doi: http://dx.doi.org/10.11606/issn.1679-9836.v100i5p504-507 Rev Med (São Paulo). 2021 Sept-Oct;100(5):504-7.

Fahr' s syndrome after thyroidectomy: a case report

Síndrome de Fahr após tireoidectomia: relato de caso

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Kzam ASL, Cunha AFC, Cunha RPC, Aguiar MAS. Fahr´s syndrome after thyroidectomy: a case report / *Síndrome de Fahr após tireoidectomia: relato de caso*. Rev Med (São Paulo). 2021 set.-out.;100(5):503-7.

ABSTRACT: Fahr´s syndrome is a rare disease characterized by bilateral symmetrical calcification over the basal ganglion of the brain and nuclei dentate of cerebellum mainly resulting from disorders in calcium-phosphorus metabolism. Neuropsychiatric changes and extrapyramidal symptoms are one of the main clinical manifestations described. Computed tomography is the imaging method used for visualizing brain calcifications and when associated with clinical history and laboratory findings is a triad for definitive diagnosis. Therefore, the objective of this was to report the clinical case of a patient with Fahr Syndrome presenting cranium brain computed tomography with symmetrical calcifications associated with a cognitive deterioration and extrapyramidal signs resulting from changes in calcium metabolism, having hypoparathyroidism as an etiological cause.

Key-words: Fahr's syndrome; Calcification; Basal nucleus; Hypoparathyreoidism.

RESUMO: A síndrome de Fahr é uma doença rara caracterizada pela calcificação simétrica bilateral dos núcleos basais do cérebro e do núcleo denteado do cerebelo decorrente principalmente de desordens no metabolismo cálcio-fósforo. Alterações neuropsiquiátricas e sintomas extrapiramidais são uma das principais manifestações clínicas descritas. A tomografia computadorizada constitui o método de imagem para a visualização das calcificações cerebrais e, quando associada à história clínica e achados laboratoriais, formam a tríade para o diagnóstico definitivo. Diante disso, objetivou-se relatar o caso clínico de um paciente com Síndrome de Fahr apresentando tomografia computadorizada crânio-cerebral com calcificações simétricas associadas à um quadro clínico de deterioração cognitiva e sinais extrapiramidais decorrentes de alterações no metabolismo do cálcio, tendo como causa etiológica o hipoparatireoidismo.

Palavras-chave: Síndrome de Fahr; Calcificação; Núcleos da base; Hipoparatireoidismo.

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INTRODUCTION

Fahr's syndrome is a rare neuropsychiatric disorder characterized by bilateral symmetrical calcification of the basal cerebral nuclei and dentate nuclei of the cerebellum, secondary to inflammatory, infectious diseases, exposure to toxins and radiation, but mainly due to metabolic disorders of calcium and phosphorus as occurs in hypoparathyroidism and pseudohypoparathyroidism[1,2,3,4].

Described by Karl Theodor Fahr in 1930[3], this condition presents a series of possibilities of clinical manifestations that encompass extrapyramidal symptoms, chronic and progressive cognitive deterioration and psychiatric alterations[3,5].

Because it is a rare syndrome, with few reports in the world literature and so far none in the literature of Pará, the objective of this study was to report the clinical case of a patient with Fahr's syndrome admitted to the general Intensive Care Unit (ICU) of a private hospital in Belém do Pará, Amazon, Brazil.

A CASE REPORT

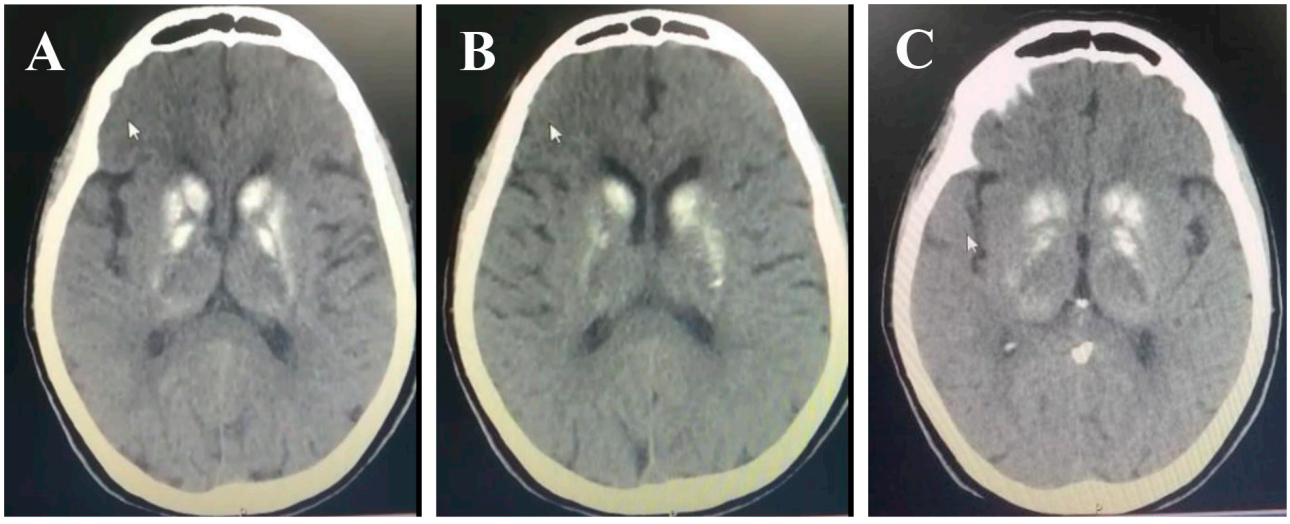
A 68-year-old woman is brought by family members to the emergency department of the hospital, who reported a fall from their own height, producing facial trauma and loss of consciousness for about five minutes. Also in the emergency department, she presented three generalized epileptic episodes, with clonic-tonic component.

