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Case Report

Fahr's Syndrome: Rare Cause Of Parkinsonism- A Case Report

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Abstract: A 63 years old male presented with progressive decline of motor functions and characteristic resting 'pill rolling' tremor of bilateral upper limbs. CT scan brain showed diffuse cerebral atrophy with bilateral cerebellar vermis, basal ganglia and parietal lobar calcification. The clinical and radiological features in this case suggested the diagnosis of Fahr's syndrome.

Keywords: Fahr's syndrome, Basal ganglia calcification, Cerebellar calcification

INTRODUCTION:

Fahr's syndrome, also known as idiopathic basal ganglia calcification, cerebrovascular ferrocalcinosis, Fahr's intracerebral calcinosis, or Fahr's disease[(1)], is a rare neurological disorder of unknown etiology characterized by Parkinsonian or choreoathetotic type movement disturbance, neuropsychiatric abnormalities and symmetrical calcification of basal ganglia and dentate nucleus of cerebellum. The basic pathology in Fahr's syndrome involves progressive calcific deposits in the blood vessels of the basal ganglia and the dentate nuclei of the cerebellum. [(2)]. It was first noted by Karl Theodore Fahr in 1930. The disease is often familial, with autosomal dominant inheritance, but few cases have been reported to have autosomal recessive inheritance [(3)].

CASE REPORT:

A 63 years old male patient, non-smoker and not a known case of hypertension or diabetes mellitus presented with resting 'pill rolling' tremor involving both upper limbs and lower limbs for the last 5 years and gradual deterioration of motor functions causing slow initiation of movement and rigidity of muscles of limbs and trunk for the last 3 years. There was history of aggressive behavior with increased irritability and reduced sleep. Family members reported frequent use of sedatives for this reason. This was associated with gradual deterioration of speech and shuffling gait. Patient was bed ridden for 20 days and in altered sensorium for 3 days prior to admission. There was no family history of mental illness or dementia.

On examination**,** patient was normotensive and afebrile. Voice was low volume with poorly discernible speech. Patient was conscious but laid supine with minimal limb movements, had a staring look and mask like facial expression. Patient was conscious and drowsy but able to follow verbal commands.

There was no cranial nerve dysfunction. Motor system examination showed normal muscle mass, power and tone with normal deep tendon reflexes. 'Cog wheel' rigidity was noted in muscles of bilateral upper and lower limbs. There was no sensory deficit noted. Bladder and bowel continence was maintained.

Investigations revealed Hb- 10.8 g%; ESR- 30 mm/ hr; TLC- 8700/ mm3; Serum Na+ 138.7 meq/L; Serum K+ 3.62 meq/L; RBS- 77 mg/dL; Total Bilirubin: 0.73 mg/dL; Serum Albumin 3.8 g/dL; AST- 25 IU/ L, ALT 32 IU/ L; Serum urea 29 mg/dL; Serum Creatinine 0.82 mg/dL. Serum Phosphorus 4.3, Serum Calcium 7.1. CT Scan Brain showed bilateral symmetrical calcification of cerebellar vermis, parietal lobe and basal ganglia with diffuse cerebral atrophy.

The typical history, characteristic clinical features, supporting radiological findings and normal blood biochemistry suggested the diagnosis of the rare disorder of Fahr's syndrome.

DISCUSSION:

Idiopathic basal ganglia calcification is also known as Fahr's disease or Fahr's syndrome. It is a rare inherited or sporadic neurological disorder with a prevalence of less than 1/ 1,000,000 [(4)]. It is characterized by abnormal deposition of calcium in areas of brain that control movement including basal ganglia, thalamus, dentate nucleus, cerebral cortex, cerebellum, subcortical white matter, and hippocampus [(5)].

Most cases present with extra pyramidal symptoms initially. Additionally they may present with cerebellar dysfunction, speech difficulty, dementia and neuropsychiatric symptoms.[6] The disease typically presents between 4th to 6th decades of life.

Symptoms of this disorder may include- deterioration of motor function, dementia, seizures, headache dysarthria, spasticity, visual impairments and athetosis. Fahr's syndrome can also include symptoms of Parkinsonism such as tremors, muscle rigidity, mask like facial expression, shuffling gait and pill rolling motion of the fingers. Neurological evaluation reveals Parkinsonism to be present in 57% of the cases. [4]

Making a clinical diagnosis of Fahr's syndrome relies on clinical features, brain imaging along with exclusion of other causes of intracerebral calcifications. Normal serum levels of calcium and phosphorus in our patient helps to rule out endocrine causes like hyperparathyroidism. Other causes like tuberous sclerosis, toxoplasmosis, systemic lupus erythematosus must also be ruled out in suspected cases.

There is no cure for Fahr's syndrome and neither is there a standard course of treatment. Treatments target symptomatic support. Levodopa was prescribed in our case for control of Parkinsonism features. Antipsychotics and anticonvulsants can also be used to control the respective symptoms.

Optimal time for determination of genetic risk is before pregnancy. Brain CT scan serves as a pre- symptomatic screening test in at-risk individuals, but is not useful for predicting age of onset, severity, type of progression in asymptomatic individuals. Consensus holds that individuals lesser than 18 years of age who are at risk but asymptomatic should not be tested as it may remove future autonomy, harm parent child relationship, cause anxiety and guilt, as there are limited treatment options [7]

CONCLUSION:

Idiopathic basal ganglia calcification or Fahr's syndrome is a rare neurodegenerative disorder that is passed on in families as an autosomal dominant trait, and is a cause of Parkinsonism. So, detailed family history and imaging studies are indicated to rule out Fahr's disease in patients with Parkinsonism features. Also, radiological screening may be employed in first-degree relatives of patients of Fahr's syndrome in order to determine their susceptibility. The role of genetic counselling before conception in susceptible individuals is also of paramount importance.

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