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Dementia Due to Hippocampal Sclerosis: Clinical Features and Comparison to Alzheimer's Disease

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Baltimore, MD
2:30 PM

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Abstract S03.003

OBJECTIVE: To define the clinical and neuropsychological features of dementia due to hippocampal sclerosis (HS) and to compare it to Alzheimer's disease (AD).

BACKGROUND: Several autopsy studies have identified HS as an etiology of dementia in the elderly. More recently, dementia due to HS was found to be frequently misdiagnosed as AD. In this study, we attempt to differentiate HS dementia from AD using data collected from subjects enrolled at the Johns Hopkins Alzheimer's Disease Research Center.

DESIGN/METHODS: Eight cases of HS and 84 cases of definite AD were included in the study. All 84 AD cases were diagnosed using CERAD criteria and were free of additional pathology. HS was defined by a combination of neuronal loss and gliosis in the hippocampus in the absence of senile plaques, neurofibrillary tangles, or other potential causes of dementia. All subjects were followed longitudinally until death. Data was collected at 6-month intervals. Mean scores on several clinical and cognitive measures from both groups were compared at study entry using the Wilcoxon Rank Sum (nonparametric) test. A group of 23 AD subjects, matched with the HS group on MMSE at study entry, were used for analysis of rate of decline. A weighted regression procedure was used for the latter analysis.

RESULTS: The HS group did not differ significantly from the AD group in age at onset, duration of illness prior to study entry, age at death, or education level. At study entry, the HS group had significantly ($p < 0.05$) better test scores on several measures including the Mini-Mental State Exam (MMSE), verbal fluency task, Boston Naming Test, and the orientation subscale of the Psychogeriatric Dependency Rating Scale (PGDRS). There were no significant differences on the physical and behavior subscales of the PGDRS, nor on the Hamilton Depression Scale. Analysis of rate of decline showed significantly slower cognitive decline over 5 years in the HS group, notably on the MMSE (-0.90 points/yr in HS vs. -2.80 points/yr in AD, $p < 0.01$). Surprisingly, the HS group slightly improved on the Boston Naming Test during follow-up (0.26 points/yr in HS vs. -3.2 points/yr in AD, $p < 0.01$).

CONCLUSIONS: Dementia due to hippocampal sclerosis appears to be clinically different from Alzheimer's disease. Particularly, patients with HS appear to have slower cognitive decline than AD patients. Larger studies are needed to further characterize HS dementia.

Afternoon Session; Session No. 3; AGING AND DEMENTIA: CLINICAL-PATHOLOGIC CORRELATION; Tuesday, April 28; 2:00 PM-4:00 PM; Room 101-F-H

Co-chairs: Neil Graff-Radford, Jacksonville, FL; Norman Relkin, New York, NY

2:00-2:30 PM

Presentation of the Potamkin Prize for Research in Pick's, Alzheimer's and Related Diseases

Michel Goedert, PhD; Virginia Lee, PhD; John Trojanowski, MD, PhD

Neurology

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