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Case Report
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Frontotemporal Dementia Associated with Behavioral and Psychotic Symptoms: Case Series

Davranışsal ve Psikotik Semptomlarla İlişkilendirilen Frontotemporal Demans: Olgu Serisi

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Abstract

Aim: Frontotemporal dementia (FTD) is a subtype of degenerative dementia characterized by prominent impairments in personality, behavior, and cognition. This study aims to evaluate three cases presenting with acute psychotic symptoms who were later diagnosed with FTD, focusing on their behavioral and neuropsychiatric features.

Case: The first case was a 68-year-old woman who had been diagnosed with schizophrenia for 40 years and was reported to have exhibited forget fulness, disorganized behavior, and social inappropriateness in recent years. Despite antipsychotic treatment, disinhibition persisted. The second case is a 79-year-old woman who presented with memory impairment and psychotic symptoms. Cognitive and behavioral symptoms did not improve despite treatment. The third case was an 85-year-old male patient who presented with complaints of disorganized behavior, psychotic symptoms, and forgetfulness. While antipsychotic treatment reduced aggression, disinhibition persisted.

All three cases demonstrated disinhibition, disorganized behaviors, cognitive deficits and neuroimaging results indicated marked atrophy in the frontal and temporal lobes. The persistence of symptoms despite treatment supports the diagnosis of the behavioral variant of FTD. Before the diagnosis of FTD, the first case had recurrent episodes associated with schizophrenia, while no psychiatric diagnosis was present in the other two cases. In all three cases, disinhibition, disorganized behavior, and personality changes were predominant, aligning with the behavioral variant of FTD.

Conclusion: Advanced imaging techniques and long-term neurocognitive monitoring play a crucial role in confirming the diagnosis of FTD and understanding its clinical course. Further research is needed to determine whether psychosis is a risk factor for FTD development.

Keywords: Psychotic disorder; frontotemporal dementia; disinhibition; behavior change

Öz

Amaç: Frontotemporal demans (FTD), dejeneratif demansların bir alt türü olup, özellikle kişilik, davranış ve bilişsel bozukluklarla karakterizedir. Bu çalışma, akut psikotik belirtilerle başvurup FTD tanısı alan üç vakayı, davranışsal ve nöropsikiyatrik belirtileri açısından değerlendirmeyi amaçlamaktadır.

Olgu: İlk vaka, 68 yaşında ve 40 yıldır şizofreni tanısıyla izlenmiş bir kadın olup son yıllarda unutkanlık, dezorganize davranışlar ve sosyal uygunsuzluklar gösterdiği öğrenildi. Antipsikotik tedaviye rağmen disinhibisyonun devam ettiği görüldü. İkinci vaka, 79 yaşında kadın olup, unutkanlık ve psikotik belirtilerle başvurdu. Tedaviye rağmen bilişsel ve davranışsal belirtiler gerilemedi. Üçüncü vaka, 85 yaşında erkek hasta olup dezorganize davranışlar, psikotik belirtiler ve unutkanlık şikayetleri ile başvurdu. Antipsikotik tedaviyle agresyon azalmasına karşın disinhibisyon sürdü.

Üç vakada da disinhibisyon, dezorganize davranışlar, bilişsel bozukluklar ile nörogörüntülemede frontal ve temporal loblarda belirgin atrofi bulunmaktaydı. Tedaviye dirençli semptomlar, davranışsal alt tip FTD'yi desteklemekteydi. Frontotemporal demans tanısı öncesinde, ilk olguda şizofreni tanısı ile takip edilen ataklar bulunmakta iken diğer olgularda bir psikiyatrik tanı bulunmamaktaydı. Her üç vakada da disinhibisyon ve dezorganize davranışlar ile kişilik değişimleri ön planda olup FTD davranışsal alt tipte yer almaktaydı.

Sonuç

İleri görüntüleme teknikleri ve uzun dönemli nörokognitif izlem, FTD tanısının kesinleşmesinde ve klinik seyrin anlaşılmasında önemli rol oynamaktadır. Bu tür vakaların, psikozun FTD gelişiminde bir risk faktörü olup olmadığını anlamak için daha fazla araştırmaya ihtiyaç vardır.

