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Fahr's Syndrome: A Rare Case of Bilateral Basal Ganglia Calcification Presenting with Neurological Symptoms

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ABSTRACT

Fahr's syndrome is a rare neurological condition characterized by symmetrical and bilateral calcification of the basal ganglia associated with neuropsychiatric disorders. Calcification may also be found in other brain regions such as the dentate nucleus, thalamus, and cerebral cortex. Patients typically present with neuropsychiatric and movement disorders. We present a case of a 59-year-old female who came to the emergency department with tremors, hands and face rigidity, slurred speech, and headache. Laboratory findings revealed hypocalcemia, and a head CT scan revealed bilateral calcification of the basal ganglia. The patient was treated with calcium replacement therapy, and her symptoms were rapidly improved.

Keywords: Fahr's syndrome; basal ganglia calcification; hypocalcemia; hypoparathyroidism; movement disorders.

INTRODUCTION

Fahr's syndrome is a rare neurological condition characterized by symmetrical and bilateral calcification of the basal ganglia associated with neuropsychiatric disorders. Calcification may also be found in other brain regions such as the dentate nucleus, thalamus, and cerebral cortex.[1] The calcification is hypothesized to be due to abnormal brain calcium metabolism or an altered blood-brain barrier. Fahr's syndrome may present as a primary or secondary disease to hypoparathyroidism. Fahr's syndrome commonly affects people between the 40 to 50 years of age.[1,2]

The disease has an insidious onset with patients presenting with neurological and psychiatric symptoms such as bradykinesia, rigidity, tremors, mask-like facies, fatigue, slurred speech, muscle cramps, and dementia. Fahr's syndrome is diagnosed by the finding of neurologic dysfunction and radiographic evidence of bilateral calcification of the basal ganglia.[2,3] Currently, there is no specific treatment to limit the progression of calcification in the basal ganglia. The treatment is symptomatic to improve the neuropsychiatric symptoms.[1,4]

CASE PRESENTATION

We report a case of a 59-year-old female who came to the emergency department with tremors, hands and face rigidity, slurred speech, and a headache. She stated that her symptoms have been relapsing about three times a year for the last 8 years. She had been hospitalized every time the symptoms relapsed for electrolyte correction. Past laboratory workup had always revealed hypocalcemia by the time the disease relapsed. This year alone, she had been admitted to the hospital for 4 times despite taking daily calcium and vitamin D3 supplementation. The patient denied any prior surgery or working in a place with high exposure to radiation in the past.

The patient was conscious, her vital sign examination was within the normal range as follows: blood pressure was 120/80 mmHg, heart rate was 90 beats per minute, respiratory rate was 20 times a minute, body temperature was 36.8 °C. Physical examination shows hand tremor and hand rigidity; the patient's hands were frozen in a closed position, and any attempts to open her hands failed. Her face was also rigid, which caused slurred speech. Due to a suspicion of electrolyte imbalance, the Chvostek and Trousseau test was done, and both tests elicited a positive result, which suggests hypocalcemia. Laboratory workup was done, and the laboratory examination revealed hypocalcemia and hypokalemia as shown in the table below.

TABLE 1: Laboratory findings on admission.

Test	Result	Unit	Normal Range
Hematology			
Hemoglobin	12.0	g/dL	12.3-15.3
Leukocyte	8,560	/mm^3	4,500-10,000
Thrombocyte	390,000	/mm^3	150,000-450,000
Hematocrit	38	%	35-47
Clinical Chemistry			
Creatinine	0.55	g/dL	0.5-1.1
Electrolyte			
Sodium	141	mmol/L	135-148
Potassium	3.58	mmol/L	3.5-5.1
Calcium	5.22	mmol/L	8.1-10.4

Hypocalcemia is thought to occur as a manifestation of hypoparathyroidism. Head CT scan revealed bilateral calcification of the basal ganglia, as shown below.

