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Fahr's syndrome in a patient with no history of the disease: A case report

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Case Report

Abstract

Fahr syndrome which presents with various signs and symptoms has a familial predisposition and is characterized by symmetric calcification of basal ganglia. It may present with neuropsychiatric, extrapyramidal, and cerebellar symptoms. The etiology has not been defined yet. A 38-year-old woman referred to the psychiatric clinic of 5th Azar Hospital, Gorgan, Iran, with neuropsychiatric presentation.

KEYWORDS: Fahr's Syndrome, Psychosis, Basal Ganglia Calcification

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Introduction

Fahr's syndrome is idiopathic calcification of the basal ganglia.[1] Clinical features are important since basal ganglia calcification may be viewed as a secondary finding. Other manifestations are: headache, vertigo, movement disorders, paresis, and stroke such as events, cognitive impairment, psychiatric disorders, pyramidal signals, and seizures.[2] When regions other than globus pallidus are involved pathological calcifications occur.[3] Calcification may occur secondary to abnormalities in calcium metabolism or radiation therapy.[4] Bilateral calcification of basal ganglia can be found both on plain skull X-rays (SXR), in 70-80% of cases associated with hypoparathyroidism, and in necropsy specimens.[5] The syndrome has a sporadic idiopathic form and a familial form, which is associated with progressive mental deterioration and growth retardation chorea, dementia, and dystonia.[6] Another finding is levodopa-resistant Parkinson syndrome.[7] Computerized tomography (CT) make it possible to detect calcifications which otherwise are not noticeable on SXR.[5] For instance, normal SXR was reported in 14 cases of basal ganglia calcification, without any evidence of calcium abnormalities, or neurological disease due to calcification.[7] Furthermore, several reports have described familial Fahr's syndrome.[6] Some literatures have described idiopathic familial basal ganglia calcification without clinical manifestations.[1] An array of clinical manifestations in familial Fahr's syndrome may be presented depending on age, level of calcium deposits in the brain, and due neurological deficit.[1] In general, progressive neurological deterioration may results in disability and death.[8] No cure or typical treatment has yet been

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recognized for Fahr's syndrome and treatment of individual cases have shown that haloperidol or lithium carbonate may relieve psychotic symptoms.[8]

Case Report

A 38-year-old woman referred to the psychiatric clinic of 5th Azar Hospital, Gorgan, Iran, in April 2011 due to headache, anemia, feeling of isolation, self-talk, and excessive movements of the upper limbs. She was hospitalized for further investigation in the psychiatric ward due to the severity of symptoms. Eight months before admission the patient was hospitalized and treated with the electric shock therapy because of symptoms such as depression, suicide attempt, loss of energy and insomnia, and diagnosis of psychotic depression. Then she was discharged after improvement of symptoms with perphenazin prescription and amitriptyline. There was no major problem until about 20 days before the current admission which after discontinuing the medication he had insomnia, headache, and self-talk. The patient complained of continuous non-pulsatile headache in the occipital region without nausea and vomiting. Except one of the patient's brothers who had familial Mediterranean fever, other family members had no history of psychiatric diseases. The patient had severe reduction of intellectual content and auditory hallucinations, cognitive impairment in attention, and concentration. The brain CT scan without contrast injection showed bilateral calcifications in the basal ganglia and left cerebellar (Figure 1). Electrocardiography, calcium, phosphorus, alkaline phosphatase, and albumin were within normal ranges. Ophthalmology consultation was normal. Radiographs obtained from the patients' hands showed no other calcium disorders. According to the above findings, Fahr's induced psychotic syndrome was revealed, and treatment began with perphenazin 16 mg/day about 3 weeks later, pain and auditory hallucinations disappeared completely and patient's cognitive weakness improved. Six months later, patient was on maintenance treatment with no psychotic features.

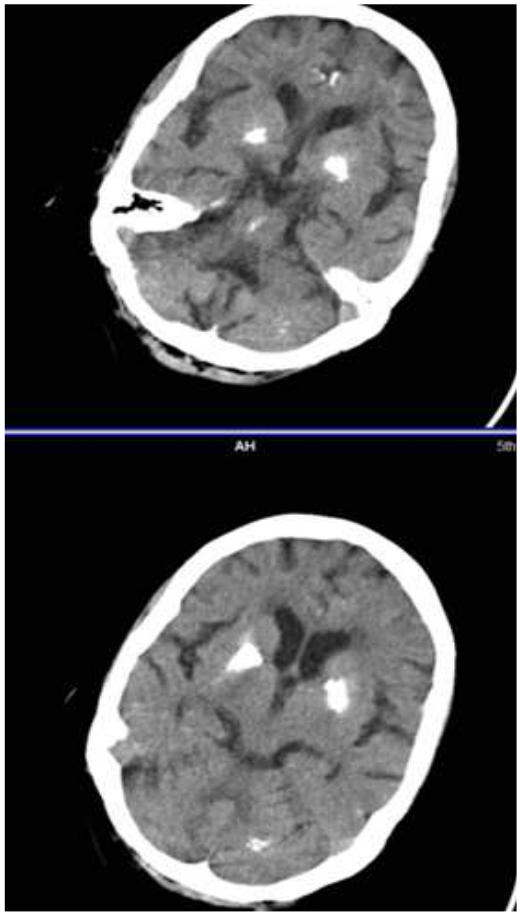

Figure 1. Computerized tomography scan of Fahr's syndrome showing bilateral basal ganglia calcification in a 38-year-old woman

Discussion

Although the etiology of calcification of the basal ganglia is rarely defined, abnormalities in vascular supply, calcium metabolism, and alkaline phosphatase activity appear to be involved. Clinical expression of Fahr's disease (FD) can vary greatly.[2] Patients with basal ganglia calcification may present initially with psychiatric features.[9] In our case, there was no evidence of hypocalcaemia and hyperphosphatemia in the patient. There was no evidence of extrapyramidal symptoms or a metabolic disorder and neurological

examination was normal. However, psychosis can induce differential diagnosis of schizophrenia or acute transient psychotic disorder.[2] Psychiatric presentations including cognitive, psychosis, and mood disorders are common in FD.[9] Other studies indicate the presence of symptoms such as auditory hallucinations, perceptual distortions, and paranoid delusions associated with FD.[9] As in our case schizophreniform psychoses have been reported. Psychosis due to FD responds variably to treatment and is sometimes unresponsive.[8] Imaging diagnosis could be the starting point to guide the clinician for the possibility of Fahr's syndrome.[1,5] The differential diagnosis includes but not limited to: Parkinson's disease, Huntington's disease, progressive supranuclear palsy, Wilson's disease, spasmodic torticollis, oligodendroglioma, low-grade astrocytoma, and arteriovenous malformation.[6] Etiology of Fahr's syndrome is not directly correlated with image calcification pattern, except for some differences noticed in calcifications site in dystrophic senile ones. Topographic image studies are promising to predict neurological deficits. Their recognition by CT is easy, has maximum sensitivity and may be contributing to early treatment.[1]

Conclusion

Our case shows that schizophreniform symptomatology presentations must be investigated in detail with due importance to family history and considering disorders with other clinical manifestations like FD.

Conflict of Interests

Authors have no conflict of interests.

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