

Traumatic Brain Injury Presenting as Paraparesis Revealing Fahr's Disease: A Case Report

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1. Abstract Background

Fahr's disease is a progressive neurological degenerative disease characterized by abnormal calcification of basal ganglia. The prevalence of this disease is < 1/1.000.000 people of the general population with those in the age of 40 - 60 being the most prevalent age group. The diagnosis is usually made incidentally as it has unspecified neurological and psychiatric symptoms. We reported a case of a 64-year-old female with headache and lump on her head following a trauma on her head. Her past medical history includes hypertension. The physical examination revealed a 5 cm-lump on her head, with a motoric strength scale of 3 for both of her lower extremities. Non contrast brain CT scan showed multiple calcifications in basal ganglia. Diagnosis of Fahr's disease was made. She was then treated with neuroprotectants, vitamins, analgesics, and antihypertensive drugs. During the hospitalization, she showed an improvement on her motoric strength scale. In conclusion, Fahr's disease revealed after traumatic brain injury is a rare case. The management is focused on symptomatic relief and treating underlying causes.

2. Introduction

Fahr's disease is a progressive and rare idiopathic neurological degenerative disease which commonly has an autosomal dominant inheritance [1]. The prevalence of this disease is <1/1.000.000 people of the general population with those in the age of 40 - 60 being the most prevalent age group [2,3]. It is characterized by abnormal calcification of basal ganglia which consists of sub-cortical nuclei responsible for motor control, learning, executive

functions, behavior and emotions [4,5]. Therefore, the clinical manifestations encompass neurological, mental, psychiatric, and cognitive impairment. As a result of its wide variety of neurological as well as psychiatric symptoms, the diagnosis is usually made after excluding other causes of these symptoms [1]. Furthermore, it is frequently incidental on computed tomography (CT) imaging in patients with different diagnoses [3,6] This report presents a case of traumatic brain injury presented as paraparesis which later diagnosed as Fahr's disease.

3. Case Report

A 64-year-old female came to the emergency department of Annisa Cikarang hospital with a main complaint of headache and lump on her head after she fell down and hit her head in a bathroom. She also complained that both of her legs felt weak and numb. Three days prior to the accident, she had begun to feel numbness and stiffness in both of her lower extremities. She was unaware of her past numbness or stiffness. Complaints of nausea, vomiting, loss of consciousness, seizure, hemiparesis, mood disturbance, and other neuropsychiatric symptoms were denied. Her past medical history include hypertension, there was no history of diabetes, stroke, autoimmune disease, or endocrine abnormality. None of her family has similar complaints. The patient was fully conscious with good orientation. General examination revealed that her blood pressure was 160/80. She had a lump on her head with a diameter of 5cm. While neurological examination showed a decrease on her motoric movement score with a score of 3 for both of her lower extremities. Other physical and neurological examinations were within

normal range.

Laboratory test showed complete blood tests, random glucose test, electrolytes, kidney and liver function tests, were all within normal range. Serum level of magnesium, phosphorus and parathyroid hormone have not been measured due to economic limitation. Non contrast brain CT scan revealed multiple calcification in basal ganglia, there was no hemorrhage or intracranial fracture (Figure 1).

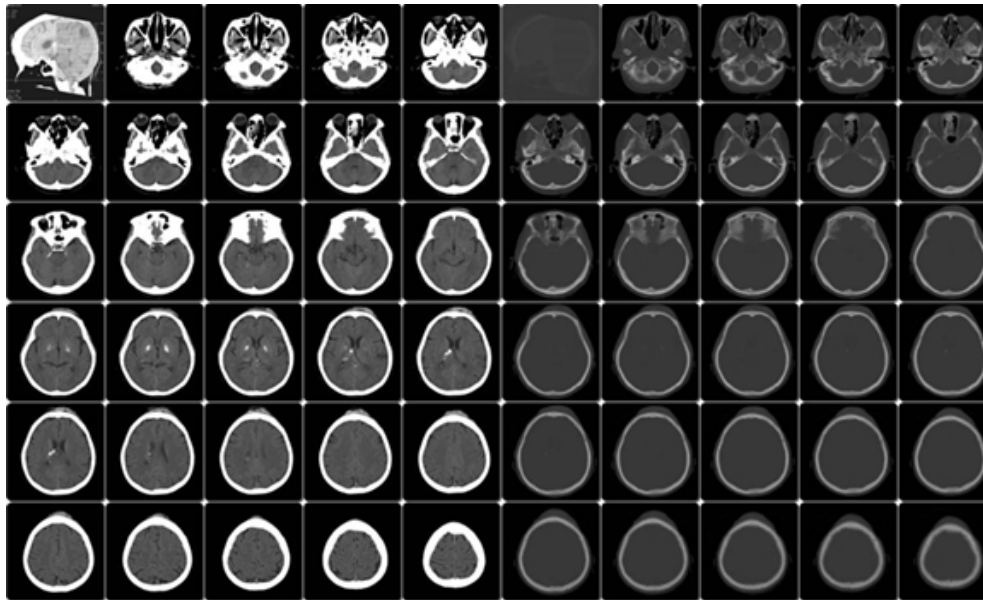


Figure 1: Non-contrast brain CT scan showing multiple calcification in basal ganglia.

