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Case Report

Fahr's Syndrome: A Typical Entity Presenting as Psychosis

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I N F O

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A B S T R A C T

Fahr's syndrome is a rare neurological entity characterized by bilateral and symmetrical intracranial deposition of calcium mainly in the basal ganglia. Motor and Psychiatric symptoms are clinically relevant features in this syndrome. We report a case where psychiatric symptoms and features of delirium were more prominent as compared to motor symptoms. Radiological findings depicted bilateral basal ganglia calcification and oedema in parietal region mainly on right side. Parathyroid hormone and calcium levels were mildly low with no significant past or family history. Most behavioural symptoms showed significant improvement on treatment with Risperidone, Amisulpride, Lorazepam, Sodium Valproate, Calcium, Folic acid and Trihexyphenidyl but some symptoms like auditory hallucinations remained unresponsive.

Keywords: Basal Ganglia Calcification, Fahr's Syndrome, Parathyroid Hormone, Hypocalcemia, Psychiatric Symptoms

Introduction

Fahr's syndrome is a rare Neurological condition characterized by deposition of calcium in Basal Ganglia.[1] The most common region in the brain structure involved is lenticular nucleus and the internal globus pallidus.[2] Calcified regions' circulation in brain is mainly correlated with clinical signs.[3] The disease usually presents in later ages but may appear in childhood also with early presentation in less than 30 years of age and late presentation after 49-50 years of age.[4] Neuropsychiatric symptoms generally presented are mild difficulty in concentration, amnesia, behaviour abnormalities, frank psychosis, motor symptoms, and dementia.[5] In clinical scenarios, 40% of patients with Basal Ganglia Calcification (BGC) may come into attention with psychiatric symptomatology in from of mania, apathy or psychosis. The available treatment is mainly symptom control and treating underlying causes of calcification. Prognosis is unpredictable with neurological deterioration as main cause of further disability or sometimes death in such rare conditions.[6]

Case Description

A 33-year old married female with ten years of formal education belonging to middle socio-economic status presented with her mother to Outpatient Unit of Department of Psychiatry Lady Harding Medical College, Delhi with acute onset symptoms of last 15 days duration characterised by diffuse headache, abnormal behaviour, reduced sleep and one episode of generalised tonic clonic seizure and occasional abnormal jerky movements in left lower limb. Symptoms of decreased appetite, hearing voices of a man, undue suspiciousness towards parents and unprovoked aggression were present since 3 days prior to presentation. No apparent precipitating factor or past history or family history of neuropsychiatric illness was

evident. In view of marked violent aggressive behaviour patient was admitted. Physical examination revealed bilateral intentional tremors, poor cortical sensation in sensory examination with no evidence of rigidity or cerebellar dysfunction. Mental state examination revealed a thin built female ill-kempt having increased psychomotor activity, irrelevant speech, loosening of association, delusion of persecution, auditory hallucinations second person type with absent insight. Blood investigations, haemogram, liver function tests, renal function tests, thyroid function tests, electroencephalogram, and electrocardiogram did not reveal any abnormality.

Patient was not accepting oral medications and was given injection Haloperidol 5 mg and Promethazine 50 mg IM route on day 1. On day 2, patient appeared disoriented to time and person during the afternoon with no signs suggestive of extrapyramidal side effects. Her serum electrolytes showed sodium level as 132 mEq/ L and chloride 94 mEq/L. Patient electrolytes were corrected by fluid therapy and electrolytes were reviewed on repeated values which were normal, her delirium also improved on day 3.

In view of persisting headache MRI Brain was done which revealed bilateral basal ganglia calcifications mainly in caudate nucleus and oedema in the right parietal region. Further work up revealed mild decrease in serum PTH level (8.9 pg/Ml) and serum calcium level (7.4 mg/Dl).

Diagnosis of Fahr's syndrome was made and patient was started on tab Sodium Valproate 500 mg twice daily. For psychotic symptoms adequate trial of risperidone upto 6mg was tried with no significant improvement over 3 weeks. Risperidone was cross tapered to Amisulpiride and was optimised upto 200mg and there was significant improvement in delusion and related violent behaviour, frequency of auditory hallucination decreased but persisted.

Discussion

Study by Nicolau et al. reported that calcium and phosphate metabolism was mainly disturbed in these rare-neurological-disorders which leads to hypocalcemia and hypoparathyroidism which was seen in investigation profile of our patient also.[7,8] Other etiologies of Fahr's syndrome may also include infections, metabolic, and genetic diseases.[9] Kazis et al. in his review reported that the neurological findings of basal ganglia calcifications were found to be around 0.3% to 1.3% as the patient had basal ganglia bilateral calcifications and the right side parietal area oedema.[10,11] General clinical features are mainly headache, tremors, motor symptoms, psychosis, mania**,** seizures and memory changes as reported by Modrego et al., we found similar findings in our patient reported here.[12] Simone et al. have reported a case of Fahr's syndrome with syncope, delirium, motor symptoms and hypocalcemia with psychiatric issues. Our patient showed similar features of delirium and psychotic features in the form of, disorganized behaviour, pacing around, and irritability. Early onset psychotic presentation in Fahr's syndrome is generally presented with mean age of approx. 30.7 years with less involvement of movement disorder and late onset with mean age of 49.4 years associated with dementia, amnestic changes and movement disorders, our patient age was 33 years which seems to be early onset type. The treatment of Fahr's syndrome is focussed on treating underlying causes. There is variable response to drugs in Fahr's syndrome management and sometimes psychiatric symptoms are unresponsive to drugs as we found auditory hallucination very less responsive to drugs in our patient.[13]

Conclusion

Psychiatrists should always keep in consideration of comorbid medical or neurological causes for psychiatric manifestations. We highlighted a rare entity of Fahr's syndrome as an underlying diagnosis in clinical picture of psychosis associated with motor abnormalities. This case gives us the insight of important role of neuroimaging and the relevant endocrinological hormone assays for psychosis in clinical workup of a psychiatric patient presented with psychosis. Symptomatic management proved to be helpful in controlling some behavioural and motor symptoms of this entity and some are unresponsive to management like we found hallucinations in our case. So a thorough medical and neurological knowledge is essential to evaluate other causes of psychosis before reaching a diagnosis of primary psychiatric illness.

Conflict of Interest: None

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