When a close person suddenly changes their behavior – dementia in young people. A case report

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ABSTRACT

The aim of our study is to present a case report of a young patient with rapidly progressive behavioral changes. A 39-year-old well functioning man in a short period of time began to show symptoms like aggressive towards his family members, binge eating, apathy, difficulties with social interactions, reduced verbal fluency and poverty of speech, disorders in executive functions and restlessness and anxiety. This behavioral changes appeared in a short period of time (6 months). The concerned family, alarmed by this unusual behavior not previously observed, sought medical help. The patient was disoriented, not receptive to arguments about his atypical behavior, and unaware of the existing problem. A comprehensive diagnostic evaluation (including laboratory, imaging, and neuropsychological assessments) was conducted during the hospital stay. MRI scan showed substantial cortical atrophy in frontal and temporal lobes. Based on the clinical status, typical MRI and neuropsychological examination, the patient was diagnosed with frontotemporal dementia (FTD).

The patient received a diagnosis in a very short period of time, which is not typical in establishing a diagnosis for this particular medical condition. The symptoms are often nonspecific, which delays the diagnosis or leads to an incorrect preliminary diagnosis of another medical condition. Difficulties in diagnosing also arise from the non-specific symptoms, which are often overlooked by the patient themselves. It is important to emphasize the crucial role of family members and close associates in recognizing the initial symptoms and making attempts to diagnose the disease. It is also important to provide support and care for the patient's family, for whom the diagnosis of FTD becomes challenging and burdening.
INTRODUCTION

Frontotemporal dementia (FTD) is a group of neurodegenerative disorders, being the second most common cause of dementia, typically diagnosed between the ages of 45 and 65 [1]. FTD is characterized by progressive disturbances in behavior, language, and executive functions. There are two clinical forms that can be distinguished:

1. Frontal (behavioral) variant: It is characterized by antisocial behavior, stereotypes, perseveration, compulsions, disinhibition, impulsivity, hyperorality, voracity, and increased sexual drive.

2. Temporal variant - dividing into SD- semantic dementia (loss of word meaning and ability to match meaning to images and words) and PNFA- progressive nonfluent aphasia (apraxia of speech, agrammatism, effortful, halting speech with sound errors). [2,3]

The majority of FTD cases occur between 45 and 65 [1]. An average age of onset is 57. That’s why FTD is frequently misdiagnosed as Alzheimer’s disease. The progression of symptoms varies from 2 to over 20 years, it varies by individual. For all FTD patients decline in functioning, behaviour, language and movement is inevitable. Average life expectancy is 7 to 13 years after the start of symptoms. [4] The most common cause of death is pneumonia. [5]

Currently there is no known cure for frontotemporal dementia. Medicines are used to help manage some of the symptoms – antidepressants to help control the loss of inhibition, overeating and compulsive behaviors, antipsychotics for preventing behaviour that is putting the patient and others at risk of harm. Medications for Alzheimer’s disease are avoided due to possibilty of worsening cognition and behaviour in FTD patients. [6]

CASE REPORT

A 39-year-old well functioning man in a short period of time began to show behavioral changes which included impulsive behavior. He was agressive towards his family members, even his young children. Binge eating led him to gaining 10 kgs. He was also apathetic and had difficulties with social interactions. Those disturbing symptoms forced the family to refer the patient to the hospital. Neurological and psychological examination showed e.g. reduced verbal fluency and poverty of speech, disorders in executive functions and restlessness and anxiety. Laboratory tests of blood, urine and CSF didn’t reveal any
abnormalities, same as EEG. But significant changes were visible in MRI. MRI scan showed substantial cortical atrophy in frontal [Figures 1 and 2] and temporal lobes [Figure 3 and 4]. These areas in the brain have important functions when it comes to behavior, planning, problem-solving, emotional control, and speech.

After excluding other possible causes, based on the clinical status (behavioral and speech disorders), typical MRI (atrophy within the lobes) and neuropsychological examination, the patient was diagnosed with frontotemporal dementia (FTD).

Figure 1: Patient’s MRI scan - substantial cortical atrophy in frontal lobes.
Figure 2: Patient’s MRI scan - substantial cortical atrophy in frontal lobes.

