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Epileptic Seizure Revealing Fahr Syndrome With Phosphocalcic Metabolism Abnormalities. A Case Report

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Abstract:
Fahr syndrome (FS) is a rare entity, characterized by phosphocalcic metabolism disorders and clinical polymorphism of which neuropsychological symptoms are the most common. We report the case of a 42-year-old woman presented for a generalized tonic-clonic seizure and for whom the diagnosis of FS secondary to primary hypoparathyroidism with severe hypocalcemia was retained.

Key Word*: Fahr syndrome; Epileptic seizure; Brain imaging; Hypocalcemia; Hypoparathyroidism.*

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I. Introduction

Fahr syndrome (FS) was described by Theodor Fahr in 1930 by the presence of bilateral and symmetrical, non-arteriosclerotic intracerebral calcifications located in the basal ganglia **[1]**. These calcifications can also be located at the thalamus, the cerebral cortex and the dentate nucleus of the cerebellum **[2]**.

FS is a rare entity, characterized by phosphocalcic metabolism disorders. It occurs preferentially in patients with dysparathyroidism, mainly hypoparathyroidism **[3]**. FS is characterized by the predominance of neuropsychiatric manifestations which are the consequences of either hypocalcaemia or perivascular cerebral calcifications testifying to the duration of the phosphocalcic disorders or more particularly the direct action of Parathormone on the basal ganglia **[4-5]** .

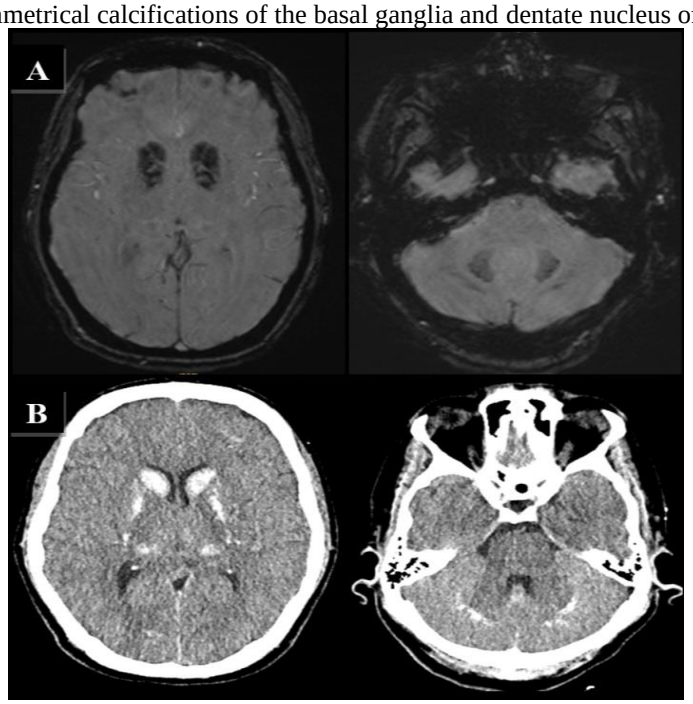
Epileptic seizures could reveal a Fahr syndrome. We report a case of generalized seizure revealing a Fahr syndrome with severe hypocalcaemia and hypoparathyroidism.

II. Case presentation

We present a case of 42-year-old woman admitted to the emergency room after a generalized tonic-clonic seizure. She has no known personal or family, surgical or medical history. Neurological examination as well as the rest of the somatic examination was normal.

The brain MRI revealed the presence of bilateral and symmetrical calcifications of the basal ganglia and dentate nucleus of the cerebellum, suggesting FS (**Figure 1A**). A complement with a cerebral computed tomography (CT) scan is carried out showing the same aspect and location of the calcifications (**Figure 1B**).

