## Case report

# Rapidly progressive young onset dementia: an atypical presentation of a confounding clinical scenario

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**Summary**

Rapidly onset progressive dementia representing frontotemporal variety had a relentless debilitating impact which sometimes presented with psychological and behavioral symptoms leading to diagnostic dilemma. An adult male had been admitted into the hospital with the complaints of progressively odd and inappropriate social behavior, occasional self-muttering, confusion regarding his surroundings and lack of spontaneous speech for around 9 months. Initially, the patient was diagnosed as schizophrenia comorbid with delirium and treated accordingly. However, no improvement was apparent and the diagnosis was reviewed. Magnetic resonance imaging (MRI) of the brain showed significant cerebral atrophy particularly in the frontal and temporal lobes and finally the case was diagnosed as early onset frontotemporal dementia. The clinician should be cautious about the psychiatric manifestations of frontotemporal dementia to avoid diagnostic error and to manage the patients with more specification from the beginning.

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### Introduction

Rapidly progressive onset dementia referred to disorders in which the progress is devastatingly fast often within 1-2 years.[1] These heterogenous group of disorders includ Alzheimer's disease[2] as well as Lewy body dementia[3]. Creutzfeldt-Jakob disease is also included in the aforementioned group of disorders.[4] The diagnosis of dementia can be incredibly upsetting for patients and the patient party but it can prove to be particularly emotionally challenging if the patient is very young.

The author herself recently felt emotionally taxed as she dealt with a rapidly progressive case of young onset dementia. While such cases are rare in clinical presentation, extensive literature might prove to be of some use to identify a pattern with such patients which might lead to improved rapidity in diagnosis and treatment. Frontotemporal dementia might initially present with nonspecific behavioral complaints that might lead to misdiagnosis. Additionally, in such cases such as the one discussed herein, the course of deterioration might be rapid. Hence, there might be a need to repeat imaging studies despite the cost.

### Case summary

A 40 years old married man having a child who was a van driver had been suffering from gradual onset of psychiatric problems for the 9 months. According to the statement of his father, he was introverted in nature. Initially, he began to speak less and his family members assumed he was becoming more introverted than before. He would begin to avoid having sexual intercourse with his wife and he showed a lack of interest in his wife. He also began to exhibit some odd, socially inappropriate behavior which was unlike himself e.g. he would suddenly clap in an empty room or abruptly leave when someone would try to have a conversation with him. At times, he was seen to muttering by himself and he felt confused when people would ask him simple questions. A few times, he would claim that his father was actually his uncle. His family then took him to some physicians; following consultations and medication, there was no amelioration in his behavior and rather his problems seemed to worsen. Investigations such as complete blood count and liver function tests showed nothing contributory and the computed tomography (CT) scan of the brain was shown to be normal. Following the patient's admission, the treating physicians had trouble establishing rapport with the patient and neurological examinations were carried out with difficulty. The patient showed some behavioral problems during the initial part of his hospital stay i.e. he would steal food from the bed of his fellow patients and eat indiscriminately, he would also try to suddenly leave the ward and despite being shown the direction of the toilet many times, he would forget the way. Initially, it was suspected that the patient might have had schizophrenia with comorbid delirium as he had been prescribed multiple antipsychotics. Later, the diagnosis was set as delirium due to use of psychotropics. The patient's previous medication was stopped and he was then treated with low dose risperidone, but no improvement in excessive and inappropriate eating habits took place. Low dose

aripiprazole was then added which lead to end to his inappropriate behavior but not the other inappropriate behaviors. Then mini mental state examination (MMSE) was performed as the patient became less restless and the score was very low. The decision was then taken to repeat the neuroimaging studies despite the fact that around 6 months back the patient's CT scan showed no abnormal findings. When the patient's magnetic resonance imaging (MRI) was done, there was remarkable shrinkage of the gyri and deepening of the sulci i.e. cerebral atrophy in the frontotemporal regions, the lateral ventricles were also much dilated. The human immunodeficiency virus (HIV) antibody test and the venereal disease research laboratory (VDRL) tests were done but they were negative. Other relevant investigations showed no remarkable findings. Genetic testing was advised but refused by the patient party. Finally the case was diagnosed as early onset frontotemporal dementia. It was to be noted that the patient's father did not report any family history of any kind of psychiatric illness including dementia.

## Discussion

Young onset dementia had been described as dementia that occurs before 65 years of age; variants such frontotemporal dementia had even reported of having an onset after 40 years of age.[5] Neuroimaging studies were invaluable and require repetition as the course of early onset dementia might be rapid, which might be rare as in cases associated with Creutzfeldt Jacob disease[6] or HIV[7]. Furthermore, rapidly progressive dementia seen under the age of 65 had been observed due to neurosyphilis and due to hereditary causes [8] i.e. genetic mutations.[9] However, more importantly as it was likely in this particular case, frontotemporal dementia might be idiopathic[10] in nature which might be difficult and confounding factor which also might delay proper diagnosis ultimately.

Frontotemporal dementia presented with behavioral disturbances,[11] and it was rare condition[12] which was even rarer in younger people[13]. Thus, as seen in this case, it might lead to very unfortunate misdiagnosis as psychiatric disorder of a wide variety could present with behavioral disturbances including psychotic disorders[14] and delirium[15]. Reports of the patient's investigation profile, sexual history and family history did not reveal anything of note; and the patient party was not interested in doing further genetic testing.

The rarity of rapidly progressive dementia in the young leaded to delay in the diagnosis and treatment of the patient who might have derived some benefit, no matter how little, from pharmacological and nonpharmacological interventions. Thus, the topic of rapidly progressive young onset dementia absolutely had more awareness in the society of physicians as the disease caused tragic devastation not just for the patient in question but also the respective caregivers who themselves required support.

## Conclusion

Rapidly progressive young onset dementia was still a life-threatening clinical term where specific management was so far to be developed. The diagnosis was crucial for the physicians to design plan to handle the situation where psychiatric manifestations might create confusion. Clinicians as well as mental health professionals should have to be cautious about the phenomenon.

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