Anahtar sözcükler: Psikotik bozukluk, frontotemporal demans; disinhibisyon; davranış değişikliği

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INTRODUCTION

Frontotemporal dementia (FTD) is a neuro-psychiatric condition accounting for 12.5–16.5% of degenerative dementias. It constitutes 3–10% of all dementias and typically begins between the ages of 45 and 65 (1). Frontotemporal dementia has three subtypes: the behavioral variant, characterized by personality, behavioral, and cognitive changes; semantic dementia, marked by a loss of word meaning; and progressive non-fluent aphasia, associated with difficulty in word retrieval and reduced fluency and spontaneity in speech (2).

Key features of the behavioral variant include psychiatric and behavioral symptoms that occur without significant cognitive impairment, insidious personality changes, emotional dysregulation, and social dysfunction in interpersonal relationships (3). Literature highlights that in the neurodegenerative dementia process, temporal lobe involvement can lead to emotional dyscontrol, interpersonal disconnection, and elevated mood states resembling hypomania. In contrast, frontal lobe involvement is associated with apathy and a noticeable reduction in social engagement (4).

In the clinical course of FTD, symptoms such as apathy, repetitive compulsive-like behaviors, changes in sleep and appetite, and lack of insight into the illness can act as confusing and complicating factors for early diagnosis. As a result, these cases are often misdiagnosed with late-onset psychotic disorders, depression, obsessive-compulsive disorder, or late-onset bipolar disorder (5). Although it becomes easier to differentiate FTD from other types of dementia as the disease progresses, distinguishing the behavioral variant of FTD from other psychiatric disorders remains particularly challenging. Due to disorganized behaviors and psychotic symptoms, which can occur not only in the behavioral variant but across all FTD subtypes, these cases may be misdiagnosed as schizophrenia.

In this article, three cases of late-onset FTD diagnosed after presenting with acute psychotic episodes are examined within the framework of current literature. Informed written consent was obtained from all patients prior to inclusion.

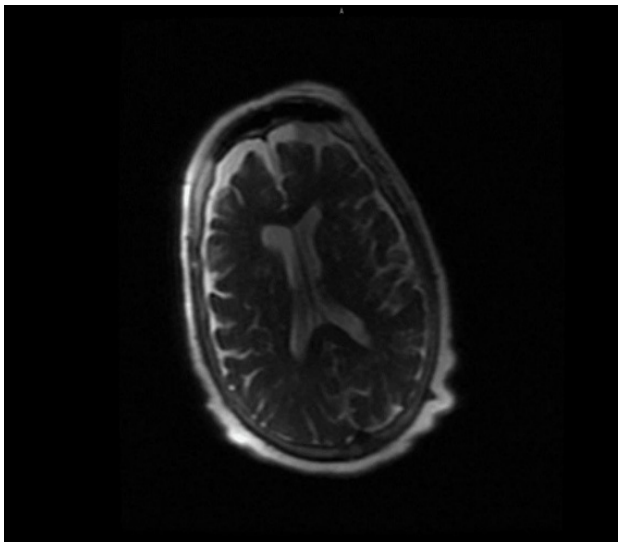
CASE

Written consent was obtained from the patients that their medical data could be published.

Case 1: A 68-year-old female with a 40-year history of schizophrenia, requiring recurrent hospitalizations every 1.5 to 2 years, presented with progressive cognitive and behavioral changes. Despite partial remission for five years on olanzapine, her symptoms worsened over the past two years, including forgetfulness, urinary incontinence, and disorganized behaviors such as screaming, drinking from mud puddles, peeling paint, and ingesting fabric. She also exhibited disorientation, nonsensical speech, insomnia, socially inappropriate behaviors, and refusal to eat, resulting in a 10-kilogram weight loss. Episodes of binge eating further complicated her condition.

Admitted for differential diagnosis, her evaluations revealed stable vital signs, normal laboratory findings, and unremarkable physical and neurological examinations. A family history of dementia suggested potential neurodegenerative processes. On September 26, 2024, MRI showed significant diffuse cerebral volume loss in the bilateral frontal and temporal regions with ventricular enlargement (Figure 1).

Figure 1. MRI imaging shows ventricular enlargement, diffuse cerebral volume loss in bilateral frontal and temporal regions



The Frontal Behavior Inventory (FBI) score was 47, with high disinhibition sub-scores indicative of predominant behavioral symptoms. Despite continued treatment with olanzapine (20 mg/day) and quetiapine (300 mg/day), psychotic symptoms, aggression, and disinhibition persisted. Eight sessions of electroconvulsive therapy improved aggression but had no effect on disinhibition. Memantine (10 mg/day) was added following a neurology consultation.

