



Fahr's Syndrome in A Patient with Secondary Hypoparathyroidism - Case Report

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Abstract

Introduction: The calcifications of the the basal ganglia (BGC) have been described since 1850, providing a common finding in neuroimaging exams, ranging from asymptomatic to more severe clinical manifestations such as seizures and extrapyramidal symptoms. Calcifications must be analyzed for the etiology, location, extent and composition, being the basal ganglia symmetric calcification (BGSC) a very important condition, which may be caused by idiopathic form, genetic, infectious or metabolic diseases. To describe such a condition, authors have used the eponyms: Fahr's Syndrome, Bilateral striatopallidodentate calcinosis, Fahr's Disease. Fahr's Syndrome is a degenerative nature rare neurological disorder, characterized by the presence of bilateral and symmetrical intracranial calcifications of the basal ganglia and cerebellar dentate nucleus, with consequent neurological manifestations mainly associated with disorders of calcium and phosphorus metabolism, particularly hypoparathyroidism.

Objectives: The objective of this article is to describe by a case report the diagnosis and treatment carried out in a case of Fahr's Syndrome and hypoparathyroidism due to thyroidectomy performed 10 years ago, with neurological manifestations due to hypocalcemia.

Method: Methodology: D.L.S., 64 years old, underwent to thyroidectomy 10 years ago, in use of L-Thyroxine since then. She was admitted at Veneza Emergency Unit due to generalized tonic-clonic seizure frame and tetany.

Results: Laboratory and cranial computed tomography findings revealed calcification areas in lenticular and dentate nuclei, corona radiata, semioval centers and caudate nucleus secondary to postoperative hypoparathyroidism. The patient was treated with oral calcium and vitamin D.

Conclusion: Neurologic findings were controlled with the treatment of hypoparathyroidism.

Keywords: Fahr's Syndrome; Hypoparathyroidism; Basal Ganglia Calcification

Introduction

Basal ganglia calcifications (BGC) have been known since 1850, however it was in 1930 that the eponymous "Fahr's Disease" (FD) arised, in honour od the german pathologist Karl Theodor Fahr, after he reported the case of a diseased man with a previous stage of convulsions, dementia, extrapyramidal symptoms and symmetrical and diffused calcification of the basal nuclei visualized at a post-mortem anatomopathological examination. Ever since, authors have been referring to FD as a primary, symmetrical and diffused of calcification of the basal ganglia, originated from idiopathy or familiarity, whilst the cases of pathological calcification of the basal ganglia, secondary to many etiologies are called Fahr's Syndrome (FS).

FS, also know as Non-Atherosclerotic Cerebral Calcification or Bilateral Perforated-Pale-Toothed Calcinosis, is a rare neurological disorder, with degenerative character, characterized by the presence of bilateral and symmetrical intracranial calcification of the basal ganglia and the cerebellar nucleus, with recurring neurological manifestations, associated mainly with calcium and phosphorus metabolism disorder, with hypoparathyroidism in 70% of the cases.

Clinically, FS could be manifested in neuropsychiatric disorders (schizophrenia, depression, mood disorders and OCD), cognitive and neurological symptoms, with convulsions, motor deterioration, dementia, speech disorders and extrapyramidal disorders such as parkinsonism, chorea, athetosis, myoclonus, gait disorders, dystonias or tics. BGC diagnosis can be made through anatomopathological examination, simple cranial x-ray and mainly computed tomography scans of the head (CT).

The objective of this paper is to relate, through a clinical case, the diagnosis and treatment realized in a case of Fahr's Syndrome and secondary hypoparathyroidism due to a thyroidectomy, done 10 years ago with neurological repercussions because of hypocalcemia.

Patients and Methods

64-year-old patient of the female sex who was submitted to thyroidectomy 10 years ago and ever since has been using L-Thyroxine. The patient was checked in the Emergency Care Unit Veneza in May of 2015, due to a generalized tonic-clonic convulsive crisis and tetanus she was medicated with diazepam and phenytoin causing regression of the convulsive condition. The patient remained hospitalized, waiting to be transferred to the Hospital São Lucas - Cascavel/PR, being transferred after 10 days with "neurological condition to be clarified".

