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A Brief Overview on Causes and Diagnosis of Frontotemporal Lobar Degeneration

Xiuyun Liu*

Department of Physiology Nursing, University of California, San Francisco (UCSF), USA

Frontotemporal lobar degeneration (FTLD) is a clinically and pathologically heterogeneous syndrome, characterised by progressive decline in behaviour or language associated with degeneration of the frontal and anterior temporal lobes. While the seminal cases were described at the turn of the 20th century, FTLD has only recently been appreciated as a leading cause of dementia [1], particularly in patients presenting before the age of 65 years. Three distinct clinical variants of FTLD have been described: (i) behavioural-variant front temporal dementia, characterized by changes in behaviour and personality in association with frontal-predominant cortical degeneration; (ii) semantic dementia, a syndrome of modern loss of knowledge about words and objects related to anterior temporal neuronal loss; and (iii) progressive no fluent aphasia, characterized by effortful language yield, loss of grammar and engine speech deficits in the setting of left perisylvian cortical atrophy.

What causes FTD?

Scientists describe FTD using the designs of alter in the brain seen in an autopsy after death. These changes include loss of neurons and abnormal amounts, or forms of proteins called tau and TDP-43. These proteins arise clearly in the body and help cells work properly [1-3]. When the proteins don't work properly, for reasons not but fully caught on, neurons in specific brain regions are damaged.

FTD that runs in a family is often related to mutations (changeless changes) in certain qualities. Genes are fundamental units of heredity that tell cells how to make the proteins the body wants to function. Even small changes in a gene may produce an abnormal protein, which can cause changes in the brain and, eventually, disease.

Types and symptoms of FTD

In the early stages, it can be tough to know which type of FTD a person has because symptoms and the order in which they appear can vary from one character to another.

There are 3 types of frontotemporal disorders (FTD): behavioral variant frontotemporal dementia (bvFTD), primary progressive aphasia (PPA), and movement disorders.

Behavioural variant frontotemporal dementia

The most common FTD, bvFTD, involves changes in personality, behaviour, and judgment. People with this disorder may have issues with cognition [4], but their memory may live highly intact. Symptoms can include:

Problems planning and sequencing (thinking through which steps come first, second, and so on)

Difficulty prioritizing tasks or activities

Repeating the same pastime or saying the same word over and over

Primary progressive aphasia

PPA includes changes in the ability to communicate — to utilize language to talk, read, type in, and understand what others are saying. This includes issue utilizing or understanding words (aphasia) and issue talking properly (e.g., slurred speech). People with PPA may have one or both of these symptoms. They may become quiet or unable to speak. Movement disorders

Two rare neurological movement disorders related to FTD, corticobasal syndrome and progressive supranuclear palsy, occur whilst the parts of the brain that control movement are affected. The disorders may have an effect on thinking and language abilities, too.

Symptoms

Signs and symptoms of frontotemporal dementia may be one of a kind from one individual to the next. Signs and symptoms get gradually worse over time [2], usually over years. Clusters of symptom sorts tend to occur together, and people may have more than one cluster of symptom types.

Behavioural changes

The most common signs of frontotemporal dementia contain extreme changes in conduct and personality. These consist of:

- Increasingly inappropriate social behavior
- Loss of empathy and other interpersonal aptitudes, such as having sensitivity to another's sentiments
- Lack of judgment
- Loss of inhibition
- Lack of interest (unresponsiveness), which can be mixed up for depression
- Repetitive compulsive behavior, such as tapping, clapping or smacking lips
- A decline in personal hygiene
- Changes in eating habits, as a rule overeating or creating an inclination for desserts and carbohydrates
- Eating inedible objects
- Compulsively wanting to position things in the mouth

Diagnosis

A diagnosis of frontotemporal degeneration is based upon

***Corresponding author:** Xiuyun Liu, Department of Physiology Nursing, University of California, San Francisco (UCSF), USA, E-mail: liuxiuyun@gmail.com

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identification of characteristic symptoms, a detailed patient and family history, a thorough clinical assessment and a variety of specialized tests. A neuropsychological assessment involves an interview and certain tests, often pencil and paper type tests. This evaluation will allow a doctor to survey behavior, language, memory, visual-spatial and other cognitive functions. Early in the route of the disorder, people with the behavioral variant of frontotemporal degeneration generally tend to attain very well on neuropsychological testing.

Clinical Testing and Workup

Specialized imaging techniques may include attractive resonance imaging (MRI), single-photon emission computed tomography (SPECT) and positron emission tomography (PET) scans. An MRI uses a magnetic field and radio waves to supply cross-sectional images of particular organs and bodily tissues consisting of the brain. In SPECT [1-4], physicians use a radioactive substance and a special camera to create 3-d images of internal areas of the frame, consisting of the brain. SPECT can monitor characteristic changes such as reduced blood flow in sure areas of the brain. In PET scans of the brain, a radioactive atom is joined to glucose (blood sugar). This allows doctors to see the chemical activity (digestion system) within the brain. Individuals with frontotemporal degeneration exhibit reduced chemical activity (hypo metabolism) in the frontal and transient regions of the brain, a finding that might distinguish the clutter from Alzheimer sickness. More these days, an amyloid PET tracer has been developed that can bind to misfolded amyloid proteins accumulated in the mind. This can assist or rule out a diagnosis of Alzheimer's disease which can then be ruled out during the diagnostic process for FTD. A spinal tap (lumbar puncture) may be performed to measure amyloid and tau proteins. This test can moreover back or decrease the probability of an Alzheimer's diagnosis.

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