Figure 3: Patient’s MRI scan - substantial cortical atrophy in temporal lobes.
Figure 4: Patient’s MRI scan - substantial cortical atrophy in temporal lobes.

DISCUSSION

This patient was diagnosed with FTD in just a few months, but in most patients this process takes much longer: an average of 3.6 years, but it may take even 10 or 12 years. [Table 1.] [7,8]

Table 1. Gender, age at earliest FTD symptoms, age at FTD diagnosis, and years between first symptoms and FTD diagnosis observed by caregivers. [7]

<table>
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<tr>
<th>Nr</th>
<th>Gender of person with FTD</th>
<th>Age at earliest FTD symptoms observed in person with FTD</th>
<th>Age at FTD diagnosis in person with FTD</th>
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Relationships summarized

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<td>Child</td>
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Dementia diseases have an ambiguous onset and the first symptoms are often unnoticed or ignored. Affected person does not see the changes in himself or herself, which makes it difficult to talk about it and also to make the person go see a doctor. That is why it takes so long to diagnose frontotemporal dementia. FTD symptoms are non-specific, which can lead to a misdiagnosis of depression, midlife crisis, marital conflict, stress, menopause, manic psychosis, obsessive- compulsive disorder, or sociopathic personality disorder. [9] Even physicians may overlook that neurodegenerative disease can affect younger patients. While in older individuals, with longer duration of symptoms, the diagnosis can be mistakenly identified as Alzheimer's disease or vascular dementia.

Unfortunately, there is no treatment that would stop or slow the course of the disease. But there are treatments that can help manage some of the symptoms. [10] Therefore we use SSRI that may help control the loss of inhibitions, overeating and compulsive behaviours. If SSRIs are insufficient, antipsychotic can help control severely challenging behaviour that is putting the person with dementia or others around them at risk of harm. It is important that medications for Alzheimer’s disease are generally avoided, as they may worsen cognition and behavior in FTD. [11]

CONCLUSION

To conclude, this case is worth to analyse because of an unexpectedly young age of onset, rapid development of symptoms and efficient diagnostics. In cases similar to our patient's history, FTD is often misdiagnosed or ignored, which frequently delays the diagnosis.

Studies [Beber, 2012; Bahia, 2007] show that FTD has the highest rate of misdiagnosis among neurological patients with behavioral and cognitive complaints. Most commonly, it was mistaken for Alzheimer's disease (30%), psychiatric disorder (40%), mania (20%), and depression (20%). As a consequence, this delayed the admission of patients to appropriate specialized centers and hindered the initiation of treatment, resulting in a faster decline in functioning for FTD patients. Therefore, it is important to emphasize awareness of the symptoms of FTD in order to avoid misdiagnosis, which frequently occurs in clinical practice. [12,13]

This case shows, similar to the study by Rasmussen et al. that is important for clinicians to pay attention when spouses or other family members are concerned about
personality and behavioral changes, even if the symptoms are difficult to pinpoint and describe. Families of FTD patients reported that not knowing diagnosis is particularly psychologically stressful as it does not allow to explain the causes of unacceptable behavior changes and they cannot accept the loss of a relationship with a loved one. Unfortunately getting the diagnosis does not bring much relief, because the most difficult task belongs to the family, as it will be the family who will have to face new difficulties that will increase with time. As the person with FTD starts to change and become increasingly helpless, the family is forced to take on a new role in the relationship: a caregiver role. As in the case of this patient, he will not be able to act as a father for his young children or provide for the family. [7] That is why we should provide support and care for the patient's family, for whom the diagnosis of FTD becomes challenging and burdening.

These findings indicate the need to raise awareness among both healthcare professionals and the general public about FTD and organic behavioral and personality changes, as well as the necessity to develop guidelines for diagnosing and for patient care.

FOOTNOTES

Author's contribution

Conceptualization, K.T. J. W-S.; writing—original draft preparation, K.T. J. W-S. A.Hapon.; writing—review and editing, K.C. M.Z., K.K., A.Hunek.; visualization A.Hapon; project administration M.D.

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Conflicts of Interest

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