

**FAHR SYNDROME IN HYPOPARATHYROIDISM SECONDARY TO PARATHYROID
GLAND SURGERY: A CASE REPORT****Najoua Belhaj^{*1}, Khalil Hjaouj¹, Hanaa Rahim¹, Razika Bencheikh², Mohammed Anass Benbouzid², Leila Essakalli Houssyni²**¹Resident physician in Otorhinolaryngology, Department of Otorhinolaryngology, Head and Neck Surgery, Ibn Sina University, Hospital, Faculty of Medicine, Mohammed V University, Rabat, Morocco.²Professor of Otorhinolaryngology, Department of Otorhinolaryngology, Head and Neck Surgery, Ibn Sina University Hospital, Faculty of Medicine, Mohammed V University, Rabat, Morocco.***Corresponding Author: Najoua Belhaj**

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ABSTRACT

Fahr syndrome is a rare anatomico-clinic disease whose most common cause is primary or postoperative hypoparathyroidism. It is characterized by bilateral and symmetrical intracerebral calcifications located in the central gray nuclei, most often associated with phosphocalcium metabolism disorders. We here report the case of a 28-year old patient who underwent surgery for two parathyroid adenomas, presenting with neuropsychiatric symptoms revealing Fahr syndrome secondary to hypoparathyroidism.

KEYWORDS: Fahr syndrome, intracerebral calcifications, postoperative hypoparathyroidism, neuropsychiatric symptoms.

INTRODUCTION

Fahr's syndrome is a rare anatomical and clinical entity, characterised by the presence of bilateral and symmetrical, non-arteriosclerotic, intracerebral calcifications.^[1]

This condition is often associated with phosphocalcic metabolic disorders, mainly due to primary or postoperative hypoparathyroidism.^[2] Fahr's triad is determined by the association of symmetrical basal ganglia calcifications, neuropsychiatric symptoms and hyperparathyroidism.^[3]

This work underlines, through the case of a patient who underwent surgery for two bilateral parathyroid adenomas in the ENT & CCF department of the Avicenne University Hospital in Rabat, the interest of monitoring the neuropsychiatric state postoperatively and considering brain imaging in the event of neuropsychiatric manifestations, in order to identify a Fahr's syndrome and thus adopt the most appropriate therapeutic measures.

OBSERVATION

Mr. B.L is a 28 years old patient with a chronic kidney disease secondary to an undetermined nephropathy,

treated with dialysis for 8 years now, and a hypertensive dilated cardiomyopathy.

The patient was admitted to our department for treatment of two bilateral parathyroid adenomas, discovered fortuitously during a routine blood test prescribed by his nephrologist.

The preoperative phosphocalcic blood tests revealed a hyperparathyroidism marked by hypercalcaemia at 112ng/l (VN: 84-102), a very high PTH level at 1260 pg/ml (VN: 5-65), alkaline phosphatase=882 IU/L (VN: 40-150) and a normal 25 OH vitamin D at 22 µg/L (VN: 9-52). The assessment of renal function showed high levels of urea=0.6 (VN: 0.15-55) and creatinine=46 (VN: 5.7-12.5). Blood count, blood serum ionogram and liver function tests were normal.

Cervical ultrasound showed two nodules, measuring 17mm in diameter each and located on the posterior aspect of the lower poles of the right and left lobes.

A parathyroid scan with MIBI-Technétium99 showed two zones of hyperfixation projecting on the two thyroid lobar bases, appearing early and persisting on the late acquisition, contrasting with an almost complete washout of the rest of the thyroid parenchyma.

The patient underwent surgery to remove both parathyroid adenomas.

The post-operative course was marked by the onset of hypocalcaemia with tremors and muscle cramps, both Chvostek and Trousseau signs were present. Subsequently, the patient presented neuropsychic manifestations such as oro-facial dyskinesia, abnormal movements, a depressive syndrome associating slow and inarticulate speech, bradypsychia, anhedonia as well as affective anesthesia.

The electrocardiogram did not show any heart rhythm disturbances.

Post-operative phosphocalcic metabolism assessment showed severe hypocalcaemia secondary to hypoparathyroidism due to surgery: Calcaemia= 41mg/l, PTH= 4pg/ml, PAL=965UI/L, 25 OH vitamin D =19ng/ml. The renal and ionogram assessments showed a normal evolution of the serum levels according to the haemodialysis sessions.

In view of the neuropsychological signs presented, a cerebral CT scan was performed, showing calcifications in the basal ganglia, which were bilateral and symmetrical.

