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Fahr's Syndrome Misdiagnosed As Delusional Disorder: A Case Report

Sanrısal Bozukluk ile Giden Fahr Hastalığı: Olgu Sunumu

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ABSTRACT

Fahr's disease is a rare neurological disorder that is characterized by bilateral basal ganglia calcification. In the present study, a 49-year-old male patient presented with delusional beliefs. He had normal neurologic examination. Follow-up mental status examination and clinical findings revealed delusional disorder. After three weeks, the patient presented to the clinic with postural tremor in the hands, and gait difficulties. A cranial CT scan showed that he had bilateral basal ganglia, thalamus, and centrum semiovale calcifications. The case illustrates the importance of considering organic etiologies before diagnosing a patient, particularly one who has late-onset presentation of psychosis.

Keywords: Fahr's disease, delusional disorder, calcification, basal ganglia

ÖZ

Fahr hastalığı bilateral bazal gangliyon kalsifikasyonu ile karakterize nadir bir nörolojik bir tablodur. Bu makale de, sanrısal bozukluk ile giden 49 yaşında erkek hasta sunulmaktadır. Hastanın, ilk başvuru sırasında nörolojik muayenesi normaldi. Ruhsal durum muayenesi ve klinik bulgular neticesinde sanrısal bozukluk tanısı kondu. Antipsikotik tedavisi başlanan hasta, üç hafta sonra halsizlik ve yürümede güçlük şikâyeti ile tekrar başvurdu. Bilgisayarlı tomografide, bilateral bazal gangliyonlar, talamus ve sentrum semiovale'de kalsifikasyon ile uyumlu dansite artışı saptandı. Bu vaka, özellikle geç yaşta psikotik semptomlar ile başvuran hastalarda tanı koymadan önce mevcut tabloya sebep olabilecek organik etiyolojilerin dışlanmasının önemini vurgulamaktadır.

Anahtar Kelimeler: Fahr hastalığı, sanrısal bozukluk, kalsifikasyon, bazal gangliyon

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INTRODUCTION

Fahr's disease or "familial idiopathic basal ganglia calcification (IBGC)" is a rare neurological disorder that is characterized by bilateral calcification of the basal ganglia, cerebellum and centrum semiovale. Calcification of the basal ganglia can be associated with neurological, cognitive and psychiatric abnormalities and motor symptoms (1). Neurological and psychiatric mental status examination, along with imaging, are necessary for the differential diagnosis of Fahr's disease (2). In this disease, the characteristic neuroimaging finding is bilateral basal ganglia calcifications on cranial CT scans of patients (3).

Although familial idiopathic basal ganglia calcification presents most commonly with motor symptoms, about 40% of the patients with this disease are seen with psychiatric features including cognitive and psychotic symptoms, and mood disorders (1, 4). Also, anxiety disorders and obsessive-compulsive disorders are seen in some patients with Fahr's disease (1). Atypical antipsychotics should be preferred due to the co-existent extrapyramidal symptoms in patients with Fahr's disease who exhibit psychiatric symptoms. Neurological symptoms seen in Fahr's disease include delirium, dementia, and mental impairment (5). Psychotic symptoms can present with auditory and visual hallucinations, paranoid ideation, delusions, and fugue states (1). Here, the case of a man with pure delusions and Fahr's disease is presented.

CASE

"Mr. O," a 49-year-old married man was brought by his family to the psychiatry out-patient ward in our hospital with complaints of being suspicious for the last three months. At the time of admission to our hospital, the patient was suffering from paranoid ideas (believed his enemy was spying on him and intending to kill him). He lacked insight into his mental state. His wife reported that the patient became suspicious that his enemy was spying on him and he became increasingly agitated at home and started closing the curtains to avoid being killed. His wife also reported that the patient had good functionality in areas not related to his delusions. He did not have a history of mental health problems. The patient had no history of significant medical problems, alcohol or substance abuse. There was no familial psychiatric history. Neurological examination showed normal muscle tone, with no ataxia or tremor at first admission.

