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Neuropsychiatric Manifestations Indicative of Fahr's Syndrome Secondary to Hypoparathyroidism after Total Thyroidectomy

Tall CT[1,2*], Fall AK[3], Ndiaye M[3] and Gning SB[3]

[1]*El Hadj Ibrahima Niasse Private University, Saint Christopher Iba Mar Diop School of Medicine, Pharmacy and Dental Surgery, Dakar.*

[2]*Dakar University Polyclinic.*

[3]*Service Médecine Interne/Hôpital Principal de Dakar.*

[*]Correspondence:
Cheikh Tidiane TALL, El Hadj Ibrahima Niasse Private University, Saint Christopher Iba Mar Diop School of Medicine, Pharmacy and Dentistry, Dakar, Call : 00221 77 459 33 69.

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ABSTRACT

Introduction: *Fahr's syndrome is defined by the existence of cerebral calcifications affecting the basal ganglia. Clinical manifestations are variable, dominated by tetany attacks and severe neuropsychiatric manifestations.*

Clinical Case: *We report the case of a 36-year-old female patient transferred to internal medicine for management of behavioural disorders associated with confusional syndrome and cognitive impairment, without fever. Her history included a total thyroidectomy in 2009. A phosphocalcic work-up and an intact parathyroid hormone (PTH) assay revealed hypoparathyroidism marked by severe hypocalcaemia. Cerebral computed tomography (CT) revealed calcifications in the caudate and lenticular nuclei. The diagnosis of Fahr's syndrome secondary to post-total thyroidectomy hypoparathyroidism was therefore accepted in view of these anamnestic, clinical, biological and radiological argu ments.*

Conclusion: *Fahr's syndrome is a rare neurological disease, characterised by severe non-specific symptoms and a simple and effective treatment, particularly in the context of total thyroidectomy.*

Keywords
Fahr syndrome, Hypoparathyroidism, Cerebral calcifications.

Introduction
Fahr syndrome (FS) is an anatomoclinical entity characterised by the presence of bilateral and symmetrical non-arteriosclerotic intracerebral calcifications located in the basal ganglia. It is a rare condition with a prevalence of less than 0.5%. Clinical manifestations vary from simple behavioural disorders to tetany attacks and severe neuropsychiatric manifestations. This condition is often associated with disorders of phosphocalcium metabolism, mainly secondary to primary or postoperative hypoparathyroidism [1]. The Fahr triad is defined by the association of symmetrical calcifications of the basal ganglia, neuropsychiatric symptoms and hypofunction of the parathyroid gland [2].

We report the case of a 36-year-old woman with Fahr syndrome secondary to hypoparathyroidism following total thyroidectomy.

Observation
Mrs S.C, aged 36, was transferred to the internal medicine department for management of behavioural disorders associated with confusional syndrome and cognitive impairment, without fever, after an 11-day stay in intensive care. The reasons for hospitalisation, after admission to the emergency department, were non-febrile convulsions, followed by post-critical confusion, with no sign of neurological localisation or meningeal irritation, which had benefited from neuropsychiatric management. The family also reported that the patient had experienced sudden convulsive seizures with altered consciousness a few days previously, and then became agitated and insomniac, with delusions of persecution. This situation necessitated a consultation at the psychiatric department

of the FANN hospital, with a prescription for neuroleptics. Her history included a total thyroidectomy in 2009 for an unspecified thyroid condition treated with levothyroxine. On admission, the patient was found to be disorientated in time and space, with a fixed, perplexed gaze and no response to questions. There were no signs of meningeal irritation or sensory-motor deficits. Osteotendinous reflexes were present and symmetrical, tonus normal; there was no coordination disorder or damage to the cranial pairs. Trousseau's sign was positive and Chvosteck's sign negative. The patient was apyretic. Cerebral computed tomography (CT) revealed calcifications in the caudate and lenticular nuclei (Figure 1).

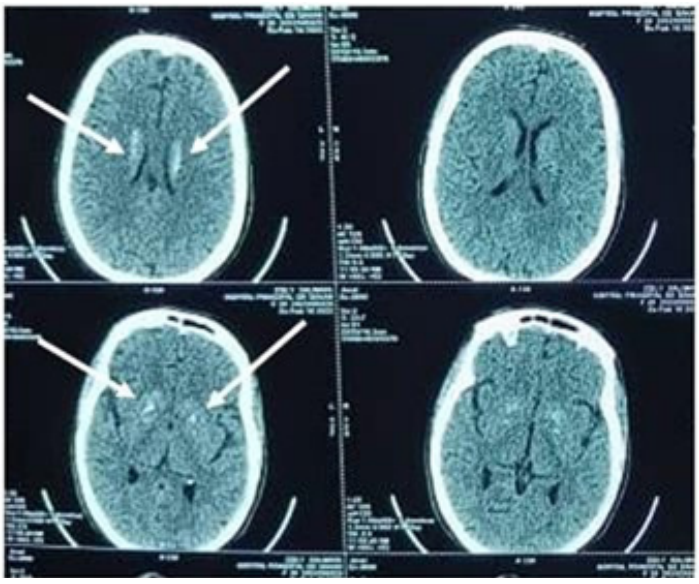

Figure 1: Extensive calcifications of the caudate and lenticular nuclei.

