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(CASE REPORT)

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Depression revealing Fahr's syndrome in association with primary autoimmune hypoparathyroidism: A case report

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Abstract

Fahr's syndrome is a rare entity characterised by the presence of intracerebral calcifications, which are mainly located in the basal ganglia, and are bilateral and symmetrical. It is associated with phosphocalcium metabolism disorders, such as hypoparathyroidism. It can manifest as a variety of neurological or psychiatric disorders. We report the case of a 60-year-old female patient with primary autoimmune hypoparathyroidism, who presented with a depressive syndrome. Paraclinical investigations indicated the presence of Fahr's syndrome, which was associated with chronic hypocalcemia due to digestive malabsorption, and aggravated by therapeutic non-adherence due to her depression.

Keywords: Fahr's syndrome; Hypoparathyroidism; Depression; Autoimmune

1. Introduction

Fahr's syndrome, first described by Theodor Fahr in 1930, is defined by the presence of bilateral, symmetrical, non-arteriosclerotic intracerebral calcifications, which are located mainly in the basal ganglia (BG) and serrated nucleus [1]. This is a rare entity whose pathophysiological mechanisms are the subject of debate. It is characterised by clinical polymorphism, with an underlying etiology dominated by disorders of the phosphocalcic metabolism, occurring in the context of dysparathyroidism [2].

2. Case report

We report the case of a 60-year-old female patient with a history of rheumatic fever since the age of 8, vitiligo since the age of 9, well-balanced type 2 diabetes with dietary hygiene measures for 8 years, hypertension for 8 years with a triple therapy, and dilated cardiomyopathy for 8 years, witch the etiological work-up revealed a primary hypoparathyroidism. The diagnosis of primary hypoparathyroidism of autoimmune origin was retained, given the association with vitiligo and the negative results of the remaining etiological work-up. The patient was placed on a treatment plan comprising calcium and alfacalciferol. Over the past three years, the patient has required multiple hospitalisations due to severe hypocalcaemia. The patient was admitted to our department with a two-month history of tetany attacks. The clinical examination revealed moderate obesity with generalized vitiligo, a positive Chvostek and Trousseau sign, and a prolonged QT interval on the electrocardiogram. Furthermore, we observed a depressive behaviour pattern, associated with episodes of unexplained crying. This was subsequently confirmed by the psychiatric investigation as a depressive syndrome. Biological tests indicated severe hypocalcaemia at 70 mg/l, which was promptly addressed. A subsequent investigation into malabsorption identified non-atrophic, non-active chronic gastritis, with negative celiac disease antibodies, and negative anti-parietal cell, anti-21 hydroxylase, anti-TPO and anti-GAD antibodies. Given the chronic hypocalcaemia associated with a depressive syndrome, a cerebral Computed Tomography (CT) scan was commissioned,

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which revealed calcifications of the white matter and basal ganglia, consistent with the diagnosis of Fahr's syndrome (Figure 1). In collaboration with the psychiatrist, the patient's comprehensive care plan was initiated with the objective of enhancing her psychological well-being and adherence to treatment, which would consequently influence the depressive syndrome.

