Navigating The Cognitive Internet: Introduction

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What’s so hard about diagnosing dementia?

- Diagnosis relies on synthesis of information from many sources
  - Time consuming
  - Requires training / specialized skill set
- Multiple dementia syndromes
- Most dementias are mixed disorders
- Heterogeneity of phenotypes
Colors of the Palette

- History & Functional Status (aka symptoms)
  - First symptoms, temporal evolution, functional impairments, sleep, appetite, falls etc...

- Medical History
  - Cancer, autoimmune, diabetes, liver failure, medications

- Family History
  - Cognitive disorders, seizures, mental retardation

- Cognitive Exam
  - Deficits in memory, attention, language, visuoperceptual abilities

- Neurologic Exam
  - Parkinsonism, eye movements, ataxia, dystonia

- Diagnostics
  - Labs, MRI, EEG, PET,
Alzheimer’s Disease
NINCDA-ARDA Criteria for AD

Probable AD
- Dementia established by clinical examination
- Dementia confirmed with cognitive testing
- Deficits in 2 or more cognitive domains
- Progressive decline in memory or other areas of cognition
- Preserved consciousness
- Onset between the ages of 40-90
- Absence of systemic illness or other brain disease that could otherwise account for the symptoms

AD History & Functional Status (Symptoms)

- Repetitions
- Misplacing
- Geographic disorientation
- Word Finding
- Delusions

- Ask same question, repeat same statement
- Lose glasses, keys, purse
- Lost while driving
- That whatchamacallit
- Someone stole my purse. Spouse is having an affair
AD History & Functional Status (Symptoms)

- Bill paying
  - Forget to pay or mail bills, pay twice, can’t write out checks

- Cooking / Appetite
  - Leave burners on, forget well known recipes, confused with the steps of cooking, spoiled food in the refrigerator, weight loss, forgets to eat

- Driving
  - Get lost, ask which way to turn

- Socialization
  - Quieter in a group, wants to stay home
Medical History

Family History

By definition, medical comorbidities should not be of a sufficient magnitude to explain the dementia

Increased risk with family history of

- A first degree relative with AD
- Down’s syndrome
Cognitive Exam

- Memory loss
  - Amnestic or retrieval deficit (early disease)
- Attention preserved or mildly reduced
- Visuospatial deficits proportional to memory loss
- Deficits in executive functioning, present but usually not prominent
  - Planning, organizing, prioritizing, focusing attention, sustaining attention, shifting attention
Cerad in AD

- Ticket
- Letter
- Arm
- Grass
- Pole
- Cabin
- Letter
- Shore
- Engine
- Butter

Trial 1: 2
Trial 2: 3
Trial 3: 3

Free recall: 0
Recognition: 2 Targets
10 Foils

Fruits & vegetables: 6
Letter F: 8
AD Neuro Exam

- No focal abnormalities
- Absence of parkinsonism
Diagnostics

- MRI
- EEG
- PET/SPECT
- Lumbar puncture

- Temporal lobe atrophy
- Independent bitemporal slow waves.
- Absence of seizures.
- Hypometabolism in bilateral temporal/parietal lobes
- Tau/a-beta ratio
- Useful biomarker
Lewy Body Disease
Chronic delirium
LBD History & Functional Status (Symptoms)

- Visual hallucinations
- Visual perceptual deficits
- Fluctuations
- Milder memory problems
- REM sleep disorder
- Word finding
- Delusions
- See animals, little people
- Scrape the car when backing out, can’t stay in lane,
- Fine one moment/ bad the next. Fluctuations scale
- Forget why they went into a different room
- Kick & punch the bed, dreams seem real, walk or talk in sleep
- Search for word, hesitancies
- Complex, stereotyped
Medical History

- Autonomic Dysfunction: Supportive Feature
  - Postural hypotension
  - Syncope
  - Arrhythmias
  - Temperature dysregulation

- There should be no other medical condition that in and of itself could explain the patient’s symptoms
Patient’s with LBD have an increased incidence of a family history of dementia compared to normal controls
Cognitive Exam

- Poor attention (digits forward, digits backwards)
- Disproportionate deficits in executive functioning (poor letter fluency, clock drawing)
- Visuospatial deficits out of proportion to memory impairment
Cerad LBD

- Ticket
- Letter
- Arm
- Grass
- Pole
- Cabin
- Letter
- Shore
- Engine
- Butter

  Trial 1: 2
  Trial 2: 5
  Trial 3: 7
  Free recall: 3
  Recognition: 10 Targets
  9 Foils

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Fruits & vegetables: 13
Letter F: 4
House Drawing

FRUITS 4, VEG 12
LETTER F 1

TRIALS 3, 2, 4
FREE RECALL 3
RECOGNITION TARGETS 3
FOILS 4
Simple Figures
Boston Naming Test
Comparison of AD to LBD

- Flat learning
- Amnestic or retrieval deficit
- Fairly well preserved attention
- Poor category fluency
- Anomia
- Visuospatial deficits in keeping with memory impairment

- Learning curve
- Retrieval or source memory deficit
- Poor attention
- Poor letter fluency
- Visual errors with confrontational naming
- Visuospatial deficits out of proportion to memory impairment
LBD Neuro Exam

- Parkinsonism
- Increased tone, gait changes, decreased facial expression, rest tremor, bradykinesia, bradyphrenia, hypophonia, hypoknietic dysarthria
Diagnostics