Lumbar puncture with cytochemical and bacteriological study of the cerebrospinal fluid ruled out meningitis. The phosphocalcic profile, intact parathyroid hormone (iPTH) and 25-hydroxy-vitamin D assays revealed hypoparathyroidism marked by severe hypocalcaemia with corrected calcaemia at 58 mg/l (VN: 85-100), hyperphosphaemia at 57.3mg/l (VN: 27-45), low PTH at 1 pg/ml (VN: 15-65) and a collapsed 25 OH vitamin D at 9 µg/L (VN: 29-50). TSHus was normal at 4 mIU/L. Blood count, blood ionogram, liver function tests and renal function tests were normal. The electroencephalogram was also normal. The cervical ultrasound showed total thyroidectomy. Characteristic ECG changes were observed, with QT and ST segment prolongation. The diagnosis of Fahr's syndrome secondary to post-total thyroidectomy hypoparathyroidism was made on the basis of these anamnestic, clinico-biological and radiological arguments. Calcium and vitamin D replacement therapy (IV calcium gluconate, vitamin D) was started. Over time, Phosphocalcic metabolism disorders and neuropsychiatric signs were corrected. The follow-up CT scan remained unchanged. The patient was discharged from hospital with a prescription for calcium combined with vitamin D (2 tablets twice daily), a calcium-rich diet and continued levothyroxine supplementation (100µg/day). Outpatient follow-up was indicated.

Discussion

Fahr's syndrome, defined by the existence of bilateral and symmetrical cerebral calcifications involving the basal ganglia (GBN), occurs preferentially in patients with dysparathyroidism, first and foremost hypoparathyroidism [3,4]. It is important to differentiate it from Fahr's disease, which is an autosomal inherited disorder with clinico-radiological similarities and no disorders of phosphocalcium metabolism. The pathophysiological mechanisms involved in the occurrence of intracerebral calcifications in Fahr's syndrome are poorly understood, and are probably multifactorial.

Most of authors suggest a metabolic disorder of the oligoglial cells with mucopolysaccharide deposits and the secondary appearance of vascular and perivascular lesions and calcareous encrustations [2,5]. These cerebral calcifications involve the small vessels of the GBN (caudate and lenticular nuclei) as illustrated in our observation, the thalamic and sometimes the dentate nuclei. Exceptionally, they may involve the white matter/grey matter junction, sometimes with asymmetry of the lateral ventricles [3,6]. Fahr's syndrome may remain asymptomatic and be discovered during cerebral radiological investigations. It may also present with polymorphous and non-specific neurological manifestations [3,4]. These are generally neuropsychiatric disorders, such as behavioural problems, or even a confusional or delusional syndrome, as in our patient. Other neurological manifestations are possible but less common, such as cognitive disorders, extrapyramidal involvement and generalised or partial seizures [3,4]. The latter were observed in our disease. The diagnostic test of choice for Fahr's syndrome is cerebral CT (computed tomography) which, as in our patient, shows intracerebral calcifications (in the form of spontaneous hyperdensities), bilateral and symmetrical, involving the basal ganglia. Injection of contrast medium was not necessary and there was no enhancement of the calcified areas. When performed, cerebral magnetic resonance imaging (MRI) reveals the presence of hyposignals on T2-weighted sequences [7]. Hypoparathyroidism is the most common cause of hypocalcaemia in Fahr's syndrome. The hypocalcaemia caused by hypoparathyroidism accounts for the majority of clinical signs (cataract, malabsorption, neuromuscular hyperexcitability, various neurological and neuropsychological signs, psychiatric disorders that can go as far as psychosis, various cardiovascular disorders) [8,9]. Hypoparathyroidism is the most frequent cause of hypocalcaemia associated with Fahr's syndrome. The hypocalcaemia caused by hypoparathyroidism accounts for the majority of clinical signs (cataracts, malabsorption, neuromuscular hyperexcitability, various neurological and neuropsychological signs, psychiatric disorders that can go as far as psychosis, various cardiovascular disorders) [8,9]. It is important not to confuse Fahr's syndrome with other conditions that can cause intracerebral calcifications, in particular endocrinopathies (hypothyroidism, hypogonadism), systemic pathologies (systemic scleroderma, systemic lupus erythematosus, celiac disease), infections (eg, celiac disease), infections (toxoplasmosis, neurocysticercosis, rubella), various diseases (chronic renal failure, vitamin D intoxication, mitochondrial cytopathies) and primary or secondary calcified brain tumours. However, the intracerebral calcifications observed in these different pathologies have different locations

and appearances [1]. In contrast to the severity of the symptoms for which it may be responsible, Fahr's syndrome has a good prognosis and correction of phosphocalcium metabolism disorders often leads to significant improvement [10].

Conclusion

Our case highlights the importance of looking for abnormalities of phosphocalcic metabolism and intracerebral calcifications in the presence of psychiatric disorders associated with neurological signs (seizures, parkinsonian syndrome, etc.), particularly in the context of total thyroidectomy. Fahr's syndrome is a rare neurological disease, characterised by severe non-specific symptoms and a simple, effective treatment.

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