Managing Behavioral FTD (bvFTD)

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Overview of bvFTD

- Behavioral variant (predominant) – Frontotemporal dementia (bv-FTD; in short, frontotemporal dementia – FTD): one kind of presentation of “frontotemporal lobar degeneration (FTLD)”. The other kind is the language variant (predominant) – FTLD (so called Primary Progressive Aphasia – PPA)
- A relatively ***rare***, ***young-onset*** (younger than 65) ***neurodegenerative*** disorder (progressive brain cell loss predominantly affecting frontal and temporal lobes)

Clinical Symptoms

- Cognitive symptoms – impaired executive function (dysexecutive syndrome) – “frontal lobe syndrome”
- Behavioral symptoms – outstandingly bizarre actions; out of character
- Emotion and affect – blunt; lacking sympathy and empathy – personality change
- Unusual eating behavior or sexual behavior
- Changing from one extreme to the other
- Lacking self-regulation and insight

Diagnostic Evaluation

- Every patient with bv-FTD has a unique story because of his/her education, job and social status.
- Although there is a set of diagnostic criteria (revision after revision over the past 30 years), it is not easy to diagnose patients with bv-FTD. Often patients would take 2 to 3 years to receive the diagnosis.
- On the other hand, some patients can be given the wrong diagnosis of bv-FTD if there are conditions that can be mimicking bv-FTD.

Diagnostic Evaluation

- In order to make a proper diagnosis of bv-FTD, the clinician has to spend time to interview the family members/friends/colleagues who have known the patient well over the years.
- It is very important to survey or rule out common medical conditions that mimic bv-FTD, such as alcohol/substance abuse, traumatic brain injury, stroke, brain tumor and psychiatric disorder.
- Brain scans (head CT or brain MRI) are crucial for establishing the diagnosis of bv-FTD --- objective evidence of atrophy (shrinkage; brain cell loss) in frontal and temporal lobes.
- Functional brain studies, like SPECT or PET, can be used but they often are NOT covered by insurance plans. And they are NOT always reliable.

Diagnostic Evaluation

- Some patients have main features of bv-FTD; yet their brain MRI and/or head CT studies do not show objective evidence of atrophy in the frontal and/or temporal lobes over the years of follow-up. Be cautious! Need a second opinion.
- Professor Hodges reports “bv-FTD phenocopy syndrome” or so-called “non-progressive FTD”.

Complex Terminology

- Over the years, there are following terminology – frontal lobe syndrome, Pick's disease, dysexecutive syndrome, frontotemporal dementia, bv-FTD/bv-FTLD, temporal lobe predominant – FTD, Semantic dementia and Primary progressive aphasia (PPA).
- Based on research, majority of patients with bv-FTD are "sporadic" in nature (meaning unknown cause of several altered proteins in brain cells). Few bv-FTD patients are caused by the particular mutations of several genes.
- At times, it is also difficult to tell bv-FTD from young-onset Alzheimer's disease.

Progression

- Because bv-FTD is a neurodegenerative disorder, the brain will continue to lose brain cells and progressively spread the changes into other brain regions over the years. Therefore bv-FTD patients will continue to lose cognitive skills, including language, memory, motor and visual functions.
- Although bv-FTD does not directly affect heart rate, blood pressure and breathing, this disorder inevitably makes the patients to be vulnerable to dangers, to lose ability to take care of themselves and to cause care-providers a lot of stress because of lacking empathy and impaired insight.

Management

- There is NO cure for bv-FTD at the present time.
- Management – the family and caregivers need to learn about this disorder so that they can form their way and plan to provide the care.
- Patients with bv-FTD need a principal “custodial” person to protect and provide the care (legal guardian or legally-appointed representative) .
- There are some psychiatric medicines that can be prescribed selectively by psychiatrists or neurologists for some behavioral issues or neurological symptoms.