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Frontotemporal Dementia

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Dementia is a common problem for older adults, and roughly 2/3 of cases are diagnosed as Alzheimer's disease. One third of patients, therefore, have other etiologies for their dementia, the most common including vascular dementia, dementia with Lewy bodies, and frontotemporal dementia (see Figure 1). This issue of Elder Care will review frontotemporal dementia (FTD), a condition that is sometimes misdiagnosed as Alzheimer's disease.

Frontotemporal dementia is the current term for a dementia syndrome that used to be known by several different names, including Pick's Disease, progressive aphasia, semantic dementia, and frontal dementia of the non-Alzheimer's type. The common pathophysiological finding in these syndromes is brain atrophy that is most marked in the frontal and temporal lobes – hence the name frontotemporal dementia.

About 40% of patients with FTD have family histories of a similar dementia syndrome; many of these individuals have a mutation of the tau protein gene on chromosome 17. Other mutations also occur. The remaining 60% of cases are sporadic.

Prevalence and Age of Onset

The true prevalence of FTD is unclear, as it is often not considered in the differential diagnosis of dementia by providers, and as a result is often confused with and mislabeled as Alzheimer's disease. One important clue to the diagnosis of FTD, however, is its earlier onset than Alzheimer's disease, with most cases appearing before age 70 (and usually appearing before age 65). Among people with dementia in their early 60s, the relative prevalence of FTD and Alzheimer's disease is about the same. With increasing age, Alzheimer's becomes the more common diagnosis. Overall, FTD is thought to account for about 2-5% of all cases of dementia (see Figure 1), though some estimates go as high as 10-15%.

Recognizing Frontotemporal Dementia

As well as an earlier age of onset, FTD has several other characteristics that distinguish it from Alzheimer's Disease. These are shown in Table 1.

Most importantly, personality, behavior, and language changes predominate (Table 2), especially in the initial stages of FTD. Executive function is also impaired early on in the course of the illness, and only later does memory impairment develop.

Personality and Behavior Changes fall into one of three initial patterns. Over time, these patterns blend and patients may demonstrate aspects of all three. The most common pattern is **disinhibition**, in which patients lose learned social behaviors. They may steal or use foul language. They may lose a sense of safety (people with FTD have been known to try to exit moving automobiles). They may demonstrate bizarre behavior, poor financial judgment, and compulsive buying, leading to an incorrect diagnosis of bipolar disorder or schizophrenia.

Another pattern of personality change is **withdrawal**, in which patients become apathetic and don't participate in daily activities. They may need prompting even for basic activities like personal hygiene.

The third pattern is **repetitive/compulsive behaviors**. An individual might wash repeatedly, read the same book over and over, or eat excessively.

Language Changes (semantic dementia) are less common. These patients have difficulty expressing themselves, finding correct words, and naming objects, even though understanding of words is preserved. Over time, they may become mute.

Diagnosis

FTD is suggested by the clinical presentation. When evaluated with the Mini-Mental State Exam, patients

TIPS TO DIAGNOSE FRONTOTEMPORAL DEMENTIA (FTD)

- Consider FTD when dementia occurs in individuals younger than age 65-70, or in older individuals with dementia when there is a family history of early-onset dementia.
  Consider FTD when behavior disturbances, such as disinhibition or withdrawal, outweigh memory impairment.
- Consider FTD when impairments of executive function predominate over memory impairment.
- Order brain imaging in case of suspected FTD to detect atrophy of the frontal and/or temporal lobes.

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perform more poorly on items that test executive function and better on items related to memory. Clock-drawing, a measure of executive function, is impaired early.

CT or MRI often shows the typical atrophy of the frontal and/or temporal lobes.

Treatment

There is no cure for FTD. Drug treatment is aimed at controlling behavioral symptoms. Selective serotonin reuptake inhibitors may help with withdrawal or compulsive behaviors. The newer "atypical" antipsychotics may help with aggressive behaviors or disorganized thinking, but there has been limited study of their use. Prescription of the older, "typical" antipsychotics is not recommended. Cholinesterase inhibitors, widely used for Alzheimer's disease, have no benefit in FTD and have caused worsening symptoms in some patients. The role of memantine is being studied. Overall, none of these medications has a major influence on the course or outcomes of FTD. The Food and Drug Administration has not approved any medications for treating FTD.

Prognosis

FTD is a fatal illness. Life expectancy from time of diagnosis ranges from 2-17 years (mean 8 years).

Table 1. Distinguishing Frontotemporal Dementia from Alzheimer's Disease

Characteristic	Frontotemporal Dementia	Alzheimer's Disease
Age of Onset	< 70 yrs usually <65 yrs	Usually > 65 yrs
Memory Deficits	Late	Early
Executive Function Deficits	Early	Late
Behavior Disturbance	Early	Late
Brain Imaging	Frontal/temporal atrophy	Diffuse atrophy
Response to cholinesterase inhibitors	None	Some

Table 2. Various Symptoms of Frontotemporal Dementia (adapted from Vossel Reference)

Uninhibited and inappropriate social behaviors – Uncharacteristic behavior, such as stealing or using foul language in public
Inappropriate sexual behaviors
Loss of awareness about changes in behavior
Loss of concern about appearance and hygiene
Increase in appetite that leads to constant eating and weight gain
Apathy, loss of drive, social withdrawal
Loss of speech and language (many people become mute as the disease progresses)
Compulsive or repetitive behaviors, such as pacing collecting things, or hand washing

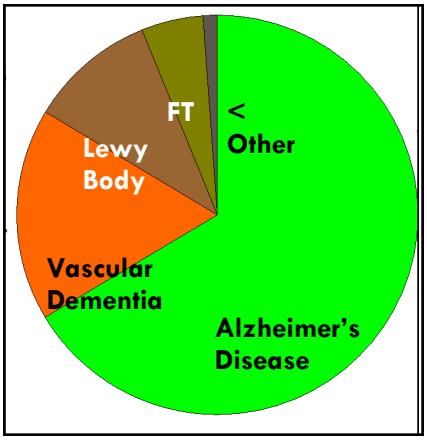

Figure 1:
Relative Prevalence of Common Dementias

FT = Frontotemporal Dementia

"Other" includes normal pressure hydrocephalus, HIV related dementia, Creutzfeldt-Jakob disease, Huntington's disease and more

References and Resources

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