JURNAL KEDOKTERAN DIPONEGORO
(*DIPONEGORO MEDICAL JOURNAL*)
Online http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : http:// 10.14710/jkd (dmj).v12i2.37711
JKD (DMJ), Volume 12, Number 2, March 2023 : 88-91

CASE REPORT : FAHR'S DISEASE WITH ACUTE DOUBLE HEMIPARESE MANIFESTATION

Vania Ayu Puspamaniar[1]*, Giam, Billy Putera Kurniawan[1], Bonfilio Neltio Ariobimo[1], Lisa Puspitorini[2]
[1]General Practitioner, Ibnu Sina General Hospital, Kabupaten Gresik, Indonesia
[2]Department of Neurology, Ibnu Sina General Hospital, Kabupaten Gresik, Indonesia
* Corresponding Author : E-mail: puspamaniar06@gmail.com

ABSTRACT

Fahr's disease is a very rare disease with a prevalence of 1/1,000,000 individuals. We present a case report of a male with sudden double hemiparese and basal ganglia calcification leading to the diagnosis of Fahr's disease. A 59 years old male presented to the emergency department with a chief complaint of unable to move his upper and lower extremities in a sudden. The complaint begins with weakness of the right side of the body, followed by weakness of the left side of the body the day after. The serum levels of calcium, magnesium, phosphorus, and Parathyroid Hormonal (PTH) have not been measured due to the patient's financial problems. CT scan of the brain showed bilateral, symmetrical, wide areas of calcification over the fossa posterior, basal ganglia, periventricular, and parietal area, which were suggested as Fahr's disease. The patient is being treated with the injection of neuroprotectant, antibiotic, vitamin, neuropathic analgesia, and fluids. He is also being consulted with medical rehabilitation to get some physical treatments. Treatment goals include: increase and or maintain ROM, prevent contractures, strengthens weak muscles that may be underutilized, improvement and maintenance of postural stability in static postures and during mobility, and fall prevention. Our case highlight sudden double hemiparese which different from previous literature which says that neurological deficit symptoms appear with a chronic nature and the importance of combining pharmacological therapy and physical therapy as in this patient to reduce the patient's morbidity.

Keywords: *Bilateral basal ganglia calcification, Double hemiparese, Fahr's disease, Neurodegenerative disease*

INTRODUCTION

Fahr's disease is a condition that causes basal ganglia to calcify. Theodor Fahr, a neurologist, made the initial discovery of this disease in 1930. The prevalence of this condition is 1/1,000,000 people in general population.[1] The most prevalent age group is between 40 and 60, and man having a higher incidence than woman.[2,3] Although several hypotheses suggest that it can be inherited genetically, mostly through autosomal dominant or autosomal recessive inheritance, the precise reason is still unclear, and it can also happen sporadically.[1] Platelet-Derived Growth Factor Receptor Beta (PDGFRB) and Solute Carrier Family 20 Member 2 (SLC20A2) are two genes that are suspected to be mutated and are connected to the occurrence of Fahr's diseases.[4,5]

The basal ganglia, thalamus, dentate nucleus, cerebral cortex, cerebellum, subcortical white matter, and hippocampus are among the brain regions that control movement, as are the cerebellum and subcortical white matter. Abnormal calcium deposition causes calcification in both hemispheres of the brain.[1,6] The third decade of life is when calcifications often start, however they have occasionally been discovered as early as childhood.[7] Extrapyramidal syndrome is the most typical sign of Fahr's disease, however it can also include cerebellar abnormalities, speech problems, and neuropsychiatric symptoms.[1]

We present a case report of a male with sudden double hemiparese which different from previous literature which says that neurological deficit symptoms appear with a chronic nature and basal ganglia calcification leading to the diagnosis of Fahr's disease.

