Case Report

A Case of Schizophrenia Like Psychosis Due to Fahr's Disease

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ABSTRACT

Fahr's disease (FD) is a rare idiopathic degenerative neurological disorder, which can be present in different heterogeneous manifestations and characterized by bilateral symmetrical cerebral calcification. We present a case of a 55-year-old male who presented with the psychotic feature, bilateral tremors of hand and bilateral symmetrical calcification of basal ganglia. Hence our case suggests that psychiatrists should evaluate the cases of psychosis thoroughly when the age of presentation is atypical, and they should consider the diagnosis of FD when psychosis presents with motor abnormalities.

Key words: *Fahr's disease, psychosis, schizophrenia*

INTRODUCTION

Fahr's disease (FD) also known as idiopathic calcification of the basal ganglia, is a rare sporadic or familial neurological disorder of unknown etiology characterized by extensive symmetrical calcification of the basal ganglia and is associated with neuopsychiatric manifestations. However, Fahr's syndrome is calcification of the basal ganglia secondary to endocrinological causes. FD usually manifests itself in the third to fifth decade of life, but may appear in childhood or later in life. Neuropsychiatric symptoms can be the first or the most prominent manifestations. The most common neurological presentation of FD are parkinsonism, chorea, dystonia, tremor, gait disturbance, dysarthria, seizures, headache, vertigo and myoclonus[1] etc. About 40% of patients with FD present initially with psychiatric features.[2] Psychiatric manifestations include psychotic features, mood symptoms, personality changes, anxiety, dementia, apathy and amnesia etc. We present a case of a 55-year-old male who presented with the psychotic feature, bilateral tremors of hand and bilateral symmetrical calcification of basal ganglia.

Access this article online	
Website: www.ijpm.info	**Quick Response Code**
DOI: 10.4103/0253-7176.178813	

CASE REPORT

Mr. A, a 55-year-old male who was premorbidly well-adjusted and without past and family history of neurological and psychiatric illness presented with complaints of fearfulness, suspiciousness, irritability, hearing of voices decreased sleep for last 2 years. The onset of illness was insidious and course was progressive. From last 1-year his psychotic symptoms increased in intensity and he further developed muttering to self and smiling to self. From last 2 months, he developed bilateral tremors of hand. He has not received any treatment for the above-mentioned symptoms. On mental status examination, he has the delusion of persecution, auditory hallucination of commenting type. On physical examination, he was having bilateral tremors of hand (both resting and intentional). Other than this no other abnormal finding was observed in physical examination. Laboratory investigations, including hemogram, blood biochemistry including serum calcium levels, parathyroid hormone levels, liver

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function tests, renal function tests, thyroid function tests, electroencephalogram, and electrocardiogram were within normal limits. As the age of onset of psychosis was late, noncontrast computerized tomography of the brain was done which demonstrated bilateral symmetrical calcification of globus pallidus. Considering the atypical age of onset of psychosis with presence of movement disorder, basal ganglia calcification on computed tomography scan, normal parathyroid functioning, lack of family history of basal ganglia calcification a diagnosis of FD was considered. He was started on tablet olanzapine 5 mg/day and tablet procyclidine 5 mg/day. The patient's psychotic symptoms gradually improved over a period of 8 weeks.

DISCUSSION

In FD psychotic presentation includes auditory hallucinations (sometimes musical), complex visual hallucinations, paranoid delusions or nondelusional trends, and fugue states. Ideas of reference or influence and catatonia also have been observed. There are two patterns of psychotic presentation in FD, including early onset (mean age 30.7 years) with minimal movement disorder and late onset (mean age 49.4 years) with by dementia and movement disorder.[3] FD may present neurologically "asymptomatic" that is, lacking movement disorders, seizures, or stroke-like events, but with pronounced rapidly progressive cognitive and behavioral abnormalities. Our patient at the age of 55 years presented with psychotic features and tremors only. Symptomatic features in FD may change over time. More extensive calcification and subarachnoid space dilatation correlate with the presence of psychiatric manifestations, but calcific distribution and etiology do not.

Clinical diagnosis of FD was done on the combination of clinical features, brain imaging, and exclusion of other causes of intracranial calcification. The most common radiologic feature of FD is the presence of small bilateral intracranial calcifications which are usually restricted to the globus pallidus, but may also affect the putamen, caudate nucleus, thalamus, dentate nucleus and white matter of the cerebral hemispheres.[4] There is no cure for FD or any standard course of treatment. Treatment is only symptomatic support. Psychotic features can be managed symptomatically. However, studies show that the response to antipsychotics is variable.[3] Patients with FD are more susceptible to neuroleptic malignant syndrome when treated with antipsychotic drugs. Atypical antipsychotics or those with less extra-pyramidal side-effects are the drug of choice because the disease itself causes extra-pyramidal symptoms.

Hence, our case suggests that psychiatrists should evaluate the cases of psychosis thoroughly when the age of presentation is atypical and they should consider the diagnosis of FD when psychosis presents with motor abnormalities. We also advocate the appropriate use of neuroimaging in the diagnosis of different psychiatric disorders.

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How to cite this article: Mohapatra S, Satapathy A. A case of schizophrenia like psychosis due to Fahr's disease. Indian J Psychol Med 2016;38:155-6.

Source of Support: Nil, **Conflict of Interest:** None declared.