

Neuropathology of Dementia with Lewy bodies



안 태 범

경희대병원 신경과

Tae-Beom Ahn, MD, PhD

Department of Neurology, Kyung Hee University Hospital

Dementia with Lewy bodies (DLB) is a neurodegenerative disorder characterized by dementia and other clinical manifestations such as cognitive fluctuation, parkinsonism, visual hallucination, et al.

Pathologic substrates of DLB are centered on the accumulation of α -synuclein (aSyn) as Lewy bodies (LBs) or Lewy neuritis. However, as pathologies related to Alzheimer disease (AD) such as neurofibrillary tangles and β -amyloid plaques (bAmp) can coexist with aSyn pathologies, it is mandatory to exclude AD in pathologic diagnosis. Other pathologies such as vascular lesions or argyrophilic granules are also found in the patients with DLB. A recent study reported that aSyn pathologies were more positively associated with the clinical diagnosis of DLB than AD pathologies.

Clinicopathologic correlation between aSyn pathologies and key clinical symptoms of DLB is commonly studied in relation with Parkinson disease (PD). The correlation between cortical LBs or bAmp and cognitive decline was more clearly described in PD dementia. In case of visual hallucination, aSyn pathologies in neocortex and limbic structures were underscored along with thalamic involvement in functional studies.

Spreading aSyn pathologies is actively studied as a common pathomechanism of PD and DLB. Asymptomatic aSyn pathologies were found in healthy subjects, which were known as incidental Lewy body disease (ILBD). Interestingly, in some cases with ILBD, LBs were more abundant in the limbic or cortical areas than the brainstem, which resembled the distribution pattern of DLB than that of PD. Actually, these cases were prodromal DLB but incongruent with Braak hypothesis. Recent studies reported diverse strains of aSyn. Molecular behavior of different strains of aSyn was dissimilar. It remains to be studied whether the clinical and pathologic diversity of Lewy body diseases such as PD and DLB could stem from the variety of aSyn.

Future studies are needed to understand the clinicopathologic correlation, in which dissecting the role of concomitant multiple pathologies is essential, and pathomechanism related to spreading aSyn pathologies.
