



Case Report

Pseudotumor cerebri secondary to hypoparathyroidism and association to Fahr's Syndrome: a case report

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Abstract

The authors present the case of a patient who presented an initial headache attack and seizure, who developed intracranial hypertension secondary to hypoparathyroidism. Her complementary clinical investigation was compatible with Fahr's syndrome.

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Introduction

Headaches are extremely common complications in emergency departments, accounting for up to 16% of visits, according to some studies¹ and are observed in almost all patients with idiopathic intracranial hypertension (IIH), which is characterized by signs and symptoms that include, in addition to headache, nausea, vomiting, diplopia, decreased visual acuity, pulsatile tinnitus, among other possible manifestations. However, the clinical presentation is highly variable, which can delay diagnosis.²

Idiopathic intracranial hypertension is predominantly found in obese women and of childbearing age, with a worldwide incidence of around 12-20 cases per 100,000 people in this most affected group, but only 0.5-2 cases per 100,000 people in the general population.³

Systemic disorders and some drugs have been associated with increased intracranial pressure, such as thyroid disorders, hyper- or hypothyroidism⁴ and hypervitaminosis A, as well as isotretinoin and tetracyclines, being in this occasion defined as a condition of secondary intracranial hypertension.

Regarding Fahr's disease, a rare neurological disorder with a prevalence of 1/1.000.000, it is known that it is characterized by abnormal deposition of calcium, of idiopathic origin, in brain areas that control movement.⁵

Below, we present the case of a patient who presented an initial headache attack and seizure, who developed intracranial hypertension secondary to hypoparathyroidism. Her complementary clinical investigation was compatible with Fahr's syndrome.

Case report

A 33-year-old woman, BMI of 40.17 kg/m², with grade 3 obesity, Caucasian, single, with Down syndrome. Presents a history of postoperative hypothyroidism, whose thyroidectomy was performed 10 years ago, due to nodular goiter. This patient presented subacute nausea, visual turbidity, diarrhea and pulsatile holocranial headache, which worsened in the horizontal position and presented slight relief in the orthostatic position. This clinical picture lasted for about three days and evolved with complaints of loss of visual acuity and the occurrence of two generalized tonic-clonic seizures, which led to the emergency care in her city. At the time, the patient was referred to a tertiary service for specialized evaluation.

Upon neurological examination, the patient presented on the campimetry of confrontation, loss of peripheral visual field and preservation of the central field, and visual acuity of 20/200 in the right eye - measured by the Rosenbaum card - and counted fingers on the left eye. In the fundoscopy, bilateral papilledema was noticed. In the other neurological systems, no abnormalities were detected. This patient was then submitted to complementary exams, where hypocalcemia and hypomagnesemia were noted, thus being compatible with secondary, postoperative hypoparathyroidism. On computerized tomography examination of the skull, calcifications were identified in the basal ganglia, bilaterally, and these findings are characteristic of Fahr's syndrome. Due to the clinical finding of visual changes and headache, the patient underwent lumbar puncture with spinal manometry; it is important to note that she had no contraindications to the procedure. Following are the exam data: opening pressure (OP) = 84 cm H₂O and closing pressure (CP) = 15 cm H₂O. Proteins, glycorrachia and lactate without changes. No evidence of pleocytosis. With these findings, the proposed diagnostic hypothesis was intracranial hypertension secondary to hypoparathyroidism, presenting visual impairment and symptomatic convulsive crisis, secondary to hypocalcemia.

The patient was submitted to the correction of metabolism disorders, with replacement of calcium, phosphate and magnesium, and additional tests were requested to rule out other clinical conditions that promote intracranial hypertension. Five days after these measurements, the patient was again submitted to a lumbar puncture with spinal manometry, for control, being as follows: OP = 54 cm H₂O and CP = 12 cm H₂O. Thus, lumboperitoneal shunt was considered, due to the refractoriness of intracranial hypertension, visual loss and side effects of medications (topiramate, acetazolamide and furosemide) in high doses in an attempt to control the production of cerebrospinal fluid (CSF) - that is, to reduce the elevation of intracranial pressure. After these measures, the patient started clinical treatment directed at obesity and the surgical procedure was scheduled, which happened without complications. The patient presented a satisfactory clinical response, controlling for metabolic disorders with endocrinology and there was adequate control of intracranial pressure. Even so, the patient presented with loss of peripheral visual field as a sequel, but much less when compared to the admission examination.



Discussion

Headaches in a general context are classified into primary and secondary. Although primary headaches are the most common, there are other serious and potentially fatal conditions in 5-15% of cases, these being secondary. Therefore, the priority is to establish an accurate diagnosis and distinguish between them which can be benign and severe situations (subarachnoid hemorrhage, meningitis, intracranial hypertension).⁶ In IHH, headache is progressively more severe and frequent.⁴ In addition, it has a pulsatile characteristic, which may be associated with photophobia and phonophobia, in addition to worsening with decubitus and Valsalva maneuver.

