Frontotemporal dementia

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Dementia has different causes

- Alzheimer’s disease
- Vascular dementia
- Frontotemporal dementia
- Lewy Body dementia
Frontotemporal dementia - the basics

- Neurodegeneration affects *frontal* and *temporal* lobes
- Affects behaviour and language
Frontotemporal dementia - the basics

- Neurodegeneration affects *frontal* and *temporal* lobes
- Affects behaviour and language
- Previously known as ‘Pick’s disease’
- Second most common cause of dementia <65 yrs
- 20,000 people with FTD in UK

Arnold Pick (1851-1924)
Frontotemporal dementia - the basics

Frontal lobe

Temporal lobe

Frontotemporal dementia

Behavioural variant (bvFTD)

Primary Progressive Aphasia (PPA)

Semantic dementia (SD)

Progressive non-fluent aphasia (PNFA)
Behavioural variant FTD

- 'Susan' - 58 year old taxi driver
- Careless driving, 'road rage'
- Singing in public
- Disinterested in others
- Obsessive Sudoku
- Eating chocolate biscuits
- Mother died of MND

Slowly progressive changes in behaviour
Often misdiagnosed initially
Family history is common
Motor features (MND, parkinsonism) common
Semantic dementia

- John - 56 year old engineer
- 4 years progressive language problems
  - finding words
  - understanding speech
  - recognising objects/people
- No difficulties driving or doing finances
Semantic dementia

- 'John' - 56 year old engineer
- 4 years progressive language problems
  - finding words
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  - recognising objects/people
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Progressive loss of ‘semantic’ knowledge
Memory for events and navigation OK
Left temporal lobe of the brain
Changes in behaviour come later
Progressive non-fluent aphasia

- 'Margaret': 68 year old
- 3 years progressive problems:
  - speaking is 'hard work'
  - mispronunciation of words
  - shorter sentences
- Otherwise completely independent

Progressive speech difficulty
Understanding of speech OK
Left hemisphere damage
Frontotemporal dementia - the basics

- Frontal lobe
- Temporal lobe

Frontotemporal dementia

- Behavioural variant (bvFTD)
- Primary Progressive Aphasia (PPA)

- Semantic dementia (SD)
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What causes FTD?

Frontotemporal dementia

- Tau: 40%
- TDP-43: 50%
- FUS: 5-10%

PNFA

SD

behavioural variant FTD

40% 50% 5-10%
Genetics

Behavioural variant FTD

• 1 in 10 cases is clearly familial

• 3 key genetic mutations identified:
  • Chromosome 17: Tau (MAPT): <65 yrs at onset
  • Chromosome 17: Progranulin (GRN): often >65 yrs at onset
  • Chromosome 9: C9ORF72 expansion

• C9ORF72 especially in FTD-MND cases
Summary

• FTD is a common cause of younger-onset dementia

• Diagnosis can be difficult

• Patients and carers have specific needs

• Recent advances in FTD research uncovering neurodegeneration pathways