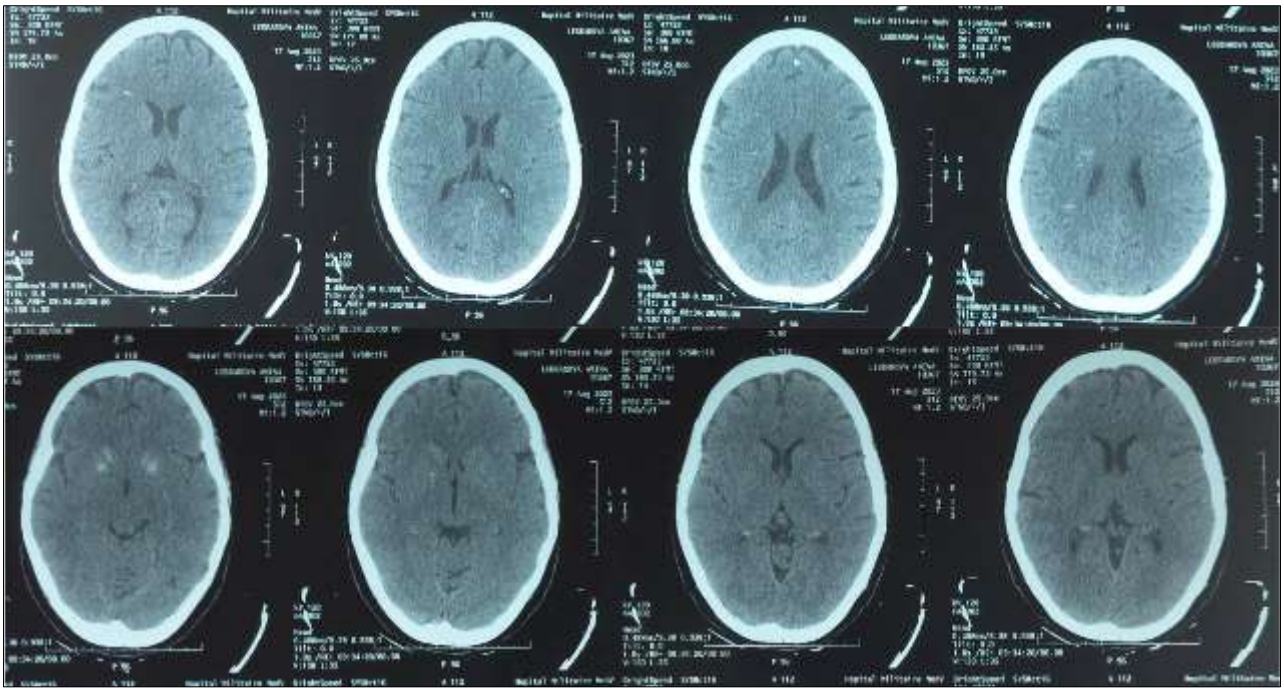

Figure 1 T1 coronal plane of brain MRI showing calcifications of the basal ganglia

3. Discussion

Fahr's syndrome is a rare anatomo-clinical condition characterised by bilateral, symmetrical intracerebral calcifications in the basal ganglia. It is most often associated with disorders of phosphocalcium metabolism. It should be differentiated from bilateral strio-pallido-dentate calcinosis, known as Fahr's disease, which is a genetic form of basal ganglia calcifications not associated with endocrine disorders [1].

Primary hypoparathyroidism is an endocrine disease defined by insufficient secretion of parathyroid hormone (PTH), a crucial hormone in phosphocalcium metabolism. The clinical manifestations are diverse and secondary to chronic hypocalcemia [3]. A biological diagnosis is made on the basis of the characteristic association of hypocalcemia, hyperphosphatemia, hypocalciuria, hypophosphaturia and a decrease in PTH serum levels.

The most common cause of primary hypoparathyroidism is cervical surgery, which accounts for 75% of cases [4]. The next most common cause is autoimmune origin, either isolated or associated with other autoimmune disorders in the context of autoimmune polyglandular syndrome (APS-1) [5]. In this case, we retained the autoimmune origin hypothesis due to the association with vitiligo alone, with no further evidence of other autoimmune disease.

The pathophysiological mechanisms involved in the development of intracerebral calcifications in Fahr's syndrome are not yet fully understood. The majority of authors have proposed a metabolic disorder of oligoglial cells with mucopolysaccharide deposition, which leads to calcification of the small vessels of the basal ganglia. The biochemical analysis revealed the presence of deposits of mucopolysaccharides, acids and minerals, including calcium, phosphorus, iron, sulfur, magnesium, aluminum and zinc [6].

Other conditions that can lead to Fahr's syndrome include pseudohypoparathyroidism, hypothyroidism, hypervitaminosis D [7], coeliac disease, systemic diseases (such as systemic lupus erythematosus) and certain infections (such as toxoplasmosis, rubella, AIDS and so on). Additionally, mitochondriopathies can also be a contributing factor [8].

The clinical presentation of Fahr's syndrome is complex and variable, with no discernible correlation between the affected brain regions and the observed clinical manifestations. The syndrome may manifest in a range of neurological disorders, including seizures, gait disturbances, bradykinesia, rest tremors, and cognitive decline. In 40% of cases, it may also present as psychiatric manifestations, such as psychosis, anxiety, apathy, or manic and depressive disorders [9].

A cerebral CT scan is the recommended examination for diagnosing of basal ganglia calcifications [10]. Magnetic resonance imaging (MRI) can show T1 and T2 hypersignals, but is of limited use in cases of Fahr's syndrome. Functional imaging using a SPECT scanner (single-photon emission computed tomography) represents a promising new avenue for investigating basal ganglia and the pathophysiological mechanisms of Fahr's syndrome, as evidenced by studies that have identified perfusion abnormalities in calcified regions [2].

4. Conclusion

Our case study demonstrates the importance of monitoring and managing chronic hypocalcaemia secondary to autoimmune hypoparathyroidism. This requires a multidisciplinary approach to address malabsorption due to chronic atrophic gastritis and potential therapeutic non-adherence. The depressive syndrome in the context of Fahr's syndrome, confirmed by cerebral CT, further complicates the management of hypoparathyroidism. Our findings emphasise the necessity for effective hypoparathyroidism management and prompt detection of complications. This is crucial for improving the functional and vital prognosis of patients

Compliance with ethical standards

Disclosure of conflict of interest

We have no conflicts of interest to disclose.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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