

Crampes musculaires chez un épileptique : penser au syndrome de Fahr

Muscle cramps in a patient with epilepsy: think of Fahr's syndrome

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Case

A 49-year-old man with history of cataract surgery, presented with a 30-month history of cramps and stiffness in lower limbs. He had medical history of epilepsy not controlled by antiepileptic therapy including carbamazepine and phenobarbital even with gradually increasing dose. Physical examination revealed a reduction in jerk reflex but Chvostek and Trousseau signs were negative.

Laboratory examinations showed low serum calcium level at 42 mg/L (normal: 85-108 mg/L), elevated se-

rum phosphorus level at 64 mg/L (normal: 24-45 mg/L) and deficiency of 25-hydroxyvitamin D at 10 ng/mL (normal: 30-60 ng/mL). Serum levels of albumin and magnesium were within the normal range. Renal and liver function tests were unremarkable. The serum level of parathyroid hormone was 0.33 pg/mL (normal: 15-65 pg/mL). The thyroid hormone levels and the rate of magnesium were normal.

Cerebral CT scan revealed diffuse and symmetric calcifications involving basal ganglia (Fig. 1) and cerebella (Fig. 2). The diagnosis of Fahr's syndrome due to idiopathic hypoparathyroidism was established.

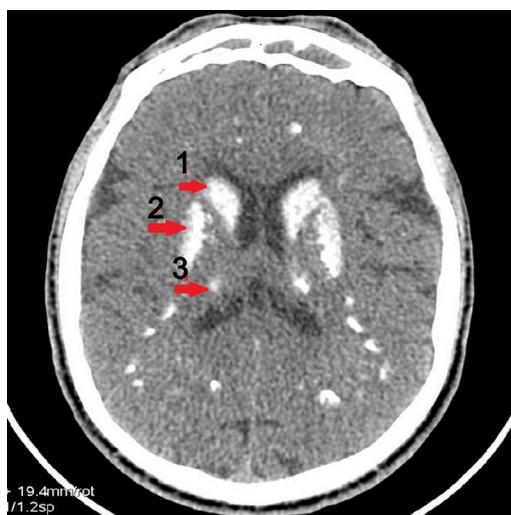


Figure 1. Axial non-contrast cerebral CT-scan. Calcifications of caudate nucleus (1), lentiform nucleus (2) and thalamus (3).



Figure 2. Axial non-contrast cerebral CT-scan. Calcifications of cerebella.

He underwent a treatment including oral calcium supplementation (2 g daily) and 1-alpha-hydroxy vitamin D3 (1 µg daily). Improvement of cramps and decrease in the frequency of seizure were obtained. Calcium level after 6 months of follow-up was at 80 mg/L.

Fahr's syndrome or Fahr's disease is a rare disorder characterized by bilateral and symmetric calcifications involving basal ganglia, thalami and cerebella [1]. It is commonly diagnosed in patient at the third and fourth decades of their lives [1]. The etiology of Fahr's syndrome is not yet identified. However, it can be associated with various conditions especially endocrine disorders. Parathyroid disturbances particularly idiopathic hypoparathyroidism is the most common disease associated with Fahr's syndrome. This condition is characterized by reduced parathyroid hormone associated with hyperphosphatemia and hypocalcemia. It has been demonstrated that basal ganglia calcification occurred in 73.8% of patients with idiopathic hypoparathyroidism and correlated with the duration of hypocalcemia [2].

Clinical expressions of Fahr's syndrome are characterized by a broad spectrum of clinical manifestations. Neurological and psychiatric abnormalities are the

most common. Cataract and cramps are due to chronic hypocalcemia related to hypoparathyroidism [3]. On the other hand, hypovitaminosis D could have significant implications for patients with Fahr's syndrome [1]. Cerebral CT scan is considered as the investigation of choice to diagnose Fahr's syndrome. It shows diffuse and symmetric calcifications involving basal ganglia and cerebella [3].

Fahr's syndrome is yet incurable. However, proper early symptomatic treatment based on calcium supplementation may prevent further neurophysiological disorders of Fahr's syndrome related to idiopathic hypoparathyroidism.

References

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