THE BEHAVIORAL VARIANT OF FRONTOTEMPORAL DEMENTIA PRESENTING AS A PRIMARY PSYCHIATRIC DISORDER: challenges in diagnosis and management

A. Ono Baraniuk¹, J.A. Morales Saute², R. Machado de Castilhos³, M.P. Loayza Hidalgo¹.

¹Hospital de Clínicas de Porto Alegre, Psychiatry, Porto Alegre, Brazil.

²Hospital de Clínicas de Porto Alegre, Medical Genetics Service, Porto Alegre, Brazil.

³Hospital de Clínicas de Porto Alegre, Neurology, Porto Alegre, Brazil.

OBJECTIVE

The aim of the research is to highlight the difficulties in the diagnostic process and management of frontotemporal dementia when psychiatric symptoms dominate the clinical picture.

BACKGROUND

The behavioral variant corresponds to the majority of frontotemporal dementia cases, accounting for nearly 60% of patients. It is marked by progressive decline in social functioning and changes in personality. Early symptoms typically include mood changes such as apathy, depression, withdrawal and some individuals experience obsessive compulsive behavior. As a result, this neurological condition is commonly misdiagnosed as a primary psychiatric disorder. This barrier probably underestimates the real prevalence of this neurodegenerative disease.

METHODS

We report a case of a 24-year-old male patient who was admitted to a psychiatric setting in order to investigate psychotic and behavioral symptoms and was diagnosed with early onset frontotemporal dementia after a cautious investigation. A literature review was conducted in the PubMed database, including studies from 2000 to 2018.

CASE DESCRIPTION

The patient had no previous history of psychiatric diagnoses or treatments. One year prior to psychiatric hospitalization, he started with behavioral changes: depressed mood, apathy, social isolation, insomnia and loss of interest. He also presented repetitive behaviors, performing checking rituals and repeating a particular phrase many times. Over the subsequent months, his behavior became increasingly bizarre and inappropriate (inducing vomiting after eating, urinating indoors, etc.), with episodes of visual hallucinations. He had received mood stabilizers, antidepressants and many different antipsychotics in effective doses. He was subsequently initiated on Clozapine, which was increased up to 550 mg/day, but his psychiatric state remained unchanged. A significant impairment in cognitive functioning was also noted and he developed mutism over the months at the hospital. Considering the refractoriness of pharmacotherapy, he received a total of 15 sessions of electroconvulsive therapy, with no clinical improvement.

An extensive investigation was performed. He had been previously scanned with magnetic resonance imaging (MRI) of the brain five years prior to our investigation, which showed no abnormalities (figure 1). On admission, he underwent a new MRI, revealing mild brain atrophy with enlarged ventricles (figure 2). No other abnormalities were evident. After three months, he was submitted to a third MRI scan, which demonstrated increased brain volume loss with prominent atrophy over the frontotemporal area (figure 3).

DISCUSSION

A probable diagnosis of an early-onset and fast progressive form of frontotemporal dementia was considered the final diagnosis, based on the neuroimaging findings, together with the clinical history.

Frontotemporal dementia causes dramatic reduction in life expectancy and there are currently no treatments to slow or stop the progression of the disease.¹ Interventions consist mainly in symptomatic management. An individualized therapeutic plan should be established, including non-pharmacological approaches that target environmental adaptations, psychoeducation, behavioral strategies and caregiver training.^{2,3}

Figure 1

Figure 2

Figure 3

CONCLUSIONS

Our study emphasizes the need of awareness, since many cases of the behavioral variant of frontotemporal dementia may suffer from a heterogeneous presentation. A careful assessment is crucial to achieve the proper diagnosis, since the symptomatology sometimes may be indistinguishable from primary psychiatric conditions. This scenario could be avoided if physicians rely on some warning signs in the clinical history. Rapid-onset of disease with no prior psychiatric history, poor response to standard treatments and presence of atypical symptoms should raise further attention to the etiology of symptoms and prompt physicians to consider the presence of underlying organic diseases.⁴

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