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Semantic Variant Frontotemporal Dementia: A Case Report

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Abstract

Background: Frontotemporal dementia (FTD) is a neurodegenerative condition marked by progressive impairments in behavior, executive function, or language, and is a leading cause of early-onset dementia, affecting 4% of the dementia population and up to 30% of cases before age 65. FTD presents significant diagnostic challenges due to its neuropsychiatric symptoms mimicking psychiatric disorders. It manifests in three variants: behavioral-based, primary progressive aphasia, and semantic-type dementia. Typically, the disease spans 6-8 years from onset to death, with symptoms like apathy, disinhibition, and agitation.

Case report: The case report details a 55-year-old woman who displayed initial symptoms of forgetfulness and disorganized behavior. Despite normal MRI results, further evaluations indicated impaired verbal memory and significant hypometabolism in brain regions consistent with FTD, leading to a diagnosis of the semantic variant of FTD. Diagnosing FTD is complex due to symptom overlap with psychiatric disorders, especially in early stages where cognitive deficits may not be pronounced. Advances in imaging and molecular characterization have improved diagnostic accuracy. The prevalent manifestation of the semantic variant includes language abnormalities and word-related memory loss, while executive dysfunctions are typical. Misdiagnosis can delay effective treatment, highlighting the importance of considering FTD

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in middle-aged patients with new neuropsychiatric symptoms.

Conclusion: The intricate relationship between FTD and other neurodegenerative or psychiatric conditions further complicates diagnosis, underscoring the necessity for thorough neurodegenerative disease assessments in relevant patients.

INTRODUCTION

Frontotemporal dementia (FTD) is a neurodegenerative condition characterized by progressive impairments in behaviour, executive function, or language (1). FTD stands out as one of the primary causes of early-onset dementia, being present in 4% of the dementia population, and accounting for 20-30% of dementia cases before the age of 65 (1, 2). FTD usually manifests through a gradual alteration in behaviour, personality, and/or language abilities. Neuropsychiatric symptoms can mimic psychiatric disorders, making diagnosis difficult and potentially leading to misdiagnosis (3, 4). Each variant can present its own challenges in diagnosis. Three clinical variants exist: behavioural-based, primary progressive aphasia, and semantic-type dementia. The average duration of the disease from symptom onset to death is 6-8 years (5, 6). FTD is characterized by a range of neuropsychiatric symptoms such as apathy, disinhibition, agitation, aggression, eating disorders, and other behavioural abnormalities (7, 8). The following case report discusses a patient who was referred to our outpatient clinic due to depression with psychotic features and was recently seen at a neurological outpatient clinic. The discussion will be conducted with the consent of the patient's relatives.

CASE REPORT

The report details the symptoms displayed by a 55-year-old, housewife, woman who had not previously been admitted into psychiatric care. The woman first showed symptoms, such as forgetfulness and frequent use of the term "thing," approximately 9 months ago. At the suggestion of her son, the woman sought professional help from a psychologist based in the city where she resided. The patient was referred to the neurology outpatient centre of the same hospital as he did not score well enough in the neuropsychological test. Neurologic evaluation performed in neurology department inpatient clinic of hospital. A brain MRI scan was performed, and he received confirmation that there was no pathology found. Upon receiving information from his son about the patient's recently increased disorganised behaviour and suspicions regarding his relatives, he was referred to the psychiatry outpatient centre. She was hospitalised in the psychiatry service for a differential diagnosis of dementia and major depressive disorder with psychotic features because she spoke less and had difficulty in finding words.

During the admission interview, it was observed that she had difficulty in naming objects but her word fluency was not affected much. Repetition was preserved in the patient who was able to take single orders. Neuropsychological testing was performed and verbal memory was found to be impaired with a minimum test score of 14/30. A brain PET scan has shown symmetrical, diffuse hypometabolism in both parietotemporal lobes and the prefrontal cortex. The FDG brain PET/CT imaging displayed the maximum intensity

projection of both lateral MIP images, highlighting areas of hypometabolism in the frontal, temporal and parietal lobes with yellow, blue and red stars, respectively. A semantic variant of frontotemporal dementia was considered as the most likely diagnosis and the patient was referred to a dementia outpatient clinic.

