

Fahr Syndrome seen with aneurysms : A case report

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Introduction

First, in 1930, an adult with progressive neurological symptoms patients described by Fahr(4). Fahr's Syndrome is characterized by the presence of intracerebral, bilateral and symmetrical non-arteriosclerotic calcifications, 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.(2) The etiology often not known for certain. Fahr's syndrome is often occur due to disorders of calcium and phosphorus metabolism. Also described the disease may develop as a result of genetic damage (3).

Case:

Forty two years old female presented with complaint of headache, nausea and depressive complaints. We learned from her history she had thyroidectomy operation, she had been using antidepressant. The patient had normal calcium levels in blood tests. (Ca: 9,4mg/dL) Neurologic examination of the patient was intact. According to the standing complaints first she has taken cerebral CT . There was the calsification of bilateral basal ganglia at the cerebral CT. Kranial MRI studies, DSA and CT angiography examinations she had aneursyms on MCA , ICA , Ophtalmic artery and calsification of bilateral basal ganglia . She underwent operation with left pterional craniotomy , clipping of the MCA aneurysm. ICA and ophtalmic artery aneurysms was treated by endovascular team in different times. The patient has been following 11 months with no neurological deficits.

Discussion

Fahr's Syndrome is characterized by the presence of intracerebral, bilateral and symmetrical calcifications, located bilateral basal ganglia, thalamus, and cerebellum (1,5). Although there are a lot of research on the etiology of Fahr's syndrome is still not fully elucidated. Clinical features are important because basal ganglia calcification may be viewed as an incidental finding. Globus pallidus is a most common area for calcifications in Fahr Syndrome.

Headache, vertigo, movement disorders, paresis, stroke like events, cognitive impairment, psychiatric disorders, pyramidal signs and seizures are the most common manifestations(5). A patient presented with some of these symptoms must be evaluated for a Fahr Syndrome with an aneurysm. This patient has diagnosed aneurysms and Fahr Syndrome after headache, after that taking cerebral CT and CT angiography. The diagnosis of the patient must be distinguished to etiologies and clinical features. This kind of abnormality is related with many etiologies that can be classified as inflammatory (CMV infection, neurocysticercosis, toxoplasmosis, neurobrucellosis, tuberculosis, HIV infection), tumoral (astrocytomas), hypoxic and vascular (arteriovenous malformations calcified infarct, ischemic encephalopathy), endocrine (hypoparathyroidism, pseudohypoparathyroidism, hyperparathyroidism), toxic (CO and Pb intoxication, hypervitaminosis D, radiotherapy), metabolic and degenerative (senility, mitochondrial encephalopathies, leukodystrophic diseases, idiopathic familial, motor neuron disease, myotonic muscular dystrophy, carbonic anhydrase deficit, biotin deficiency) and others. (malabsorption, Down Syndrome, lupus, tuberous sclerosis, arthrochondrodysplasia)(6,7). This patient had any of these clinical features, but no hypoparathyroidism, pseudohypoparathyroidism even though she had thyroidectomy.

Fahr's syndrome familial cases have been reported rarely. Most of the cases where an autosomal dominant genetic transfer, in some cases, developed in the autosomal recessive disorder has been reported (8). The genetic defect of the short arm of 14 chromosome is estimated to be developing Fahr Syndrome (10).

Firstly outer layer of the vessels become storage of calcium, and then spreading the intimal area and end of that obliteration of the cranial micro-vascular vessels (9). Calcification is not like Intracranial aneurysms that the formation of intracranial aneurysms starts with endothelial injury at the apical intimal pad (Stage I); this leads to the formation of

an inflammatory zone (Stage II), followed by a partial tear or defect in the inflammatory zone. Expansion of this defect forms the nidus of the intracranial aneurysm (Stage III) (11). Intracranial aneurysms are pouch-like pathological dilatations of intracranial arteries that develop when the cerebral artery wall becomes too weak to resist hemodynamic pressure and distends. Some intracranial aneurysms remain stable over time, but in others mural cells die, the matrix degenerates (12).

There are different pathologies with each other aneurysms and Fahr Syndrome. There are no any knowledge that elucidate the mechanism of an aneurysm and Fahr Syndrome occurs together.

