Case Report

Fahr's Syndrome: A rare case- Presented as Acute Ischaemic Stroke

Sarkar DN[1], Sarkar D[2], Zahin AKM[3], Ohab MA[4], Miah MS[5], Haque MM[6]

Abstract

Fahr's syndrome refers to a rare syndrome characterized by symmetrical and bilateral intracranial calcification. We present a 65-year-old female with Fahr disease, presenting with headache with acute ischaemic stroke with left sided hemiplegia. CT scan of brain reveals irregular variable size hyperdense areas are noted in both basal ganglia regions and in both cerebellar hemisphere.

1. Corresponding Author:
Prof. Dr. Devendra Nath Sarkar
MD, MCPS, FCCP, FRSM, FACP
Professor
Department of Medicine
Rangpur Medical College Hospital, Rangpur.
e-mail: dnathsarkar@yahoo.com

2. Dr. Devavrata Sarkar
Medical Doctor
Enam Medical College Hospital, Savar, Dhaka.

3. Dr. A K M Zahin
Assistant Register
Department of Medicine
Rangpur Medical College, Hospital, Rangpur.

4. Dr. M A Ohab
MD
Assistant Professor
Department of Medicine
Rangpur Medical College Hospital, Rangpur.

5. Dr. Md. Shahin Miah
Register
Department of Medicine
Rangpur Medical College Hospital, Rangpur

6. Dr. Md. Mahmudul Haque
Assistant Register
Department of Medicine
Rangpur Medical College Hospital, Rangpur.

Introduction

Fahr's disease (FD) is a rare, degenerative, neurological condition characterized by idiopathic calcification of the basal ganglia. This condition has been known since the middle 1800s. The clinical manifestations of Fahr's disease vary. One definition proposed by, requires bilateral calcifications with neuropsychiatric and extrapyramidal disorders with normal calcium and phosphorus metabolism[1]. Another definition which had seizures, rigidity and dementia with characteristic calcification of the basal ganglia[2]. Others opined that radiologists may view basal ganglia calcification (BGC) as an incidental finding so clinical findings associated with Fahr's disease are important[3].

The basal ganglia and dentate nucleus are the most common site of involvement and most cases present with extra pyramidal symptoms. This disease is mostly associated with a phosphocalcic metabolism disorder, especially to hypoparathyroidism. Defective iron transport and free radical production may damage tissue, initiating calcification[2]. In adult-onset FD, calcium deposition generally begins in the third decade of life, with neurological deterioration two decades later[4]. Reduced blood flow to calcified regions correlates with clinical signs.

Case Report

Our patient, 65 years old normotensive, non diabetic woman presented with headache for last three months which is diffuse, mostly occur throughout the day without any history of nausea, vomiting, aura or altered level of conciousness. She has no history of trauma convultion or fever. Before the day of admission patient had a history of sudden fall on the ground with vomiting and altered level of conciousness. On clinical examination pulse 80 b/min, BP: 140/90 mm of Hg, temperature: 99 degree farenhite, respiratory rate 18 breath/min. Neurological examination reveals. GCS: 9 out of 15. Bulk and tone of muscle is normal. Power of muscle of both left upper and lower limb is 2/5. Jerks are exaggerated of both left upper and lower limb. Planter is extensor on left side. All modalitys of sensation is intact, co ordination is not possible. Sign of meningeal irritation was absent. Fundoscopy shows no abnormality.

Laboratory tests including serum calcium, phosphorus, and parathyroid hormone (PTH) levels are within normal limits. CT scan of brain reveals irregular variable size hyperdense areas are noted in both basal ganglia regions and in both cerebellar hemisphere which suggestive calcification. So ischemic stroke due to FD is diagnosed and treated conservatively with physiotheraphy.

CT scan of brain showing irregular variable size hyperdense areas are noted in both basal ganglia regions and in both cerebellar hemisphere (figure-I).