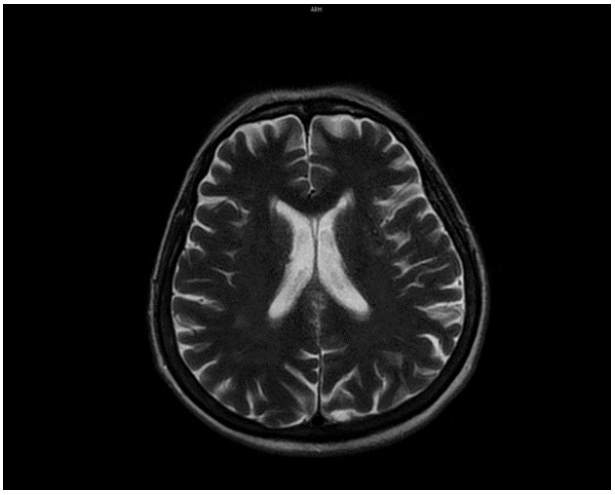
After three months of follow-up for FTD, her disinhibited behaviors, such as tearing and ingesting clothing, reduced in severity, but residual psychotic symptoms and limited cooperation remained. She requires ongoing assistance for self-care and daily needs.

Case 2: A 79-year-old female presented with a one-month history of psychotic symptoms, including self-directed speech, auditory, and visual hallucinations. Over the past two years, she exhibited progressive cognitive and functional decline, such as forget fulness, repetitive questioning, inability to prepare meals, and episodes of staring while talking to herself. Previously living independently, she became reliant on assistance for self-care, raising concerns of neurodegenerative processes like FTD.

According to her caregiver, the patient displayed socially inappropriate behaviors inconsistent with her premorbid personality, such as public undressing and offensive language. Examination revealed clear consciousness but disorientation and inability to cooperate. Aggression necessitated close observation. Laboratory tests and physical and neurological examinations were unremarkable. The patient had no history of psychiatric or medical conditions, nor alcohol or substance use. Due to psychotic symptoms and aggression, olanzapine 5 mg/day was initiated. However, the patient refused medications and food, and increasing the dose to 10 mg/day over two weeks yielded no significant improvement in symptoms, including nonsensical speech, food refusal, and memory deficits.

On September 24, 2024, MRI revealed bilateral frontal and temporal gyri thinning with ischemic changes in the frontal and parietal lobes (Figure 2).

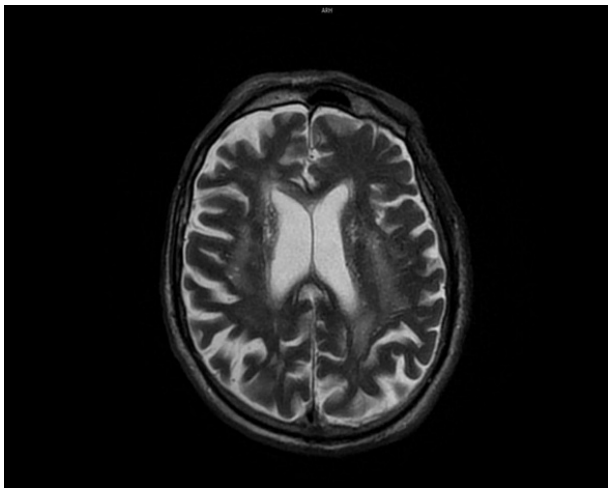
Figure 2. MRI imaging shows ischemic changes in the frontal and parietal lobes, thinning of bilateral frontal and temporal gyri



Behavioral evaluation using the Frontal Behavior Inventory (FBI) resulted in a score of 46, highlighting prominent disinhibition and negative behaviors. Despite continued olanzapine treatment, disinhibition and memory deficits persisted. Following neurology consultation, memantine 10 mg/day and acetylsalicylic acid 100 mg/day were added to her regimen.

