Dusunen Adam The Journal of Psychiatry and Neurological Sciences 2017;30:266-268
DOI: 10.5350/DAJPN2017300312
Letter to the Editor / Editöre Mektup

A Case of Frontotemporal Dementia in the Shadow of Schizophrenia Diagnosis

Solen Arslan[1], Bengu Yucens[1], Cengiz Celebi[2]

[1]Afyon Kocatepe University, School of Medicine, Department of Psychiatry, Afyonkarahisar - Turkey
[2]Afyonkarahisar State Hospital, Department of Psychiatry, Afyonkarahisar - Turkey

How to cite this article: Arslan S, Yucens B, Celebi C. A Case of frontotemporal dementia in the shadow of schizophrenia. Dusunen Adam The Journal of Psychiatry and Neurological Sciences 2017;30:266-268. https://doi.org/10.5350/DAJPN2017300312

Address reprint requests to / Yazışma adresi: Bengu Yucens, Afyon Kocatepe University, School of Medicine, Department of Psychiatry, Afyonkarahisar, Turkey
E-mail address / Elektronik posta adresi: dr.bengubaz@yahoo.com
Date of receipt / Geliş tarihi: July 28, 2017 / 28 Temmuz 2017
Date of the first revision letter / İlk düzeltme öneri tarihi: August 6, 2017 / 6 Ağustos 2017
Date of acceptance / Kabul tarihi: August 9, 2017 / 9 Ağustos 2017

Dear Editor,

Frontotemporal dementia (FTD), is the second most frequent type of early onset dementia, constituting about 13% of all dementia cases (1,2). Typical age of onset is 45-60 years of life, it is more common in men and about 50% of the patients have positive family history (3). These cases are misdiagnosed more often than late-onset dementias; they mostly emerge with neuropsychiatric presentation. Clinical presentation often includes deficiency in performing daily activities, decrease in self-care, decrease in human relations, and change in eating habits. Disorientation, distractibility, disinhibition, perseverations, compulsive and stereotypical behaviors, and lack of insight can be observed in the psychiatric examination (4,5).

Imaging studies will depict medial and anterior temporal lobe degeneration (6). Different psychiatric symptoms can be seen in FTD cases related to the affected neuroanatomical regions. Personality and behavioral changes, apathy, and psychotic symptoms prevail in frontal region involvement, whereas, decrease in emotional processing, interpersonal coldness, and hypomania-like behavior prevail when temporal region is affected (7). The frequency of delusions is 14%, and most common are paranoid and somatic delusions (8). FTD patients can be misdiagnosed as having a psychiatric disorder because of the symptoms that could be seen in schizophrenia such as obsessional thoughts, typical or bizarre compulsions, and delusions in the early stages and; mutism, inappropriate social behavior, impaired social relationships, lack of insight, stereotyped behavior and speech, in the late stages (9).

Our case is a 47-year-old male, married, college graduate, and had resigned from his job as a civil servant at a library. The patient was admitted to the psychiatric outpatient clinic for the first time 10 years ago with complaints of unhappiness, reluctance, and weakness. He was hospitalized with the diagnosis of depression, received antidepressant treatment and was discharged after recovery. Six months after being discharged, the patient was once again followed up with the diagnosis of psychotic disorder when he had the complaints of sadness, unhappiness, skepticism, hearing voices that others did not hear while falling asleep, problems at work and with family members, and difficulties in working. The patient who showed partial improvement

with various antipsychotic treatments did not adequately respond to medical treatment, so electroconvulsive therapy (ECT) was performed for 13 and 8 sessions during 2 separate hospitalizations. A partial reduction in psychotic symptoms was achieved after ECT, but due to an increase in paranoid delusions and a decrease in self-care, he was diagnosed with treatment-resistant schizophrenia and clozapine was added to the existing amisulpride treatment. One year ago, cranial magnetic resonance (MR) imaging—performed as a result of additional symptoms, such as marked reduction in communication, increased introversion, and inappropriate social behaviors—showed significant atrophy of the frontal and temporal lobes. The patient's mini mental test score was 24; the results of Wisconsin Card Sorting Test (WCST), Trail-Making Test, Stroop Test, and Oktem Verbal Memory Process Test (VMPT) (10) showed significant deterioration in; frontal lobe functions of complex (executive) attention, executive functions, feature identification, conceptualization, and abstract thinking skills; and short-term and long-term memory functions, the ability to learn new information, focusing and sustaining attention. Patient was diagnosed with frontotemporal dementia and treatment was changed to clozapine 250mg/day and fluoxetine 40mg/day.

It may take long to discover etiologic factors that we encounter less often and that predominantly present with psychiatric symptoms, like in FTD. The presence of a family history and prevailing personality change is important in the differentiation of FTD from psychiatric diseases (11). The patient described in Grewal et al.'s (12) case report of 2011, who had obsessive compulsive symptoms at the age of 30, presented psychotic symptoms after 3 years, has only been diagnosed with FTD after receiving 8 years of various antipsychotic treatments and rapid worsening after ECT. Although there was no negative response to ECT in our case, it is not a treatment with anticipated efficacy in FTD. In the study conducted by Landqvist Waldö et al. (13) in 2015, when the initial diagnosis of 97 FTD patients was investigated, 33 were found to have other types of dementia, 21 depression, 13 psychotic disorder, and 7 other psychiatric disorders. Those initially diagnosed with psychiatric disease were younger than the others. In an analysis of 17 cases with early-onset FTD, 5 of the patients were diagnosed with schizophrenia before FTD diagnosis, and the average age of onset of psychotic symptoms of these 5 patients were found to be 35.6 years and mean lifetime was 45.4 years (7). In a recent study, 95.5% of patients with the behavioral variant of FTD were found to have at least one psychotic symptom; and negative symptoms and formal thought disorders occurred more frequently rather than positive symptoms; and it was concluded that, this was the main cause of misdiagnosis in FTD (14).

Psychotic symptoms emerging before the age of 30-35 years are considered in favor of primary psychiatric disorder, but it may be difficult to distinguish whether psychotic symptoms starting after age 35 are due to psychiatric illness or FTD (9). Significant positive psychotic symptoms accompanied by apathy and cognitive impairment are more pronounced for psychiatric disorders. Psychotic disorders are mostly accompanied by intermittent exacerbations in between stable phases, whereas progression of deficits without a remission in FTD is important in differential diagnosis (15). In our case, the onset age of the disease being 37 years old, the insidious onset, the progressive course without remission, and the positive family history could be considered in favor of FTD, whereas the presence of predominant positive psychotic symptoms at the beginning was in favor of psychotic disorder. Our case has also been treated initially for schizophrenia and then for treatment-resistant schizophrenia for about 8 years. When diagnosing schizophrenia—as in all psychiatric diagnoses—general medical conditions and substance use should be investigated first. The necessary tests should be performed not only in the initial diagnosis process but also during follow-up when the expected response to the treatment cannot be achieved. Since patients are considered to be resistant to treatment when the desired response to antipsychotic medication is not accomplished in spite

of the use of two different antipsychotic medicines at adequate dose and for adequate duration in the treatment of schizophrenia, our case was also considered as resistant to treatment and clozapine was started. However, it has been considered that schizophrenia may have been a misdiagnosis when adequate response to clozapine was not achieved and rapid deterioration was observed. The delay in the diagnosis and in treatment may be due to the confounding effect of the early onset, as well as because of the low awareness of FTD as a clinician. One of our main aims in presenting this case is to increase the awareness. In the literature, generally there are case reports on the differential diagnosis of psychotic disorders and FTD, and there is a need for studies with large samples on this topic.

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