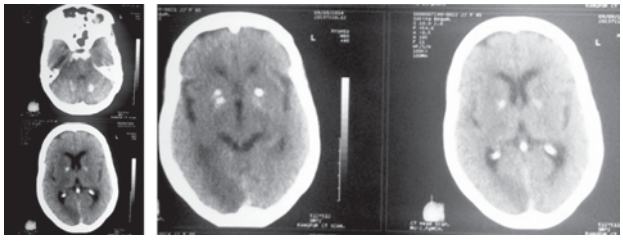

Figure-I: CT scan of brain

Discussion

Fahr's syndrome is a very rare disease with unknown prevalence, characterised by symmetrical intracranial calcification with a predilection for the basal ganglia and dentate nuclei. As the basal ganglia and dentate nuclei are always involved in a symmetrical pattern, the term Bilateral Striopallidodentate Calcinosis (BSPDC) was suggested[5]. Typical age of presentation is in middle-age between the 4th - 6th decades of life (as seen in our case), although an early onset type may also occur[6]. 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 Parkinson's disease such as tremors, muscle rigidity, mask-like facial appearance, shuffling gait, and a "pin-rolling" motion of the fingers. Our case presented with headache with acute ischemic stroke. The brain metabolism in a person with Fahr's disease who presented with predominant frontal lobe syndrome and dementia[7]. There was a massive reduction in the glucose metabolism in both the basal ganglia and frontal lobes which correlated with the clinical picture of disinhibition and personality change. The involvement of frontal-subcortical circuits provides a hypothetical framework for the interpretation of cognitive and psychotic problems in Fahr's disease. Making a clinical diagnosis of Fahr's syndrome relies on the combination of clinical features, brain imaging and exclusion of other causes of intracranial calcification. Symmetrical and extensive calcification of basal ganglia, dentate nuclei and centrum semiovale are typical and conspicuous of Fahr's syndrome. Normal serum levels of calcium, phosphorus, alkaline phosphatase and parathormone can help in differentiating it from the endocrine disorders like hyperparathyroidism, pseudo-hypoparathyroidism and pseudo-pseudo-hypoparathyroidism. Other rare causes of scattered basal ganglia and dentate nuclei calcification are tuberous sclerosis, toxoplasmosis, syphilis and inflammatory illness such as systemic lupus erythematosus and all these must be ruledout in suspected cases. There is no cure for Fahr's syndrome, nor is there a standard course of treatment. Treatment targets symptomatic support. The response to Levodopa in those with Parkinson's features is reportedly poor. Antipsychotics may be indicated in those with psychotic symptoms and behavioural problems, and anticonvulsants for the control of seizures.

In conclusion, though Fahr's syndrome is a rare idiopathic neurodegenerative disorder, yet, with proper knowledge of its clinical manifestations and more vigilant approach, a diagnosis can be made in time. More research is required to locate and understand the action of the genes involved in this disorder. Finding these genes could lead to effective ways to treat and prevent Fahr's syndrome.

References

l. Trautner RJ, Cummings JL, Read SL. Idiopathic basal ganglia calcification and organic mood disorder. Am J Psychiatry. 1988;145:350-353.

2. Beall SS, Patten BM, Mallette L. Abnormal systemic metabolism of iron, porphyrin and calcium in Fahr's syndrome. Ann Neurol. 1989;26:569-575.

3. Flint J, Goldstein LH. Familial calcification of the basal ganglia: a case report and review of the literature. Psychol Med. 1992;22:581-595.

4. Manyam BV, Bhatt MH, Moore WD. Bilateral striopallidodentate calcinosis: cerebrospinal fluid, imaging and electrophysiological studies. Ann Neurol. 1992;31:379-384.

5. Manyam BV. Bilateral strio-pallido-dentate calcinosis: a proposed classification of genetic and secondary causes. Mov Disorder. 1990;5 (supl): 94S.

6. Lam JSP, Fong SYY, Yiu GC, Wing YK. Fahr's disease: a differential diagnosis of frontal lobe syndrome. Hong Kong Med J. 2007;13:75-7.

7. Benke T, Karner E, Sappi K. Subacute dementia and imaging correlates in a case of Fahr's disease. J Neurol Neurosurg Psychiatry. 2004;75:1163-5.