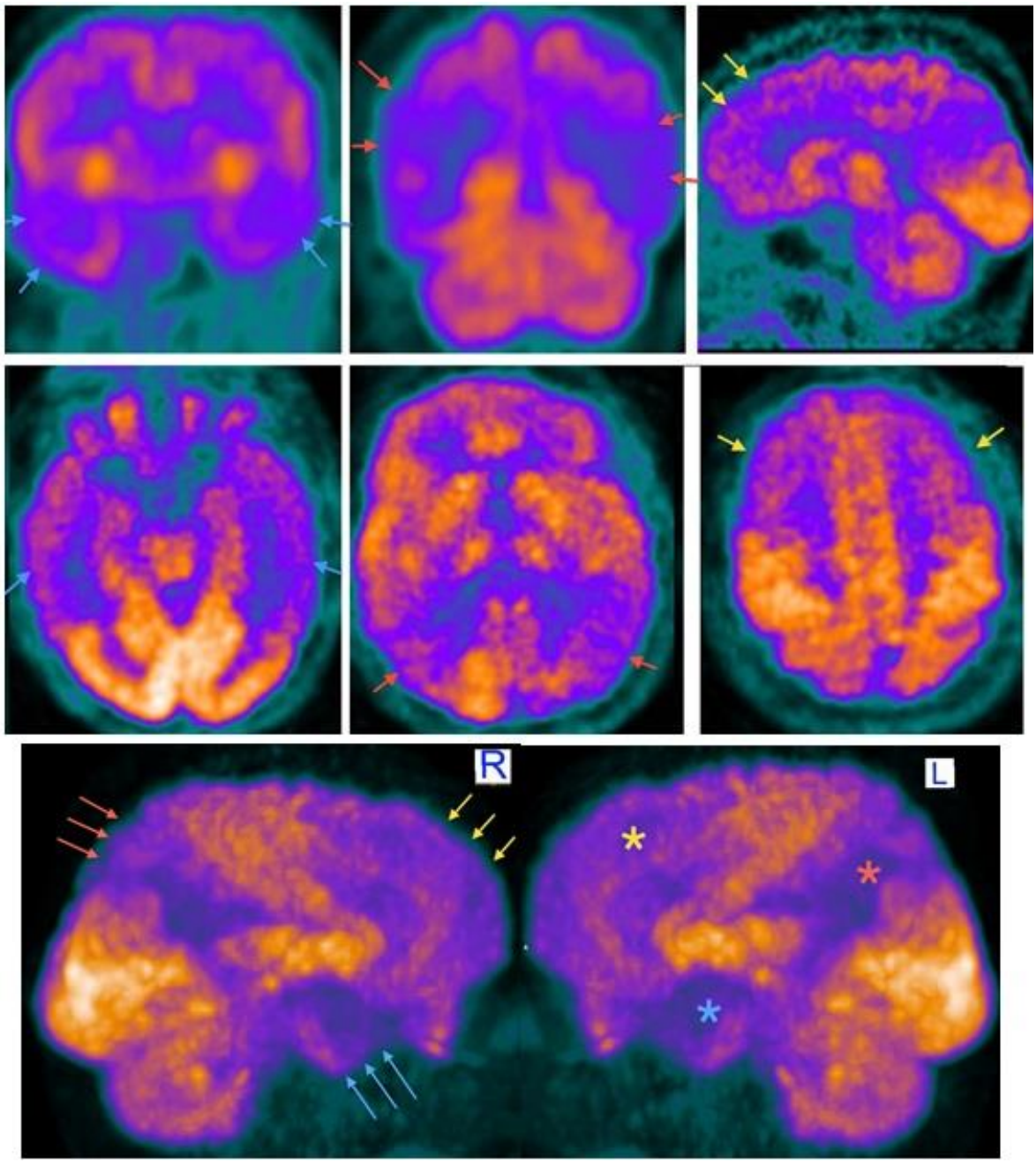

Figure 1. PET images of patient with Frontotemporal Dementia
Cross-sectional image: in coronal, sagittal and axial sections, arrows point to areas of hypometabolism; Yellow: Frontal, Blue: Temporal, Red: Parietal lobe.

DISCUSSION

The assessment and differential diagnosis of FTD pose considerable difficulties owing to the overlapping of clinical symptoms with primary psychiatric illnesses. Progresses in clinical, imaging and molecular characterisation have enhanced the precision of FTD diagnosis, enabling discrimination between FTD and psychiatric disorders (1). he challenge of diagnosing dementia in patients with severe psychiatric disorders across the lifespan is particularly complex in cases of behavioural variant frontotemporal dementia (bvFTD). This is because patients may not display gross cognitive deficits, especially in the early stages (9). Additionally, the differential diagnosis of FTD encompasses other neurodegenerative dementias, vascular and other brain-affecting conditions, and psychiatric disorders (10).

The prevalent manifestation of semantic-type FTD involves a language abnormality with word meaning or word-related memory loss, associated with bilateral atrophy of the mid-inferior temporal cortex. The executive dysfunctions typical of FTD comprise inadequacies in planning, reasoning, problem solving, organisation, attention, abstraction, and mental flexibility. The fundamental proficiencies of language, elementary visual perception, and memory remain well-preserved. However, frontal executive function test results suggest poor performance (5).

Patients with young-onset FTD may receive a diagnosis of a psychotic disorder years prior to being diagnosed with dementia, highlighting the intricacy of differential diagnosis in psychiatric settings (11). The loss of social emotions and severely disordered social behaviour in FTD could be advantageous factors in the differential diagnosis (2). The diverse clinical manifestation of FTD, particularly its behavioural variant, possibly coincides with not only other neurodegenerative ailments,but also primary psychiatric ailments, resulting in further complications in the differential diagnosis (3). Misdiagnosis in psychiatry can cause postponed and ineffective therapy, underscoring the significance of conveying the patients exhibiting new-onset neuropsychiatric symptoms for neurodegenerative disease assessment (12).

Efficient organisation of the brain's functional network has been proposed as a means of supporting cognitive wellbeing, even when there are presymptomatic reductions in both atrophy and connectivity in FTD. Such research has the potential to inform diagnostic approaches (13). Furthermore, there is evidence to suggest that amyotrophic lateral sclerosis (ALS) and FTD comprise a range of disorders that encompass both motor and cognitive dysfunctions. This highlights the intricacies involved in making a differential diagnosis for FTD (14).

Psychiatric symptoms may occur first in these patients, and the disease progresses rapidly. Therefore, it is important to carefully consider the possibility of FTD in middle-aged patients who present with mood and behavioural changes, and not dismiss this diagnosis early on.

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