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Clinical and pathological features of cognitive-predominant corticobasal degeneration

CBD is a rare, progressive neurodegenerative disorder, originally associated with a movement disorder, however, it is recognized that the clinical phenotypes are actually various. The heterogeneous combination of motor, sensory, behavioral and cognitive symptoms in CBD makes its antemortem diagnosis difficult. Although cognitive deficits are common in CBD, it is not widely recognized that some patients would present with predominantly cognitive dysfunction with minimal or no motor findings. To evaluate this atypical CBD phenotype, we studied the clinical and pathological characteristics of CBD with cognitive predominant problem (CBD-Cog) during the disease course, as presenting with cognitive predominant syndromes resembling AD or bvFTD.

In terms of the clinical aspect, we found prominent executive dysfunction, language problems, visuospatial problems and apathy as characteristic features of CBD-Cog. Neuropathological features of CBD-Cog were greater tau pathology in frontal and temporal cortices and less tau pathology in motor cortex. The significant tau pathologies were neuronal tau, tau threads and oligodendroglial tau. Also greater frequency of argyrophilic grain disease (AGD) was suggested in CBD-Cog. Whilst the astrocytic plaques are the histopathologic hallmark of CBD, the number and distribution of astrocytic plaques did not correlate with clinical syndrome. Our study suggests that CBD should be in the differential diagnosis of patients who present with dementia and executive dysfunction, especially apathy, even if they do not have overt extrapyramidal or motor features.

Additionally, I would like to make some comparison between PSP-FTD and CBD-Cog, based on our previous clinicopathological studies.