Conclusion :

Fahr's Syndrome is characterized by the presence of intracerebral, bilateral and symmetrical non-arteriosclerotic calcifications, located in the central grey nuclei. Approximately About 40 % of the patients with Fahr's Syndrome are seen with primarily cognitive and other psychiatric findings. This report is the first case an aneurysm and Fahr Syndrome occurs together until now. The patient presented with headache, vertigo, movement disorders, paresis, stroke like events, cognitive impairment, psychiatric disorders, pyramidal signals and seizures with aneurysms; Fahr Syndrome must be kept in the mind.

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Figure captions:

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: Three-dimensional computed tomography angiography shows the left MCA aneurysm (green arrow) and the bilateral calcifications (yellow arrows)

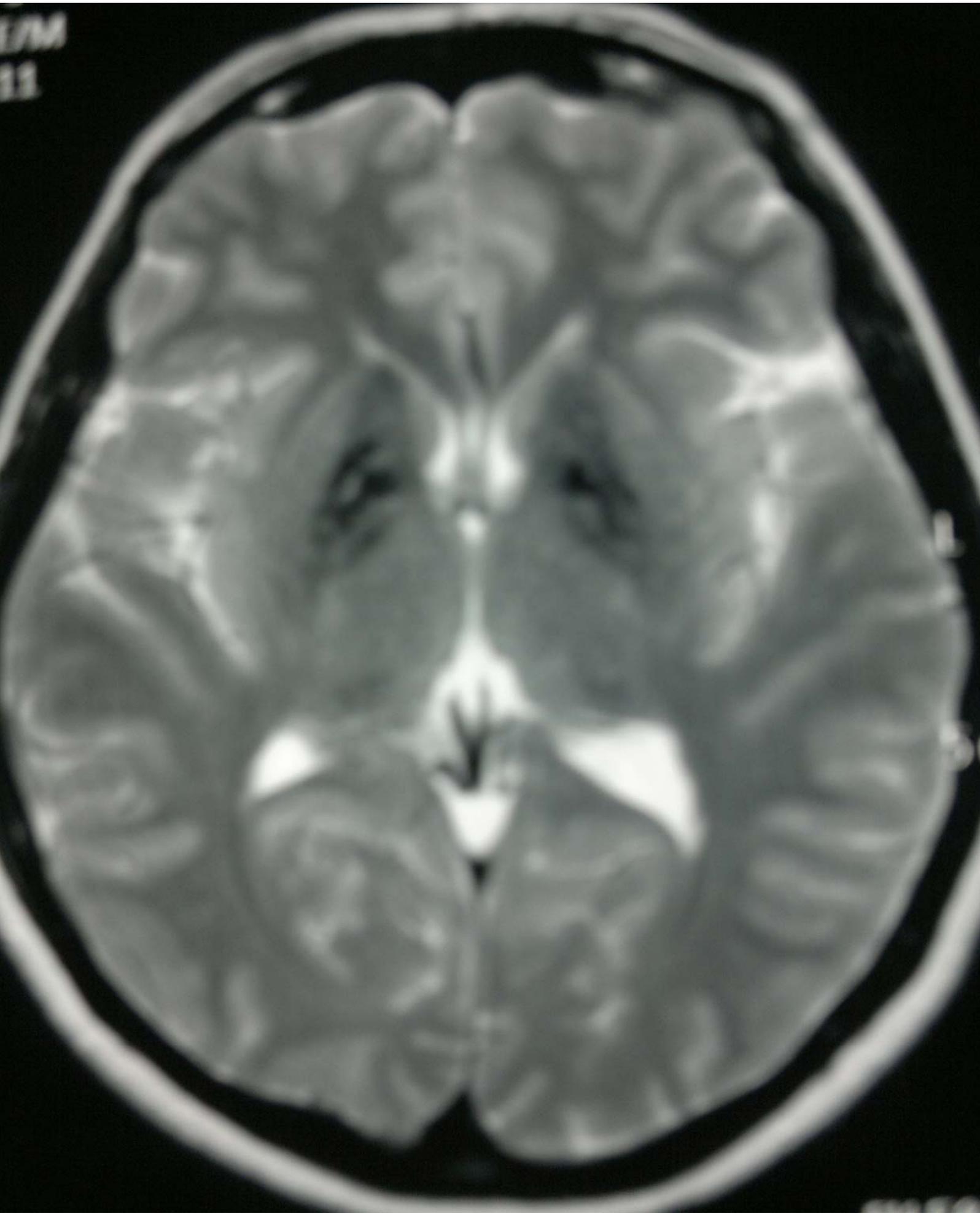
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