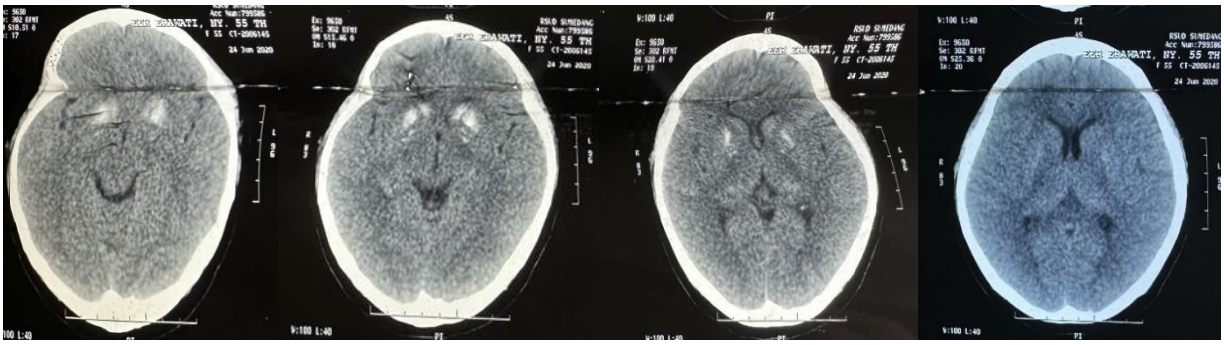

FIGURE 1: Head CT scan revealing bilateral calcification of the basal ganglia.

Due to the clinical findings of neurological symptoms such as tremors, muscle rigidity, and slurred speech, and a head CT scan revealed bilateral calcification of the basal ganglia, the patient was diagnosed with Fahr's syndrome. The laboratory workup revealed hypocalcemia, which suggests that the disease might occur secondary to hypoparathyroidism. The definitive diagnosis of hypoparathyroidism was not established because of the limited ability of the laboratory to measure parathyroid hormone.

The patient was treated with intravenous calcium gluconate replacement therapy. Seven cycles of calcium gluconate replacement were given to the patient until the calcium level returned to normal. In each cycle, 4 grams of calcium gluconate within 100 ml of NaCl 0,9% was given within 1 hour, with 2-hour intervals between each cycle. After the calcium level returned to normal, the symptoms rapidly improved. The patient was discharged from the hospital and was prescribed calcium lactate 500 mg three times a day and vitamin D3 5000 IU twice a day.

DISCUSSION

Fahr's syndrome is a rare neurological condition characterized by symmetrical and bilateral calcification of the basal ganglia associated with neuropsychiatric disorders. Calcification may also be found in other brain regions such as the dentate nucleus, thalamus, and cerebral cortex.[1] Bilateral basal ganglia calcification are incidental finding in up to 15% of patients who undergo a head CT scan.

The presence of bilateral basal ganglia calcification associated with the presence of neuropsychiatric symptoms is referred to as Fahr's syndrome[5]. The neuropsychiatric symptoms that have been documented in various literature included psychiatric features such as dementia, hallucination, delusion, depression, manic symptoms, and anxiety, somatic symptoms such as Parkinsonism, seizures, headache, vertigo, paresis, dysarthria, and tremors.[1] Fahr's syndrome should be differentiated from Fahr's disease, in which basal ganglia calcification is caused by an unknown etiology, while Fahr's syndrome basal ganglia calcification is secondary to a metabolic condition, infection, or genetic[2,4].

In our case, the most possible cause of Fahr's disease is secondary idiopathic hypoparathyroidism, which explains why the disease has been commonly recurring over the year with constant findings of hypocalcemia every time she was hospitalized. The pathophysiology of basal ganglia calcification in Fahr's syndrome secondary to hypoparathyroidism is thought to be caused by imbalances of calcium and phosphorous levels regulated by the parathyroid gland. Parathyroid hormone is responsible for maintaining calcium levels in cerebrospinal fluid and preventing phosphorous deposition in periventricular regions.[6]