Thus, according to the consensus of IHH¹, the diagnostic criteria are: papilledema, normal neurological examination (except for VI cranial nerve palsy), imaging tests without structural lesions (without hydrocephalus, mass, structural lesion or meningeal enhancement, cerebral and cervical venous thrombosis). In the study of CSF, the biochemical assessment and serology for fungi and tuberculosis are innocent. However, OP is ≥ 25 cm H₂O. For cases in which papilledema does not occur, the presence of high OP and another clinical alteration are necessary, be it paralysis of the VI cranial nerve (unilaterally or bilaterally) or alteration of the neuroimaging.¹ Assessing the case described here, we observed that the patient also meets diagnostic criteria for IHH, but with a known underlying clinical cause-metabolic dysfunction-being thus defined as a case of secondary intracranial hypertension.

Some patients with IHH complain of persistent headache after normalization of CSF pressure, so it is necessary to evaluate the possibilities of comorbidities, such as an underlying primary headache.² In turn, it is possible to attribute the appearance of this clinical condition to the hypofunction of parathyroid glands, the latter resulting, in the vast majority, from a surgical procedure, in the others, it arises due to genetic, autoimmune or idiopathic etiologies.^{7,8}

As for hypothyroidism, some reports address patients who met the criteria for the pseudotumor cerebri associated with hypothyroidism. It was then assumed that hypothyroidism primarily caused intracranial hypertension (IH) and with the replacement of the thyroid hormone, without any other measures to treat the pseudotumor, the condition would normalize. However, the relationship between normalization of thyroid function and improvement in neurological status was not observed.^{1,6,9-10} In addition, in another case study there was a report of a female patient, with clinical hypothyroidism, where persistent elevated CSF pressures were observed for four months, even with the normalization of thyroid status.

Therefore, it is more consistent evidence with the assumption that the two conditions have no causal relationship, with obesity exacerbated by thyroid hormone deficiency having more importance, as a precipitating factor for pseudotumor.¹¹ It is likely that occasional reports of simultaneous hypothyroidism and cerebri pseudotumor reflect the incidence of these diseases in a young female population.⁵

Regarding obesity, it is one of the clinical contexts most often associated with the development of pseudotumor cerebri, affecting about 80% of patients, showing itself to be fundamental in the pathophysiology of this disorder.²

Another important point is that hypocalcemia is a disorder present, mostly, due to surgical hypoparathyroidism. The rapid onset of hypocalcemia in the post-surgical setting can be acute, a fact that requires immediate and aggressive intervention. However, patients can evolve with chronic hypoparathyroidism, and are almost asymptomatic, even when there is loss of metabolic compensation to trigger symptomatic hypocalcemia. Despite this reality, patients with cerebral calcifications in the basal ganglia may evolve with IH, when this condition is attributed to calcium metabolism.⁸

So, in this context, Fahr's syndrome is defined by a triad: association of symmetrical calcifications of the central gray nuclei, neuropsychiatric symptoms and hypofunction of the parathyroid gland. In addition, hypocalcaemia caused by hypoparathyroidism explains most of the clinical signs, which may possibly arise as a result of this deficiency, such as: cataracts, calcium malabsorption, neuromuscular hyperexcitability, neurological and neuropsychic signs, psychiatric disorders that can even lead to psychosis, in addition to several cardiovascular disorders.¹²

Conclusion

With all these findings, and ruling out the possible differential diagnoses, the diagnosis of pseudotumor cerebri secondary to hypoparathyroidism due to total thyroidectomy, with a radiological picture of Fahr's Syndrome, was closed.

Given the above, it appears that hypothyroidism itself had no direct cause relationship with the development of the patient's IH, which is a condition resulting from hypoparathyroidism, which generated a metabolic dysfunction, reducing serum calcium levels, and also calcifications of the basal ganglia bilaterally. This glandular dysfunction of parathyroid glands was also responsible for neurological symptoms, resulting in intracranial hypertension, manifested by progressive and



pulsatile holocranial headache, accompanied by nausea and visual changes.

The conduct of Fahr's syndrome, in this case, was focused on the treatment of hypoparathyroidism and which consists of correcting metabolism disorders of phosphorus and calcium, which naturally resulted in clinical and radiological improvement, however, without definitive resolution of the condition.

Conflicts of Interest: The authors declare no conflicts of interest.

Author's contributions: PGGG- Data Collection, Project Management, Research, Writing; HDM-Project Management, Writing, Reviewing and Editing; FLAB- Writing, Proofreading and Editing; VGNF- Writing; NMAM- Writing

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