Case 3: An 85-year-old male presented with a three-year history of progressive forgetfulness, visual hallucinations, impulse control issues, nonsensical speech, and disorganized behaviors. Over the past year, these symptoms worsened, including disrupted sleep, confusion in his home, binge eating, urinary incontinence, increased sexual drive, and disinhibition. His behaviors were inconsistent with his premorbid personality. On examination, he exhibited visual hallucinations, persecutory delusions, disorganized speech, impaired orientation, and dependence on others for daily care. Laboratory tests were normal except for folic acid deficiency, for which supplementation was initiated. Neurological and physical exams showed no pathological findings, and there was no significant psychiatric or medical history. Treatment began with olanzapine at 5 mg/day for psychotic symptoms and behavioral issues. Neuropsychological tests revealed severe deficits in memory, frontal-executive functions, global attention, abstraction, immediate recall, and visual-spatial abilities. The Frontal Behavior Inventory score was 49, indicating severe behavioral symptoms. MRI showed frontal and temporal cortical atrophy, supporting a diagnosis of FTD (Figure 3).

Figure 3. Frontal and temporal cortical atrophy on MRI imaging supporting the diagnosis of FTD



Despite increasing olanzapine to 10 mg/day, aggression improved slightly, but hallucinations, disinhibition, orientation disturbances, and memory impairments persisted. Motor symptoms like urinary incontinence and autonomic dysregulation,

including hypotensive episodes, remained. Memantine at 10 mg/day was added after neurology consultation. The patient required full-time assistance, was placed under guardianship, and continued with close follow-up.

DISCUSSION

Frontotemporal dementia is a neurodegenerative and neuropsychiatric disorder characterized by significant impairments in behavior, language, and executive functions, primarily due to atrophy in the left temporal and frontal regions caused by neuropathogenic processes and genetic factors. While initial psychiatric symptoms may present as obsessive-compulsive disorder, compulsive behaviors, depression, or anxiety, cognitive impairments invariably emerge and become a prominent feature in the later stages of the disease (7).

This article discusses three cases of patients who presented with acute psychotic episodes and were initially considered for differential diagnoses of schizophrenia and dementia, ultimately receiving a diagnosis of FTD. In the first case, the patient had a history of psychotic episodes managed under a diagnosis of schizophrenia, while the other two cases had no prior psychiatric diagnosis. In all three cases, disinhibition, disorganized behaviors, and personality changes were prominent, aligning with the behavioral variant of FTD.

Similar to our first case, the literature reports instances of patients initially presenting with psychosis but later being diagnosed with FTD (8). These cases are characterized by an acute onset of psychotic symptoms accompanied by behavioral changes. Detailed histories reveal that, while psychosis may emerge acutely, symptoms such as amnesia, disinhibition, disorganized behaviors, and signs of autonomic dysregulation often manifest earlier and progressively worsen over time, indicating an advancing clinical course. A distinguishing factor separating these cases from late-onset schizophrenia is the generally favorable prognosis of late-onset schizophrenia (9). In contrast, the clinical course of our cases progressed destructively and with significant worsening over time. Similarly, in the literature, a study involving five cases of FTD presenting with psychosis-like symptoms found that all patients demonstrated prominent right frontotemporal involvement on imaging studies (10). In these cases, the use of antipsychotics should be approached cautiously, as they may exacerbate movement disorders, particularly in FTD. For this reason, atypical antipsychotics are preferred. Advanced imaging techniques, such as Pozitron Emisyon Tomography (PET) scans, are valuable in understanding the presentation of FTD with psychotic symptoms. Specifically, FDG-PET scans can highlight metabolic abnormalities and frontal/temporal lobe involvement characteristic of FTD (11). These methods can be employed in cases where diagnostic uncertainty is high. Metabolic changes in regions such as the anterior cingulate cortex have been shown to be associated with the behavioral and psychotic symptoms accompanying FTD (12). Neuroanatomical and functional studies of these areas could contribute to a deeper understanding of FTD.

CONCLUSION

Further research is needed to explore the relationship between psychotic episodes and the development of FTD. Specifically, prospective studies with longterm follow-up, supported by clinical tests of neurocognitive functions, are required to determine whether psychosis acts as a risk factor for the onset of FTD.

Author's Contribution

Written consent was obtained from the patients that their medical data could be published.

The authors declare no conflict of interest.

The authors disclose that no grants or support resources were used.

All authors declared their contribution to the study at all stages and approved the final version of the manuscript.

All authors declared that this manuscript has not been published before and is not currently being considered for publication elsewhere.

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