

Updates on Clinical, Neurophysiological and Neuroimaging Advances in Cortico-Basal Syndrome

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Introduction

Corticobasal degeneration (CBD) is a neurodegenerative condition characterized by 4R-tau protein deposition in several brain regions that clinically manifests as a heterogeneous atypical parkinsonism. The prototypical phenotype of CBD is corticobasal syndrome (CBS). Important insights into the pathophysiological mechanisms underlying motor and higher cortical symptoms in CBS have been gained by using advanced neuroimaging and neurophysiological techniques. Neuroimaging and neurophysiological abnormalities in multiple brain areas reflect the asymmetric neurodegeneration, leading to the asymmetric motor and higher cortical symptoms in CBS.

Methods

We analyzed the most recent and advanced updates on clinical, neurophysiological and neuroimaging of tauopathies. Analyzing tauopathies, we focused on CBS, stressing the most relevant differences between clinical, neurophysiological and neuroimaging characteristics of CBS and other neurodegenerative tauopathies, such as Frontotemporal Degeneration (FTD) and Supranuclear Progressive Palsy (PSP).

Results

Recent neuroimaging studies demonstrated a number of structural, functional and metabolic abnormalities leading to a better understanding of pathophysiology of CBS. Structural studies have demonstrated that asymmetric degeneration in fronto-parietal cortex is likely responsible for unilateral symptoms such as alien limb phenomena, cortical sensory loss and apraxia in CBS. Furthermore, neurodegeneration of subcortical brain areas and intra- and inter-hemispheric structural disconnection processes are likely to be involved in motor symptoms such as dystonia, myoclonus and parkinsonian features. Asymmetric DTI abnormalities in several associative fiber bundles and in the cortico-spinal tract may also contribute to specific CBS features. SPECT and PET studies found asymmetric metabolic changes possibly contributing to parkinsonism and other asymmetric motor signs and symptoms in CBS (1, 2). Recent neurophysiological studies have also led to a better understanding of the asymmetric neurophysiological abnormalities in specific brain networks in patients with CBS. It is reasonable to assume that abnormal M1 excitability and LTP/LTD-like plasticity play a crucial role not only in rigidity and bradykinesia but also in focal motor symptoms such as dystonia, thereby reflecting the structural and functional impairment of cortico-basal ganglia-thalamo-cortical motor loops (3).

Conclusions

We focused on neuroimaging and neurophysiological findings in CBS highlighting the important advances that have recently been made in our understanding of this disease and that point to asymmetry as a relevant feature of CBS.

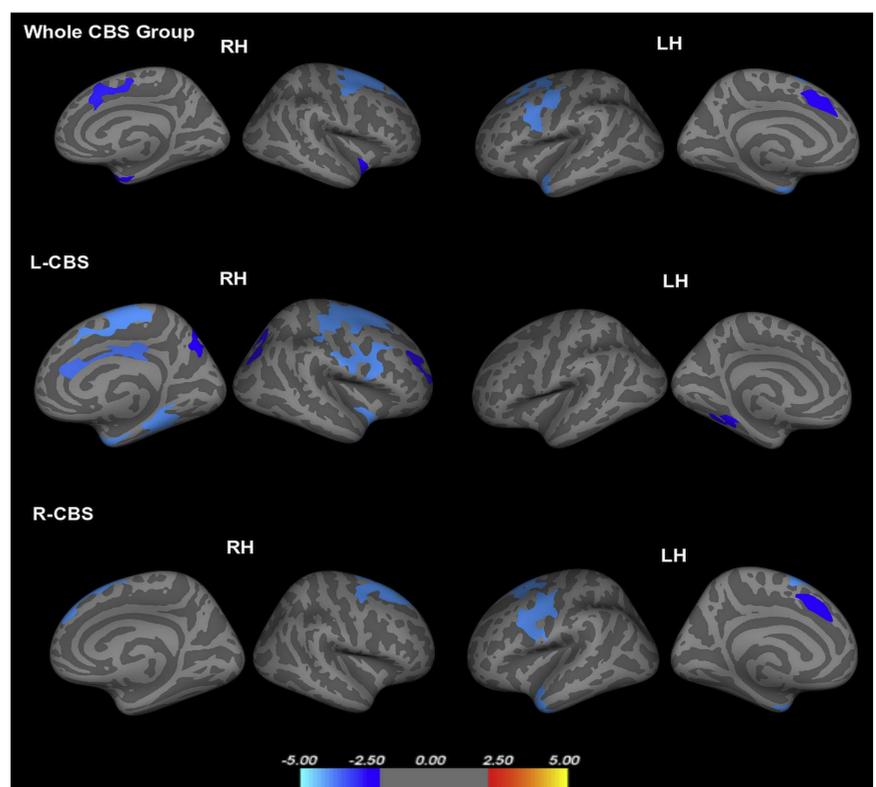
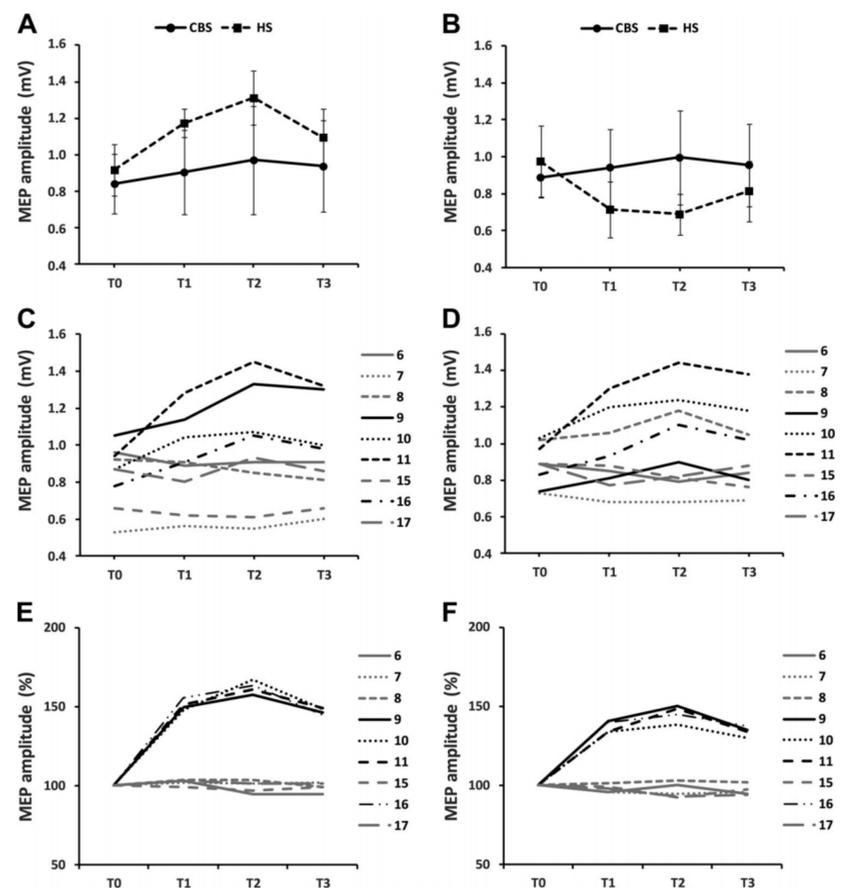


Fig.1. Neuroimaging in CBS.

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