New onset of epileptic seizures induced by Fahr`s syndrome secondary to idiopatic hypoparathyroidism.

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Introduction

Hypocalcemia due to hypoparathyroidism leads to a large spectrum of clinical manifestations but a rare and unusual presentation is onset or aggravation of epilepsy.
Physiological intracranial calcification occurs in about 0.3-1.5% of cases. It is asymptomatic and detected incidentally by neuroimaging. Pathological basal ganglia calcification is due to various causes, such as: metabolic disorders, infectious and genetic diseases. Hypoparathyroidism and pseudohypoparathyroidism are the most common causes of pathological basal ganglia calcification. We present a representative case study of a spectacular extensive bilateral brain calcifications in the basal ganglia in a patient with idiopathic hypoparatiroidism with neurological manifestations.

Clinical case

We report a 77-year-old man who was found to have profound hypocalcaemia and idiopatic hypoparathyroidism when investigated for epileptic seizures and loss of consciousness. He had affective disturbances and only mild neurocognitive disorders. Cataract was present. The neurological examination showed an extrapyramidal syndrome with postural tremor and cerebellar ataxia. The deep tendon reflexes were normoactive in all four limbs. Chvostek's sign was present but Trousseau sign was not observed.

Laboratory analysis showed: low concentration of serum ionized calcium at 2.9 mg/dL (normal: 4.2-5.4 mg/dL), total calcium at 5.9 mg/dL (normal: 8.8 to 10.0 mg/dL), hyperphosphoremia at 6.4 mg/dL (normal: 2.3 to 4.7 mg/dL) and 2.5 pg/mL intact-parathyroid hormone (normal: 11.0 to 67.0 pg/mL). Alkaline phosphatase, magnesium, calcitonin, serum thyroxin and thyroid-stimulating hormone levels were normal.

Brain computed tomography demonstrated a symmetric, extensive, bilateral calcification of the basal ganglia, centrum semiovale, and bilateral dentate nuclei of the cerebellum, typical for Fahr's syndrome. The red nucleus and substantia nigra appeared normal. The diagnosis of Fahr's syndrome, secondary to hypoparathyroidism was posed. A prepontine meningioma was also found (Figure 1 A and B).

The electrocardiogram showed normal QTc interval and the interictal electroencephalography and electromyography were normal.
Bone densitometry showed increased bone mineral density.

After the patient was treated with oral calcium and active vitamin D (1-alphahydroxy vitamin D3), serum calcium levels returned to normal and seizure attacks ceased progressively resulting in stopping antiepileptic drugs.

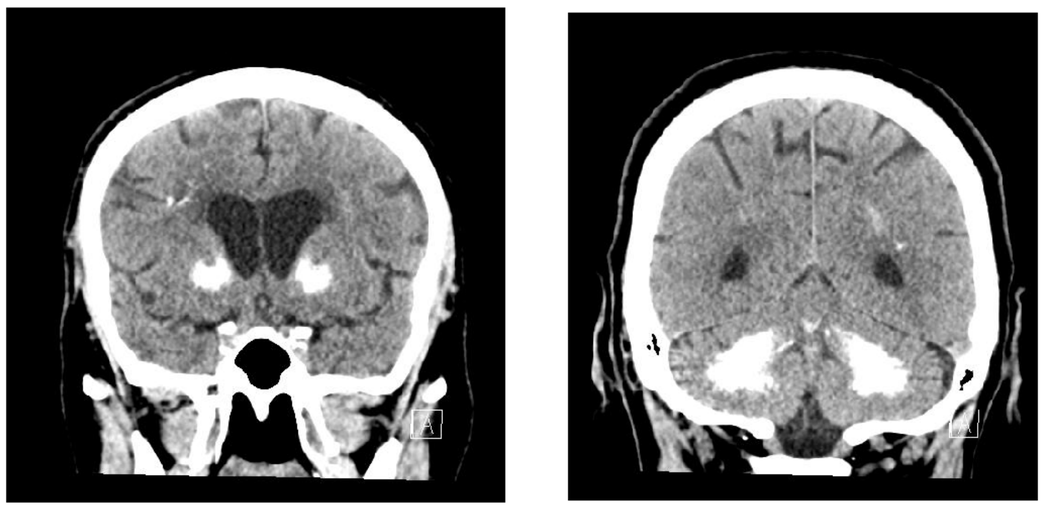

Fig 1. Brain computed tomography demonstrated a symmetric, extensive, bilateral calcification of the basal ganglia, centrum semiovale (A) and bilateral dentate nuclei of the cerebellum, (B).

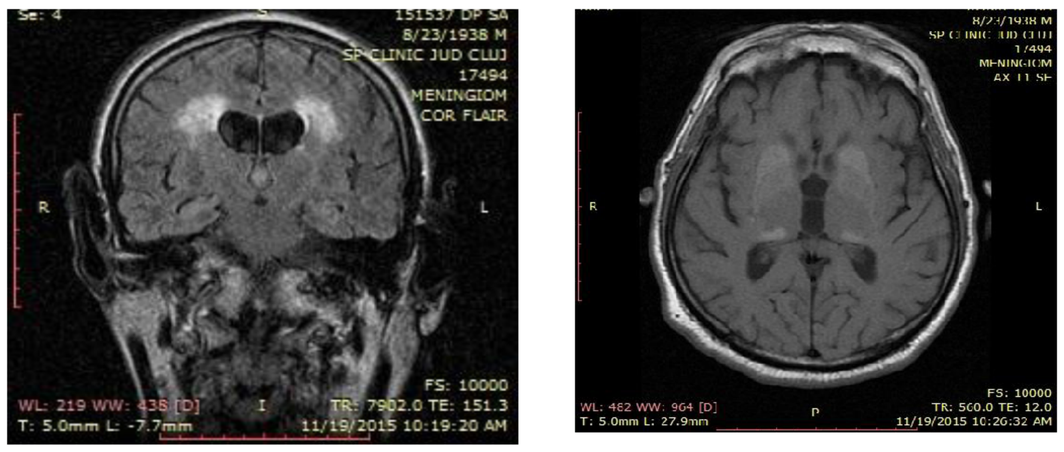

Fig 2. Multiple and symetrical areas of calcification: periventricular (A) , intraparenchymal, basal nuclei, thalamus and bilateral semiovale centers(B)

Discussions:

Many studies have described an association between parathyroid diseases and brain calcifications, especially in primary idiopathic and autoimmune hypoparathyroidism. Cerebral calcifications occur after years of evolution of the disease and could cause extrapyramidal manifestations.
The association between hypocalcaemia and low levels of PTH exclude other causes of cerebral calcifications (pseudohypoparatiroidism, hyperparathyroidism, CO poisoning, encephalitis, Fahr disease, Cockayne syndrome, tuberous sclerosis, neurofibromatosis, vascular disease, cerebral parasitosis). Hypocalcemia and hyperphosphatemia cause widespread occurrence of symptomatic brain calcifications/ asymptomatic, through a mechanism incompletely elucidated. The symptoms and clinical signs can often be reversible by administration of calcium. The evolution is usually favorable once the level of calcium is restored. Hypoparatirodism patients described an increased bone mineral density. Lack of parathyroid hormone results in low bone turnover and increased bone mineral density with associated increased bone mineralization. Whether these changes affect risk of fracture is not yet clear. Nevertheless, treatment with parathyroid hormone appears to improve the structural quality of bone.

Conclusions:

These cases illustrate the importance of search for disrupted phosphocalcic metabolism but also emphasizes the importance of the role of neuro-imaging in patients with new-onset epileptic seizures in order to detect hypocalcemia secondary to hypoparathyroidism.

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