“The eye doesn’t see what the mind doesn’t know.”

Sir William Osler

Lewy Body Dementia

“Atypical Dementia”
The Lewy Body Spectrum

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Progressive loss of intellectual and social function involving several spheres. Memory loss is common. Others include language, calculating ability, judgment and insight, visuospatial / perceptual ability, behaviors interfering with ADL’s, such as resistance to care.

Dementia
It is estimated that there are over 80 causes of dementia, some reversible; most not.
Alzheimer’s disease is the most common cause, 60 – 70% in most autopsy series.
Lewy Body Dementia is the second most common cause, 15 – 20%, and the most often misdiagnosed.

Etiologies of Dementia

Prevalence doubles every five years after age 65, from <5% to 50% after age 90.
Alzheimer’s Disease accounts for most dementia. Male / Female 50/50.
Lewy Body Dementia is next, at 20%. Male / Female ratio is 2:1. Progress is a bit faster, but variable. It is nonfamilial.

Demographics of Dementia

Post-Rolandic
(temporal, occipital cortex)
Alzheimer’s / Lewy Body
- Memory loss
- Aphasia, apraxia, agnosia
- Preserved personality
- Preserved social skills
- MMSE is a valid measure

Pre-Rolandic
(Frontal / Subcortical)
FT / ETOH / Vascular / NPH
- Memory loss
- Loss of ambition, initiation
- Personality change
  - Cynicism
  - Apulia (apathy)
  - Incontinence
  - MMSE useless

Where did it start?
Deficit in 2 or more cognitive domains
• Progressive decline, affecting function
• Preserved level of consciousness
• Onset after age 40
• Absence of other brain pathology
• Absence of parkinsonism

Definition of Probable AD

Post-Rolandic type dementia
• Begins in hippocampus in temporal lobe, spreads to occipital lobe
• Characterized by beta amyloid plaques and neurofibrillary tangles (tau)
• 65% also have Lewy bodies (LB variant)

Alzheimer’s Disease Pathology

Spectrum of Lewy Body Dementia
Lewy Bodies first described by Dr. Lewy in 1912 in the substantia nigra in PD.
- Eosinophilic granules in cytoplasm with a halo, easy to see. (cytoplasmic inclusion bodies)
- Composed of ubiquitin, alphasynuclein, without a halo in the cortex.
- First described in cortex in 90’s through immunoflorescence.

**Alpha-synucleinopathy**

Lewy bodies in Substantia nigra only
- Parkinson’s Disease or Level 1

Lewy bodies in basal ganglia
- “Parkinson’s plus” or Level 2

Lewy Bodies throughout cortex
- Lewy Body Dementia or Level 3

**Location dictates clinical picture**

**Spectrum of Pathology**

- Tau aggregation
  - Alzheimer’s
  - PSP
  - CBD

- Amyloid deposits
  - Alzheimer’s
  - [Lewy Body]

- A-synuclein inclusion body deposits
  - Lewy Body Dementia
  - Parkinson’s
  - Multisystem Atrophy
  - [Alzheimer’s]
Progressive cognitive decline
- Frontal, subcortical, or visual may dominate

Core features
- Fluctuations in cognition, sensorium
- Visual hallucinations
- Parkinsonism

Supportive features
- Falling, syncope, neuroleptic sensitivity, delusions

**LB Dementia Criteria**

Tremor: slow, resting
Rigidity: cogwheeling quality
Akinesia: loss of amplitude of both voluntary and automatic movements
Postural instability: forward posture, tendency to fall backward

**Definition of Parkinsonism**

"Up and down" cognition and sensorium
- Agitation one minute, sleeping the next
- Walking one day, falling the next
- "pseudo-delirium"
  - Need to rule out usual causes of delirium
  - May last hours, days, weeks
  - Light touch with medication

**Core Feature of LB Dementia Fluctuations**
Rigidity is prominent.
Tremor is absent or mild.
Onset within a year of dementia.
Forward posture and poor righting reflex confers a high fall risk!
Autonomic symptoms may be prominent, leading to orthostasis, syncope, constipation and urinary retention.