4. Discussion

Basal ganglia calcification can be associated with physiological aging or secondary to other causes such as infections, metabolics, and intoxications [7,8]. Fahr's disease is a condition in which calcium deposition in the brain, or most frequently basal ganglia has unknown etiology [7-9]. Although it is frequently defined as idiopathic, several conditions have been noted to have been associated with Fahr's disease, such as endocrine disorders (most commonly parathyroidism abnormalities), and other underlying genetic causes [1,7]. However, in our patient we did not perform diagnostic tests to assess parathyroid hormone abnormalities due to financial limitations, and the patient's unknown family history nor other genetic causes.

The specific pathophysiology of Fahr's disease is still unknown. Researches showed that some patients have a locally altered Blood-brain barrier which produces metastatic deposition of calcium with or without an abnormal calcium metabolism. Progressive calcification compresses nearby vessels reducing blood flow and hence continuing the vicious cycle of decreased blood flow, tissue injury, and mineral deposition [1,7,9]. In our patient, we did observe mild hypocalcemia and possible local blood brain barrier disturbances due to the patient's history of head trauma which might contribute to the pathophysiology of this case. Fahr's disease has a wide variety of symptoms, ranging from neurological to psychiatric symptoms as the most common manifestations. Thus,

During the hospitalization, the patient was given neuroprotectants, vitamins, analgesic, and antihypertensive drugs. In the follow up process, thoracolumbar x-ray was performed to exclude another cause of paraparesis and it showed normal result. During 5 days of hospitalization, the patient showed an improvement on her motoric strength scale (score of 4 on both of her legs), and then she was discharged from the hospital.

it frequently is an incidental finding in 15-20% of patients undergoing a CT scan [1,7,9]. Our patient presented with mild traumatic brain injury and paraparesis which later revealed Fahr's disease as an incidental finding on her CT scan.

Careful history-taking, physical examination, and further diagnostic workup should be performed to exclude other potential diagnosis. The most frequently used diagnostic criteria for Fahr's disease is adapted from modified Moskowitz et al, Ellie et al, and Mayam et al studies, as follows: Bilateral calcification of the basal ganglia visualized on neuroimaging or other brain regions, progressive neurologic dysfunction, which generally includes a movement disorder and/or neuropsychiatric manifestations, age of onset is typically in the fourth or fifth decade, although this dysfunction may also present in childhood, absence of biochemical abnormalities and somatic features suggestive of a mitochondrial or metabolic disease or other systemic disorder, absence of an infectious, toxic, or traumatic cause, and family history consistent with autosomal dominant inheritance [10]. Nicolas et al. in Melissa et al developed a rating scale of the brain calcifications which ranged from 0 (no calcification) to 5 (severe and confluent) for numerous locations in the brain (lenticular, caudate, thalamus nuclei, subcortical white matter, cortex, cerebellar hemispheres, vermis, pons, and medulla). The total calcification score is 80 [11]. In our patient, the findings include bilateral calcification of the basal ganglia on brain CT scan and progressive neurologic dysfunction which is paraparesis

in both of her legs. There is no specific evidence of biochemical abnormalities or systemic disorder, infectious, toxic, and family history. Despite her history of head trauma, the CT scan showed no other abnormalities except for calcification which is likely the cause of our patient's condition. CT imaging can be used to detect calcium depositions, as calcium has a very high density on CT [11].

To our knowledge, the literature reporting Fahr's disease revealed by traumatic brain injury is rare. A similar case was reported by Yakhya Cisse, et al, they presented a case of Fahr's disease revealed in a 14-year-old boy with history of cranioencephalic trauma, then the diagnosis was made after CT scan showed diffuse calcification at periventricular, lenticular nuclei, the head of the caudate nuclei, and both thalamuses [12]. Another study by Rahul Singh, et al, they found of 7344 brain CT scans done for head injury patients in 2 years, 13 (0.18%) patients were found to have bilateral symmetrical striopallidodentate calcification [13].

There is no specific treatment for Fahr's Disease. Therefore, the management is focused on eliminating underlying cause and treating symptomatic relief, such as antiepileptic for seizure, pain killers, physical rehabilitations, and neuroleptic for movement disorders [1,7]. Currently, no effective treatment can limit nor slow down calcification in the basal ganglia [14]. However, a study mentioned a first potential treatment for patients with Fahr disease is bisphosphonate therapy. Newer nitrogen-containing bisphosphonates, such as alendronate, predominantly inhibit osteoclasts [11]. A second candidate therapy could be vitamin D. Keasey et al, suggested that vitamin D reduced calcifications in the brain cause vitamin D deficiency was found in patients with Fahr disease [15]. Overall, the prognosis of fahr's disease is variable and unpredictable, as it is determined by the underlying causes. However, there is a study report from which fahr's disease presenting as head injury were having good recovery [1,13].

5. Conclusion

Fahr's disease revealed after traumatic brain injury is a rare case. Fahr's disease has a wide variety of neurological and psychiatric symptoms. Thus, it frequently is an incidental finding. Complete examination is necessary to raise the diagnosis. There is no specific treatment, the management is focused on symptomatic relief and treating underlying causes.

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