Introduction

Frontotemporal lobar degeneration (FTLD) is a clinically and pathologically heterogeneous syndrome, characterized by progressive decline in behaviour or language associated with degeneration of the frontal and anterior temporal lobes[1]. The original cases described by Arnold Pick and Alois Alzheimer demonstrated neuronal inclusions that were later shown to be tau-positive at histopathology[2]. FTLD has only recently been appreciated as a leading cause of dementia, particularly in patients presenting before the age of 65 years.

Classification and symptoms

Three distinct clinical variants of FTLD have been described: (i) Behavioural-variant frontotemporal dementia, characterized by changes in behaviour and personality in association with frontal-predominant cortical degeneration; Behaviour deficits can be either impulsive (disinhibited) or bored and listless (apathetic) and includes inappropriate social behaviour; lack of social tact; lack of empathy; distractability; loss of insight into the behaviours of oneself and others; an increased interest in sex; changes in food preferences; agitation or, conversely, blunted emotions; neglect of personal hygiene; repetitive or compulsive behaviour, and decreased energy and motivation. (ii) Semantic dementia, also referred to as the temporal-variant of FTLD, is characterized by a fluent, anomic aphasia and behavioural changes in the setting of marked, often asymmetric degeneration of the anterior temporal lobes, also a syndrome of progressive loss of knowledge about words and objects associated with anterior temporal neuronal loss; and (iii) progressive nonfluent aphasia, characterized by effortful language output, loss of grammar and motor speech deficits in the setting of left perisylvian cortical atrophy, language disturbance. Spatial skills and memory remain intact[3].

Diagnosis

Structural MRI scans often reveal frontal lobe and/or anterior temporal lobe atrophy but in early cases the scan may seem normal. Atrophy can be either bilateral or asymmetric. Registration of images at different time points of time (e.g., one year apart) can show evidence of atrophy that otherwise (at individual time points) may be reported as normal. Many research groups have begun using techniques such as magnetic resonance spectroscopy, functional imaging and cortical thickness measurements in an attempt to offer an earlier diagnosis to the FTD patient. Fluorine-18-Fluorodeoxyglucose Positron Emission Tomography (FDG-PET) scans classically show frontal and/or anterior temporal hypometabolism, which helps differentiate the disease from Alzheimer's disease. The PET scan in Alzheimer's disease classically shows biparietal hypometabolism. Meta-analyses based on imaging methods have shown that frontotemporal dementia mainly affects a frontomedial network discussed in the context of social cognition or 'theory of mind'. This is entirely in keeping with the notion that on the basis of cognitive neuropsychological evidence, the ventromedial prefrontal cortex is a major locus of dysfunction early on in the course of the behavioural variant of frontotemporal degeneration. The language subtypes of frontotemporal lobar degeneration (semantic dementia and progressive nonfluent aphasia) can be regionally dissociated by imaging approaches in vivo[4].

Treatment
Currently, there is no cure for FTLD. Treatments are available to manage the behavioural symptoms. Disinhibition and compulsive behaviours can be controlled by selective serotonin reuptake inhibitors (SSRIs).

**Prognosis**

The outcome for people with FTLD is poor. The disease progresses steadily and often rapidly, ranging from less than 2 years in some individuals to more than 10 years in others. Eventually some individuals with FTLD will need 24-hour care and monitoring at home or in an institutionalized care setting.

**Further Information and Supports**

National Dementia Helpline on 1800 100 500.

http://www.fightdementia.org.au/content/fronto-temporal-lobar-degeneration

RACGP - Australian Family Physician

Frontotemporal dementia Features, diagnosis and management


Neuroscience Research Australia

https://www.neura.edu.au/health/frontotemporal-dementia

UCSF Memory and Aging Centre

http://memory.ucsf.edu/ftd/

**References**


