Fahr’s Syndrome: A Rare Case Report

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ABSTRACT
Fahr’s syndrome is a rare neurodegenerative disorder characterized by bilateral basal ganglia calcification. The exact incidence of Fahr’s syndrome is still unknown but estimated to be <1/1,000,000. On routine non-contrast-CT scan examination, 0.3-1.2% cases of intracranial calcification can be incidentally detected. It is caused by several medical conditions, with hypothyroidism as the most common one. Patient may come with neurologic or psychiatric symptoms. CT scan is the main supportive examination; it is used to determine the location and extension of calcification. Therapy is directed to the underlying etiology and usually symptomatic. A female, 46 years old, came with stiffness on both hands. This symptom appeared multiple times during past decade but with mild intensity. CT scan revealed calcification in both cerebral hemisphere and cerebellum. Blood test showed low calcium concentration. Patient was diagnosed with Fahr’s syndrome and given anticonvulsant and calcium supplementation. She was getting better during hospitalization and then discharged 4 days later with oral home medication.

Keywords: Fahr’s syndrome, brain calcification

INTRODUCTION
Calcification of basal ganglia can be idiopathic or secondary due to other medical condition. Fahr’s disease is defined as idiopathic bilateral basal ganglia calcification while Fahr’s syndrome is referred to the secondary one. This disorder is found by Fahr, a German neurologist, which found an 81-year-old patient with dementia, fever, and immobility. Brain autopsy was performed and revealed some calcifications in striate.

The exact incidence of Fahr’s syndrome is still unknown but estimated to be <1/1,000,000. On routine non-contrast CT-scan examination, 0.3-1.2% cases of intracranial calcification can be incidentally detected. In term of age, Fahr’s syndrome is usually found in fourth or fifth decade.

Therapy of Fahr’s syndrome is usually symptomatic and adjusted based on the etiology. The prognosis is varying, unpredictable, and depends on the extension of calcification.

CASE REPORT
A 46 years old female was admitted to emergency room of Mangusada Hospital with chief complaint of stiffness on both hands since yesterday. Stiffness was felt from wrist to fingers and made her unable to move them. She also felt some tingling sensation on same location. No stiffness was felt on other part of her body. Other complaints such as headache, fever, disorganized speech, or history of unconsciousness was not found. She had been experiencing this symptom since the age of 6. Stiffness was occurred multiple times, within short duration, with unpredictable triggering factor. She never visited a neurologist due to this symptom. No history of neurologic or other chronic disease was found.

Physical examination was performed; she was alert, showed elevated blood pressure (160/90 mmHg), while her heart rate, respiratory rate, and axillary temperature were within normal value. No abnormality was found on her eyes, ENT, neck, chest, and abdomen. Neurological examinations revealed some positive finding on both of her upper extremity especially wrist and fingers, such as increased tonus, decreased power (2255/5522), and spasm. Other neurological test showed normal finding.

Patient was checked for some routine laboratory examinations, such as complete blood count, serum blood glucose, liver function test, urea, creatinine, albumin, and electrolytes including calcium. The results showed increased white blood cell count with neutrophil dominance, low calcium and potassium. Non-contrast CT-Scan was also performed and showed significant finding: hyperdense lesions with density of 89-127 HU in bilateral temporal lobes, basal ganglia, lateral paraventricles, occipital, parietal, and vermis. The
Conclusion was calcification in right and left cerebral hemispheres and cerebellum (Figure 1).

Patient was then diagnosed with Fahr’s syndrome and given therapy of IVFD with NaCl 0.9% 20 drips per minute, oral divalproex sodium (Depakote®) 2x125 mg, oral clobazam 2x25 mg, and oral calcium supplement (Osteocal®) 2x500 mg. Patient showed improvement during hospitalization and discharged 4 days later. She was instructed to continue the course of those three oral medications until the next follow up at neurologic polyclinic.

DISCUSSION

Fahr’s syndrome is a rare neurodegenerative disorder characterized by bilateral brain calcification.1 The deposits are composed of carbonate calcium and phosphate calcium. They usually found in basal ganglia, thalamus, hippocampus, cerebral cortex, white matter, cerebellar cortex, and dentate nuclei.6

The clinical manifestation mostly includes neurologic symptoms, such as rigidity, gait disorder, Parkinsonism, speech disorder, chorea, dystonia, tremor, and myoclonus.7 A progressive neurological deterioration may cause disabilities and mortalities. The other manifestation is psychiatric symptoms which found in 40% of cases. The psychiatric symptoms may vary from only difficulties to concentrate, personality or behavior disorder, psychosis, and dementia.8 Patient in this case was showing rigidity as well but no psychiatric symptom was noticed.

There are several medical conditions associated with Fahr’s syndrome. The most common one is endocrine disorder, which is hypoparathyroidism. Zisimipoulou et al have found Fahr’s syndrome patients with idiopathic and secondary hypoparathyroidism with percentage of 23.3% and 15.3% respectively.9 The other medical conditions which also related are vitamin D disorder, Type I Kenny Caffey syndrome, mitochondria myopathy, lipid proteinosis, etc.5,10 However, serum parathyroid hormone concentration was not able to be acquired in this case due to limited resources in the hospital.

The most important examination to diagnose Fahr’s syndrome is brain CT-scan. This radiologic examination will reveal the location and extension of calcification. The calcification is bilateral and mostly found in lenticular nuclei, especially internal Globus Pallidus.5 Other laboratory examinations may be performed as well, such as serum calcium, phosphate, magnesium, ALP, calcitonin, and PTH. These examinations are usually directed to rule out the underlying etiology of this syndrome.

Management of Fahr’s syndrome is directed to the etiology and the appearing symptoms. Seizure and movement disorder related to PTH disorder can be treated with anticonvulsant and correction of serum calcium and phosphate concentration through supplementation of calcium and alpha hydroxyl vitamin D3.11 Antipsychotic can be given for patient with psychiatric symptoms. Our patient was receiving anticonvulsants and calcium supplementation due to rigidity and low serum calcium concentration.

SUMMARY

Fahr’s syndrome is a rare neurological disorder. The symptoms are unspecific, stay for long period and progressive over time. Patient with some of chronic relapsing neurologic and psychiatric symptoms should be checked for brain CT scan if possible to rule out this syndrome. The early detection of the underlying etiology will not only give better outcome but also avoid unnecessary intervention to the patient. Although there was some limitation in patient management including inability to check serum PTH concentration, patient was successfully discharged with good clinical status.

Figure 1. The Non-contrast CT-Scan
REFERENCES

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