The family reports a history of total thyroidectomy and chemotherapy for twenty years due to thyroid neoplasia and, more recently, behavioral change, with episodes of mental confusion and memory lapse, as well as retracted and melancholic posture. Additionally, they witnessed three other episodes of fall from their own height, but without the same consequences as those observed at the time of hospitalization.

When admitted to the ICU, she was comatose (AO 2 RV 2 RM 4), with no locator signs. She breathed with difficulty, noting laryngeal stridor and SO2: 79%, extremity cyanosis and RF: 30 irpm, use of accessory respiratory muscles, beating of nasal wings, resolving by orotracheal intubation with orotracheal tube (TOT) nº 6.5 due to narrowing of the glot and stenosis of the upper trachea and invasive mechanical ventilation. During intubation, mucopurulent secretion was observed by TOT.

Cardiocirculatory conditions revealed normophonetic heartbeats in 2 times, with regular rhythm, without murmur, 100 bpm heart frequency and 80x60mmHg blood pressure.

After stabilization, she was referred to the computed tomography (CT) sector, where the craniocerebral examination revealed relatively symmetrical parenchyma calcifications (Figure 1), involving the deep gray matter, the white medullary centers of the brain and the dentate nuclei of the cerebellum, the latter with a grossly striated nonspecific aspect. Cervical and thoracic CT showed steosis of the upper rings of the trachea and consolidation in the right pulmonary base, respectively.



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Figuras 1A, 1B e 1C – Cranial-cerebral CT scan. Axial section showing calcifications in the caudate, striated and dentate nuclei.

Laboratory tests for admission indicated Hb 11.8g/dL, Ht 31%, leukocytes of 9,480/µL, platelets 230,000/µL; PCR 4.4mg-dL; urea 36mg-dl, creatinine 0.7mg-dl, Na 136mEq-L, K 3.6mEq-L, Mg 1.4mEq-L and ionic Ca 0.7mmol/L (Table 1). Additional tests such as cardiac enzymes, electrocardiogram and Doppler echocardiogram

were within normal limits.

The patient remained seed with midazolan hydrochloride and received analgesia with fentanyl citrate in continuous infusion in the first 24 h. Sodium ceftriaxone 2g-day EV was started, maintained with anticonvulsant (phenytoin), venous hydration with SF0.9% 30ml/Kg/24h, replacement with calcium gluconate 10% and magnesium sulfate 10%.

It was requested opinion of the neurosurgery that defined by the maintenance of anticonvulsants. After 24 hours of ICU, considering the stability of the condition, a sedation pause was performed but the awakening was disorganized, and there was a need to start dexmedetomidine and digestive risperidone. After another 24 hours, although she was awakened in a more organized way, she did not meet physiological conditions that allowed a gradual reduction of the pharmacological doses previously administered, opting for early tracheostomy, something that was only authorized by family members after the 4th day of hospitalization. Before that, on the 3rd day, she presented high-response atrial fibrillation and chemical cardioversion was initiated with amiodarone EV. This allowed the reversal of clinical condition and the adjustmente of the drug dose for digestive administration in the next 48 hours.

Throughout evolution, ionic calcium remained at lower than normal levels, even with daily and fixed replacement in prescription. The investigation was complemented with the following laboratory analysis: phosphorus sérum 4.59mg/dL, PTH < 4.6pg/mL, vitamin D 14.87ng/mL, TSH: 1.75mU/L, Free T4 1.49ng/dL, T3 75ng/dL, Total Ca 0.45mmol/L (Table 1).