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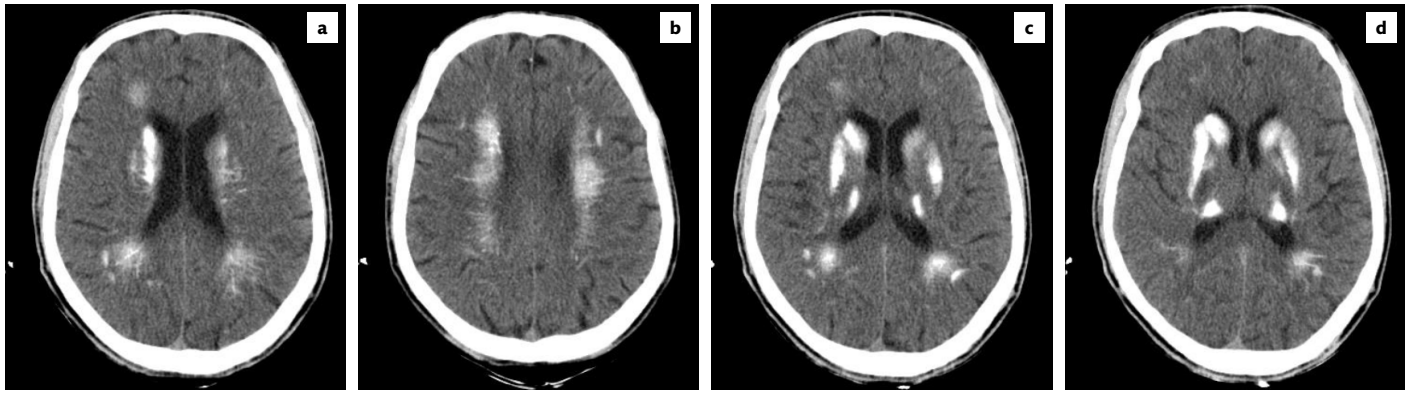

Figure 1. a–d. Computed tomography of the patient. Patient has calcified hyperdense areas in bilateral basal ganglia, thalamus and centrum semiovale.

After overall clinical features and a team meeting with his family members, the patient was provisionally diagnosed with delusional disorder and put on paliperidone (6 mg once daily). After two weeks, due to poor compliance with oral medications, a regimen of paliperidone palmitate LAIs at a dose of 100 every month was initiated. After one week, the patient presented to the clinic with weakness and difficulty walking. The results of laboratory investigations revealed normal serum chemistry levels and normal levels of thyroid-stimulating (TSH) and parathyroid (PTH) hormones, vitamin D, B12, folate, and phosphate. A formal neurological consultation was obtained. The neurological examination revealed extrapyramidal symptoms with cerebellar ataxia, postural tremor of the hands, and weakness. Because of his neurological symptoms, head computed tomography (CT) scan was performed on the patient, with a finding of bilateral basal ganglia, thalamus and centrum semiovale calcifications (Figure 1). The patient was diagnosed with Fahr's disease based on neuroimaging, clinical presentation, and laboratory investigations.

DISCUSSION

The true prevalence of familial idiopathic basal ganglia calcification is unclear, but incidental findings of calcifications in the basal ganglia range from 0.3 to 1.2% in routine radiological examinations (6, 7). This illness is characterized by various clinical features and can be asymptomatic (mostly among middle-aged patients) or associated with some psychiatric disorders like mania, psychosis, anxiety, depression and anorexia nervosa. About 40% of Fahr's disease cases present mainly with psychiatric features; among these psychotic, cognitive symptoms and mania are common (4, 8).

The calcified hyperintensities in the basal ganglia (usually restricted to the globus pallidus) on CT scans of these patients are the characteristic neuroimaging finding in Fahr's disease, but caudate nucleus, thalamus, dentate nucleus, the putamen, centrum semiovale, and white matter may also be affected (9, 10). The differential diagnosis of Fahr's syndrome is broad, including vascular lesions like angiomatous malformations and arteriovenous malformations; metabolic diseases like parathyroid disorders and diabetes mellitus; inflammatory illnesses; infectious diseases like TORCH; and neoplasms like astrocytomas, oligodendrogliomas, and metastatic tumors (11, 12).

Delusional disorder (DD) is one of the psychotic disorders which presents mainly with delusional beliefs. The mean age at onset of delusional disorder is about 40 years. Age of onset of Fahr's disease is typically in 3rd and 4th decades of life (13). This patient presented at 49 years with paranoid delusions, without hallucinations. Late-onset psychosis, with a mean age of 49.4 years, maybe associated with organic brain disorders like tumors, dementia and movement disorders like Parkinson's disease, and essential tremor (14). Positive psychotic symptoms, hallucinations, and paranoia were considered to be caused by cortico-subcortical disconnection mediated by the basal ganglia, especially involving the frontostriatal and limbic circuits (9, 10). Psychiatric symptoms in Fahr's disease (and also other disorders that can cause calcifications of basal ganglia) are usually mediated by inadequate functioning of the basal ganglia rather than other consequences of the primary disorder (15).

In the present study, we report the case of a man with Fahr's disease who presented with only persecution and referential delusions. There were no neurological and cognitive symptoms or motor deficits at the time of first admission to our hospital. In the literature, our case is the first in which delusional disorder is associated with Fahr's disease. The case illustrates the importance of considering organic neurological etiology before diagnosing a patient, particularly one who has late-onset presentation of psychosis.

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