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(CASE REPORT)

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Hemiplegia revealing Fahr's disease: A case report

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Abstract

Fahr's disease or idiopathic basal ganglia calcification is a rare pathology. The most common clinical manifestations are movement disorders, psychiatric disorders, cerebellar syndrome, seizures and cognitive disorders. We report the case of a 47-year-old female patient with Fahr's disease revealed by brutal right hemiplegia.

Keywords: Fahr's disease; Idiopathic calcifications; Hemiplegia; Case report

1. Introduction

Fahr's disease or idiopathic calcification of the basal ganglia is a rare pathology. This idiopathic form corresponds to sporadic or familial forms with autosomal dominant transmission. Its prevalence remains unknown until today [1]. Fahr's disease is characterized by an accumulation of calcium phosphate deposits around the wall of the cerebral microvessels, which is responsible for its clinical expression [1, 2].

Clinical manifestations are polymorphous, associating cognitive disorders, psychiatric disorders, delirious episodes, epileptic seizures, dysarthria, and abnormal movements [1].

We report the case of a 47-year-old female patient hospitalized for sudden right hemiplegia leading to the suspicion of a stroke, in whom the brain CT scan was in favor of Fahr's disease.

2. Clinical case

A 47-year-old female patient with no particular medical or surgical history presented two months before her admission brief loss of consciousness (two episodes), for which no exploration or treatment had been undertaken. The current episode that justified her hospitalization dates back to three hours before her admission by the sudden onset of the heaviness on the right side of the body, without any other associated signs.

On clinical examination, the patient was conscious, and well oriented, with right hemiplegia 2/5 and central facial palsy involvement without sensory disturbance. The blood pressure was 120/70 mmHg, and the BMI was 23kg/m^2.

The brain CT scan without contrast injection in axial view showed: bilateral and symmetrical spontaneous hyperdensities of the basal ganglia, the subcortical white matter, and the cerebellar hemispheres, evoking calcifications

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related to Fahr syndrome (Figure 1). The electrocardiogram showed an atrioventricular block with sinus rhythm. Chest X-ray and thyroid and parathyroid ultrasound were normal. Magnetic resonance imaging (MRI) was not performed. The blood calcium level was 98 mg/l, the blood phosphorus level was 42 mg/l, the blood creatinine level was 10 mg/l, the blood glucose level was 0.80 g/l and the blood ionogram was normal. The rest of the biological checkup was unremarkable, including blood count, transaminases, magnesium, cholesterol, triglycerides, uric acid, treponematosis, and HIV retroviral serologies.

The diagnosis of Fahr's disease was retained. The patient was put on an anxiolytic and functional rehabilitation sessions were prescribed. A genealogical investigation was not performed.

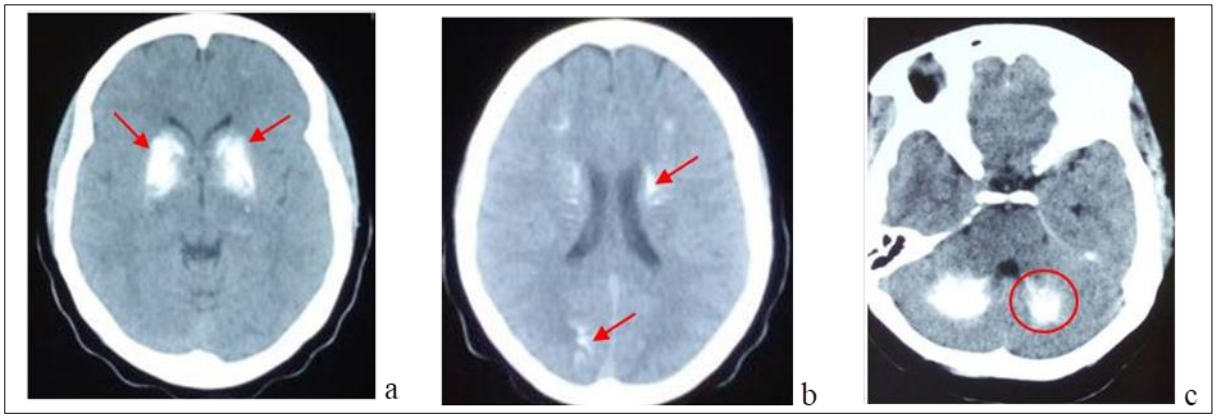

Figure 1 Brain CT scan without contrast injection in axial view showed: bilateral and symmetrical spontaneous hyperdensities of the basal ganglia [red arrow (a)]; the subcortical white matter [red arrow (b)]; and the cerebellar hemispheres [red circle (c)]

3. Discussion

The occurrence of a neurological deficit in Fahr's disease is a rare clinical manifestation. In the literature, few studies report a neurological deficit revealing Fahr's disease [2, 3, 4].

The physiopathology of Fahr's disease is not well known. It has been suggested that the accumulation of calcium deposits in the vascular walls of capillaries, arterioles, venules, and perivascular spaces would generate an inflammatory process in the brain. This slowly progressive phenomenon would probably be responsible for the neurological deficits observed. [5]. This could explain the installation of the deficit in our patient.

Clinical manifestations occur between the ages of 20 and 60. The neurological signs most often encountered are cognitive disorders, psychiatric disorders, delirious episodes, epileptic seizures, dysarthria, and abnormal movements [2-6].

The diagnosis is essentially radiological. The brain CT scan shows symmetrical calcifications of the basal ganglia, the subcortical white matter, and the cerebellar hemispheres [7]. This was the case with our patient.

Until nowadays, there is no specific treatment for Fahr's disease. The therapeutic strategy is based on symptomatic treatment and the quality of life of the patients [7].

4. Conclusion

Fahr's disease is a rare neurodegenerative disorder. Although rare, Fahr's disease must be included in the differential diagnosis of patients with a sudden neurological deficit. The brain scan remains the gold standard of diagnosis.

Compliance with ethical standards

Acknowledgments

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Disclosure of conflict of interest

The authors declare that they have no conflicts of interest.

Statement of informed consent

Informed consent and verbal permission were obtained from the patient and her family before submission of this article.

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