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RESEARCH ARTICLE

FAHR SYNDROME REVEALED BY A CONVULSIVE CRISIS ASSOCIATED WITH IATROGENIC HYPERPARATHYROIDISM: CASE REPORT

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Abstract

Fahr's syndrome is a rare anatomoclinical entity whose main etiology is primary or postoperative hypoparathyroidism. It is characterized by bilateral and symmetrical intracerebral calcifications, localized in the basal ganglia. Hypoparathyroidism, primary or postoperative, is the most classic anomaly. Hyperparathyroidism is exceptionally reported as a cause of Fahr syndrome. We report the case of a 30-year-old woman who presented with an inaugural convulsive seizure revealing Fahr's syndrome secondary to iatrogenic hyperparathyroidism.

Introduction:-

Fahr's syndrome is a rare anatomo-clinical entity, characterized by bilateral and symmetrical intracerebral calcifications, localized in the central gray nuclei, most often associated with calcium phosphate metabolism disorders. We report the case of a 30-year-old patient who presented with an inaugural convulsive seizure revealing Fahr's syndrome with hyperparathyroidism.

Patient and Observation:-

This is a 30-year-old patient, anemic for 3 years, thyroidectomized in 2011 for nodular goiter under LT4 100ug complicated by iatrogenic hyperparathyroidism under Unalpha 0.25ug per day and calcium 2 to 3 g per day without any notion of discontinuation of treatment.Admitted to shock for unique and inaugural generalized tonic clonic crisis with post-critical confusion. A cerebral CT scan was performed, showing bilateral and symmetrical calcifications of the central gray nuclei (Figure 1), subsequently benefiting from a cerebral MRI objectifying a bilateral signal anomaly and symmetrical, related to calcifications revealing fahr syndrome of metabolic origin

The phosphocalcic balance, marked by severe hypocalcemia at 51 mg/l (VN: 86-100). The complete blood count objectifying an emia at 7.6g/dl, the blood ionogram, the liver test, the kidney test and the hemoglobin A1c were normal. The diagnosis of Fahr Syndrome was retained and a substitution treatment with Unalpha and calcivitamin D was started in association with neuropsychiatric treatment. The evolution was marked by the correction of phosphocalcic metabolism disorders.

Discussion:-

Fahr syndrome (FS) is defined by the presence of cerebral, non-arteriosclerotic, bilateral and symmetrical calcifications affecting the basal ganglia. 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 [1-2-3].

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Hypoparathyroidism is the most common cause of FS-related hypocalcemia. Hypocalcaemia caused by hypoparathyroidism explains the majority of clinical signs (cataract, malabsorption, neuromuscular hyperexcitability, various neurological and neuropsychic signs, psychiatric disorders that can go as far as psychosis, various cardiovascular disorders). [4-5]

Pseudohypoparathyroidism is a familial condition defined by peripheral resistance to parathyroid hormone. On the biological level, we then find hypocalcemia, hyperphosphatemia, but a serum level of normal or high parathyroid hormone. Hyperparathyroidism is exceptionally reported as a cause of FS [6-7].

The analysis of the literature made it possible to collect less than ten observations of hyperparathyroidism during this syndrome [8-9].

Other causes give intracerebral calcifications such as endocrinopathies, system diseases, celiac disease, infections, primary or secondary calcified brain tumors and other various diseases, however, during these different pathologies, calcifications intracerebral cells have different locations and aspects [10].

The prognosis of Fahr's syndrome is good, because the clinical and neuropsychic signs regress after correction of the phosphocalcic disturbances [6].

Conclusion:-

FS is a rare neurological disease, creating a contrast between severe non-specific symptomatology and a simple and effective treatment. The correction of phosphocalcic metabolism disorders allows a marked improvement in clinical symptoms, hence the interest of systematically looking for these disorders in the face of any neuropsychiatric manifestation associated with calcifications of the central gray nuclei.

Competing interests

The authors declare no competing interest.

Authors' contributions

All authors listed are contributors to this article

Figures

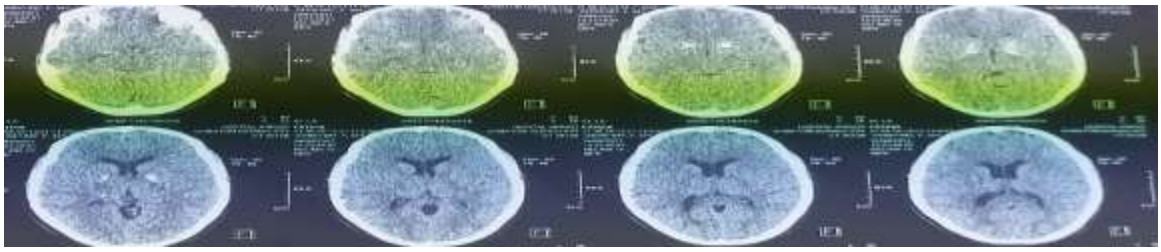

Figure 1:- Axial CT scans illustrating images of dense, symmetrical and bilateral calcification in the central gray nuclei.

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