Frontotemporal dementia and its variants: What to look for

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Frontotemporal dementia (FTD) is a neurologic disease that affects the frontal and the temporal lobes of the cerebral cortex.[1] This disorder is observed most often in people between age 45 to 65, but also can manifest in younger or older persons.[1] The cause varies among a range of pathologies affecting the anterior portions of the brain.[2]

Presentations

FTD presents with changes in personality, social skills, ability to concentrate, motivation, reasoning, and language abnormality.[3] Memory loss is less prominent in this condition compared with other dementias; therefore, identification may be a diagnostic challenge. FTD can be misdiagnosed as a psychiatric illness or not recognized because social symptoms dominate over cognitive dysfunction. As the disease progresses, patients may become increasingly unable to plan or organize activities of daily living, behave appropriately, and react normally in social interactions.[1]

FTD has 3 diagnostic variants[1-4]:

Behavioral variant. Known as Pick disease or the "frontal variant,"[1,2] this type of FTD manifests as changes in personality, improper behavior in social settings, personal neglect, or impulsivity, such as shoplifting or hypersexuality.

Primary progressive aphasia. Two types of language dysfunction are observed in FTD:

- Semantic dementia (SD)[3]: Left-sided SD presents with "meaningless speech" or "word substitutions" (eg, "chair" instead of "table"). Right-sided SD, however, is characterized by forgetting the faces of familiar people or objects.
- Primary nonfluent aphasia[3]: Language fluency is compromised. Persons with such language dysfunction cannot produce words easily, and their speech is stumbling and nonfluent.

FTD with motor neuron disease.[4] The most common type of motor neuron disease associated with FTD is amyotrophic lateral sclerosis. Afflicted patients exhibit muscle weakness, spasms, and rigidity. This leads to difficulty in swallowing or breathing because the diaphragm and pharynx are paralyzed. Other diseases associated with FTD include corticobasal degeneration and progressive supranuclear palsy.

Diagnosis

In DSM-5, FTD has been renamed "frontotemporal lobar degeneration" under the category of "Major and Mild Neurocognitive Disorders."[5] The workup begins with a history, physical examination, and mental status assessment. Physical signs can include frontal-release, primitive reflexes. Early in the disease course, a palmomental reflex often is observed; later, as disease progress, the rooting reflex or palmar grasp may become apparent.[1,5]

Diagnosing FTD requires recognizing its symptoms and ruling out conditions such as Alzheimer's disease, depression, and schizophrenia.[6] Laboratory studies may help identify other conditions. Brain imaging, such as MRI, can depict frontotemporal pathology and rule in or exclude other diseases.[3,5]

Psychometric testing can evaluate memory or cognitive ability, which might be unremarkable during the initial phases of

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FTD.[4] Further psychological assessments may provide objective verification of frontal lobe deficiencies in social skills or activities of daily living.[3] Positron emission tomography and single-photon emission computed tomography may demonstrate areas of decreased activity or hypoperfusion in frontal and temporal lobes.[7]

Interventions

Treatment of FTD is limited to symptomatic therapy[8]; there are no specific, approved countermeasures available. Comorbid conditions, such as diabetes mellitus or hypertension, should be treated medically. Social interventions such as day care, increased supervision, and emotional support from the family can be effective adjuvants.[2]

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