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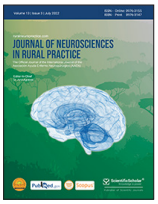

Letter to Editor

Catatonia revealing a Fahr syndrome

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Dear Editor,

Fahr syndrome (FS), since described by Theodor Fahr in 1930, is defined radiologically by the occurrence of striato-pallido-dentate, non-arteriosclerotic, bilateral, and symmetric calcifications.[1] It is a rare condition with controversial pathophysiological mechanisms that are evidenced by the multiple assigned denominations.[2] The clinical manifestations of FS are very polymorphic with dominated etiologies by dysparathyroidism.[1,3] Vitamin D deficiency has rarely been reported as the cause of this syndrome.[4]

Inhere patient was a 31 year old without any particular medical or surgical history. He presented a progressive installation of severe depression with psychotic features resisting to psychotropic treatments using antipsychotic and antidepressant. The case was complicated by acute installation of a psychomotor inhibition state with food refusal and deterioration of the general state. In clinical examination, the patient presented a catatonic syndrome according to Diagnostic and Statistical Manual of Mental Disorders, version 5 (DSM-5) diagnostic criteria including mutism, stupor, postural maintenance, rigidity, catalepsy, and waxy flexibility. The brain computed tomography (CT) scan was performed and showed a symmetrical bipallidal calcifications [Figure 1].

Parathyroid hormone (PTH) and 25 hydroxy (OH)-vitamin D assays showed PTH at 196 pg/mL (Normal value (NV): 15–65) and a 25 OH vitamin D collapsed to 9 µg/L (NV: 29–50). Phosphocalcic assessment, blood count, blood ionogram, liver function assessment, renal function assessment, and hemoglobin A1c were all normal. Thus, the diagnosis of FS secondary to hypovitaminosis D was retained and a substitution treatment with alpha-OH vitamin D3 was initiated while associating a neuropsychiatric supplementary medication using lorazepam, selective serotonin reuptake inhibitors antidepressant, and low dose of atypical antipsychotic. The evolution was marked by the correction of the metabolic disorder and neuropsychiatric signs and a complete return to the premorbid state maintained over a 2-year follow-up.

Actually, FS is known to be a rare neurological entity.[5] However, the analysis of population genome demonstrated that it is not really a very rare disease, in fact, it is rather an underdiagnosed underestimated disease so far.[6]

Indeed, Fahr's syndrome diagnostic is generally difficult to suggest based on clinical manifestations because it might remain asymptomatic and express polymorphic signs and symptoms without corresponding to a specific profile of disease. The neuropsychiatric symptoms are mainly reported and occurring gradually;[5,7] they include dementia syndrome, confusion, hallucinations, delirium, psychosis, mood disorders, panic attacks, irritability, and also aggressiveness. Other somatic symptoms, such as Parkinsonian-like movement disorder, headache, seizures, and syncope, might also occur.[6] There are very few reports of catatonia associating this condition. These symptoms would rise the basal ganglia role in originating the reported psychiatric symptoms.[8] Sometimes, the clinical manifestations are summarized in hypocalcemia signs including tetany, Chvostek's sign and Trousseau sign, or symptoms of the causal pathology of Fahr's syndrome.[9]

The diagnostic test of choice for locating and evaluating calcifications is brain CT-scan. However, imaging is rarely allowing to evoke a diagnosis immediately.[2]

Three main etiological axes are underlined comprising hypoparathyroidism, pseudo-hypoparathyroidism, and hyperparathyroidism that are uncommonly reported as a cause of Far's syndrome.[3]

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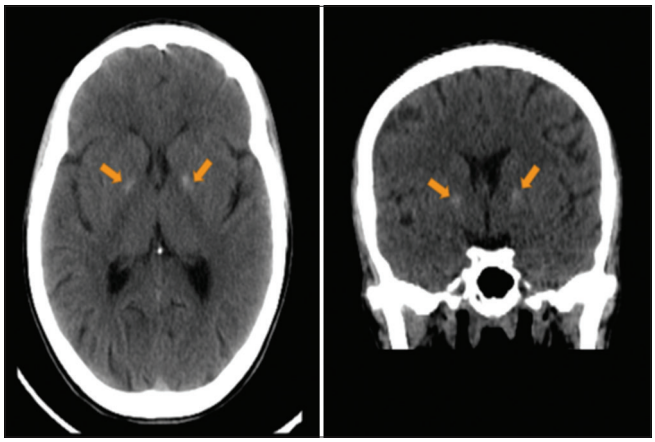

Figure 1: Brain computed tomography-scan of the patient in axial and coronal slice selections is clearly showing symmetrical bipallidal calcifications (See the pointing arrows).

Although bilateral strio-pallido-dentate calcinosis is referred as "Fahr's disease," there are 35 supplementary names in the literature which are used to denominate the same state. Besides, secondary bilateral calcification is also reported in the literature to describe a variety of infectious, metabolic, genetic, developmental, and several other conditions.[2] There is therefore a confusing literature between FS and Fahr's disease whether they are identical or not. Hence, some authors used denominations such as "primary" and "secondary." Actual the literature is reporting two entities associated with calcifications of the gray nuclei including Fahr's syndrome and Fahr's disease. Indeed, FS represents a secondary form of cerebral calcifications which are caused by another known disease.[10] Other conditions might originate intracerebral calcifications such as endocrinopathies with hypothyroidism and hypogonadism, systemic lupus erythematosus, infectious with toxoplasmosis and rubella, and tumor pathologies.[2]

Other diagnostic difficulties of this entity are as follows:

- The unknown proportion of asymptomatic carriers and the prevalence of age-related calcifications of the gray nuclei in the general population
- The absence of anatomoclinical corollary, in particular between the extent of the lesions and the severity of the clinical profile and between the localization of the calcifications and the type of symptoms encountered, moreover, patients with extensive calcifications might be pauci or asymptomatic[11]
- and the absence of a threshold delineating the classification these calcifications as pathological or not according to patients' age.

Therefore, the causal association remains difficult to demonstrate, especially in the absence of clear pathophysiological evidences.[12,13]

The care process should essentially consist of ruling out any potential organic brain disorder before classifying the patient, especially in young subjects without a history of psychiatric disorders.[14] The treatment is mainly symptomatic. Thus, the improvement of phosphocalcic metabolism disorders would often direct to a significant improvement amelioration. However, the response rate to medication is various. Psychiatric symptoms are associated with bigger variability in the response rate during the care stage.[15]

Furthermore, there are few data reporting the use of psychotropic drugs, particularly antipsychotics.' It is assumed that patients with calcifications of the basal ganglia could be more vulnerable to extrapyramidal side effects. Hence, it is importance to be cautious in the treatment prescription in case of FS with psychiatric symptoms.[16] The prognosis is generally good.

In conclusion, the interest of performing a vitamin D assessment in case of any neurological and/or psychiatric manifestation that is refractory to treatment and/or associated with basal ganglia calcifications even if the phosphocalcic balance is normal, this allows adopting the most appropriate therapeutic measures.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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