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CE - MEDICAL ILLUSTRATION

A case of brain calcifications in postsurgical hypoparathyroidism

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A 69-year-old woman presented to our inpatient clinic to evaluate a 2-month history of widespread joint pain, muscle cramps, slowness of movements, tremor of the head, and difficulty in speech. She had suffered from arterial hypertension, secondary hypothyroidism, and hypoparathyroidism for 30 years, due to a papillary thyroid cancer for which she had undergone thyroparathyroidectomy and a radioactive iodine therapy (I-131). There was no history of preexisting neuropsychiatric manifestations, mood disturbances or seizure, and her family history was not contributory. Her drug treatment was amlodipine 10 mg/die, oral levo-tiroxin 125 μg/die, and calcitriol 0.25 μg/die, and she had discontinued the supplemental calcium salts intake for the 2 prior years, because of the findings of joint and kidney calcifications.

At admission, she was conscious and oriented to the time and place, with a normal memory and intelligence. Vital signs were within normal limits, excepting a poor drug control of the blood pressure (181/103 mmHg). Neurological examination confirmed the intentional tremors of the head and the upper limbs, a gait disorder with mild impairment of balance and substantially normal coordination, and dysarthric speech with "punctuated voice". Otherwise, she had neither motor weakness nor cranial nerve involvement, presented only mild bradykinesia without rigidity or stooped posture, and absent primitive reflexes. Her remaining clinical examinations were unremarkable.

Routine laboratory tests revealed normal hemogram, renal and liver function, serum electrolytes such as sodium, potassium, chloride and magnesium, protidogram, and standard urinalysis. Laboratory investigations of thyroid and parathyroid function and calcium–phosphorus metabolism showed hypocalcemia (total calcium 6.7 mg/dL, ionized calcium 3.03 mg/dL) and hyperphosphatemia (4.9 mg/dL), with normal values of thyroid-stimulating hormone (TSH, 0.83 μUI/mL), free thyroxine (fT4, 1.18 ng/dL), parathyroid hormone (PTH, 9.1 pg/mL) and vitamin D (22 ng/mL). Moreover, blood tests for rheumatic diseases such as rheumatoid factor, antinuclear antibody test, anti-cyclic citrullinate peptide antibody and extractable nuclear antigen panel were all unremarkable.

To clarify the neurological involvement, an unenhanced brain magnetic resonance imaging (MRI) was performed, showing homogeneously altered signal intensity in the putamen, globus pallidus, caudate and cerebellar hemispheres (Fig. 1a). To rule out a chronic calcium deposition disease, an unenhanced brain computed tomography (CT) was carried out, confirming diffuse calcified concretions of basal ganglia and cerebellum (Fig. 1b, c). Based on these findings, a diagnosis of Fahr's syndrome was made.

The patient's treatment was implemented with oral calcium and vitamin D3 supplements 1000 mg + 880 UI/die, levodopa and benserazide 100 + 25 mg/die, and alprazolam 0.5 mg/die, observing an improvement in neurological signs and symptoms.

Bilateral striatopallidodentate calcinosis (BSPDC), otherwise known as Fahr's syndrome, is a rare neurodegenerative condition characterized by symmetric, non-atherosclerotic abnormal calcification of the areas of the

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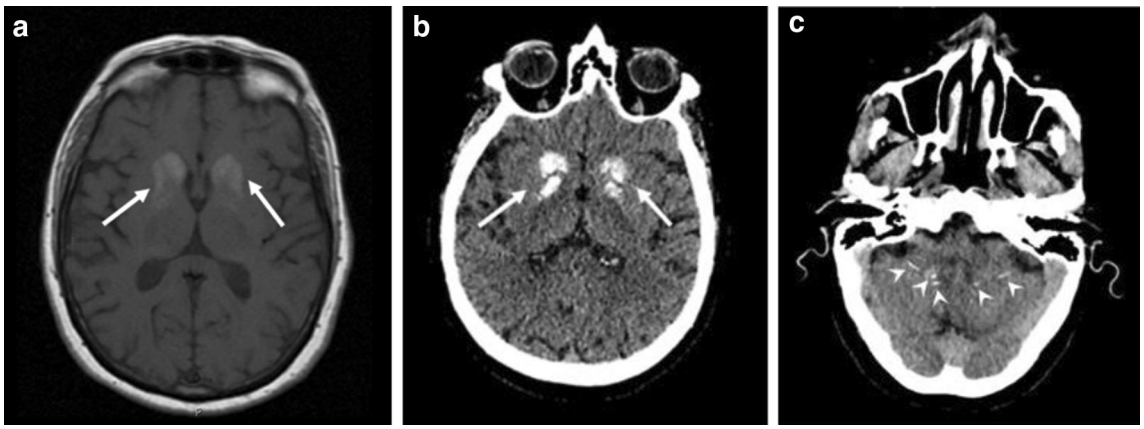