At the hospital admission, the patient was unresponsive, confused in time, but space-oriented, she was complaining of fatigue, drowsiness, cramps, and she showed positive Chvostek and Trousseau signs. At the neurological examination, she presented dysarthria, global decrease of the force, hypotonia of upper limbs and lower limbs, rest tremor, hypokinesia, bilateral dysidiadochokinesia, marching of small steps and abolished deep reflexes. Folstein Mini Mental State Examination: 17/20 (illiterate). Cranial computed tomography scan showed symmetrical calcification of the fibers of the radiated crown / semioval centers, caudate head, bilateral lentiform nucleus and dentate nuclei (Figure 1, 2 and 3). Laboratory studies showed: Calcium (Ca^{+2})=5.8 (8.5-10.2 mg/dL); Phosphor (P)=7 (2.5-4.5 mg/dL); Parathyroid Hormone (PTH)=0.3 (10-65 pg/mL); Thyroid-Stimulating Hormone (TSH)=17.09 (0.5-5 mU/L); Creatine Phosphokinase (CPK)=617.5 (22-199 U/L); Creatinine=0.73 (0.5-1.1 mg/dL); Alkaline Phosphatase (AP)=93.6 (30-120 U/L); Aspartate Aminotransferase (AST)=292.7 (<31 u/L); Alanine Aminotransferase (ALT)=432 (<31 U/L); normal plasma albumin. Venereal disease research laboratory (VDRL), Antinuclear Antibodies (ANA), HBsAg, Anti-HCV non-reactives.

In the previous history, the patient presented convulsions for about 12 months and 6 months after the convulsions started, she began to present cognitive deficits, behavioral and mood disorders, being diagnosed as Depressive Disorder and she was prescribed Escitalopram.

In view of the results of the requested tests and evaluation of Endocrinology, the patient was diagnosed with secondary hypoparathyroidism, then she was prescribed calcium carbonate at the dose of 1500mg/day, Vitamin D 50,000 IU at the dose of one capsule/week for 06 weeks and L-Thyroxine 75mg, continuous use.

She kept taking Escitalopram 10mg for improvement of the affective and behavioral condition. She is currently under follow-up, and she is indicating a significant improvement in the quality of life, remaining stable until then.

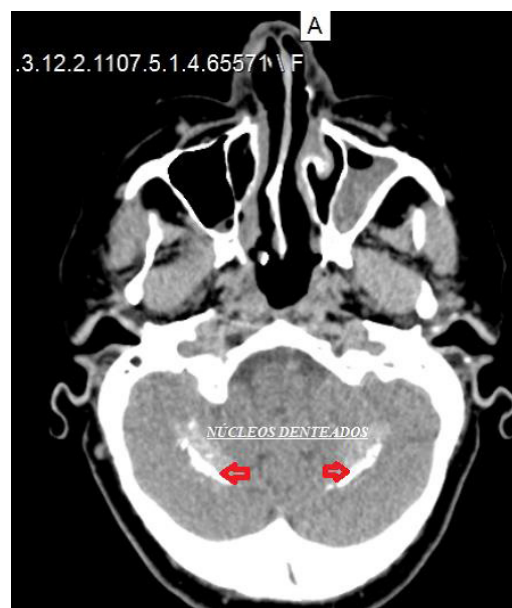


Figure 1: Cranial computed tomography scan showed symmetrical calcification of the fibers of the dentate nucleus

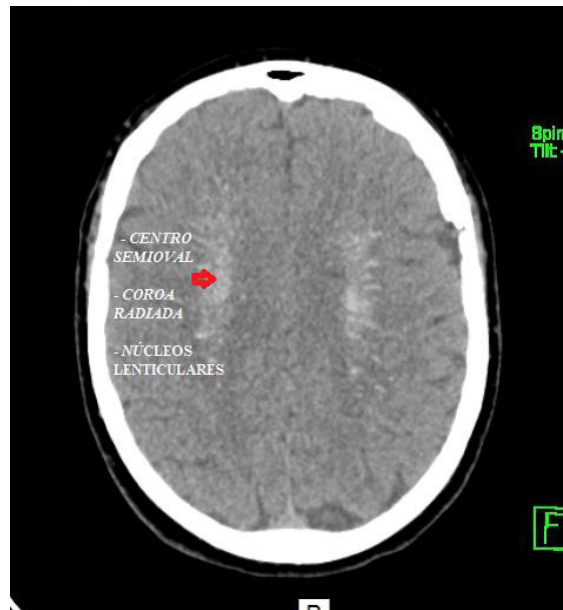


Figure 2: Cranial computed tomography scan findings demonstrate bilateral calcification of the radiated crown, semioval centers, and lentiform nucleus

