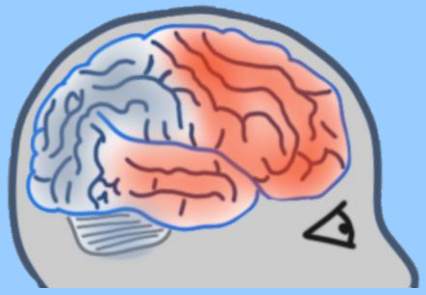
Frontotemporal Dementia

An uncommon form of dementia which causes problems with behaviour and language



Epidemiology

Typically ***earlier onset*** than other dementias (mean 53-58 years)

Aetiology

Focal degeneration of the frontal and temporal lobes

Positive family history in many

Genetic links found:

- Microtubule associated protein tau ***(MAPT)*** and progranulin ***(PGRN)*** gene mutations
  - <20% cases
  - Both Chr 17

Linked with ALS

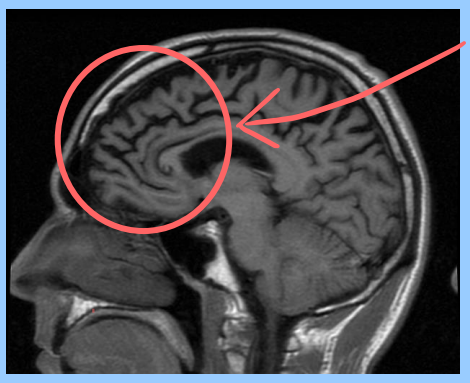
- Gene on Chr 9
- Expanded repeat of a non-coding region in familial ALS often associated with FHx of FTD

Pathophysiology

Not fully understood

Definitive diagnosis only possible at post-mortem exam

- Patterns of neuronal injury with intra-neuronal and glial cell ***inclusions***
- Inclusions can be ***tau*** positive (FTD-Tau) or tau negative (FTD-U)
- FTD-U falls under the family of TDP-43 proteinopathies
  - Associated with mutation in the PGRN gene



Advanced frontotemporal atrophy with thinned cortex

Diagnosis

A ***third-party*** personal and family history should be obtained

Common features:

- ***Insidious onset*** of coarsened personality and social behaviour
- Progressive loss of language fluency or comprehension
- Progressive self neglect and abandonment of work, activities and social contacts

Changes in personality/ language may precede memory impairment

Cognitive testing should be performed at baseline then at 6 month intervals:

- ***MMSE*** / MOCA
- May be normal at presentation
- Useful for prognosis

If associated with muscle wasting and weakness may be ALS

CT, MRI or PET will show characteristic regional atrophy

Post mortem examination if familial link suspected

Classification

2 main classifications:

- ***Behavioural FTD***
- ***Primary progressive aphasia***

Can present ***+/- parkinsonisms***

Soton Brain Hub

Management

Similar to other dementias

- Ensure ***safety*** of the patient and others
- Provide reassurance, guidance and supervision to the patient ***and their family***

Respite and residential care

Supportive care

Education and planning

- Early planning maximises the patient's ability to participate

Pharmacological interventions

Largely untested

- Only considered ***if non-pharmacological approaches fail***

May be required for specific behavioural problems

End of life care

FTD will eventually result in profound disability and death

Monitoring

Carer burden and stress will be high and should be assessed at each visit

Comprehensive cognitive and behavioural evaluations should be completed ***every 6-12 months***

Prognosis

Progression is ***variable***

Behavioural variants ***progress rapidly***

Median survival is 80 months from diagnosis

Shorter survival and ***faster decline than Alzheimer's***