A Case of Rapidly Progressing Frontotemporal Dementia

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ABSTRACT

Frontotemporal dementia commonly presents as a gradual change in personality, social conduct, and language ability, often sparing memory loss in early stages. We report a case of a 55-year-old female with rapid progression of memory impairment and other cognitive functions in a span of 2 years.

Key words: Dementia, frontotemporal dementia, rapid decline, rapid progression

INTRODUCTION

Frontotemporal dementia (FTD) is a leading cause of early onset dementia found in 4% of general dementia population and is present in 20%–30% of dementia patients younger than 65 years. FTD most commonly presents as a gradual change in behavior, personality, and/or language ability. Three distinct clinical variants have arisen: the behavioral based (behavioral variant FTD [bvFTD]), the language-based primary progressive aphasia, and semantic dementia. The mean disease duration from onset of symptoms till death is 6–8 years. The various neuropsychiatric symptoms associated with FTD are apathy, disinhibition, agitation and aggression, eating disturbances, and other behavioral abnormalities include repetitive stereotypical behaviors such as verbal perseveration, hoarding, and rituals.

CASE REPORT

A 55-year-old married female, premorbidly well adjusted, presented to outpatient department with a 2-year history of insidious onset of altered behavior such as wandering away from home to a nearby lake and coming back with a bag full of waste items.

She also had inappropriate smiling whenever she was asked something or spoken to.

From anecdotal evidence, she was suspected to have obsessive and compulsive behavior and treated in view of obsessive-compulsive disorder (OCD) for a brief period of 2 weeks; however, further details were unavailable.

In the following months, her husband reported increased progression of unusual behavior, errors while doing daily

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household chores such as cooking and washing clothes, and difficulty in self-care. She struggled to wear sari or recollect where she kept her belongings.

Her symptoms worsened over the next 18 months, and she depended completely on her husband to perform domestic work. She was unable to recollect her husband’s name and could not recognize her children.

For the past 6 months, she had completely stopped doing household chores and required assistance for self-care such as brushing, bathing, and dressing. She also had reduced speech.

There was no family history of similar disorder. On examination, primitive reflexes, apraxia, agnosia, and executive dysfunction were present.

On mental status examination, she was alert, rapport was established with difficulty, attitude toward examiner was playful and had increased nongoal directed psychomotor activity. She had impaired attention, orientation, immediate, recent, and remote memory. In language, her fluency, comprehension, naming, repetition, reading, and writing were impaired. She also had impaired social conduct, thought impoverishment, perseveration, inappropriate affect, and lack of insight.

A magnetic resonance imaging of brain showed neuroparenchymal atrophic changes seen involving the frontal, temporal, and parietal lobes bilaterally with opening up of sulci and volume loss of gyri. Atrophic changes were also noted within the temporal lobe with widening of choroid fissure, widening of temporal horns of both lateral ventricles, and hippocampal volume loss. Both lateral ventricles, third and fourth ventricles were dilated.

**DISCUSSION**

The most common presentation is with a change in personality and impaired social conduct (bvFTD). The profound impairment in social functioning, decline in personal hygiene and mental rigidity, and distractibility also point toward bvFTD. Stereotypies, hoarding, or compulsive acts are, sometimes, among the presenting symptoms of FTD and could be mistaken for OCD as reported in a previously published case report.

As a rule in FTD, memory loss is mild in the early stages, unlike this case where the patient had profound memory impairment which worsened within a short span of 2 years. Most studies show that FTD is steadily progressive, with declining function in everyday life and accumulation of social, cognitive, and neurological disabilities leading to complete dependency requiring institutional care over a course of 6–8 years. In our case, the cognitive decline progressed rapidly within short span of 2 years since symptom onset.

Prion diseases, in particular, Creutzfeldt–Jakob disease is reported to be the most common cause of rapidly progressive dementia. Other common causes are vasculitis, sarcoidosis, encephalopathies, infections, and neoplasms, all of which were unlikely however further evaluation was not possible due to the patient’s financial constraints.

From this case, we conclude that FTD may present with memory impairment early in the course with rapid progression and complete loss of cognitive functions within an unusually short span.

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**Conflicts of interest**

There are no conflicts of interest.

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