1 | INTRODUCTION

Fahr syndrome combines bilateral and symmetrical nonarteriosclerotic intracerebral calcifications, with phosphocalcic metabolic disturbances due primarily to hypoparathyroidism.\(^1,2\)

Diagnosis requires a brain scan and treatment is often symptomatic based on the improvement of the calcium metabolism.\(^3\)

2 | CASE REPORT

A 45-year-old male patient with no pathological history was admitted to the operation room for treatment of a lumbar herniated disk.

The preoperative evaluation was normal: The patient was conscious with GCS 15/15 with no sensitivity-motor deficit, blood pressure at 125/72 mm Hg and heart rate at 82. The respiratory examination found the patient eupneic at 18 and with ambient air saturation at 98%, he was classified ASA I with metabolic equivalent of task (MET) >7.

For induction, 200 mg of propofol, 25 µg of sufentanil, and 40 mg of rocuronium were given. Sevoflurane with minimal alveolar concentration at 2.5% was used for anesthesia maintenance. The monitoring of the anesthesia depth was provided by bispectral index (BIS), and it varied between 35 and 65, and at the end of surgery between 55 and 78.

The patient was placed in ventral decubitus position. The surgery lasted for 2 hours, and the per-operative course was uneventful.

Once sedation was stopped, the patient showed no signs of awakening despite antagonization with 0.4 mg of naloxone. He was admitted to the intensive care unit. In the postoperative period, analgesia and sedation were provided by propofol at the dose of 2 mg/kg/h and morphine 2 mg/h for 48 hours. The biological assessment showed a pseudohypoparathyroidism with hypocalcemia at 50 mg/L, hyperphosphoremia at 56 mg/L, and a normal serum PTH level at 34 pg/mL. Table 1 presents the main biological results.

A brain CT was performed and showed bilateral and symmetrical calcifications of the central gray nuclei (Figure 1) corresponding to Fahr syndrome. Replacement therapy with
calcivitamin D was started (calcium 2 g per day and vitamin D2 1500 UI per day).

The outcome was favorable, and the patient was extubated on the third day following his admission.

3 | DISCUSSION

Fahr syndrome is a condition rarely reported in the literature. It is most often due to disturbances of the phosphocalcic metabolism. It is to be distinguished from Fahr’s disease, which corresponds to intracerebral calcium deposits of idiopathic origin that may be hereditary or sporadic.4

Is usually occurs in patients with dysparathyroidism, mainly hypoparathyroidism.5

Postmortem studies with histochemical analysis of patients with FS have found microscopic basophilic mineral deposits in the vascular walls (arterial, capillary, and venous): calcium, iron, magnesium, zinc, copper, potassium, and aluminum.6

The pathophysiological mechanism behind these deposits remains poorly elucidated. Most authors suggest a metabolic disorder of oligogial cells with muco-polysaccharide deposits and secondary appearance of vascular, perivascular lesions and calcareous encrustations. In Fahr’s disease, some authors suggest an exaggeration of a normal process of calcium or ferrous deposits in the gray central and serrated nuclei.7

| TABLE 1  The main biological results |
|-----------------------------|--------|
| Hemoglobin (g/dL)          | 12.5   |
| Platelet (G/L)             | 195    |
| Natremia (mmol/L)          | 137    |
| Kalemia (mEq/L)            | 4.1    |
| Creatinine (μmol/L)        | 9      |
| Urea (mmol/L)              | 0.2    |
| Glycemia (g/L)             | 1.4    |
| Albumin (g/dL)             | 4.2    |

FIGURE 1 Nonenhanced brain CT showing calcifications of the caudate nuclei, lenticular nuclei, and thalami

Fahr syndrome is generally difficult to suspect clinically because it can remain asymptomatic or result in polymorphic manifestations that do not correspond to a specific pattern.8 The clinical signs that can be encountered are convulsions, dementia, syncope, vertigo, dysarthria, headache, orthostatic hypotension, and neuropsychiatric disorders such as hallucinations, delusions, anxiety, mania, and depression. Sometimes FS associates with cerebrovascular diseases and myopathies.9,10

Before the advent of CT scan, the diagnosis of FS was based on X-rays of the skull and autopsy. Today, brain CT is the investigation of choice for the detection of intracerebral calcifications. The most common locations are striated nuclei, thalamus, serrated nuclei, and semi-oval center.6

MRI, which remains of limited value in the context of FS, usually shows hypersignals in T1 and T2; however, the presence of a T1 hyposignal is possible.11