Table 1 – Laboratory findings

Initial Laboratory Analysis	**Results**	**Reference values**
Hemoglobin	11,8g/dL	12 – 16g/dL
Hematocrit	31%	35-47%
Leukocytes	9.480/µL	4.500 – 10.000/µL
Platelets	230.000/µL	150.000 – 400.000/µL
PCR	4,4mg⁄dL	< 0,8mg/dL
Urea	36mg⁄dL	15 – 45mg/dL
Creatinine	0,7mg⁄dL	0,6 – 1,35mg/dL
Na	136mEq⁄L	135 – 145mEq/L
K	3,6 mEq⁄L	3,5 – 5,5mEq/L
Mg	1,4mEq⁄L	1,5 – 2,5mEq/L
Ionic Ca	0,7mmol/L	1,17 – 1,32mmol/L
Complementary Laboratory Analysis		
Serum P	4,59mg/dL	2,5 – 4,5 mg/dL
PTH	< 4,6pg/m	10 – 65pg/mL
D vitamin	14,87ng/mL	20ng/ml
TSH	1,75mU/L	0,3 – 4,0mU/L
T4 livre	1,49ng/dL	0,7–1,8ng/dL
T3	75ng/dL	80-180ng/dL
Serum Ca	0,45mmol/L	2,1 – 2,5mmol/L

PCR – C reative protein; Na – sodium; K – potassium; Mg – magnesium; Ionic Ca – ionic calcium; Serum P – serum phosphorus ; PTH – parathormone; TSH – thyroid stimulation hormone; Serum Ca – serum calcium.

On the 6th and 7th day of hospitalization, the serum calcium value was closer to the lower level of normality and the patient was less agitated, tolerating ERT in PSV, reversed AF and no new epileptic events. On the 8th day, spontaneous breathing started only with oxygen supplementation and on the 10th he was discharged from the ICU, with a serum calcium value equal to 1.01mmol/L.

The patient's metabolic profile of calcium was altered, with reduced ionic and serum calcium, reduced 24-hour urinary calcium, serum phosphorus at the upper limit of normal, reduced D vitamin levels and decreased PTH, leading to hypoparathyroidism.

DISCUSSION

We present a case of an elderly female patient with a history of total thyroidectomy and exposure to adjuvant therapy, manifested by neuropsychiatric alterations, notably behavioral changes, episodes of syncope and epileptic seizures. The radiological findings compatible with bilateral calcifications in the basal ganglia and other brain structures associated with laboratory findings guided the suspicion and diagnosis of Fahr's syndrome.

Although there is still no consensus established in the literature regarding the conceptualization, the term

Fahr Syndrome seems to be related to neuropsychiatric manifestations associated with calcifications of the basal nuclei due to changes in the functioning of the parathyroid glands and, consequently, to the reduction in serum calcium, while that Fahr's disease is related to idiopathic calcifications of the basal ganglia, accompanied by neurological signs without a previously defined cause[2,4,6].

The clinical manifestations of the disease are varied and are closely related to areas with calcification. Thus, there is an influence of the basal ganglia on brain functions such as movement coordination, cognition and mood. The most common symptoms of Fahr syndrome in adults include parkinsonism, dystonia, ataxia, chorea and extrapyramidal syndromes, although some patients also present cognitive impairment, psychosis, hallucinations, anxiety, depression and personality change[2,3,7, 8.]

Furthermore, studies also point to a positive relationship between neurological manifestations and hypoperfused calcified areas. Thus, epileptic seizures would be closely related to the hypoxemic event. Also, hypocalcemia is known to be the cause of clinical neurological manifestations such as epileptic events and, similarly, it is well established that its treatment can stop the crises. Studies show a close relationship between post-total thyroidectomy hypoparathyroidism and extensive radiotherapy in the cervical region, with mental deterioration being more severe in these patients[9,10.]

Clinical manifestations may be due to different etiology such as vascular conditions (cerebrovascular accident, calcifications, arteriovenous malformations), degenerative (senility), infectious and inflammatory conditions (cytomegalovirus, toxoplasmosis, tuberculosis, HIV, neurocysticercosis), neoplastic (astrocytomas), toxic (radiotherapy, lead poisoning), resulting from trauma (chronic subdural hematoma)4 and endocrine (hypo/hyperpatiroidism, pseudohypoparathyroidism)[11].

Craniocerebral CT is the main method to assess areas of calcification. The hyperdensity observed is due to the deposition of hydroxyapatite, similar to that found in the bone structure. As a peculiar characteristic of Fahr's syndrome and hypoparathyroidism, calcifications present symmetry with progressive evolution and good responsiveness to the treatment of the underlying cause[1,2,3,5,7,12.]

The treatment aims to correct changes in calcium-phosphorus metabolism, in order to significantly improve symptoms, in addition to favoring the prognosis. The administration of elemental calcium and vitamin D replacement are necessary measures to maintain serum calcium and urinary calcium within normal limits[9].

Therefore, it is important that a rare entity, with multifaceted and non-specific clinical manifestations, ranging from mild muscle events to more severe and life-threatening brain and cardiocirculatory disorders, deserves citation and report, warning and instructing the medical community, and also the patient, for diagnostic possibilities, as well as for targeted treatment.

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Submeted: 2020, February 29
Aceito: 2021, February 16