Case Report

A case of Fahr's Disease - A bright brain

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Abstract

Background: The term "Farh's disease" was first coined by Karl Theodore Fahr in 1930. As seen in medical literature, this disease is predominantly autosomal dominant in inheritance as compared to autosomal recessive1.Fahr's disease is a rare neurological disorder often characterized by neuropsychiatric behavior, Parkinsonian type movement disturbances and symmetrical calcification of the basal ganglia and dentate nucleus in the cerebellum2. We report a case of Fahr's Disease in a 60 year old female.
Key Word: Fahr's Disease.

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CASE REPORT

A 60 year old, South Indian female (non alcoholic, non smoker) presented with progressive slurring of speech, ataxia and chronic headache. She also reported one suicidal attempt in the last 4 months. There was no history of seizures, trauma, or any previous history of any psychiatric or other illness. Disorientation to time, place and person was occasionally present and obeyed verbal commands.

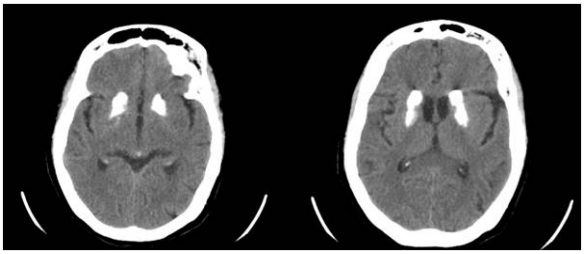

Figure 1: Showing bilateral calcification of the basal ganglia (axial view)

Blood pressure recorded was within normal limits. Cranial nerve examination showed no deficit. No sensory loss was present. Motor examination revealed normal power, with no muscle wasting and no exaggeration of reflexes. Patient had a wide stepping gait, with slurring of speech which was incoherent. Other cerebellar signs were tested negative. Remote memory was lost. Patient also reported history auditory hallucinations.

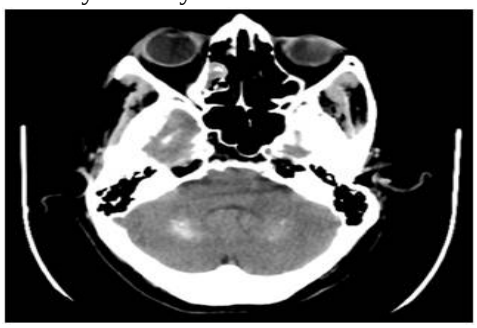

Figure 2: Shows bilateral calcification of dentate nucleus of cerebellum

Patient's blood counts were in normal limits, renal profile, lipid profile, serum calcium, serum parathyroid hormone, thyroid function tests were normal. Chest x-ray and ECG were normal. CT brain showed bilateral calcification of the basal ganglia and dentate nuclus of cerebellum. With the patient's clinical history and normal blood profile and CT brain findings, a diagnosis of Fahr's disease was established and was treated with benzodiazepines.

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DISCUSSION

Fahr's disease is a rare condition and often mimics symptoms of other psychiatric or neurological disorders. As the basal ganglia and dentate nuclei are always involved in a symmetrical pattern, the term Bilateral Striopallidodentate Calcinosis (BSPDC) was given[3]. Presenting symptoms like pill rolling tremors, gait disturbances, rigidity often confuse clinicians with an alternative diagnosis of Parkinson's. An effective diagnosis of Fahr's disease is made based on a thorough clinical history, neurological examination, brain imaging and blood biochemistry to rule out other differentials. Patients should be screened for toxins, serum parathyroid hormones, serum calcium levels to exclude hyperparathyroidism or pseudo-hypoparathyroidism. Our Patient was put on levodopa but was not effective, but benzodiazepenesdid help over time[4].

CONCLUSION

Though the cause of Fahr's Disease is not well established and with no definitive cure as of date, a detailed clinical history, brain imaging scans and blood investigations and a vigilant approach can help clinicians to make an early diagnosis and start with symptomatic management so that patients can lead a near to normal life as studies go on to find a cure. Typically, symptoms begin between 30-60 years of age group. The clinical evolution is that of a degenerative disorder, rather than a developmental disorder. Fahr's disease has a prognosis difficult to establish and explain.

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