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Fahr Syndrome and Psychosis: Clinical Case

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Abstract **Case Report**

Fahr syndrome is a rare neurodegenerative disease characterized by bilateral, symmetrical calcification of the basal ganglia and gray matter. We present the case of a 27-year-old woman who presented to the emergency department with febrile consciousness disturbance. Brain CT revealed extensive, bilateral, symmetrical calcifications of the basal ganglia and cerebellum. A rare syndrome, Fahr syndrome should be kept in mind and appropriate symptomatic treatment should be applied in order to stop progression and improve symptoms and clinical signs.

Keywords: Fahr Syndrome, Central Gray Nuclei, Neuropsychiatric, Fahr Disease.

INTRODUCTION

Fahr syndrome is a rare, chronic disease characterized by bilateral, symmetrical calcification of the basal ganglia, thalamus, cerebral cortex, cerebellum, and hippocampus [1-3].

The most common clinical symptoms are neuropsychiatric manifestations such as seizures, cognitive impairment and may be associated with paresthesias following hypocalcemia, headache, it is usually discovered in the fourth or fifth decade of life.

OBSERVATION

We report the case of a 26-year-old woman with a history of psychosis under neuroleptics, vaginal delivery 2 months ago, dyskinetic disorders that appeared 4 years ago, admitted with a picture of febrile consciousness disorder associated with convulsive seizures and an ionic balance with a calcemia without abnormality.

Brain CT scan without and after injection of iodinated contrast revealed extensive and bilateral and symmetrical calcifications of the central gray nuclei, cerebellum and frontoparietal region bilaterally (Figure 1).

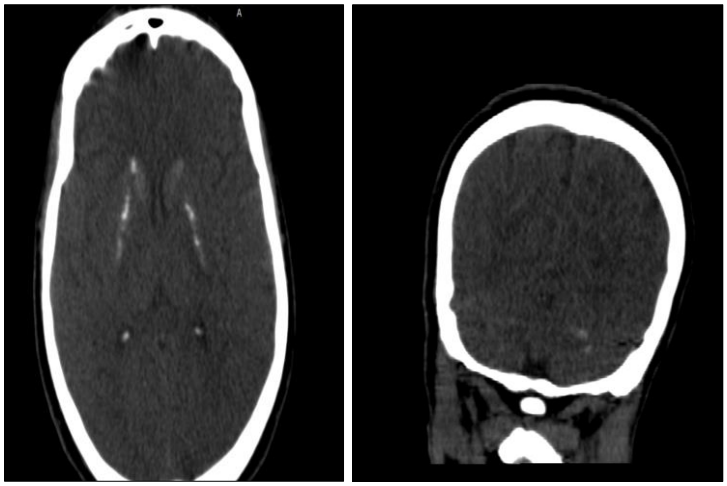

Figure 1

An ionic balance sheet did not reveal any abnormality, to eliminate the vital emergency of hypocalcemia, thus the diagnosis of Fahr syndrome due to idiopathic primary hypoparathyroidism was eliminated.

The patient was started on symptomatic treatment with levetiracetam 250 mg twice daily, which was prescribed to prevent any attacks.

During hospitalization, the patient's symptoms improved. She remained stable, without new seizures. The patient was released and referred for a neurology and psychiatry consultation for adaptation of his neuroleptic and etiological treatment.

DISCUSSION

Fahr syndrome is caused by calcification of the basal ganglia and other regions of the brain, which are bilateral and symmetrical [1, 2]. The most commonly reported metabolic disorders are hypoparathyroidism and pseudoparathyroidism.

Hypoparathyroidism is an endocrine disorder that may be iatrogenic, due to surgical removal or radiation therapy, or idiopathic.

The idiopathic form is rare and of unknown etiology, but according to several studies it is often associated with psychiatric disorders and a genetic mutation with similar cases in the family, for the other most frequent etiologies we present hypoparathyroidism by deficient secretion of PTH, with presentation of low PTH and calcium levels. In the case of pseudo-parathyroidism, PTH levels will be high in case of hypocalcemia.

There is no clear explanation of the mechanism underlying these calcifications. It is suggested that increased calcium-phosphorus complex plays an important role, depositing in the vascular wall and eventually extending to the neuron.

Patients have neuropsychiatric disorders such as seizures, headaches, cognitive decline with parkinsonian syndrome. To date, there is no specific treatment for Fahr syndrome; management of clinical manifestations is limited to supportive care using anxiolytics and anticonvulsants and a hemodynamic assessment.

CONCLUSION

Fahr syndrome, although rare, should be included in the list of differential diagnoses of neuropsychiatric disorders and epileptic seizures, can be a finding of enormous value because they can immediately guide the diagnosis and allow rapid treatment of hypocalcemia and hypoparathyroidism.

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