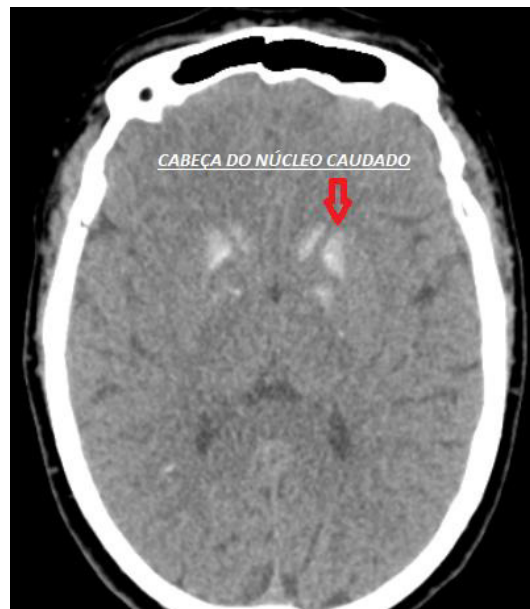


Figure 3: Cranial computed tomography scan showed calcification of the head of the caudate nucleus

Discussion

Basal ganglia calcifications (BGC) have been known since 1850, constituting a common finding in neuroimaging examinations, being found incidentally in 0.6% to 1.2% of the population, ranging from asymptomatic forms, to more severe clinical manifestations such as convulsions, neuropsychiatric disorders, speech disorders, dementia and motor deterioration with extrapyramidal manifestations, since the nuclei of the base are related to motor, behavioral and cognitive aspects.

Calcifications are not only composed of calcium, although this is the main element, but also of other minerals such as iron, zinc, aluminum, magnesium, glycoproteins and mucopolysaccharides. On anatomic-pathological examination, it is possible to witness vascular and perivascular lesions, essentially of the microvasculature, whereas neuronal and glial cells changes are verified in the more extensive lesions. The basal nucleus most affected by calcification is the pale globe, then the thalamus and eventually the brainstem. The clinical manifestations occur in middle age between the third and sixth decades of life.

Unlike FD, which has a primary course, FS has been associated with more than 30 types of diseases, which can be classified as inflammatory, tumor, degenerative, hypoxemic and vascular, toxic, and endocrine, and endocrinopathies, particularly parathyroid disorders, are the disorders most commonly associated with FS. These disorders include

mainly the primary hypoparathyroidism (HPP), the secondary hypoparathyroidism (HPS) and the pseudo-pseudohypoparathyroidism (PPH).

Postoperative HPS is the most frequent cause of hypoparathyroidism, as a result of total thyroidectomy or repeated surgeries for hyperparathyroidism, with calcifications seen on average 17 years after surgery. Hypocalcemia in postoperative HPS usually develops up to one week after the procedure, however, the diagnosis can be delayed by the asymptomatic or oligosymptomatic evolution of the patients, culminating in the appearance of the late neurological manifestations of chronic hypocalcemia.

In 1958, Bennet and Col performed a very important study in 80 patients with BGC, in which 66% had a history of calcium-phosphorus metabolism alterations, with 48% having PPH, 16% with PPHP, and only 2% with HPS. Regarding hypoparathyroidism, the goal of the treatment is to restore calcium and phosphorus levels near to normal, in order to stop the symptoms related to hypocalcemia, since this is the main consequence of endocrinopathy and also the main cause of its symptoms.

The diagnosis of BGC can be made by anatomopathological examination, simple x-ray of the skull and mainly computed tomography scans of the head. Laboratory evaluation of total and ionized serum calcium, phosphorus, magnesium and PTH is also necessary. TSH, T4, renal and hepatic function, CPK, FAN, VRDL and other serologies, in order to exclude possible etiologies of calcifications.

Serum levels of calcium, phosphorus, alkaline phosphatase and PTH, when normal, help to differentiate FD from FS due to endocrine disorders. Hypokalemia and hyperphosphatemia with normal renal function are pathognomonic of hypoparathyroidism. It is important to remember that vitamin D is a fundamental element in the metabolism of calcium and maintenance of homeostasis, and its active form is reduced in hypoparathyroidism, with consequent reduction of intestinal absorption of calcium.

The purpose of the treatment is to direct the patient to the control of symptoms, functional recovery, improvement of the patient's quality of life and prevention of complications, and when possible, the avoidance of the progression of the disease. Regarding hypoparathyroidism, the objective is to restore calcium and phosphorus levels to near normality, in order to cease symptoms related to hypocalcemia.

Final Considerations

Although some patients develop in a symptomatic or oligosymptomatic manner, there may be a correlation between the size of calcifications and the duration and severity of hypocalcemia, which are considered the main risk factors, further emphasizing the importance of early diagnosis and treatment of comorbidities associated with calcification of the basal ganglia. Treatment of hypoparathyroidism with calcium and vitamin D provided an improvement in neurological status.

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