Case Reports and Reviews

Case Report

Psychiatric Symptomatology Revealing Fahr's Syndrome: A Case Report

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Fahr's syndrome, psychosis , neuropsychiatric manifestations

Abstract

Introduction: *Fahr's syndrome is a rare anatomoclinical entity characterized by bilateral, symmetrical intracerebral calcifications, most commonly in the basal ganglia, and often associated with disturbances in phosphocalcic metabolism. These calcifications may be accompanied by a wide spectrum of neuropsychiatric manifestations.*

Methodology and Objective: *We present the case of a patient in whom psychiatric symptoms revealed underlying Fahr's syndrome. A review of the literature was also conducted to compare our findings with previously reported cases.*

Results and Discussion: *We describe a 24-year-old patient with a history of hypothyroidism since the age of 15 and chronic hypocalcemia. He was admitted to a psychiatric facility with a clinical picture characterized by a delusional syndrome, hallucinatory features, and suicidal ideation that had been evolving over one month prior to admission. Cognitive disturbances were also identified during clinical interviews.*

Neuropsychiatric disorders are frequently observed in Fahr's syndrome and may present as behavioral disturbances, confusional states, or delusional syndromes. Neurological manifestations are less common but can occur, including cognitive decline and, in some cases, intellectual disability. Cognitive impairment may range from mild deficits in memory and attention to full-blown dementia. Psychotic symptoms described in Fahr's disease include auditory and visual hallucinations, complex perceptual distortions, delusional thinking, and fugue states.

Conclusion: *Although Fahr's syndrome remains a rare clinical entity, this case highlights the importance of neuroimaging in the diagnostic evaluation of patients presenting with psychiatric symptoms, as well as the role of phosphocalcium testing in the assessment of psychotic disorders. Recognition of these associations can facilitate appropriate diagnosis and management. Further studies are warranted to evaluate pharmacological strategies and to determine the most effective therapeutic approaches for managing the psychiatric manifestations of Fahr's syndrome.*

Introduction

Fahr's syndrome is a rare anatomoclinical entity [1], characterized by bilateral, symmetrical intracerebral calcifications in the basal ganglia, most often associated with disorders of phosphocalcium metabolism [2]. These intracerebral calcifications may be associated with various neuropsychiatric manifestations [3].

Objective

To study the particularities of psychiatric symptomatology in Fahr's syndrome.

Method

We described the clinical case of a patient consulting for psychiatric symptoms revealing Fahr's syndrome, with a discussion based on a review of the literature. The keywords used were "*Fahr syndrome*," "*psychiatric symptomatology*," "*psychotic disorder*," "*depressive symptomatology*.".

Case description

We report the observation of a 24-year-old patient from a non-consanguineous marriage, whose parents had separated at an early age and whose family dynamics were very disturbed. He lives with his maternal grandmother and is the eldest of four siblings. His family history includes a 22-year-old sister who consulted a psychiatrist for irritability and intolerance of frustration.

He left school in the ninth grade after repeating two years. He has a personal history of hypothyroidism and hypocalcemia, followed since the age of 15 at Charles Nicoles Hospital. He was hospitalized in psychiatry for a delusional syndrome, a hallucinatory syndrome, and a depressive syndrome with suicidal ideation, which had been evolving for a month prior to admission. A brain CT scan performed upon admission revealed

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calcifications of the basal ganglia, leading to the diagnosis of Fahr's syndrome.

The clinical course was marked by favorable improvement under 10 mg of olanzapine.

Discussion

This case report presents interesting challenges regarding diagnosis and pharmacotherapy.

The Fahr triad is defined by symmetrical calcification of the basal ganglia, neuropsychiatric symptoms, and hypofunction of the parathyroid gland [4]. The pathophysiological mechanisms responsible for the appearance of these cerebral calcifications remain poorly understood [5]. Most authors suggest a metabolic disorder of the oligoglial cells with mucopolysaccharide deposits and the secondary appearance of vascular and perivascular lesions and calcareous encrustations. These calcifications affect the small vessels of the basal ganglia [3]. Their biochemical analysis revealed an organic matrix made up of neutral and acidic mucopolysaccharides, as well as mineral elements (calcium, phosphorus, iron, sulfur, magnesium, aluminum, zinc) [6].