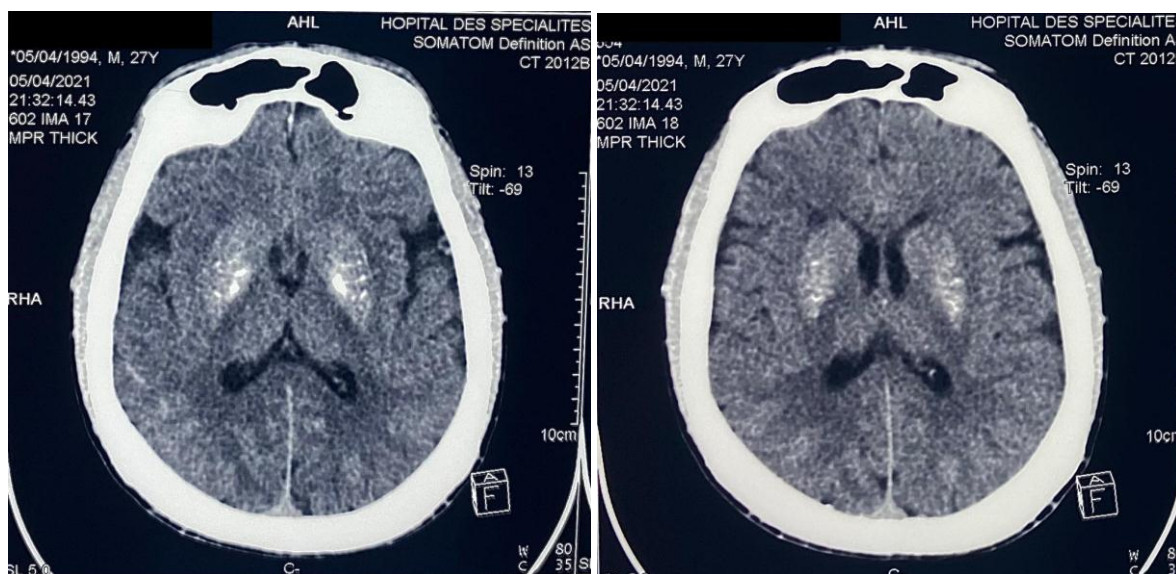


Figure 1: Axial CT sections of the thalamic region, showing hyperdensities of NGCs with bilateral and symmetrical distribution.

The diagnosis of Fahr's syndrome secondary to postoperative hypoparathyroidism was confirmed and a calcium supplementation was initiated in association with neuropsychiatric therapy.

DISCUSSION

It is a rare condition, characterized by a clinical polymorphism with a predominance of neuropsychiatric manifestations and disorders of phosphocalcic metabolism. FS occurs preferentially in patients with dysparathyroidism, mainly hypoparathyroidism.^[3-4] The pathophysiological mechanisms that contribute to the occurrence of intracerebral calcifications in FS are poorly understood.

Post-mortem studies with histochemical analysis of patients with Fahr's syndrome have found microscopic basophilic mineral deposits in the walls of small vessels of basal ganglia (Arterial, capillary and venous): calcium, iron, zinc, copper, magnesium, aluminum and potassium. While the intima of these vessels is preserved. Deposits of mucopolysaccharides constituting an organic matrix have also been identified. These

abnormalities are accompanied by minimal glial response, and varying degrees of neuronal loss. Thus, calcium is not the only element involved in the genesis of the deposits.^[5-6]

Clinically, the symptoms are rich and polymorphic, and the anatomical-clinical correlation is not always obvious. Neuropsychiatric disorders are frequently observed, such as character and/or behavioral disorders, or even a confused or delusional state. Other neurological manifestations are possible but less usual, such as cognitive disorders, mental retardation, extrapyramidal syndrome, and partial or generalized seizures, more rarely cerebellar or pyramidal syndrome, intracranial hypertension syndrome, cranial nerve damage or chorea. FS may remain asymptomatic. Manifestations can sometimes be limited to signs of hypocalcaemia (tetany, Chvostek's sign, and Trousseau sign).^[5-8]

Hypoparathyroidism remains the most common cause of FS, whether primary or postoperative. The biological diagnosis is made on the basis of the combination of these following characteristic: hypocalcaemia, decreased

serum PTH level, hyperphosphoremia, hypocalciuria and hypophosphaturia.

Before the advent of digital CT, the diagnosis of FS was based on skull X-rays and autopsy. Currently, the CT scan is the examination of choice for the detection of intracerebral calcifications. The most frequent locations are the striated nuclei, the thalamus, the dentate nuclei and the semiautomatic center. A multitude of pathologies can cause intracerebral calcifications: endocrinopathies (hypothyroidism), infections (toxoplasmosis), systemic (systemic lupus erythematosus), degenerative, congenital and tumoral pathologies..., where calcifications are not bilateral, symmetrical, nor localized to the basal ganglia UCS as observed in FS.^[7]

In contrast to the severity of the symptoms for which it may be responsible, FS has a good prognosis and correction of the disorders of phosphocalcic metabolism often leads to significant improvement.

CONCLUSION

FS is a rare neurological disease, contrasting a severe non-specific symptomatology with a simple and effective treatment. The correction of phosphocalcic metabolism disorders allows a marked improvement of the clinical symptoms, hence the interest of a systematic search for these disorders before any neuropsychiatric manifestation after parathyroid gland surgery.

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