Abstract

Objective: To describe characteristics of hippocampal sclerosis dementia. Methods: Convenienve sample of Hippocampal sclerosis dementia (HSD) recruited from the Johns Hopkins University Brain Resource Center. Twenty-four cases with post-mortem pathological diagnosis of hippocampal sclerosis dementia were reviewed for clinical characterization. Results: The cases showed atrophy and neuronal loss localized to the hippocampus, amygdala and entorhinal cortex. The majority (79.2%) had amnesia at illness onset, and many (54.2%) showed abnormal conduct and psychiatric disorder. Nearly 42% presented with an amnesic state, and 37.5% presented with amnesia plus abnormal conduct and psychiatric disorder. All eventually developed a behavioral or psychiatric disorder. Disorientation, executive dysfunction, aphasia, agnosia and apraxia were uncommon at onset. Alzheimer disease (AD) was the initial clinical diagnosis in 89% and the final clinical diagnosis in 75%. Diagnosis of frontotemporal dementia (FTD) was uncommon (seen in 8%). Conclusion: HSD shows pathological characteristics of FTD and clinical features that mimic AD and overlap with FTD. The findings, placed in the context of earlier work, support the proposition that HSD belongs to the FTD family, where it may be identified as an amnesic variant.

Keywords
dementia classification, hippocampal sclerosis, frontotemporal dementia, neuropsychiatry.