Global Surgery

Case Report ISSN: 2396-7307

Fahr's Syndrome, brain vascular disease and epilepsy: A case report

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Abstract

Calcium deposits in the basal ganglia and in the other areas of the brain can be idiopathic or secondary to metabolic disturbances including impairment of parathyroid glands. Fahr's syndrome is characterized by secondary cerebral calcification and variable clinical manifestations like movement disorder, cognitive impairment, and behavioural changes and later dementia. The clinical manifestations of this disease include a wide variety of symptoms, ranging from neurological symptoms of the extrapyramidal system to neuropsychiatric memory and concentration abnormalities to movement disorders including parkinsonism, chorea, and tremors, among others epilepsy is also described in the classification of the various clinical forms of Fahr's disease. Often this last clinical aspect remains unrecognized for a long-time giving rise to incorrect diagnoses. Diagnosis includes performing computed tomography, magnetic resonance imaging and standard brain radiography. Other investigations are blood tests and urine tests for haematological and biochemical and endocrinilogical indices. We report a 78-year-old woman with Fahr's syndrome due to idiopathic hypoparathyroidism who presented with recurrent epileptic seizures, she suffered from migraine attacks at a young age. Brain CT showed a hypodensity of the periventricular white matter from chronic ischemic cerebrovascular leukoencephalopathy, as well as a hypodense area in the right corona radiata moreover multiple small symmetrical nubecular calcifications of the basal ganglia. Brain MRI showed confluent areas expression of ischemic cerebrovascular disease in the periventricular and pontomesencephalic and areas of signal reduction compatible with calcific areas at the level of the basal ganglia. The patient entered in the outpatient's clinic for depressive syndrome and disabling dizziness not responding to common GP therapies; moreover, she seemed to be underestimating the symptoms that could be interpreted as due to epileptic disease.

Introduction

We studied a case of Fahr's disease diagnosed in a patient who presented at a young age crisis of migraine followed by lipothymia. The patient came to our out-patient clinic because of a depressive syndrome with emotional lability since she was a young age and for about twenty years, she had suffered from migraine treated with pizotifen and lipothymic episodes. In the past, the patient had undergone removal of the thyroid and uterus. The electroencephalogram showed a focal epilepsy with bilateral evolution [1] treated with lacosamide 200 mg daily. Fahr's disease can be known to be associated with epilepsy [2]. In addition, the patient was affected by cerebral vascular disease which recent studies have shown to be a relevant risk factor for epilepsy.

Case

We studied a female patient, age today 77 years, who underwent to the clinical observation in 2013. At that time, she was suffering from depressive syndrome, recurrent headache, orthostatic hypotension, rare lipothymic episodes. Fahr's disease was diagnosed in 2014 following radiographic diagnostic tests: computed axial tomography (CT) with the diagnosis of calcifications of the basal ganglia and brain magnetic resonance imaging (MRI) with the diagnosis of cerebrovascular disease due to the presence of hyperintense areas of hyperintense altered signal, chronic ischemic cerebrovascular pain expression at the periventricular and ponto-mesencephalic level.

At the time of taking charge, the patient exhibited a Single Photon Emission Computed Tomography (SPECT) already performed in 2013 which showed a focal perfusion defect in the left fronto-parietal area, site of the epileptic focus highlighted later. The patient had undergone a tiredectomy with sparing of the parathyroid glands. The blood calcium and parathyroid hormone values were normal, albeit with variations over time. The patient underwent a tilt-test to evaluate the frequently reported lipothymic episodes in the medical history. She performed both standard and dynamic 24 h EEG for the diagnosis of epilepsy. The patient was followed in the neurology clinic from 2013 to date.

Results

The diagnosis was complex and confirmed over time thanks to the investigation adopted. In particular, the following pathological conditions were detected: depressive syndrome treated with citalopram 10 mg per day, migraine treated with FANS, lipothymic episodes of unclear nature partly due to orthostatic hypotension treated with: caution in postural changes, use of elastic stockings, Trendelemburg position and intense and repeated muscle contractions, and partly due to seizures. The epilepsy was in fact documented by standard and dynamic EEG investigations treated with Lacosamide 200 mg day; cerebral calcifications of the basal nuclei compatible with Fahr's disease without familiarity or associated endocrinological alterations (blood parathyroid hormone, calcitonin, calcium) monitored over the years without alterations; neuropsychological tests performed from 2013 to 2021 with the exclusion of dementia. From 2013 to 2021 the syndromic situation remained in a condition of good compensation with respect to the various symptomatic aspects.

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Key words: *Fahr's Disease, Epilepsy, Brain Vascular Disease*

Received: June 16, 2021; **Accepted:** June 28, 2021; **Published:** July 01, 2021

Conclusion

Fahr's disease is a rare neurologic disorder with variable clinical manifestations, in which some forms are genetically determined while others occur sporadically and, in some patients, it is found to be associated with epileptic disease while in others the calcification of the basal ganglia is occasionally found through brain-TC. The age of onset and the association with endocrinological alterations are also variable (Fahr's disease secondary to hypo or hyperthyroidism). The diagnosis is linked to a syndromic picture to find which may be necessary some time. In the case of the patient studied, the main factor that led to the diagnosis was the association of epilepsy and calcifications of the basal ganglia, but the diagnosis was made more difficult by the fact that the patient presented a vascular disease which determined a secondary epilepsy as evidenced by recent literature [3,4] and the fact that she also presented orthostatic hypotension well documented by targeted investigations. In fact, the possibility of an association with a neuro-vegetative parkinsonism is envisaged.

Acknowledgement

The author wishes to thank the Nurse Coordinator Francesco Pepe and the staff, the Coordinator of Outpatient Clinic Dott.ssa Rita Bartolomei, and Director of the district 2 RM6 Dott.r Pierluigi Vassallo, for their organizing work.

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