Hypoparathyroidism is a rare endocrine disorder in which the parathyroid gland causes parathyroid hormone deficiency. Hypoparathyroidism is diagnosed by the finding of low serum calcium levels and low levels of parathyroid hormone.[7,8] Hypoparathyroidism can be familial with X-linked transmission, characterized by the absence or atrophy of the parathyroid gland, or acquired from a complication of a thyroidectomy. Less commonly, it occurs from an autoimmune disorder or metastasis of the parathyroid gland.[5]

In Fahr's syndrome secondary to hypoparathyroidism, the hypocalcemia caused by hypoparathyroidism would manifest as paresthesia, hyperreflexia, cramps, or tetany. The basal ganglia are a part of the brain that is involved in controlling voluntary movements. Bilateral basal ganglia calcification would manifest as extra-pyramidal syndrome, cognitive impairment, seizures, and psychiatric disorders [9,10]

To this date, there is no specific treatment for Fahr's Syndrome. Current treatment is based on symptoms using antiepileptic and antiparkinsonian drugs and correction of serum calcium level with intravenous calcium gluconate or oral calcium with vitamin D supplementation.[3,11] In our case, intravenous calcium gluconate with vitamin D supplementation brought the calcium serum level to normal, and the symptoms rapidly improved.

CONCLUSION

Fahr's syndrome is characterized by symmetrical and bilateral calcification of the basal ganglia associated with neuropsychiatric symptoms. Fahr's syndrome secondary to hypoparathyroidism is a rare disorder. It presents with neuropsychiatric symptoms such as dementia, hallucination, delusion, depression, manic symptoms, anxiety, Parkinsonism, seizures, headache, vertigo, paresis, dysarthria, or tremors. To this date, there is no specific treatment for Fahr's syndrome. Symptomatic treatment and calcium correction therapy with vitamin D have been proven effective in treating Fahr's syndrome in our case.

REFERENCES

[1] Mufaddel AA, Al-Hassani GA. Familial idiopathic basal ganglia calcification (Fahr's disease). *Neurosciences*. 2014;19(3):171-177.

[2] Gupta SK, Gandotra D, Singh K, Sharma R, Sharma A. Fahr syndrome. *JK Sci*. 2023;9(4):215.

[3] Costa R, Mendes T, Fontes J, Silva J. Fahr ' s Syndrome – A Case Report Síndrome de Fahr : un caso clínico. 2022;(Figure 1):44-45.

[4] Prasetya D, Sumadiono S. Fahr's syndrome in teenage girls with systemic lupus erythematosus: A case report. *J Kedokt dan Kesehat Indones*. 2020;11(3):290-294. doi:10.20885/jkki.vol11.iss3.art11

[5] Kalampokini S, Georgouli D, Dadouli K, et al. Fahr's syndrome due to hypoparathyroidism revisited: A case of Parkinsonism and a review of all published cases. *Clin Neurol Neurosurg*. 2021;202(December 2020):106514. doi:10.1016/j.clineuro.2021.106514

[6] Supit VD, Kurniawan D, Fatimah E. Fahr syndrome and neurological manifestations in hypoparathyroidism patients. *Radiol Case Reports*. 2024;19(4):1248-1253. doi:10.1016/j.radcr.2023.12.034

[7] Bilezikian JP. Hypoparathyroidism. *J Clin Endocrinol Metab*. 2020;105(6):1722-1736. doi:10.1210/clinem/dgaa113

[8] Hans SK, Levine SN. Hypoparathyroidism. [Updated 2024 Feb 24]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-.

[9] Arruda, A. C. G., Guerra, A. C. D. Z., Pessoa, C. H., Marquezine, G. F., & Delfino, V. D. A. (2022). Hypoparathyroidism and Fahr's syndrome: case series. *Jornal brasileiro de nefrologia* , *44*(4), 592–596. https://doi.org/10.1590/2175-8239-JBN-2020-0243

[10] Simonyan K. Recent advances in understanding the role of the basal ganglia. *F1000Research*. 2019;8. doi:10.12688/f1000research.16524.1

[11] Wazir MH, Ali Y, Mufti AZ, Ahmad A, Ahmad H. Fahr's Syndrome: A Rare Case Presentation. *Cureus*. 2023;15(10):8-12. doi:10.7759/cureus.. 47812