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Case Report Open Access

Behavioral-Variant Frontotemporal Neurocognitive Disorder Presenting as Insomnia

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Introduction

Frontotemporal Neurocognitive Disorder (FTNCD) is a neurodegenerative disorder with various subtypes. Behavioral symptoms of FTNCD include apathy, disinhibition and impulsivity, socially inappropriate behavior, compulsive/ritualistic behavior, and dietary changes [1]. Language deficits present as loss of fluency with obvious word finding difficulties or fluent speech with loss of semantic meaning [2]. Cognitive symptoms are executive function deficits with early sparing of visual-spatial skills [1]. Here we report a case with behavioral-variant FTNCD (bv-FTNCD) presenting as insomnia to highlight the issue of early detection for immediate management to improve prognosis.

Case History

This patient is a 57-year-old female diagnosed as having primary insomnia 8 years ago and having received Lorazepam 2 mg and Trazodone 50 mg since then. No history of major mental or physical illnesses was reported and she denied any substance abuse. She was an elementary school graduate and worked in the service industry until age 55. One year ago, she gradually became more passive and avoidant of daily activities instead of her pre-morbid dominant and egocentric traits. She would sit and watch TV for an entire day, in contrast to her previous active engagement with household and self-care tasks. Dietary changes were noted, as her variety of food choices were limited to dumplings or noodles. Irritability and emotional dysregulation was noted when her daughter encouraged her to perform physical and self-care activities. Her sleep-wake cycle changed and sleep became fragmented. She took sedatives at multiple time points in a day. One month ago, she was brought to our clinic in a general hospital. MSE (mental status exam) was performed. She was fully alert and was aware of time/person/place. Attitude was cooperative. No impairment in attention focusing/shifting/maintenance was noted. Her affect was mildly anxious and irritable. No bizarre/agitated behavior was seen. Her speech was mostly relevant, coherent, and fluent with minor delays in response latency. She denied all suicide/homicide ideation, persecutory/reference delusions, and all perceptual disturbances. No focal neurological deficits were noted on examination. Her mini-mental-status examination (MMSE) score showed 24/30 (test/overall score), with major impairment on recall (0/3) and mild on naming (1/2) and calculation (4/5). She rated a score of 1 on the Clinical Dementia Rating (CDR). Her social role at home and daily hobbies were greatly impaired as well as impairments with memory, judgment, and problem-solving. She was then admitted for further management. At admission, brain magnetic-resonance imaging showed significant prominence of the bilateral frontal-temporal sulci and bilateral Sylvian fissures (Figure 1). Other exams such as laboratory tests and Electroencephalograph (EEG) revealed findings within normal limits. She scored 69.2 on the Cognitive Abilities Screening Inventory (CASI) with obvious impairment on short-term memory, working memory and abstract thinking. On the Wisconsin Card Sorting Test, she made 5 perseverative mistakes (>99th percentile), and 8 non-perseverative errors (86th percentile). She spent 56 seconds on the trail A test (mean completion time 29 seconds, >78 seconds may indicate executive memory deficit). The Trail B test was not completed due to her lack of knowledge of the English Alphabet. She was diagnosed with probable bv-FTNCD. Psychoeducation, including diagnosis, prognosis, and management, was given to her family members.

Discussion

Frontotemporal Neurocognitive Disorder describes a spectrum of neurocognitive disorders, mainly involving the frontal and temporal lobes. Variants include behavioral-variant FTNCD (bv-FTNCD), and the semantic, non-fluent and logopenic subtypes of primary progressive aphasia (PPA) [1]. Around 85-90% of FTNCD cases have cellular inclusion bodies of either Tau protein (FTLD-Tau) or TDP-43 (FTLD-TDP), while a smaller percentage of cases have the FTLD-Fus inclusion [2,3]. However, the logopenic subtype may have differing underlying neuropathological changes compared to other subtypes. The logopenic subtype of primary progressive aphasia features predominantly Alzheimer's disease neuropathology and may evolve to Alzheimer's disease later in the disease course [2,4]. The agrammatic/

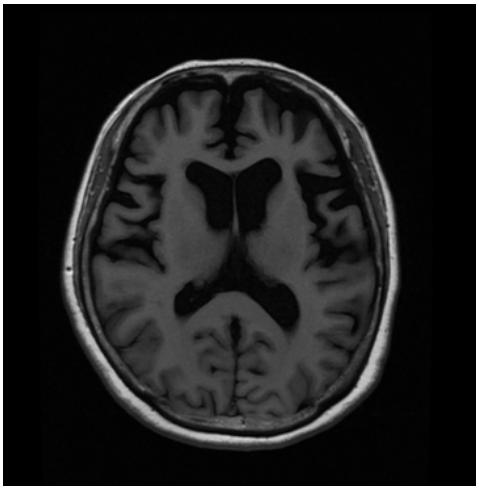

Figure 1: Axial T1-FLAIR image with predominant bilateral frontal and temporal lobe atrophy in the present case.

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nonfluent subtype of primary progressive aphasia has also been shown to progress to full FTNCD and in later stages of disease course present with signs of asymmetrical Parkinsonian features and corticobasal degeneration [5,6]. Diagnostic certainty for bv-FTNCD is defined as possible, probable, and definite based on six symptom clusters. The six clusters include behavioral disinhibition, apathy/inertia, loss of sympathy/empathy, perseverative/repetitive behavior, dietary changes, and a specific neuropsychological profile [1,7]. Neuroimaging evidence is necessary for probable FTNCD, with either frontal/temporal atrophy or hypo-perfusion/-metabolism of frontal areas. This patient did not receive histopathological confirmation and was not known to have a pathogenic mutation, therefore not meeting the criteria for definite FTNCD. The onset of bv-FTNCD is insidious and age of onset is from 50's to early 60's. In one Dutch study of 245 patients, prevalence was 3.6 per 100,000 at age 50-59 years, 9.4 per 100,000 at age 60-69 years and 3.8 per 100,000 at age 70-79 years [8]. Apathy/inertia is the most common initial presentation [1]. She became very passive toward daily and social activities near the beginning of her illness and required additional persuasion and encouragement. Patients with bv-FTNCD may have deficits in social cognition [9]. Our patient became disengaged with social activities, decreased eye contact, and became emotionally detached for the past one year. She also displayed altered food preference, increased craving for carbohydrate-rich foods, and a stereotyped meal requirement. Our patient displayed several bv-FTNCD symptoms during the early phase of her illness. Perhaps her chronic sleep disturbance was a prodromal problem. Fortunately, she did not display inappropriate, impulsive, or harmful behavior and her social support was adequate when she visited our clinic. She received prompt diagnosis and adequate pharmacological and non-pharmacological treatment. Clinicians need to be aware of the early symptoms/signs of FTNCD and management, including drug and non-drug therapies. This report not only reports the early signs of bv-FTNCD, but emphasizes to general psychiatrists and general clinicians that insomnia, a common chief problem among pre-senile patients, may be the first presentation of bv-FTNCD. This knowledge may assist with early diagnosis and may improve the outcome of patients with FTNCD.

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