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The radiological diagnosis of frontotemporal dementia in everyday practice: an audit of reports, review of diagnostic criteria, and proposal for service improvement

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AIM: To investigate how commonly valuable diagnostic information regarding the frontotemporal dementias (FTDs) may be missed on routine radiological reporting.

MATERIALS AND METHODS: The magnetic resonance imaging (MRI) examination results of a series of 39 consecutive patients in whom the diagnosis was initially thought to be a form of FTD were audited. Twenty-two patients satisfied formal diagnostic criteria for subtypes of FTD. The initial non-specialist radiological reports of the MRI examinations were compared with those of a radiologist who specifically examined the images for the possibility of atypical dementia.

RESULTS: Six of the 22 original reports provided a full and accurate description of the radiological findings, while two provided a fully accurate interpretation.

CONCLUSION: Valuable diagnostic information may be missed unless clinicians and radiologists jointly review and discuss brain imaging in cases of dementia. The use of standardised scales may enhance the reporting of MRI examinations for dementia.

Introduction

The frontotemporal dementias (FTDs) are a clinically and pathologically complex group of disorders typified by selective degeneration of the frontal and temporal lobes.[1] They account for 10–20% of cases of early-onset dementia[2] with a mean age onset of 58 years.[3] The term refers principally to four clinically distinctive syndromes: behavioural variant FTD (bvFTD), semantic dementia (SD), the fluent form of primary progressive aphasia, and two non-fluent varieties, agrammatic and logopaenic aphasia, each of which can have a range of underlying pathologies.[4] Corticobasal syndrome (CBS) and progressive supranuclear palsy (PSP) are often considered forms of FTD because of their related pathology and some overlap in clinical presentation. Motor-neuron disease (MND) associated frontotemporal dementia also belongs to this family of disorders but does not figure in the case series audited here.

BvFTD (Table 1, Figure 3)

BvFTD, also known as the frontal variant, is characterised by a progressive deterioration in social functioning and personality.[5] Patients usually present with symptoms such

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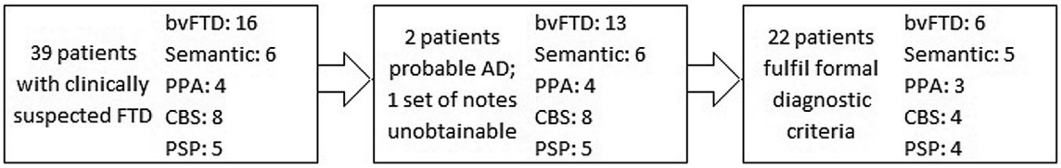

Figure 1 The diagnoses of patients included in the audit.

as loss of empathy, disinhibition, and impulsive and socially inappropriate behaviour.[6] It is associated with frontal lobe degeneration progressing into the temporal lobes, usually evident on brain MRI. Indeed bvFTD is the only variant of FTD in which characteristic imaging is required for diagnosis.

Primary progressive aphasia

SD (Table 2, Figure 4)

SD typically presents with difficulty in finding words for unfamiliar items, without loss of language fluency, progressing to more severe word-finding difficulties and loss of semantic knowledge, for the items themselves as well as for their names. It is usually accompanied by asymmetric atrophy of the temporal lobes, worse on the left.[7]

Agrammatic and logopaenic aphasia (Tables 3 & 4, Figures 5 & 6)

In contrast to SD, which gives rise to a fluent form of language disorder, FTD can also present with a non-fluent language disturbance, often with prominent problems with grammar, word-finding, and repetition.[8] Imaging may show atrophy surrounding the left Sylvian fissure ("perisylvian atrophy"). Agrammatic (also known as progressive non-fluent aphasia) and logopaenic varieties of non-fluent primary progressive aphasia are distinguished, and discussed further below.

CBS (Table 5, Figure 7)

CBS typically presents with a dyspraxic limb, usually an arm, associated with extrapyramidal features, such as a "cog-wheel" increase in tone and loss of facial expression.[9] The "dyspraxia" is typically evidenced by difficulty in copying unfamiliar hand positions and/or difficulty in performing mimes. CBS is often associated with additional cognitive features: as well as the typical presenting disorder of praxis, there are often impairments of language and executive function (the ability to organise thought and behaviour). These cognitive features can predominate at the outset. Focal cortical atrophy affecting frontal and parietal regions is usually observed on neuroimaging.[10]

PSP (Table 6, Figure 8)

PSP is characterised clinically by supranuclear gaze palsy (difficulty in initiating eye movements voluntarily with preserved reflex movements, often accompanied by slowing of pursuit movements), problems with balance and symmetric akinesia.[11] Cognitive features, including subcortical dementia and language disturbance are also common. Midbrain atrophy is the characteristic neuroimaging finding.

Audit of radiological findings

The point of departure for this audit study was the observation that subtle but relevant changes on initial brain imaging were often not mentioned in the initial radiological report and only became known in the neuroradiology review meeting. Consequently, the present study was undertaken to investigate how commonly valuable diagnostic information regarding FTDs may be missed on routine radiological reporting.

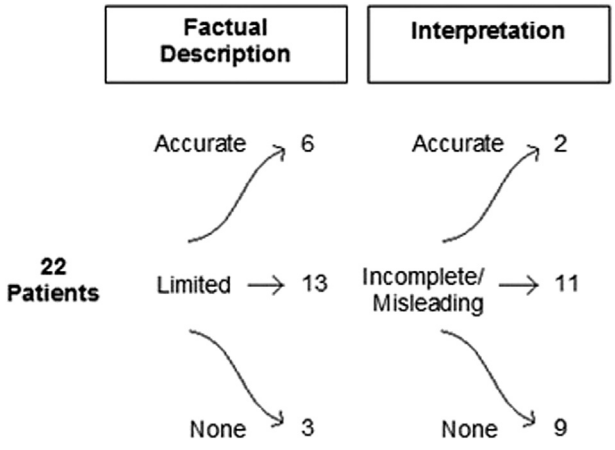

Figure 2 Reports assessed for accuracy of factual findings and helpfulness of the interpretation of findings.

Materials and methods

Thirty-nine consecutive patients were identified who attended a cognitive disorders clinic over a period of 7 years in whom the initial diagnostic suspicion was of a form of FTD, as defined in the Introduction. The clinical features of these patients were recorded using a standardised proforma. Three patients were excluded because they developed clinical features suggestive of Alzheimer's disease (2) or because the notes were unobtainable (1). In the remaining 36 patients, the clinical features were compared with accepted formal diagnostic criteria (as specified in the Discussion). Only those patients (n=22) satisfying formal diagnostic criteria for FTD were included in the neurological and radiological review (Fig 1).

The MRI reports and images from these 22 patients were obtained, which were performed at two hospitals in the southwest of England. There were some variations in the