CASE REPORT

Male 59 years old, came to the emergency room with the main complaint being unable to move the upper and lower extremities suddenly. The complaint begins with weakness on the right side of the body, followed by weakness on the left side the next day. Complaint of nausea, vomiting, headache, fever, cough, trauma, ear or nose infection, was denied. There was no history of neuropsychiatric manifestations, mood disturbances, or seizures. There are no history of hypertension, diabetes mellitus, autoimmune disease, or trauma. The patient had a history of right hemiparese and

JURNAL KEDOKTERAN DIPONEGORO
(*DIPONEGORO MEDICAL JOURNAL*)
Online http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : http:// 10.14710/jkd (dmj).v12i2.37711
JKD (DMJ), Volume 12, Number 2, March 2023 : 88-91

diagnosed as ischaemic stroke in 2008 and have experienced resolution without any sequele. None of his family had similar complaints. The patient is a non-smoker. The patient is fully conscious, has normal orientation to time and place, and has normal memory and intelligence. Vital sign was in normal limit, blood pressure 115/70 mmHg, heart rate 70x/ minutes regular, respiratory rate 20x/ minutes, temperature 36.2°C. The patient's ability to speak seems slow. There is no stiffness, tremor, or abnormal movement. Primitive reflexes are absent. The motor movement scores were 2 for the right upper limb, 3 for the left upper limb, and 0 for both lower extremities. Other systemic examinations are normal. Chest xray and electrocardiography examination were in normal limit. Laboratory tests showed complete blood tests, electrolytes, kidney and liver function tests with normal results. Serum levels of calcium, magnesium, phosphorus and parathyroid hormone (PTH) have not been measured because of the patient's economic limitations. The presenting symptom of double hemipareses mimic stroke features as a differential diagnosis and prompting the need for CT scan.

CT scan of brain showed large bilateral and symmetrical areas of calcification over the posterior fossa, basal ganglia, periventricular, and parietal areas, suggestive of Fahr's disease (Figure 1). Appearance of calcification in the patient's imaging is a hallmark of Fahr Disease thus stroke can be eliminated. Patients were given therapy in the form of injections of neuroprotectants, antibiotics, vitamins, neuropathic analgesia, and fluids. Patients are also consulted with medical rehabilitation to get physical therapy. Patient performs Bobath exercises and positions to improve motor function. Routine clinical monitoring and neurocognitive assessment must still be carried out to monitor the condition of Fahr's disease.

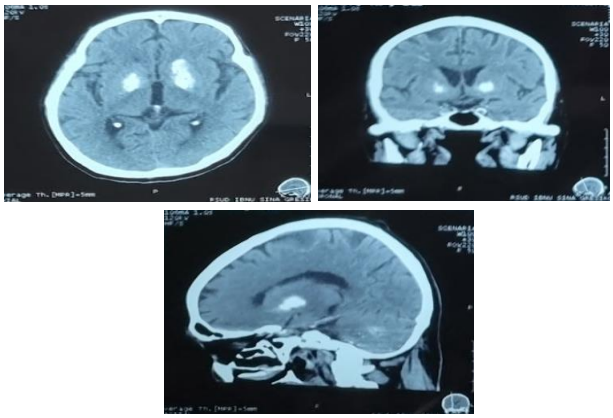

Figure 1. Brain CT Scan Examination

DISCUSSION

Idiopathic bilateral, symmetrical, and aberrant calcium deposits in the basal ganglia and other regions of the brain are the hallmarks of Fahr's disease, a neurodegenerative condition.[1,6,8,9] It is an uncommon neurological condition that is inherited autosomally dominantly, however in some recorded cases it has been diagnosed without a mutation in the causal gene or first-degree hereditary inheritance.[9]

The condition was originally recognized in 1930 by German neurologist Karl Theodor Fahr.[8] With an incidence of less than 1 in 100,000, the condition is categorized as a rare hereditary or sporadic neurological ailment.[1] Although the actual number of Fahr's disease instances is unknown, less than 200 cases have been documented. Fahr's disease affects males 2:1 more frequently than women.[10]

The criteria can be used to establish the diagnosis are:[4]

1. Bilateral calcifications are found in the basal ganglia of the brain or in other brain areas on brain imaging.
2. Progressive neurological deterioration with symptoms of motor decline and/or neuropsychiatric changes.
3. The disorder appears in the fourth or fifth decade of life but can last from childhood.
4. There are no biochemical abnormalities and somatic characteristics that suggest mitochondrial or metabolic disorders or other systemic conditions.

