Abstract N° 2859

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Abstract

Title
Neuropathological diagnostic accuracy of corticobasal degeneration: A review of 140 cases

Summary

Introduction: The clinical phenotypes of CBD are heterogeneous, but the pathological features of CBD are distinct with validated diagnostic criteria.

Methods: For a large pathological study, we collected 140 cases with a pathological diagnostic label of CBD from 12 centres internationally (4 to 28 cases per centre). The neuropathological diagnosis of each case was reviewed by evaluating a single AT8-immunostained section of the prefrontal area. In cases with equivocal findings, AT8-stained sections of additional regions and other immunohistochemical preparations using antibodies such as p62, amyloid-beta and alpha-synuclein were analysed.

Results: Of the 140 cases originally given a pathological label of CBD, 113 (80.7%) had confirmed CBD. Eleven cases (7.9%) had progressive supranuclear palsy (PSP) pathology, 5 had Alzheimer's disease (AD), 2 had FTLD-TDP, 1 had FTDP-17T, 1 had *C9orf72* repeat expansion and 1 had Lewy body disease. The remaining 6 cases had undetermined pathological diagnosis and will require further assessments such as genetic testing to

establish their diagnosis.

Conclusion: The diagnosis of a rare disorder such as CBD can be challenging because the number of cases and the neuropathologists' experience in diagnosing the condition can be limited even in large centres. In this large cohort, 20% of cases had a different pathological diagnosis from its original diagnostic label. It is likely that the tufted astrocytes in PSP, the presence of swollen neurons in coexisting agyrophilic grain disease and PSP, neuritic plaques in AD and astrocytic tangles in CTE were mis-interpreted as CBD-related features.

Presentation type : Poster
Euro CNS : No
Themes :

- 5 - Movement disorders