Fig. 1 Unenhanced brain magnetic resonance imaging (**a**) shows homogeneously altered signal intensity in putamen, globus pallidus, and caudate (*arrows*). Unenhanced computed tomography reveals diffuse calcifications of basal ganglia (**b**, *arrows*) and cerebellar hemispheres (**c**, *arrowheads*)

central nervous system (CNS) controlling movements, including basal ganglia, thalamus, dentate nucleus, cerebral cortex, subcortical white matter, and cerebellum. BSPDC is most commonly transmitted as an autosomal dominant trait, but it may also be passed as an autosomal recessive trait or occur sporadically. A locus at chromosome 14q has been suggested to be involved in its genetic transmission. The etiology of the sporadic form is not yet known, and different agents can be implicated, such as endocrine disorders, mitochondrial myopathies, dermatological, and infectious diseases. The typical age at the onset of the symptoms is 40–60 years [1–3].

Movement disorders are the most common presentation of BSPDC, accounting for about 55 % of the cases. Of these, parkinsonism is seen in the 57 %, chorea in 19 %, tremors in 8 %, dystonia in 8 %, athetosis in 5 % and orofacial dyskinesia in 3 % of cases. Other neurologic manifestations include cognitive impairment, cerebellar signs, speech disorder, pyramidal signs, psychiatric features, gait disorders, sensory changes, and pain. Overlap of signs and symptoms referable to different areas of the CNS is common [4].

Endocrine disorders are most commonly associated with BSPDC, mainly parathyroid disturbances such as both idiopathic and secondary hypoparathyroidism, pseudohypoparathyroidism, pseudo-pseudohypoparathyroidism, and hyperparathyroidism. Secondary hypoparathyroidism is generally an iatrogenic condition, occurring as a complication of thyroidectomy [1].

The diagnosis of Fahr's syndrome is based on the combination of clinical features, brain imaging and on an exclusion of other causes of intracranial calcifications.

The MRI is the best diagnostic method in detecting brain abnormalities due to its sensitivity. Nevertheless, calcifications appear with various signal intensities on conventional MRI sequences; therefore, a CT scan is generally the favorite imaging technique for detecting and localizing the extent of intracranial calcifications. Also, in cases of suspected BSPDC with brain calcifications on MRI, a CT scan should be considered for a more accurate diagnosis [1, 5].

To date, various treatments have been proposed for the management of BSPDC to obtain remission or stabilization of signs and symptoms. Although postsurgical hypoparathyroidism is a well-known condition, it is mandatory to take into account a proper dosage of the supplementary therapy with calcium and vitamin D, to obtain a correct balance between calcium and phosphorus and to avoid systemic calcium deposits. In the same way, movement disorders related to parathyroid diseases can be resolved with the correction of calcium and phosphate. Moreover, treatment includes symptomatic support such as levodopa, clonazepam, and atypical antipsychotics to treat extrapyramidal and psychiatric disorders [1, 3].

In conclusion, this report concerns a neurological involvement during a postsurgical hypoparathyroidism not well controlled by drugs. This condition requires a long-term follow-up based on clinical and biochemical data, since hypocalcemia may remain subclinical for an indefinite period of time. Nevertheless, it is essential to take into account the development of intracranial calcifications during this phase, which may be at least partially responsible for neurological symptoms. In this sense, it is worthy considering the use of imaging techniques in those cases where complementary brain alterations are suspected. Equally, an adequate treatment with calcium and hydroxy-vitamin D should be maintained, paying attention to an adequate serum calcium-to-phosphorus ratio to avoid the development of brain calcifications.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Statement of human and animal rights This article does not contain any studies with human participants or animals performed by any of the authors.

Informed consent Informed consent was obtained from the individual participant included in the study.

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