JURNAL KEDOKTERAN DIPONEGORO
(*DIPONEGORO MEDICAL JOURNAL*)
Online http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : http:// 10.14710/jkd (dmj).v12i2.37711
JKD (DMJ), Volume 12, Number 2, March 2023 : 88-91

5. No etiology of infection, poison, or trauma was found.
6. Autosomal dominant inheritance is supported by a family history of the disease.
7. In the absence of bilateral calcifications (criterion one) or progressive deterioration of neurologic and neuropsychiatric symptoms, the diagnosis is confirmed by a positive family history (criterion two).

It is still unclear which gene is the major cause. Calcium, its primary constituent, is stored in the basal ganglia and is evident on radiological scans.[10] Fahr's disease is most frequently linked to endocrine diseases, particularly parathyroid abnormalities. Hypocalcemia and hyperphosphatemia, which can speed up the calcification process, can be brought on by decreased parathormone (which occurs in hypoparathyroidism) or a renal response to parathormone (in pseudohypoparathyroidism).[1] Fahr's disease differs from atherosclerosis in that mineral deposition frequently occurs only in certain capillaries and tiny arteries in the white matter regions of the brain.[8] The endothelium, stromal vascular cells, and the interstitium are all involved in the calcification process. The process of depositing calcium and other minerals has been thought to be greatly accelerated by local circulatory abnormalities, such as ischemia. Anomalies in the metabolism of calcium or local inflammation are other recognized causes.[11] In substantial family types, whether autosomal recessive or dominant, calcifications can develop without a preceding circulatory impairment.[8]

Fahr's disease has not yet been successfully treated with a specific medicine since the calcification process is ongoing and might worsen at any time.[7] Depending on their clinical status, each person receives a distinct course of treatment to reduce symptoms. Haloperidol or lithium carbonate may be used to treat individuals with psychotic symptoms, according to many case studies.[1,7] According to earlier research, bone medications such disodium etidronate are helpful for symptom relief but not for shrinking the amount of the calcification. The medication has a strong affinity for calcification, shielding neurons from toxic calcium levels that cause excessive excitability and death.[12] Two-thirds of idiopathic basal ganglia calcification patients with parkinsonism have been observed to have a late-onset levodopa response.[13]

The two most typical infections following a stroke are pneumonia and urinary tract infections, and using preventive antibiotics may help to lower their frequency. Antibiotics, however, show no long-term advantages in terms of neurological outcomes, mortality, or morbidity. It is believed that ceftriaxone has neuroprotective properties.[14,15] Our patient has manifestation with double hemiparese that prone to UTI and pneumonia as in acute stroke, so we gave antibiotic as a prevention from infection and supportive neuroprotective.

Clinical judgment is required together with information on the treatment of overlapping symptoms in the same population due to the dearth of research on Fahr's disease and the resemblance of symptoms to Parkinson's, Huntington's, and other neurological disorders. As part of the recommended course of action, secondary problems including muscular atrophy should be avoided or delayed while concentrating on function, participation, and exercise capacity.[16,17]

Treatment objectives include: improving and maintaining postural stability in static postures and during mobility, increasing and/or maintaining range of motion (ROM), preventing contractures, strengthening weak muscles that may be underused, gait retraining and/or fall prevention, and symptom management.[1,16]

CONCLUSION

Fahr's disease is a very rare condition of bilateral abnormal calcifications in the brain, most commonly located in the basal ganglia which has a variety of clinical presentations. The clinical findings vary from no symptoms to severe neuropsychiatric signs and symptoms. Our case report highlights sudden double hemiparese which different from previous literature which says that neurological deficit symptoms appear with a chronic nature and the importance of combining pharmacological therapy and physical therapy as in this patient to reduce the patient's morbidity.