It is important not to confuse Fahr's syndrome with other conditions that can lead to intracerebral calcifications, particularly endocrinopathies (such as hypothyroidism and hypogonadism), systemic diseases (including systemic sclerosis, systemic lupus erythematosus, and celiac disease), infections (such as toxoplasmosis, neurocysticercosis, and rubella), various other disorders (including chronic kidney failure, vitamin D intoxication, and mitochondrial cytopathies), as well as primary or secondary brain tumors with calcifications [4,7]. However, intracerebral calcifications in these different pathologies have distinct locations and appearances [5].

It is plausible that the psychosis seen in both Fahr's disease and schizophrenia arises from a shared underlying pathology. While positive psychotic symptoms such as hallucinations and paranoia are not exclusively explained by the classical dopamine hypothesis involving aberrant salience attribution to internal stimuli, evidence suggests that cortical disruptions found in schizophrenia also occur in Fahr's disease, particularly within the limbic system [3].

It is important to differentiate Fahr's disease, which may also be responsible for intracerebral calcifications, from Fahr's syndrome. Indeed, in contrast to Fahr's syndrome, Fahr's disease is an autosomal dominant inherited disorder (related to mutation of the IBGC1 gene localized on chromosome 14q) and is not accompanied by disorders of phosphocalcium metabolism [8]. Three main etiological axes are to be investigated in the presence of basal ganglia calcifications: hypoparathyroidism (very often), pseudohypoparathyroidism (less frequently) [8], and, exceptionally, hyperparathyroidism [9].

Fahr's syndrome is generally difficult to suspect clinically, as it may remain asymptomatic or present with polymorphic manifestations that do not correspond to any specific picture [9]. In Fahr's syndrome, neuropsychiatric disorders are frequently observed, such as character and/or behavioral disorders, or even confusional syndrome [9].

Neurological disorders are polymorphic: generalized tonic-clonic or partial epileptic seizures, extrapyramidal syndrome, and, more rarely, pyramidal or cerebellar syndrome, intracranial hypertension, or chorea [5]. Clinical manifestations can also include paresthesias, seizures, myoclonus, tetany, and parkinsonism [10,11]. Other neurological manifestations are possible but less common, such as cognitive disorders and mental retardation [9]. Cognitive disorders can range from mild memory and attention deficits to frank dementia with a fronto-subcortical profile [8,9].

Psychiatric symptoms such as depression, irritability, manic symptoms, aggression, and cognitive impairments have been documented [12]. Psychotic symptoms include auditory and visual hallucinations, delusions, and fugue states. Psychotic symptoms associated with Fahr's disease can include auditory and visual hallucinations, complex perceptual distortions, delusional thinking, and fugue states [3,5,12,13]. Manifestations may sometimes be limited to signs of hypocalcemia (tetany, Chvostek's sign, Trousseau's sign) or to symptoms of the underlying pathology causing Fahr's syndrome [5].

From a treatment perspective, the main aim of the medical approach is to control neuropsychiatric symptoms. Despite the severity of the symptoms it may cause, Fahr's syndrome has a relatively good prognosis, and correction of phosphocalcium metabolism disorders often leads to significant improvement [4]. Given the small number of cases reported in the literature, it has not been shown that one antipsychotic is more effective than another [8]. With conventional neuroleptics, patients appear more predisposed to developing extrapyramidal symptoms [12]. Electroconvulsive therapy should probably be avoided, given the risk of aggravating tremors or intracranial hypertension [3].

Several cases of psychosis associated with Fahr's syndrome have shown resistance to haloperidol [13], while some positive responses to lithium have been reported [14]. In one case, the course was marked by resistance to both first- and second-generation antipsychotics, with significant improvement on clozapine [3]. In another report, a 58-year-old patient presenting with a first manic episode with psychotic features and basal ganglia calcifications showed good response to sequential combination therapy with valproate and quetiapine, with considerable improvement in behavior, mood, and psychotic symptoms [7].

Conclusion

Although Fahr's syndrome is a rare condition, this clinical case highlights the importance of brain neuroimaging in patients with psychiatric symptoms, as well as the relevance of assessing calcium-phosphate metabolism in the investigation of psychotic disorders, to ensure appropriate management. Further studies on pharmacological management, including the choice and efficacy of treatment, remain essential..

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