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Ischemic Stroke Revealing Fahr Syndrome

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Abstract **Case Report**

Fahr's syndrome is a rare anatomoclinical entity, characterized by bilateral and symmetrical intracerebral calcifications of the basal ganglia, generally difficult to suspect clinically because it can remain asymptomatic or result in polymorphic neurological manifestations. We report a case of Farh syndrome revealed after a cerebral infarction in the Neurology department of the CHU Gabriel Touré.

Keywords: Fahr syndrome, hypovitaminosis D, cerebral infarction.

INTRODUCTION

Fahr syndrome (FS) is defined by the presence of calcifications vascular and perivascular, bilateral and symmetrical to the gray nuclei of the base of the brain, which may extend to the dentate nuclei of the cerebellum [1]. It is a rare condition, characterized by clinical polymorphism with a predominance of neuropsychiatric manifestations and phosphocalcic metabolism disorders. FS occurs preferentially in patients with dysparathyroidism, mainly hypoparathyroidism [2].

Unlike Fahr's disease is a rare degenerative disease, characterized by the presence of calcifications of the basal ganglia. It is an autosomal recessive or dominant genetic disease with variable penetrance [1]. This gene mutation is located on chromosome 14q [3]. We report a case of Fahr's syndrome discovered during the exploration of a motor deficit of the left hemibody of sudden installation

COMMENTS

Mrs. MD, 86 years old, G1P1V1, admitted to the Neurology department for disorders of consciousness Her medical history includes arterial hypertension under nifedipine, type 2 diabetes under Insulatard for 10 years. 48 hours before her admission to the hospital, she would have presented a suspension of language, a left hemiplegia.

On admission, blood pressure was 179/95mmHg, pulse 119 beats/minute, SP02 96%, temperature 37.5°C, Glasgow sorus 8/15(E4V1M5), flaccid and proportional left pyramidal syndrome. The cerebral CT scan without injection showed hypodensity in the right deep sylvian territory and bilateral calcifications of the lenticular nuclei.

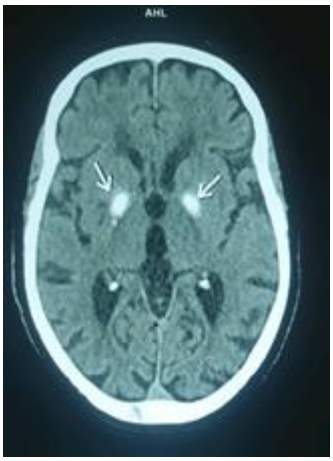

Figure 1: Bilateral and symmetrical calcifications of the lenticular nuclei

The diagnosis of Fahr's syndrome was evoked and we carried out a phosphocalcic assessment: a calcemia at 2.48mmol/L (N 2.15 and 2.5), a phosphoremia at 1.16 mmol/L (N: 0.80- 1.45), Magnesemia was 0.95 mmol / l, A Vit D dosage at 20.29 ng / ml (N: 30-100 ng / ml), while the PTH dosage was normal at 15.25 pg/ml (N: 15-65) eliminating dysparathyroidism. The thyroid balance was normal with a TSHus at 0.303 µU/ml (0.270-4.200) and the FT4 at 19.86 pmol/l (1.2-22). Serum creatinine was 73 umol/l with a clearance of 69.80 ml/min/1.73m2.

The lipid profile reveals dyslipidemia with LDL at 1.41 g/l Total cholesterol at 2.10 g/l, HBA1C at 8.90%. The ECG showed an antero-septal repolarization disorder with flattening of the R wave from V1a to V3 without cavity hypertrophy. The echocoeur was normal. Doppler ultrasound of normal supra-aortic trunks. The evolution was marked by the resumption of consciousness but with an inversion of the nychthemeral rhythm. Faced with hypovitaminosis D, we retained the diagnosis of Fahr Syndrome.

The patient received vitamin D supplementation (1500 IU per day), anticoagulation at a preventive dose and platelet aggregation inhibitor, statin and rehydration based on physiological saline. However, she came out with a Glasgow score of 15/15, left hemiplegia undergoing rehabilitation

DISCUSSION

Fahr's syndrome is a rare anatomo-clinical entity, characterized by the presence of bilateral and symmetrical intracerebral calcifications, localized in the central gray nuclei, occurring preferentially in patients with dysparathyroidism [4]. As in the case of our patient, the pathophysiological mechanisms that contribute to the occurrence of intracerebral calcifications during Fahr's syndrome are poorly understood. Most authors mention a metabolic disorder of oligoglial cells with deposits of mucopolysaccharides and secondary appearance of vascular and perivascular lesions and calcareous encrustations. In Fahr's disease (without calcium phosphate disorder), some authors mention an exaggeration of a normal process of calcium or ferrous deposits in the basal ganglia and dentate nuclei [5].

It is a rare condition, which can remain asymptomatic and be discovered fortuitously during the exploration of another pathology or be characterized by a clinical polymorphism with a predominance of neuropsychiatric manifestations and disorders of phosphocalcic metabolism [2]. In our patient, the discovery was made during the exploration of her vascular event.The pathophysiology explaining the occurrence of DALYs during Fahr syndrome remains unknown. It can be explained by extracellular calcium deposits in the walls of capillaries and small vessels leading to a decrease in tissue perfusion and cerebral ischemia [4].

Osteoporosis illustrate the crucial role of vitamin D in bone mineralization and calcium absorption. The recent discovery of its physiological role in neuroprotection, immunity, cell differentiation and proliferation justifies a growing interest in this vitamin [7]. It is important not to confuse FS with the physiological calcifications frequently observed in the elderly. They are generally located only in the pallidums, which are more discreet and punctiform [8] other conditions that can cause intracerebral calcifications, in particular endocrinopathies (hypothyroidism, hypogonadism), systemic pathologies (systemic scleroderma, systemic lupus erythematosus, celiac disease) infections (toxoplasmosis, neurocysticercosis, rubella), various diseases (chronic renal failure, vitamin D intoxication, mitochondrial cytopathies) and primary or secondary calcified brain tumours. However, the intracerebral calcifications observed during these different pathologies have different locations and different aspects. In contrast to the severity of the symptoms for which it may be responsible [9].

The analysis of the clinico-biological and radiological signs is the basis of the diagnosis. The correction of phosphocalcic metabolism disorders often leads to a significant improvement. [4] such is the case in our patient who, after correction of her vit D deficiency, improved significantly.

CONCLUSION

Fahr's syndrome is a rare condition with an essentially radiological diagnosis. Vitamin D is known for its essential role in the control of phosphocalcic homeostasis and in bone mineralization. Hypovitaminosis D in our patient could be a contributing factor to the development of cerebral calcification and thus induce Fahr's syndrome, which remains a good prognosis. Correction of phosphocalcic metabolism disorders often leads to a significant improvement

Conflicts of interest

The authors declare no conflict of interest.

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