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Short Communication

Fahr's disease: A rare case report

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ABSTRACT

Fahr's disease may be defined as a rare, degenerative neurological condition which presents itself as an idiopathic, bilateral deposition of calcium in the striopallidodentate area of the brain. Here we are presenting a case of a 38 years old female patient with complaints of fever since three days, two episodes of generalized tonic–clonic seizure (GTCS) which lasted for 10-15 minutes. Symptoms like anger outbursts, irritability, forgetfulness, tingling and numbness of extremities were noted. Chvostek sign, Trousseau sign and Café au lait spots were observed. Based on the evidences obtained from laboratory investigations and CT/USG scan the patient was diagnosed with Fahr's disease. The pharmacotherapy of this case included Phenytoin 100 mg for convulsions, Calcium gluconate injection (10%) single dose under heart rate monitoring, Ferrous sulfate, Vitamin D supplements, Thyroxine for elevated TSH levels and Ceftriaxone for prophylaxis. As with other neurodegenerative conditions, there is currently no known cure for Fahr's disease, thus symptomatic relief is the main focus of treatment. Testing calcium levels every month is mandatory to modify the treatment plan accordingly. Awareness about physical health, psychological counseling and Cognitive behavioural therapy may be beneficial in improving the quality of life and prevent further health complications.

Keywords: Fahr's Disease, Generalized tonic–clonic seizure, Thyroidectomy, Hypocalcemia, Bilateral basal ganglia calcification

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INTRODUCTION

Fahr's disease may be defined as a rare, degenerative neurological condition which presents itself as an idiopathic, bilateral deposition of calcium in the striopallidodentate area of the brain. The disease was first described in 1930 by a German pathologist named, Karl Theodor Fahr. Symptoms of Fahr's disease include degradation of motor function, seizures, dementia, headache, athetosis, eye impairments, spasticity and dysarthria. Fahr's disease may also be comprised of symptoms that are characteristic to Parkinson's Disease like a "Pill rolling" movement of the fingers, shuffling gait, tremors and muscle rigidity. These symptoms usually develop later as the disease progresses. Symptoms that are usually observed in Fahr's disease include dystonia and chorea. The disease usually develops between the age of 40-60 years [1].

Case History

A 38-year-old female patient was admitted to Vivekananda General Hospital, Hubballi with complaints of fever since three days and two episodes of GTCS which lasted for 10-15 minutes. Her past medical history revealed thyroid disorder with surgical procedure known as thyroidectomy. Symptoms like anger outbursts, irritability, forgetfulness, tingling and numbness of extremities were noted. Chvostek sign, Trousseau sign and Café au lait spots were observed. The patient had received Inj. Calcium Gluconate two months back and was on regular treatment with Calcium 500 mg. Ultrasound scan of the neck revealed total thyroidectomy. Dermatological consultation revealed multiple angiofibroma over face, wart with dermatitis neglect, wart over left ring finger. 2-D Echo findings were normal. Parathyroid glands (PTH) was intact and parathormone was 4.7 pg/d/L, Calcium 6.7 mg/dL (8.5-10.5 mg/dL). Based on the evidences obtained from laboratory investigations and CT/USG scan the patient was diagnosed with Fahr's disease. The pharmacotherapy of this case included Phenytoin 100 mg for convulsions, Calcium gluconate injection (10%) single dose under heart rate monitoring, Ferrous sulfate, Vitamin D supplements, Thyroxine for elevated thyroid stimulating hormone (TSH) levels and Ceftriaxone for prophylaxis.