- MRI
- EEG
- PET/SPECT
- Possibly parietal/occipital atrophy
- FIRDA
- Parietal/occipital hypometabolism
Lewy Body Disease
Criteria

Core clinical features

- Fluctuating cognition with pronounced variations in alertness and attention
  - daytime sleep greater than 2 hours, daytime lethargy, disorganized speech, staring spells
- Recurrent visual hallucinations that are well formed and detailed
- Spontaneous features of parkinsonism not solely due to medications

Probable LBD: 2 or more core features
Possible LBD: 1 core feature
Lewy Body Disease
Criteria

- Supportive Features
  - Repeated falls
  - Syncope / autonomic dysfunction
  - Neuroleptic sensitivity
  - Complex systematized delusions
  - Hallucinations in other modalities
  - REM sleep disorder
Frontotemporal Dementia

- Behavior variant (bvFTD)
- Semantic dementia (SD)
- Progressive nonfluent aphasia (PNFA)
MRI Frontal Atrophy
bvFTD Symptoms

- Change in personality or behavior
- Compulsions
- Executive dysfunction
- Food / Eating
- Language impairment (less prominent than personality changes)

- Disinhibited
  - Restlessness, wandering, inappropriateness, increased jocularity, sexual inappropriateness, drinking, childlike behavior, lack of empathy, remorse

- Apathetic
  - Depressed, lack motivation, hypersomnolent

- Hyper-religiousity, gambling, pornography, purchasing multiples
- Perseverations, inattention, utilization behavior
- Gluttony, unusual food cravings, cramming food, non-edible food

- Word retrieval
Progressive nonfluent aphasia

- Nonfluent speech output
- Agrammatism
- Telegraphic speech
- Phonemic paraphasias & anomia
- Single word comprehension good. May have comprehension problems for complex sentences
- Early behavior & personality changes less common
- Extrapyramidal features, behavioral problems, and executive dysfunction later
Semantic dementia

- Loss of word & object knowledge
- Sentences of normal length (>7 words /utterance), but they don’t make sense
- Poor comprehension at the single word level
- Can’t recognize faces
- Prosopagnosia
  (more common with greater L temporal lobe atrophy)
- Behavior changes
  (R temporal lobe atrophy)
Normal Pressure Hydrocephalus
Ventriculomegaly

- Idiopathic Normal Pressure Hydrocephalus
- Secondary Hydrocephalus
  - Brain surgery
  - Infection
- Medical Conditions
  - Sleep apnea
  - Diabetes
- Atrophy
NPH

Cardinal Symptoms
- Problems with walking and falls
- Bladder incontinence
- Memory loss

Diagnostic Challenge
- Who will benefit from treatment

Treatment
- Surgical
Corticobasal Degeneration

- Core features
  - Insidious and progressive
  - Cortical Dysfunction AEB at least one
    - Focal or asymmetric ideomotor apraxia
    - Alien limb
    - Cortical sensory loss
    - Visual or hemisensory neglect
    - Constructional apraxia
    - Focal or asymmetric myoclonus
    - Apraxia of speech/Non-fluent aphasia
  - Extrapyramidal dysfunction
    - Focal or asymmetric appendicular rigidity
    - Focal or asymmetric appendicular dystonia
Mixed Neuropathology

- 141 community dwelling participants.
- Annual clinical exams and brain donation.
- Examined brains for neuropathology of cerebral infarctions, AD, and PD/LBD.

- In persons with dementia
  - 30% had pure AD
  - 38% had AD & infarcts
  - 12% Vascular dementia
  - 12% AD & PD/LBD

- In person without dementia
  - 28.6% no diagnostic abnormalities
  - 24.2% had pure AD
  - 17.6% brain infarctions

- Persons with multiple diagnoses were nearly 3 times more likely to exhibit dementia compared to those with only one pathologic process

The National Alzheimer’s Coordinating Center (NACC)

Department of Epidemiology, School of Public Health, University of Washington, Seattle, WA


NACC database from >30 ADRC from 1984-2009.

8142 autopsies
<table>
<thead>
<tr>
<th></th>
<th>Normal</th>
<th>AD</th>
<th>LBD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Plaques &amp;T</td>
<td>14%</td>
<td>97%</td>
<td>70%</td>
</tr>
<tr>
<td>Lewy bodies</td>
<td>5%</td>
<td>21%</td>
<td>97%</td>
</tr>
<tr>
<td>Large stroke</td>
<td>10%</td>
<td>12%</td>
<td>8%</td>
</tr>
<tr>
<td>Micro stroke</td>
<td>18%</td>
<td>18%</td>
<td>13%</td>
</tr>
<tr>
<td>ASVD</td>
<td>18%</td>
<td>36%</td>
<td>30%</td>
</tr>
</tbody>
</table>

- Normal: n=351
- AD: n=5679
- LBD: n=1195
FTD | VaD | All
---|---|---
Plaques & T | 26% | 42% | 80%
Lewy bodies | 8% | 5% | mixed
Large stroke | 5% | 31% | path
Micro stroke | 10% | 42% | 
ASVD | 18% | 58% |
Heterogeneity of Expression

Gold Standard

- Clinical Diagnosis ?
- Genetic characterization ?
- Neuropathology ?