Lewy Body Dementias

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Zachary A. Trottier*; Morgan Farley, M.D., Samantha K. Holden, M.D.

University of Colorado

Alzheimer's and Cognition Center
UNIVERSITY OF COLORADO
ANSCHUTZ MEDICAL CAMPUS

Abstract

Lewy body dementia encompasses a spectrum of neurodegenerative disease causing progressive cognitive impairment. The disease entities included under a Lewy body dementia umbrella are dementia with Lewy bodies and Parkinson's disease dementia. These two conditions are currently best distinguished by the timing of onset and relative prominence of motor versus cognitive symptoms, yet they share many clinical and pathological characteristics. Criteria have also been developed or proposed to identify their respective prodromal stages, including mild cognitive impairment in Parkinson's disease and prodromal dementia with Lewy bodies. This article will overview the spectrum, clinical features, pathophysiologic mechanisms, diagnostic criteria, and management of LBD.

Key Points

- Lewy body diseases include dementia with Lewy bodies (DLB), Parkinson's disease dementia (PDD), and prodromal stages.
- Lewy body diseases are pathologically defined by deposits of aggregated alpha-synuclein, although co-pathology is common
- Together, Lewy body diseases may comprise up to 25% of all cases of dementia
- DLB and PDD are distinguished by the relative timing of symptom onset
- Core clinical features of DLB include parkinsonism, visual hallucinations, REM sleep behavior disorder, and cognitive fluctuations
- Treatment is symptom-based, focusing on symptoms that are unsafe or bothersome to the patient/caregiver
- Novel treatments currently being studied include tyrosine kinase inhibitors , ambroxol, and CT1812; all of which target lysosomal function

Lewy Body Diseases	Mild Cognitive Impairment	Lewy Body Dementias
Prodromal DLB	MCI-DLB	DLB
Parkinson's Disease	MCI in PD	PDD

Diagnostic Tools in Suspected LBD

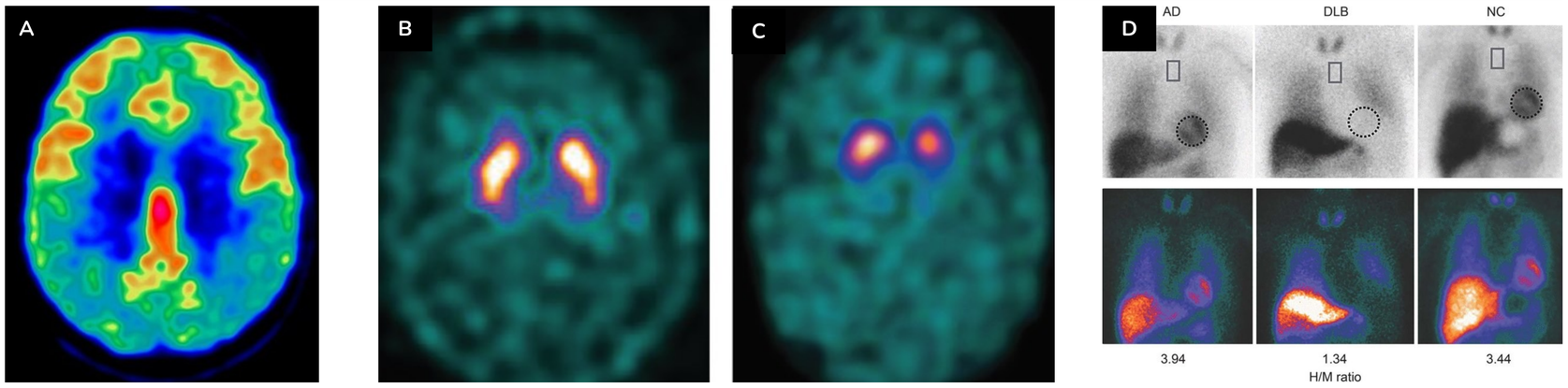

Figure 1: Advanced diagnostic modalities used in Lewy body disease diagnosis

A: [18F]Fluorodeoxyglucose positron emission tomography (18FDG-PET) imaging of a patient with DLB; prominently displaying the "cingulate island sign." B: Dopamine transporter (DaT) SPECT imaging using ioflupane (I123) tracer in a healthy patient; revealing normal uptake of tracer in dopaminergic neurons of the striatum. C: I123 DaT imaging in patient with Parkinson's disease, revealing reduced uptake in striatum. D: Cardiac [123I]metaiodobenzylguanidine (123I-MIBG) imaging in patient with Alzheimer's disease (left column), DLB (middle column) and health control patient (right column. Loss of cardiac uptake in the patient with DLB indicates loss of cardiac sympathetic innervation.

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