant polyglucosan bodies. These were most prominent within cerebellum, hippocampal CA4, cerebral white matter and subpial regions. Results from electron and confocal microscopy will be discussed as this pertains to neuronal localization as well as a comparison with age-matched controls and a case of childhood Lafora body disease.

ABSTRACT A11

Progressive ataxia and palatal tremor: 2 autopsy cases of a novel tauopathy

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Sporadic Progressive Ataxia and Palatal Tremor (PAPT) is a rare syndrome characterized by symptomatic palatal tremor and slowly progressive cerebellar ataxia. To date, there has been only 1 autopsy report, which described a novel 4-repeat tauopathy with hypertrophic olivary degeneration and tau-positive inclusions in olivary neurons and dystrophic neuritic processes termed glomeruloid bodies. We report 2 further autopsy cases.

Case 1 is a 77-year-old man who presented with blurred vision and subsequently developed ataxia and gait instability. Dysarthria and palatal tremor appeared later. MRI showed T2 hyperintensity of the pons and bilateral inferior olives.

Case 2 is an 89-year-old man who presented with dysarthria and progressed to cerebellar ataxia and palatal tremor. 9 years into his disease course, his palatal tremor spontaneously resolved. MRI showed T2 hyperintensity in the bilateral olives, left mid-brain, and right dentate nucleus.

Consistent findings in both cases included bilateral hypertrophic vacuolar olivary degeneration accompanied by tau-positive neuronal inclusions and glomeruloid bodies, along with tauopathy in the pons and midbrain. Cerebellar cortical degeneration was extensive, but involvement of the dentate was minimal. Tau and TDP-43 negative basophilic neuronal cytoplasmic inclusions in the olive and Purkinje cells were also a feature.