Clinical Neuropsychological Assessment of Frontotemporal Dementia

Nora Breen
Royal Prince Alfred Hospital
Referral: Does this patient have dementia?

- Neurodegenerative Disease?
  - YES
    - FTD
      - Behavioural FTD
      - Semantic Dementia
      - PNFA
  - NO
    - Alzheimer’s
FTD

- Behavioural Form
  - 75% FTD Cases

- Language Form
  - Primary Progressive Aphasia

- Semantic Dementia

- Progressive Non-Fluent Aphasia (PNFA)
Neuropsychological Assessment

Neuropsychologist relies equally on three components of the assessment:

1. History
2. Behavioural Observations
3. Neuropsychological Test Scores

3 cases – BvFTD, SD and AD
History

Case 1 BvFTD
• 59 woman, inpatient, morbidly obese
• Referral: poor self-care and neglect of home

Patient history :
• Denied any problems with thinking or memory
• Denied personality change
• Reports being self-caring, showering, shopping and cooking meals.
History

Case 1 BvFTD

Daughter’s history

Previously:

- Mother was immaculately groomed, meticulous about hair, nails, clothes.
- Previously boundless energy, when wasn’t working she was cleaning house or socialising.
Case 1 BvFTD
Daughter’s history continued..
• 12 months ago – stopped showering/changing clothes. No longer cared about appearance.
• 30 kilo weight gain last 12 months, binge eating
• Spends all day watching tv, smoking, eating
• Cannot be bothered to use toilet, urinates in chair
• Sits in soiled clothes until daughter forces her to change.

• Memory is good.
History

Case 2 SD
- 62 year old woman
- 3 year hx of worsening language comprehension and speech problems

Patient described:
1. Inability to name objects “I see things and recognise them but cannot put a name to them”

2. Unable to remember word meanings “I can hear a word but not know what you are talking about”
History

Case 2 SD

• Daughter’s additional history:

• Previously sophisticated vocabulary is diminished
• She no longer understands jokes or sarcasm
• Interprets spoken language very literally
• Difficulty reading because can’t remember meanings of words
• *Good memory
History

Case 3 AD

• 74 yr old woman
• Patient reported: some forgetfulness

• Husband reported:
• 12-18 mth history of increasing forgetfulness regarding everyday activities and appointments
• Sometimes disoriented to day or time
• Losing things
• Repeating same questions – up to 6 times in one hour
• Preserved personality/social skills, independent in ADL
## History Summary

<table>
<thead>
<tr>
<th></th>
<th>BvFTD</th>
<th>SD</th>
<th>AD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Insight</td>
<td>Early Loss</td>
<td>Intact</td>
<td>Intact</td>
</tr>
<tr>
<td>Behaviour Changes</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Memory Deficits</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Language Deficits</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
</tr>
</tbody>
</table>
Behavioural observations

Case 1 BvFTD

- Morbidly obese
- Unkempt and dishevelled (long nails, unwashed hair, smelled of urine)
- Brusque manner
- Bland affect
- Confabulation
- Assessed twice, 5 days apart. Remembered my name on 2nd occasion without difficulty.
Behavioural Observations

Case 2  SD

• Immaculately groomed and dressed
• Extremely pleasant and cooperative, social skills entirely intact
• Unable to follow conversation or respond to questions unless language kept very simple
• Asked if drank alcohol: “sorry, that word is familiar but I don’t know what it is”
• In spite of language difficulties, she was able to convey reasonable account of history and current difficulties.
Behavioural Observations

Case 3  AD

• Pleasant, neatly groomed and dressed, cooperative with testing, socially appropriate
• Relied on husband to give history
• Occasionally stopped half-way through task to clarify what she was supposed to be doing.
• Appropriately disappointed by her errors on testing (insight)
# Behavioural Observations Summary

<table>
<thead>
<tr>
<th></th>
<th>BvFTD</th>
<th>SD</th>
<th>AD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Behaviour</td>
<td>Impaired</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Social Skills</td>
<td>Impaired</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Affect</td>
<td>Blunted</td>
<td>Normal</td>
<td>Normal</td>
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</table>
Neuropsychological Testing

Cognitive domains assessed:

- Orientation
- Memory
- Attention
- Visuospatial and constructional skills
- Language
- Executive function (problem solving, flexible thinking, planning)
- *Mood
Cognitive profiles

- Specific patterns on testing are consistent with diagnosis of different types of dementia.

Bv FTD: executive impairment

SD: semantic knowledge loss, naming deficit

AD: memory impairment (rapid forgetting)
Memory

Expected pattern:

BvFTD - learning ⊗ recall ? recognition ⊗
SD - hard to assess, visual memory intact
AD - impaired (rapid forgetting)
## Verbal Memory

RAVLT 15 word list

<table>
<thead>
<tr>
<th></th>
<th>5th trial</th>
<th>Recall</th>
<th>Recogn</th>
<th>fp</th>
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</thead>
<tbody>
<tr>
<td>BvFTD</td>
<td>10</td>
<td>9</td>
<td>10</td>
<td>1</td>
</tr>
<tr>
<td>AD</td>
<td>6</td>
<td>1</td>
<td>13</td>
<td>9</td>
</tr>
</tbody>
</table>

SD (not appropriate)
Visual Memory
Case 1 BvFTD
Case 2 SD
Case 3: AD
Executive Function

Ability to plan, problem solve, shift mental set, adapt responses according to feedback, think flexibly.

Expected pattern:
BvFTD - impaired
SD - intact
AD - intact
Executive Function

Wisconsin Card Sorting Test
Set shifting, problem solving, mental flexibility
6 possible categories
# Executive Function

**WCST – Results**

<table>
<thead>
<tr>
<th>Case</th>
<th>Categories</th>
<th>Perseveration</th>
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<tbody>
<tr>
<td>Case 1 BvFTD</td>
<td>0</td>
<td>YES *</td>
</tr>
<tr>
<td>Case 3 AD</td>
<td>5</td>
<td>no</td>
</tr>
</tbody>
</table>

*sorted to incorrect category for entire deck!

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Case 2 SD       Unable to comprehend instructions
Language

BvFTD  altered speech output (aspon teneity or rushed speech)
  echolalia
  perseveration
  mutism

SD  fluent
  loss of word meanings
  naming and comprehension deficit
  semantic errors

AD  mild/moderate naming deficit
Language - Naming

Boston Naming Test 60 Item

- BvFTD 58 /60
- SD 5 /60
- AD 41 /60

*SD: snail

“I don’t know what that is. Is it a bird of some sort? I don’t recognise it at all.”
## Cognitive Profiles

<table>
<thead>
<tr>
<th></th>
<th>BvFTD</th>
<th>SD</th>
<th>AD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Executive Function</td>
<td>Impaired</td>
<td>Intact</td>
<td>Intact</td>
</tr>
<tr>
<td>Visual Memory</td>
<td>Intact</td>
<td>Intact</td>
<td>Impaired</td>
</tr>
<tr>
<td>Verbal Memory</td>
<td>Intact</td>
<td>n/a</td>
<td>Impaired</td>
</tr>
<tr>
<td>Language</td>
<td>Intact</td>
<td>Impaired</td>
<td>Mildly Impaired</td>
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