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Case Report

Fahr's Syndrome Associated with Multiple Intracranial Aneurysms: A Case Report

Umit EROGLU¹, Gökmen KAHİLOGULLARI¹, Altan DEMİREL¹, Anil ARAT², Agahan UNLU¹

¹Ankara University, School of Medicine, Department of Neurosurgery, Ankara, Turkey

²Ankara University, School of Medicine, Department of Radiology, Ankara, Turkey

ABSTRACT

Fahr's Syndrome is characterized by the presence of intracerebral, bilateral and symmetrical calcifications located in bilateral basal ganglia, thalamus, and cerebellum. The etiology is not exactly known. The authors reported a very rare case who had Fahr's Syndrome and intracerebral aneurysms simultaneously. The patient was female and presented with headache. Her examinations revealed aneurysms on the middle cerebral artery, internal carotid artery and ophthalmic artery. That is the first case reported in the literature having multiple intracranial aneurysms and Fahr's Syndrome together.

KEYWORDS: Fahr's syndrome, Aneurysm, Calcification

■ INTRODUCTION

Fahr's Syndrome was first described in 1930 by German neurologist Karl Theodor Fahr in an adult patient with progressive neurological symptoms (4). This syndrome is characterized by bilateral non-arteriosclerotic cerebral calcifications that are located in the central grey nuclei (1). About 40% of the patients with Fahr's Syndrome are seen with primarily cognitive and other psychiatric findings (1). Clinically it may present with an array of movement disorders, cognitive and cerebellar disorders, dementia and other behavioural disturbances (4). Sporadic and familial cases have been reported with or without calcium/phosphorus metabolism disorders (3). The etiology often not known for certain. Fahr's syndrome often occurs due to disorders of the calcium and phosphorus metabolism. The disease may also develop as a result of genetic damage (2).

■ CASE REPORT

A forty-two year-old female presented with complaints of headache, nausea and depressive complaints. We learned from her history that she had undergone a thyroidectomy operation. She had been using antidepressants. The patient

had normal calcium levels on blood tests (Ca: 9.4 mg/dL). The neurologic examination of the patient was intact. According to the present complaints, cerebral computed tomography (CT) was obtained first (Figure 1). There was calcification of bilateral basal ganglia on the cerebral CT. Cranial magnetic resonance imaging (MRI) studies, digital subtraction angiography (DSA) and CT angiography examinations showed aneurysms in the middle cerebral artery (MCA), internal carotid artery (ICA), ophthalmic artery and calcification of bilateral basal ganglia (Figures 2, 3). She underwent left pterional craniotomy, and clipping of the MCA aneurysm. ICA and ophthalmic artery aneurysms by an endovascular team at different times. The patient has been followed for 11 months with no neurological deficits.

■ DISCUSSION

Fahr's Syndrome is characterized by the presence of intracerebral, bilateral and symmetrical calcifications located in bilateral basal ganglia, thalamus, and cerebellum (1,5). Although there is a lot of research on the etiology of Fahr's syndrome, the exact etiology is still not fully elucidated.



Corresponding author: Ümit EROGLU

E-mail: umitkovikeroğlu@hotmail.com



Figure 1: Multiple calcifications are observed in bilateral basal ganglia on axial CT section.

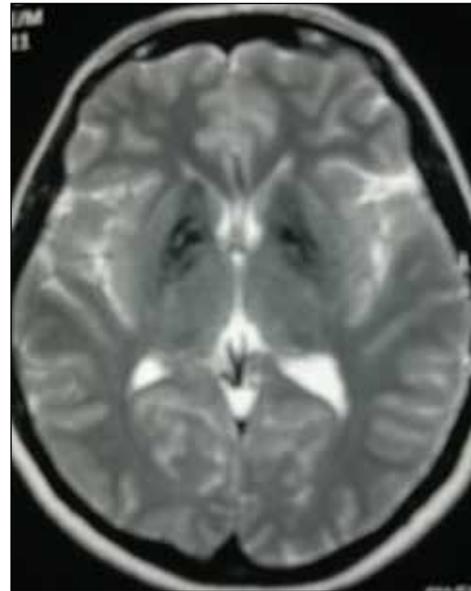


Figure 2: Gradient-echo T2 sequence MRI. This MR sequence is usually included when intracranial calcifications are suspected.



Figure 3: 3D-CT angiography shows the left MCA aneurysm (green arrow) and bilateral calcifications (yellow arrows).

The clinical features are important because basal ganglia calcification may be viewed as an incidental finding. Globus pallidus is the most common area for calcifications in Fahr's Syndrome. Vertigo, paresis, cognitive impairments, headache, movement disorders, cognitive impairment and seizures are the most common manifestations of Fahr's syndrome (5). A patient who presents with some of these symptoms must be evaluated for Fahr's Syndrome with aneurysm. Our patient was diagnosed with multiple aneurysms and Fahr's Syndrome after headache with cerebral CT and CT angiography.

This association may be related with many other disorders such as inflammatory disorders (cytomegalovirus infection, neurocysticercosis, toxoplasmosis, neurobrucellosis, tuberculosis), tumors (astrocytomas), hypoxic and vascular disorders (infarct, ischemic encephalopathy), endocrine problems

(hypoparathyroidism, pseudohypoparathyroidism, hyperparathyroidism), and metabolic and degenerative diseases (senility, mitochondrial encephalopathies, leukodystrophic diseases, motor neuron disease, muscular dystrophy, carbonic anhydrase deficit) (6,7,11,12). Our patient had the presented clinical features, but no hypoparathyroidism or pseudohypoparathyroidism even though she had undergone thyroidectomy. Familial cases of Fahr's syndrome have rarely been reported. Most of the cases concern an autosomal dominant genetic transfer but an autosomal recessive disorder has also been reported (8). A genetic defect of the short arm of 14 chromosome is thought to be responsible for Fahr's Syndrome (10). Firstly the outer layer of the vessels store calcium, and then there is spread to the intimal area and obliteration of the cranial micro-vascular vessels (9).

There are different pathologies with other aneurysms and Fahr's Syndrome. There is no knowledge that elucidates the mechanism of how an aneurysm and Fahr's Syndrome occur together.

■ CONCLUSION

This report is the first case in which an aneurysm and Fahr's Syndrome occurred together. Fahr's Syndrome should be kept in the mind in patients with headache, vertigo, movement disorders, paresis, stroke-like events, cognitive impairment, psychiatric disorders, pyramidal signals and seizures associated with aneurysms.

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