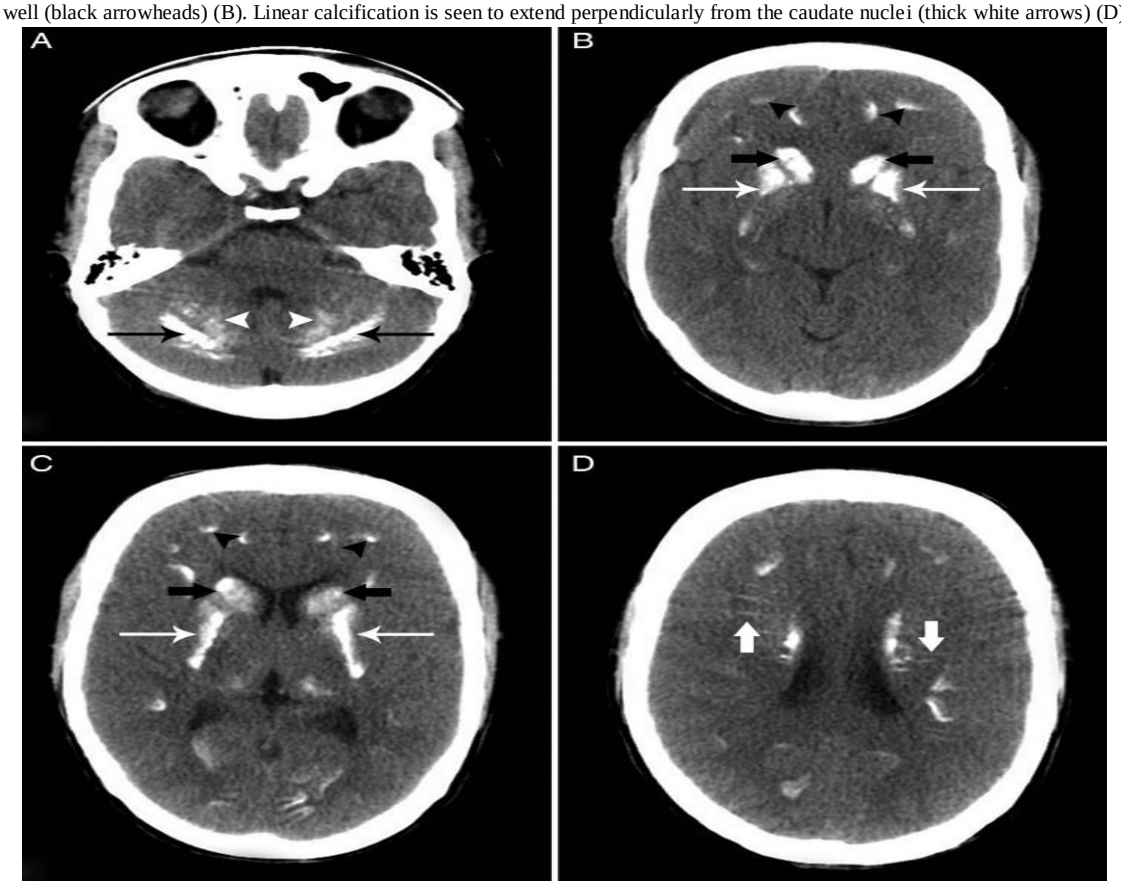
DISCUSSION

Fahr's disease is caused due to defect in iron transport and free radical production resulting in calcification. It may either affect the metabolism of calcium in brain or cause metastatic deposition. It progresses till the neuron reducing the blood flow. Bilateral basal

ganglia calcifications are most frequently caused by abnormalities of the PTH. Hyperphosphatemia and hypocalcemia are produced on by decreased PTH activity, which encourages calcification [2].

Hypocalcemia is a major post-operative complication in thyroidectomy [3]. In this case, severe hypocalcemia is observed due to both thyroidectomy and abnormal calcium metabolism from Fahr's. This can result in an increased hospitalization. Hence, the patient was treated with multiple calcium supplements to prevent worsening of symptoms.

Figure: 1 Fahr's disease (Striopallidodentate calcinosis). Cerebellar foliae on an axial non-contrast CT image show bilateral symmetrical calcifications (thin black arrows) (A). Notably, both dentate nuclei have calcification (white arrowheads) (A). There is significant bilateral calcification in the putamina, lateral globus pallidi, and head of caudate nuclei (thick black arrows in B and C), with medial globus pallidi and thalami being somewhat spared (B). There is calcification at grey–white matter junction as well (black arrowheads) (B). Linear calcification is seen to extend perpendicularly from the caudate nuclei (thick white arrows) (D) [4].



Fahr's disease is known to affect psychological behavior of the patient. Few commonly observed symptoms include auditory and visual hallucinations, complex perceptual distortions, delusions etc. [5]. In this case, the patient expressed worries regarding her clinical condition. She had borderline intelligence. Such factors worsen the symptoms and decrease quality of life of the patient. Since the primary goal is to achieve normal calcium levels, psychological intervention or anti-depressants were not used for the management. In case of severe symptoms like hallucinations, suitable anti-psychotics which have least interactions with the prescribed anticonvulsants can be recommended.

Certain dermatological manifestations terms used in this case are Chvostek sign which is increased irritability of the facial nerve [6]. Trousseau sign is considered positive when spasm of hand and wrist occurs after an individual wear a blood pressure cuff inflated for two to three minutes [7]. This is a classic sign to determine if an individual has hypocalcemia. Tuberous sclerosis was noted which is a rare genetic condition that causes benign tumors in different parts of the body [8]. In this case, since the patient has genetic disease, skin manifestations are common and are of specific type. There was no particular treatment given for Tuberous sclerosis which if uncontrolled, may become cancerous. Identification of the extent of such manifestations and planning a precise treatment algorithm is very important to prevent future complications and maintain a balanced lifestyle.

CONCLUSION

As with other neurodegenerative conditions, there is currently no known cure for Fahr's disease, thus symptomatic relief is the main emphasis of the treatment. Patients with Fahr's illness who use carbamazepine, benzodiazepines, or barbiturates may experience more gait impairment [9]. Testing calcium levels every month is mandatory to modify the treatment plan accordingly. Regular check-ups, awareness about physical health, psychological counseling and cognitive behavioral therapy can help in improving the quality of life

and prevent further health complications.

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Declaration of Patient Consent

Consent of patient to collect information was taken.

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