Core Feature of LB Dementia
Parkinsonism

- Often people, sometimes little people.
- Strictly visual and well-formed.
- Emotional reaction may go away with low dose Seroquel, without need to abolish all hallucinations.
- More social stimulation helps.
- Often caregiver and family need reassurance.

Core Feature of LB Dementia
Hallucinations

- Dopamine blockade dramatically increases parkinsonism.
  - Phenothiazines, including Compazine
  - Older antipsychotics, like Haldol, Mellaril
  - Some newer ones, including Risperdal, Abilify also effect mobility
  - Accelerates rate of decline, associated with longer hospitalizations, complications, death

Core feature of LB Dementia
Neuroleptic Sensitivity
Acetylcholine esterase inhibitors may be more effective in LBD than in AD.
Serotonin (S2) blocker Seroquel is a good choice to block hallucinations, agitation.
Depakote is often successful as a calming agent, to avoid overuse of benzodiazepines.
L-Dopa may help rigidity early on. May or may not worsen psychosis, hypotension.

Implications for treatment

Variable and hard to predict!
Comorbidities play a role.
Faster than Alzheimer’s.
Attempts at staging are ongoing.
Families have questions and need guidance.
Our health care system is overburdened.
Palliative and hospice care should be timely.

Prognosis: Compelling Need
1. No difficulties
2. Subjective forgetfulness
3. Decreased job functioning and organizational capacity
4. Difficulty with complex tasks
5. Supervision with ADL's
6. Impaired ADL's, incontinence
7A. Speech limited to 6 words
7B. Single word
7C. Loss of ambulation
7D. Inability to sit
7E. Inability to smile
7F. Inability to hold head up

Hospice Criteria for Endstage Dementia

- FAST > 7A AND one of these:
  - Aspiration
  - Upper UTI
  - Sepsis
  - Multiple stage 3 or 4 ulcers
  - Weight loss >10% in 6 mos.

Medicare uses the FAST; based on a 1987 study of 47 patients, only 12 of whom declined in stages. Overall mortality >7c was 3 mos.

Functional Assessment Staging (FAST)

<table>
<thead>
<tr>
<th>1.9 Complete dependence ADL’s</th>
<th>Risk of death in 6 months</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.9 Male gender</td>
<td>Score</td>
</tr>
<tr>
<td>1.7 Cancer</td>
<td>0</td>
</tr>
<tr>
<td>1.6 CHF</td>
<td>1 - 2</td>
</tr>
<tr>
<td>1.6 Oxygen needed within 14 days</td>
<td>3 - 5</td>
</tr>
<tr>
<td>1.5 Shortness of breath</td>
<td>6 - 840</td>
</tr>
<tr>
<td>1.5 Unstable medical condition</td>
<td>9 - 11</td>
</tr>
<tr>
<td>1.5 Bowel incontinence</td>
<td>12 +</td>
</tr>
</tbody>
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Mortality Risk Index

- ADL Dependence
- Other organ failure
- Fulltime caregiver needed
- Close medical monitoring and attempts at rehabilitation

Road to Hospice

- Falls and nonambulatory status
- Poor swallow and aspiration
- Trips to ER and Hospital for recurrent problems
- Palliative care, formal caregiving needed (board and care, SNF)
- Non-ambulatory
- Contractures and skin breakdown
- Weight loss
- Symptom control, caregiver support, resource preservation
The Lewy Body spectrum of dementias includes Dementia with Lewy Bodies, Parkinson’s Disease, and Parkinson’s Plus syndromes: Progressive Supranuclear Palsy, Multisystem Atrophy and Corticobasilar Degeneration.

It is challenging to distinguish among these illnesses because the diagnosis is a clinical one, and there is much overlap in clinical presentation.

However, it is especially important for Hospice and Palliative Care clinicians to understand subtle differences in these dementias in order to advise the family regarding treatment, expected course and prognosis.

This talk will focus on two aspects of this topic. First, a compare-and-contrast description of Alzheimer’s Disease and Lewy Body Dementia will be presented, including updates on pathology and symptomatic management. Second, an evidence-based bedside scoring system called the “Mortality Risk Index” will be introduced as a promising way to assist with prognostication.

This is a fifty (50) minute presentation.