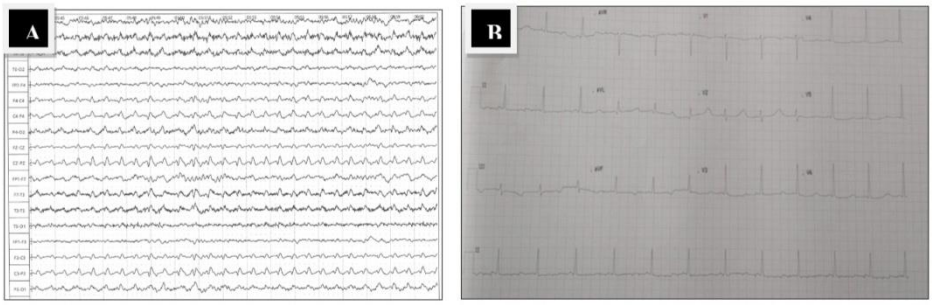
Figure no 1: Displays axial brain magnetic resonance (A) and computed tomography scan (B) images, revealing bilateral and symmetrical calcifications of the basal ganglia and dentate nucleus of the cerebellum.



The electroencephalogram revealed triphasic waves suggestive of metabolic encephalopathy (**Figure 2A**). The biological analyzes revealed severe hypocalcemia at 41 mg/L (Arsenazo technique, normal value: 86 – 100 mg/L), hyperphosphatemia at 66.5 mg/L (colorimetric dosage, normal value: 25-45 mg/L), a 25-OH-vitamin D low to 6.2 ng / mL (ECL / competition, normal value: 30 - 100 ng/mL), an intact parathyroid hormone (PTH) low to 8.8 pg / mL (ELFA Sur technique VIDAS, normal value: 9.2 – 44.6 pg/mL). Cell blood count, thyroid test, ionogram, protein and albumin levels, liver test, renal test and glycated hemoglobin were normal.

The patient underwent cardiac exploration to assess the impact of hypocalcemia. The electrocardiogram revealed QT prolongation with negative T waves (**Figure 2B**). Transthoracic echocardiography was normal.

Figure no 2: electroencephalogram revealing triphasic waves (A) and electrocardiogram revealing QT prolongation with negative T waves (B).



The diagnosis of FS secondary to primary hypoparathyroidism was retained and rapid correction of hypocalcemia was initiated by intravenous administration of calcium in association with calcivitamin D, UN-ALFA and an antiepileptic drug. The evolution was favorable and the control serum calcium level was normal at 100 mg/L.

III. Discussion

FS is a rare entity, characterized by the presence of intracerebral calcifications at basal ganglia and dentate nucleus of the cerebellum, non-arteriosclerotic, symmetrical and bilateral. FS occurs preferentially in patients with dysparathyroidism **[2-6]**. Hypoparathyroidism is the most classic abnormality which associates hypocalcaemia, hyperphosphatemia, hypocalciuria and hypophosphaturia **[7]**.

The pathogenic mechanism of these calcifications remains not well elucidated. Authors suggest a metabolic disorder of oligodendrocytes with mucopolysaccharide deposits and secondary appearance of vascular, perivascular lesions and encrustations limestone. Other authors suggest an exaggeration a normal process of calcium or iron deposits at the basal ganglia and dental nucleus. This entity must be distinguished from the genetic or sporadic calcifications of the basal ganglia, which does not associate phosphocalcic disorders and defined Fahr disease **[8]**.

FS is characterized by clinical polymorphism and most often it manifests with neuropsychological symptoms **[9]**. Hypocalcaemia resulting from hypoparathyroidism could explain the majority of clinical symptoms which are muscular hyperexcitability, malabsorption, cataract, neurological, psychiatric and cardiovascular disorders **[8]**. Epileptic seizures could also be explained by severe hypocalcaemia such as in our patient **[9]**. The diagnosis of Fahr syndrome is made by CT scan images, which show bilateral and symmetrical intracerebral calcifications, affecting the basal ganglia **[2]**.

FS should be differentiated from other affections that can cause intracerebral calcifications such as endocrinopathies, celiac disease, systemic diseases, infections, primary or secondary calcified brain tumors, hypervitaminosis D, intoxications and other affections. In these several affections, intracerebral calcifications have different appearance and are not bilateral and symmetrical **[10-11]**.

There is no specific treatment for FS. The therapeutic management consists on correcting the phosphocalcic disorder **[12]**. FS has a good prognosis and correction of disorders of phosphocalcic metabolism often allow a total regression of symptoms **[7]**.

IV. Conclusion

FS is a rare disease with clinical polymorphism. We highlights in this case report the interest of researching phosphocalcic metabolic disorders in the presence of associated epileptic seizures to calcifications of the basal ganglia, in order to adopt the appropriate therapeutic management.

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