Dementia with Lewy Bodies
Tracy Boschee, DO  PGY II
Wellmont Family Medicine Residency
October 2012

Objectives

• Describe the unique signs of Dementia with Lewy Bodies (DLB)
• Propose a treatment plan for patients with DLB
• Outline the general course of DLB
Dementia

- Progressive loss of cognitive function independent of the state of attention
- ALWAYS a pathologic process (NOT a normal part of aging)

DSM IV – Dementia Due to Other General Medical Conditions

- **A.** The development of multiple cognitive deficits manifested by both
  - 1. **Memory impairment** (impaired ability to learn new information or to recall previously learned information)
  - 2. **One (or more) of the following** cognitive disturbances:
    - **Aphasia** (language disturbance)
    - **Apraxia** (impaired ability to carry out motor activities despite intact motor function)
    - **Agnosia** (failure to recognize or identify objects despite intact sensory function)
    - **Disturbance in executive functioning** (i.e., planning, organizing, sequencing, abstracting)

DSM IV

- **B.** A1 and A2 cause significant impairment in social or occupational functioning & represent a significant decline from a previous level of functioning
- **C.** Evidence from the H&P or labs that the disturbance is the direct physiological consequence of a general medical condition other than Alzheimer’s disease or cerebrovascular disease
- **D.** The deficits do not occur exclusively during the course of a delirium
What is a Lewy Body (LB)?
- Round, eosinophilic, intracytoplasmic inclusion in the nuclei of neurons
- Main component = Alpha-synuclein protein
  - Normal synaptic protein – unknown function
  - Insoluble & aggregated form in LBs
- Other components = neurofilament proteins & ubiquitin

Lewy Bodies

History of the Lewy Body
- Described by Dr. Frederick Lewy, MD in 1914, as he was working in the laboratory of Dr. Alois Alzheimer
History of the Lewy Body

- Initially found in substantia nigra of patients with idiopathic PD
- 1960’s – LBs in neocortex; presumed rare
- 1980’s – immunocytochemical methods to identify LBs; DLB found to be far more common

Spectrum of LB disorders

- Parkinson Disease (many have LBs in neocortex)
- Parkinson’s disease dementia
- Dementia with Lewy Body (ALL have LBs in neocortex)

Dementia with Lewy Bodies
**Etiology**
- Unknown
- Possibly in part from disruption of information flow between striatum and neocortex, especially the frontal lobe
- Multifactorial – altered level of neurotransmitters (acetylcholine, dopamine), impaired cell function.

**Epidemiology**
- 2nd most common cause of degenerative dementia after Alzheimer’s disease
- 10-22% of dementias
- Disease of late middle and old age
- Most studies suggest DLB slightly more common in men

**Diagnosis = Clinical**
- 1st Clinical criteria for DLB – 1996
- Revised to improve sensitivity & specificity by the third report of the DLB consortium – 2005
- Define criteria for **Probable & Possible** DLB
#1 = ESSENTIAL

- Dementia
  - Early impairment of attention, executive & visuospatial function
  - Memory affected later in course of disease (AD typically presents with memory loss)

Probable DLB

- Dementia + at least 2 Core Clinical Features

- Dementia + 1 Core Clinical Feature + 1 Suggestive Feature

Possible DLB

- Dementia + ABSENCE of Core Clinical Features + at least 1 Suggestive Feature
Core Clinical Features

- Cognitive fluctuations
- Visual hallucinations
- Parkinsonism

Suggestive Features

- REM sleep disorder
- Severe neuroleptic sensitivity
- Low dopamine transporter uptake in the basal ganglia on SPECT or PET

Let me explain…

- Fluctuations
  - Seen in 60-80% of DLB; early sign
  - Change in level of cognition & level of alertness
  - Difficult to evaluate
  - Can last seconds to days
  - Examples:
    - "Blank out"
    - Confusion
    - Excessive somnolence
    - Speech / motor arrest
  - Daytime drowsiness, daytime naps > 2hrs, prolonged staring spells, & episodes of disorganized speech. Episodes of at least 3 of 4 = more likely in DLB than AD.
Am I seeing things???

- Visual Hallucinations
  - Seen in 50-75% of DLB; early sign
  - Simple to complex hallucinations
  - Visual misperceptions

Give me a push...

- Parkinsonism
  - Seen in 70-90% of DLB
  - Bradykinesia & akinesia
  - Limb rigidity
  - Gait disorder
  - Tremor can be present, but is less common and less severe than in PD

Stop hogging the covers!

- REM sleep disorder
  - Seen in 85% of DLB
  - Parasomnia with vivid dreams in REM sleep, without the usual muscle atonia = “acting out” dreams
  - Violent (fighting, fleeing) or non-violent (making speeches, feeding, urinating, clapping)
  - May precede other DLB symptoms by many years
  - Could be an independent, idiopathic condition
“You take the red pill - you wake up in Wonderland and I show you how deep the rabbit-hole goes”

- Neuroleptic Sensitivity
  - Seen in 30-50% of DLB
  - Severe, sometimes irreversible parkinsonism & impaired consciousness
  - Sometimes with features of neuroleptic malignant syndrome
  - May precipitate / worsen confusion or autonomic dysfunction

Supportive Features

- Common in DLB, but have no clear role in diagnosis
  - Repeated falls
  - Syncope or transient LOC
  - Autonomic dysfunction
  - Auditory / olfactory / tactile hallucinations
  - Delusions
  - Depression
**Treatment**

- Symptomatic
- Behavior strategies
- Modify stressors
- Physical therapy
- Mobility aids

**Medications**

- Cholinesterase inhibitors
  - First line
  - One RCT showed help with cognition, fluctuations, psychotic symptoms, anxiety, & parkinsonian symptoms
  - Donepezil, rivastigmine
- Memantine
  - One RCT showed clinical global impression of change

**Medications**

- REM sleep disorder
  - Clonazepam, Melatonin
- Depression
  - SSRI
  - AVOID tricyclic agents due to their anticholinergic effects
- Parkinsonism
  - Carbidopa-levodopa – start low & go slow. Less successful than in PD
The BIG issue... Neuroleptics!

- Potential for severe, irreversible reactions
- If antipsychotic therapy is required – only atypicals at very small doses
  ○ Advise patient / family / caregivers of possible risks
- Associated with increased risk of death in elderly with dementia
- AVOID older, conventional antipsychotics completely

Prognosis

- Progressive cognitive decline
- Psychotic symptoms / visual hallucinations typically persist
- Parkinsonisms worsen
- In two studies survival time after onset of cognitive symptoms = 7.7 years (9.3 years in AD)
- Death usually from complications of immobility, poor nutrition, and swallowing difficulties
References List


- AmericanMedicalAssociation.com
- www.CartonStock.com
- www.crackhospital.com
- www.elements4health.com
- www.theallroom.com