Clinical Characteristics of Pathological Early Onset Dementia with Lewy Bodies

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Background Early-onset dementia (EOD) is characterized by distinct clinical profiles and prognosis when compared to late-onset dementia (LOAD). As the second most common neurodegenerative form of dementia, little is known about the clinical profile of early-onset Dementia with Lewy Bodies (DLB).

Results This study included 363 patients with EOAD, 32 EODLB and 147 late-onset DLB. Patients with EODLB were more likely to present with psychosis, apathy, REM sleep behavioral disorder, and motor symptoms. While EOAD patients were more likely to present with cognitive symptoms as first recognized and predominant presentation and perform worse in memory assessment. Motor as first recognized presentation, slowness, visual hallucination, caregiver reporting of agitation and apathy were the significant predictors to differentiate the two. Late-onset DLB patients were less depressed and more impaired in memory and executive function related scores than EODLB. Significant number of EODLB patients were misdiagnosed as EOAD (46.9%), p<0.0001.

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Background Early-onset dementia (EOD) is characterized by distinct clinical profiles and prognosis when compared to late-onset dementia (LOAD). As the second most common neurodegenerative form of dementia, little is known about the clinical profile of early-onset Dementia with Lewy Bodies (DLB). A current challenge for clinicians when managing patients with DLB is the suboptimal diagnosis rate which will affect treatment efficacy and outcome. To address this knowledge gap, by hypothesizing early-onset DLB will have a distinct profile when compared to Alzheimer’s disease (AD), we accessed and reviewed data of patients with pathological confirmed DLB from National Alzheimer’s Coordinating Center (NACC) database.

Methods Patients with first visit that fulfill criteria for dementia of AD or DLB were analyzed. Early onset age was defined as less than 65 years old. Variables included in the analyses include baseline demographics, cognitive, behavioral, motor symptoms, neuropsychological battery scores and clinician diagnosis. Comparisons were made between early-onset AD (EOAD) versus early-onset DLB (EODLB), and early versus late-onset DLB.

Results This study included 363 patients with EOAD, 32 EODLB and 147 late-onset DLB. Patients with EODLB were more likely to present with psychosis, apathy, REM sleep behavioral disorder, and motor symptoms. While EOAD patients were more likely to present with cognitive symptoms as first recognized and predominant presentation and perform worse in memory assessment. Motor as first recognized presentation, slowness, visual hallucination, caregiver reporting of agitation and apathy were the significant predictors to differentiate the two. Late-onset DLB patients were less depressed and more impaired in memory and executive function related scores than EODLB. Significant number of EODLB patients were misdiagnosed as EOAD (46.9%), p<0.0001.