Behavioral – variant frontotemporal dementia due to spontaneous intracranial hypotension

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Behavioral – variant frontotemporal dementia

• Progressive deterioration of personality, social comportment, and cognition
• Caused by frontotemporal lobar degeneration
• Incurable
• Second most common cause of dementia in <60 years
• Genetic basis
A Core diagnostic features

- Behavioral disinhibition
- Apathy or inertia
- Loss of empathy
- Perseverative behavior
- Hyperorality
- Altered neuropsychological performance

From: Sensitivity of revised diagnostic criteria for the behavioural variant of frontotemporal dementia
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Behavioural variant frontotemporal dementia (bvFTD) diagnostic criteria.

Possible (at least three features A-F must be present)
A) Early behavioural disinhibition
   A1. Socially inappropriate behaviour
   A2. Loss of manners or decorum
   A3. Impulsive, rash, or careless actions

B) Early apathy or inertia
   B1. Apathy
   B2. Inertia

C) Early loss of sympathy or empathy
   C1. Diminished responsiveness to other people’s needs and feelings
   C2. Diminished social interest, interrelatedness, or personal warmth

D) Early perseverative stereotyped or compulsive/ritualistic behaviour
   D1. Simple repetitive movements
   D2. Complex compulsive or ritualistic behaviour
   D3. Stereotypy of speech

E) Hyperorality (a tendency to explore objects with the mouth) or dietary changes
   E1. Hyperorality and dietary changes
   E2. Binge eating, increased consumption of alcohol or cigarettes
   E3. Oral exploration or consumption of inedible objects
FTD Types

1. **Behavioral variant frontotemporal dementia (bvFTD)**
   - Also called frontal variant frontotemporal dementia (fvFTD) or Pick’s disease
   - Negatively impacts social skills, emotions, personal conduct, and self-awareness.
   - People with bvFTD might act in inappropriate ways, show a lack of judgment or inhibition, neglect to maintain personal hygiene, do something compulsively or repetitively, or feel euphoric or apathetic.
   - bvFTD is the most common form of frontotemporal dementia.
Spontaneous intracranial hypotension causing reversible frontotemporal dementia
M. Hong, G. V. Shah, K. M. Adams, R. S. Turner, N. L. Foster
Behavioral variant frontotemporal dementia-hypersomnolence syndrome due to spontaneous intracranial hypotension

- N = 31
- Insidious onset
- Sixth decade of life
- Male:Female – 4:1
- Brain sagging: 100%
- CSF leak: 3%
- Treatment: refractory; Surgery in 94%
bvFTD in spontaneous intracranial hypotension
Frontotemporal dementia in spontaneous intracranial hypotension
## Associated symptoms

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Count</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypersomnolence</td>
<td>29</td>
<td>(100%)</td>
</tr>
<tr>
<td>Headache</td>
<td>27</td>
<td>(93%)</td>
</tr>
<tr>
<td>Orthostatic</td>
<td>20</td>
<td>(69%)</td>
</tr>
<tr>
<td>Nonpositional</td>
<td>4</td>
<td>(14%)</td>
</tr>
<tr>
<td>Reverse orthostatic</td>
<td>3</td>
<td>(10%)</td>
</tr>
<tr>
<td>Auditory symptoms</td>
<td>21</td>
<td>(72%)</td>
</tr>
<tr>
<td>Dysequilibrium/gait dysfunction</td>
<td>19</td>
<td>(66%)</td>
</tr>
<tr>
<td>Tremors</td>
<td>15</td>
<td>(52%)</td>
</tr>
<tr>
<td>Posterior neck pain</td>
<td>13</td>
<td>(45%)</td>
</tr>
<tr>
<td>Nausea/emesis</td>
<td>10</td>
<td>(34%)</td>
</tr>
<tr>
<td>Dysphagia/dysarthria</td>
<td>9</td>
<td>(31%)</td>
</tr>
<tr>
<td>Orofacial dyskinesia</td>
<td>6</td>
<td>(21%)</td>
</tr>
<tr>
<td>Blurred vision</td>
<td>5</td>
<td>(17%)</td>
</tr>
<tr>
<td>Dysgeusia</td>
<td>4</td>
<td>(14%)</td>
</tr>
<tr>
<td>Impotence/erectile dysfunction</td>
<td>4</td>
<td>(14%)</td>
</tr>
<tr>
<td>Incontinence</td>
<td>4</td>
<td>(14%)</td>
</tr>
<tr>
<td>Hiccupping</td>
<td>3</td>
<td>(10%)</td>
</tr>
<tr>
<td>Abducens nerve palsy</td>
<td>2</td>
<td>(7%)</td>
</tr>
<tr>
<td>Coma</td>
<td>1</td>
<td>(3%)</td>
</tr>
<tr>
<td>Trigeminal neuralgia</td>
<td>1</td>
<td>(3%)</td>
</tr>
</tbody>
</table>

Bending at the waist
Frontotemporal dementia in spontaneous intracranial hypotension

• Good outcome after treatment

1 / 7 patients who had undergone Chiari decompression (14%)

vs

20 / 22 patients who had not undergone Chiari decompression (91%)

P=0.003
bvFTD in SIH
Surgical solutions for the recalcitrant patient

• Lumbar dural reduction surgery

• Wearable epidural saline infusion catheter system
Dural reduction surgery
Dural reduction surgery
Dural reduction surgery

- N = 52
- 40 women and 12 men (most “without” SIH)
- Age: 21 – 72 years
- Good outcome: 31 (60%)
- Complications: Pseudomeningocele: 5 (10%)
  Suicide: 1 (2%)
  Infection/sepsis: 1 (2%)
Implantation of a wearable epidural spinal infusion system
Spinal epidural infusion system

• N = 17
• 7 women and 5 men (most “without” SIH)
• Age: 34 – 79 years
• Good outcome: 15 (88%)
• Complications: Infection: 3 (18%) Hardware failure: 7 (41%)
“So, do you like working here?” the middle-aged man bellowed to the young physician at the other end of the hospital coffee shop. The woman, the object of this not-very-subtle pickup line, ignored him. The man's sister cringed. When had her younger brother, who was 49, turned into such a jerk? He had always been so quiet and shy. She was living across the country in Washington State, so she didn't see him often, but he had certainly changed.

In his 20s, he had a problem with alcohol. But back then, his drinking made him quieter. And even during the worst of his drinking days, he had always been tidy and well kempt, fastidious in everything he did. That morning, she drove from the airport to pick him up on the way to visit their father, who was in the hospital after heart surgery. She had taken a red-eye flight from Seattle to Philadelphia, but her brother looked worse than she felt: tired, disheveled, dirty. He said he had just showered, but she could tell it wasn't true.