

Primary Care and early diagnosis of front temporal dementias

Front temporal Dementias: A difficult diagnosis

Most primary care physicians do not include dementia on the list of differential diagnoses commonly considered when seeing patients under age 60. However, a growing body of research on the front temporal dementias (FTD) reveals a clinical picture illustrating that while the disorders known as FTD are still not common, they are much less rare than once thought. One tangible result of this increased awareness is a growing list of expert clinicians with substantial knowledge of FTD. Thus, there are medical centers to refer patients to for a full diagnostic workup and which can offer support to the PCP as he or she provides ongoing care. The key to limiting the impact this disease has on a family lies with the family physician and early diagnosis.

The front temporal dementias are a group of degenerative brain disorders that result from progressive damage to cells in the anterior temporal and/or frontal lobes of the brain. The clinical hallmark of FTD is a gradual, progressive decline in behavior, language, or movement that has a relatively young age of onset (average in the late 50's, but ranging from 20's to 80's). The front temporal dementias are the second leading cause of dementia in people under 65 after Alzheimer's disease.

Behavioral Presentations

FTD characterized by loss of empathy and increasingly inappropriate social behavior is known clinically as behavioral variant FTD (bvFTD) or Pick's disease. Behavioral changes are typically seen as alterations in personality, emotional blunting, or loss of empathy that result in increasingly inappropriate social behavior.

People gradually become less involved in routine daily activities and withdraw emotionally from others. Unusual behaviors may include swearing, overeating or drinking, impulsivity, shoplifting, hypersexual behavior, and deterioration in personal hygiene habits. Accompanying this is a decreasing self-awareness. The patient displays little insight into the inappropriateness of his or her behavior. Little or no concern for its effects on other people, including family and friends. Patients may also display repetitive, stereotyped behaviors, such as hand clapping, humming the same song over and over, gorging on one specific type of food, or walking to the same place day after day.

Language Disorders

Primary progressive aphasia (PPA) is a progressive deterioration in language ability, including the ability to speak fluently, understand language, read, and write. The language deficits experienced by FTD patients are distinguished by two presenting issues: problems with expression of language or difficulty with naming and word meaning. People with progressive non fluent aphasia (PNFA) become hesitant in their speech and begin to talk less, but appear to retain the meaning of words longer. In semantic dementia (SD), people experience a progressive deterioration of understanding words and recognizing objects, but retain the ability to produce fluent speech.

Movement Disorders

Corticobasal degeneration (CBD) and progressive supranuclear palsy (PSP) are neurologic disorders characterized by deterioration in motor skills and movement. CBD classically begins as a movement disorder with affected individuals showing a unilateral paucity of movement and muscle rigidity with a

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