CONFLICT OF INTEREST

The authors stated there are no conflict of interest.

JURNAL KEDOKTERAN DIPONEGORO
(*DIPONEGORO MEDICAL JOURNAL*)
Online http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : http:// 10.14710/jkd (dmj).v12i2.37711
JKD (DMJ), Volume 12, Number 2, March 2023 : 88-91

FUNDING

There is no specific funding was provided.

REFERENCES

1. Saleem S, Aslam HM, Anwar M, Anwar S, Saleem M, Saleem A, et al. Fahr's syndrome: Literature review of current evidence. Orphanet J Rare Dis [Internet]. 2013;8(1):1. Available from: Orphanet Journal of Rare Diseases
2. Asokan AG, D'souzA S, Jeganathan J, Pai S. Fahr's syndrome- An interesting case presentation. J Clin Diagnostic Res. 2013;7(3):532–3.
3. Gligorievski A. CT Diagnosis of Fahr disease, a case report. Clin Case Reports Rev. 2018;4(2):1–3.
4. Samuels CR, Khanni JL, Espinosa PS, Barkley K, Azhar M. Case Report: An Incidental Finding of Fahr's Disease in a Patient with Hypochondria. Cureus. 2018;10(12):1–6.
5. Donzuso G, Mostile G, Nicoletti A, Zappia M. Basal ganglia calcifications (Fahr's syndrome): related conditions and clinical features. Neurol Sci. 2019;40(11):2251–63.
6. Durante A, Audino N, Cristiano M, Tanga M, Martino MT, Noschese I, et al. Basal ganglia calcification: a Fahr's disease case report. Radiol Case Reports [Internet]. 2021;16(10):3055–9. Available from: https://doi.org/10.1016/j.radcr.2021.07.042
7. Sinha R, Sodhi K, John BM, Singh D. Fahr's disease: A case report. J Nepal Paediatr Soc. 2010;30(1):44–5.
8. Ma A, Sm K, Salma U. Fahr's Syndrome: A rare Neurodegenerative Disorder. 2017;29(01):39–41.
9. Ayoub B, Chakib C, Khalil M, Hicham B. Article Case report Fahr ' s disease presenting with epileptic seizures : a case report. 2020;3(55):1–5.
10. Manyam B V., Walters AS, Narla KR. Bilateral Striopallidodentate calcinosis: Clinical characteristics of patients seen in a registry. Mov Disord. 2001;16(2):258–64.
11. Şenoğlu M, Tuncel D, Orhan FÖ, Yuksel Z, Gokçe M. Fahr 's Syndrome : A Report of Two Cases. Fırat Tıp Derg. 2007;12(1):70–2.
12. Tai XY, Batla A. Fahr ' s disease : current perspectives. 2015;5:43–9.
13. Kumar N, Jog M. Fahr's Disease Presenting as Late-Onset Levodopa-Responsive Parkinsonism. Can J Neurol Sci. 2017;44(3):322–3.
14. Xi YG, Tian X, Chen WQ, Zhang S, Zhang S, Ren WD, et al. Antibiotic prophylaxis for infections in patients with acute stroke: A systematic review and meta-analysis of randomized controlled trials. Oncotarget. 2017;8(46):81075–87.
15. Rashid MH, Kabir A, Waris MU, Salman U, Zain S. Role of Prophylactic Antibiotics in Critical Care of Stroke Patients - A Preventive Approach to Post-stroke Infections? Cureus. 2020;12(3).
16. O'Sullivan S, Schmitz T, Fulk G. Physical Rehabilitation. 6th ed. Philadelphia: F.A. Davis Company; 2014.
17. Sarkar DN, Sarkar D, Zahin A, Ohab M, Miah MS, Haque MM. Fahr's Syndrom e: A rare case- Presented as Acute Ischaemic Stroke. Med Today. 2017;29(1):45–6.