Other causes can be at the origin of intracerebral calcifications such as endocrinopathies (hypothyroidism, hypogonadism...), systemic diseases (systemic scleroderma, systemic lupus erythematosus), celiac disease, infections (rubella, toxoplasmosis...), and primary or secondary calcified brain tumors. However, during these different pathologies, intracerebral calcifications have different places and aspects.12

Calcivitamin D therapy is the basis for the treatment of FS. Its outcome is favorable because clinical signs regress after once the phosphocalcic disturbances are corrected.6,13

4 | CONCLUSION

Fahr syndrome is a rare clinico-radiological entity. Its pathophysiological mechanism remains poorly elucidated. Intracerebral calcifications must be sought in the setting of phosphocalcic metabolism disorders, especially when associated with neurological or endocrine pathologies. The correction of those disorders leads to a marked improvement in clinical symptoms.

CONFLICT OF INTEREST
The authors do not declare any conflict of interest.

AUTHOR CONTRIBUTIONS
All authors have read and approved the final version of the manuscript.

PATIENT’S PERSPECTIVE
The patient expressed her satisfaction with the treatment.

ORCID
Yassine Mellagui https://orcid.org/0000-0002-4785-5116
REFERENCES

1. Rhouda H, Gaouzi A, Kriouile Y. Signes neuropsychologiques chez l’enfant: penser au syndrome de Fahr. Neuropsychiatrie de l’enfance et de l’adolescence. 2019;67(2):75-80.

2. El Hechmi S, Bouhlel S, Melki W, El Hechmi Z. Psychotic disorder induced by Fahr's syndrome: a case report. L’Encéphale. 2014;40(3):271-275.

3. Calili DK, Mutlu NM, Titiz APM, Akcaboy ZN, Aydin EM, Turan IO. Unexplained neuropsychiatric symptoms in intensive care: a Fahr syndrome case.

4. Saddoud N, Remili H, Jebali I, Moueddeb S, Gmiha B, Daghfous MH. Syndrome de Fahr secondaire à une hypoparathyrédie: à propos de deux cas. J Neuroradiol. 2017;44(2):101.

5. El Hechmi S, Bouhlel S, Melki W, El Hechmi Z. Trouble psychotique secondaire à un syndrome de Fahr: à propos d’une observation. Encephale. 2014;40(3):271-275.

6. Rafai MA, Oumari S, Lytim S, Boulaajaj FZ, El Moutawakkil B, Slassi I. Le syndrome de Fahr: aspects cliniques, radiologiques et étiologiques. Feuill Radiol. 2014;54(1):59.

7. Doumbia M, Kouassi L, Kouame-Assouan AE, Douayoua-Sonan TH, Boa-Yapo F. Maladie de Fahr révélée par des troubles de la marche et de la parole. Rev Int Sci Med. 2006;8:32-35.

8. Ait El Hadj H, Kandri Rody K, Oujennane K, Akhdari N, Amal S, Hocar O. Syndrome de Fahr à révélation dermatologique : deux cas. Ann Dermatol Vénérol. 2019;146(12):A204-A205.

9. Park S, Jee DL, Kim H. General anesthesia for patient with Fahr's syndrome: a case report. Medicine. 2019;98(17):e15390.

10. Jaworski K, Styczyńska M, Mandecka M, Walecki J, Kosior DA. Fahr syndrome – an important piece of a puzzle in the differential diagnosis of many diseases. Pol J Radiol. 2017;82:490-493.

11. Casanovaa MF, Araquec JM. Mineralization of the basal ganglia: implications for neuropsychiatry, pathology and neuroimaging. Psychiatry Res. 2003;121:59-87.

12. Khammassi N, Chrif J, Mohsen D, Abdelhedi H, Tougourtis MN, Hamza M. Fahr’s syndrome: two case report. Rev Neurol. 2010;166(4):446-450.

13. Rharrabti S, Darouch I, Benbrahim M, Belahsen F, Rammouz I, Alouane R. Un syndrome confusionnel révélant un syndrome de Fahr avec hyperparathyrédie. Pan Afr Med J. 2013;14:123.

How to cite this article: Mellagui Y, Aabdi M, Ouachaou J, Bkiyar H, Housni B. Delayed awakening from general anesthesia revealing Fahr syndrome: Case report and literature review. Clin Case Rep. 2020;8:3336–3338. https://doi.org/10.1002/ccr3.3183