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Case Report

A Case of Seizure Diagnosed with Primary Hypoparathyroidism with Fahr's Syndrome: A Case Report

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Abstract:

Fahr's syndrome is a rare neurodegenerative disorder associated with bilateral symmetric calcification in basal ganglia, thalami, deep cerebellar nuclei and subcortical white matter. The clinical presentations of Fahr's syndrome are pyramidal signs, Parkinsonism, gait disorders, cognitive disorders, movement disorders and psychotic symptoms. Here, we report a case of chronic hypocalcemia due to primary hypoparathyroidism diagnosed with Fahr's syndrome.

Keywords: Fahr's Syndrome, Bilateral Basal Ganglia Calcification, Hypocalcemia, Seizure, Hypoparathyroidism.

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Introduction

Fahr's disease was first reported by Karl Theodor Fahr in 1930 as a rare familial autosomal dominant disease with idiopathic basal ganglia calcification. It is defined as symmetric bilateral basal ganglia calcification in the absence of an explainable underlying abnormality such as endocrine disorders, biochemical disorders, infections, toxins, or history of trauma [1]. If calcifications are due to above mentioned causes then the diagnosis must be classified as "Fahr's syndrome" rather than "Fahr's disease" [2].

The pathogenesis of the calcifications is not completely understood .It manifests in age of 40-60 years old and often found incidentally on CT imaging in patients who present with non-specific symptoms such as movement disorders, gait abnormalities, dementia, cognitive deficits or seizures. The pathogenesis of the calcifications is not completely understood. There are various theories explained in literature. There is no definitive treatment for Fahr's disease [3,4].

Case Report

A 49 years old female was presented to emergency department with complaints of involuntary tonic clonic movements of all four limbs associated with frothing from mouth lasted for 2 to 3 minutes. She had similar episode 3 days back and similar 10 to 15 episodes in last 10 years but never evaluated because of financial issues. On arrival patient was drowsy but arousable and was following simple commands. She had history of mild cognitive impairment and personality changes and rest other neurological examinations were normal. Non-contrast brain tomography (CT) imaging showed calcifications in both basal ganglia (Figure 1). Electroencephalography (EEG) examination was normal.

laboratory investigation revealed hypocalcemia level of 5.40mg/dl, ionized calcium 0.49mmol/l, hyperphosphatemia serum phosphate level 6.12 mg/dl, hypoparathyroism serum parathyroid level 14.20 pg/ml, 24 hours urine calcium level 502mg, Vitamin D3 32.40ng/ml. Other electrolytes were normal. Ultrasound neck, liver function and renal function were normal. She did not have any significant past or family history. Based on above investigations, a diagnosis of Fahr's syndrome due to primary hypoparathyroidism was made. The patient received intravenous calcium gluconate to relieve symptoms and continued to take oral calcium and calcitriol for further treatment.

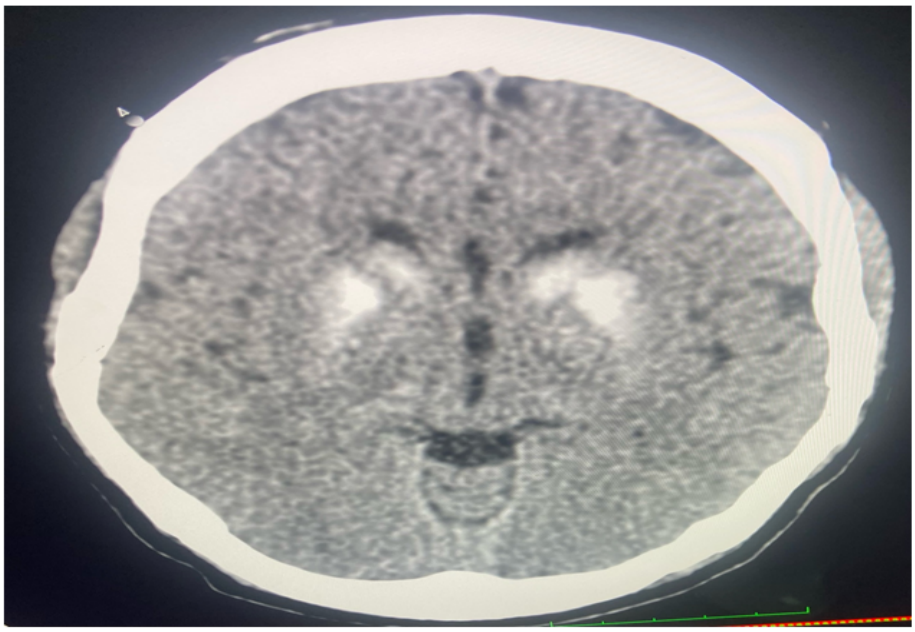

Figure 1: Image of CT SCAN showing bilateral basal ganglia calcification

Discussion

Hypoparathyroidism is caused by insufficient secretion and/or resistance to parathyroid hormone. Hypoparathyroidism can be primarily, secondary or pseudo- hypoparathyroidism. It leads to hypocalcemia, hyperphosphatemia and increased neuromuscular irritability. Patient can present with myalgias, seizures, twitching, tetany. The clinical trial of hypocalcemia, hyperphosphatemia, and hyperparathyroid are called pseudo- hypo parathyroidism because of the resistance of target organs such as the kidneys and bone to parathyroid hormone. Some patients can have special physical deformities, such as short stature, round face, short neck, short finger (toe), obesity or mental retardation, called Albright Hereditary Osteodystrophy [5]. Most common cause of hypoparathyroidism is Neck surgery followed by autoimmune diseases and genetic factors. Other factors are abnormal magnesium metabolism, invasive diseases and ionizing radiation [6].

Fahr's syndrome is a rare disorder in which calcium metabolism is affected due to hypoparathyroidism or pseudo hypoparathyroidism as well as other conditions such as developmental, infectious, genetic, sporadic, metabolic and other conditions [7].

Fahr's syndrome can present with seizures, tetany, tremors, chorea, dystonia, myoclonus, spasticity, dementia, gait disorder, speech impairment, Parkinsonism, and rarely coma can occur. Fahr's syndrome can also lead to Neuropsychiatric symptoms such as mild difficulty with concentration ,dementia, personality disorders, behaviours disorder, and psychosis [8]. Histopathological studies revealed that major element is calcium and calcium is responsible for radiological changes of the brain. Other important minerals are aluminum, iron, zinc and magnesium [9].

Conclusion

Parathyroid disorder should be considered in a patient with chronic hypocalcemia. Though there is no specific treatment for Fahr's syndrome, CT scan should be performed to look for abnormal intracranial calcification. Treatment is based on underlying cause and supportive treatment for symptoms since there is no definitive treatment for Fahr's syndrome [10].

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