

Fahr Syndrome Presenting in the Emergency Department: A Series of Eight Cases and a Review of the Literature

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Abstract

Case Series

Introduction: Fahr syndrome (FS) is characterised by bilateral, symmetrical, non-arteriosclerotic calcifications of the basal ganglia. It is a rare condition with heterogeneous clinical manifestations and is most commonly associated with parathyroid disorders. **Patients and methods:** We conducted a retrospective descriptive study of eight patients admitted to the medical and surgical emergency department of Mohammed V Military Teaching Hospital over a nine-year period, all of whom had basal ganglia calcifications on brain computed tomography (CT). **Results:** The mean age was 52 years (range, 39–60 years), and six patients were women. Clinical manifestations were non-specific and included seizures (n = 4), cognitive impairment (n = 3), altered consciousness (n = 3), psychiatric symptoms (n = 2), and signs of hypocalcaemia (n = 2). FS was detected incidentally in three patients during the evaluation of ischaemic stroke, haemorrhagic stroke, and shock, respectively. Abnormalities of calcium and phosphate metabolism were identified in seven patients, with hypoparathyroidism in four (including two following thyroidectomy) and pseudohypoparathyroidism in three. Electroencephalography, performed in four patients, showed background slowing or diffuse cerebral dysfunction in two, ictal activity in one, and normal findings in one. Calcium and vitamin D supplementation was initiated in seven patients, and six patients had a favourable clinical outcome. Two patients died of causes not directly related to FS. **Conclusion:** Bilateral, symmetrical basal ganglia calcifications should prompt a systematic evaluation for an underlying parathyroid disorder. Correction of the associated metabolic abnormalities usually results in a favourable clinical outcome.

Keywords: Fahr syndrome; basal ganglia calcification; hypoparathyroidism; pseudohypoparathyroidism; emergency department.

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INTRODUCTION

First described by Theodor Fahr in 1930, Fahr syndrome is radiologically characterised by bilateral, symmetrical, non-arteriosclerotic striatopallidodentate calcifications [1,2]. A distinction is traditionally made between Fahr disease, a primary, idiopathic or familial form, and Fahr syndrome, a secondary form predominantly associated with disorders of calcium and phosphate metabolism [2].

This rare condition has an incompletely understood pathophysiology and a broad spectrum of clinical manifestations, making diagnosis challenging, particularly in the emergency setting.

The aim of this study was to describe the clinical, radiological and aetiological characteristics, as

well as the outcomes, of eight patients with FS managed in an emergency department.

PATIENTS AND METHODS

We conducted a retrospective descriptive study of eight patients with FS managed in the medical and surgical emergency department of Mohammed V Military Instruction Hospital over a nine-year period. Diagnosis was based on the detection of bilateral, symmetrical basal ganglia calcifications on brain CT.

Data collected for each patient included demographic characteristics, medical history, clinical presentation, brain CT findings, calcium and phosphate metabolism assessment, electrocardiography (ECG), electroencephalography (EEG, performed in four patients), treatment and clinical outcome. All data were anonymised.

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RESULTS

Demographic characteristics and medical history:

The mean age was 52 years (range, 39–60 years). Six patients were women and two were men, indicating a female predominance. No family history of FS was identified. Two patients had undergone thyroidectomy, two had a history of dementia, and one had epilepsy treated with sodium valproate for more than ten years.

Clinical presentation: Clinical manifestations were heterogeneous, with no specific clinical pattern. Neuropsychiatric manifestations predominated and included:

- generalised tonic–clonic seizures: four patients;
- cognitive impairment, including intellectual decline and confusion: three patients;
- altered consciousness: three patients;
- pyramidal signs, including hemiparesis or hemiplegia, in the setting of stroke: two patients;
- extrapyramidal manifestations, including tremor, abnormal movements and parkinsonism: one patient;
- psychiatric symptoms, including anxiety and irritability: two patients;
- signs of hypocalcaemia, including tetany and positive Chvostek and Trousseau signs: two patients.

FS was detected incidentally in three patients undergoing brain imaging for ischaemic stroke, left thalamic and capsulolenticular intracerebral haemorrhage, and shock, respectively.

Imaging:

Brain CT demonstrated bilateral, symmetrical basal ganglia calcifications in all patients (Figure 1), with additional cerebellar calcifications in one patient.

Laboratory investigations and ancillary testing:

Abnormalities of calcium and phosphate metabolism were identified in seven patients, with hypoparathyroidism in four (including two postoperative cases), and pseudohypoparathyroidism in three. ECG showed QT interval prolongation in three patients and was normal in the remaining patients. Among the four patients who underwent EEG, findings were normal in one; the remaining recordings showed non-specific diffuse cerebral dysfunction in one, background slowing without an epileptiform focus in one, and ictal activity in one.

Treatment and outcomes:

Calcium and vitamin D supplementation was initiated in seven patients, with additional antiseizure medication in two. Six patients had a favourable clinical outcome, with resolution of seizures, improvement in neurological status and normalisation of follow-up EEG findings. A residual motor deficit remained in one patient. Two women died of causes not directly related to FS: septic shock secondary to ventilator-associated pneumonia (VAP) in one, and multiple organ dysfunction syndrome (MODS) following shock associated with intrauterine fetal demise in the other. Individual patient characteristics are summarised in Table 1.

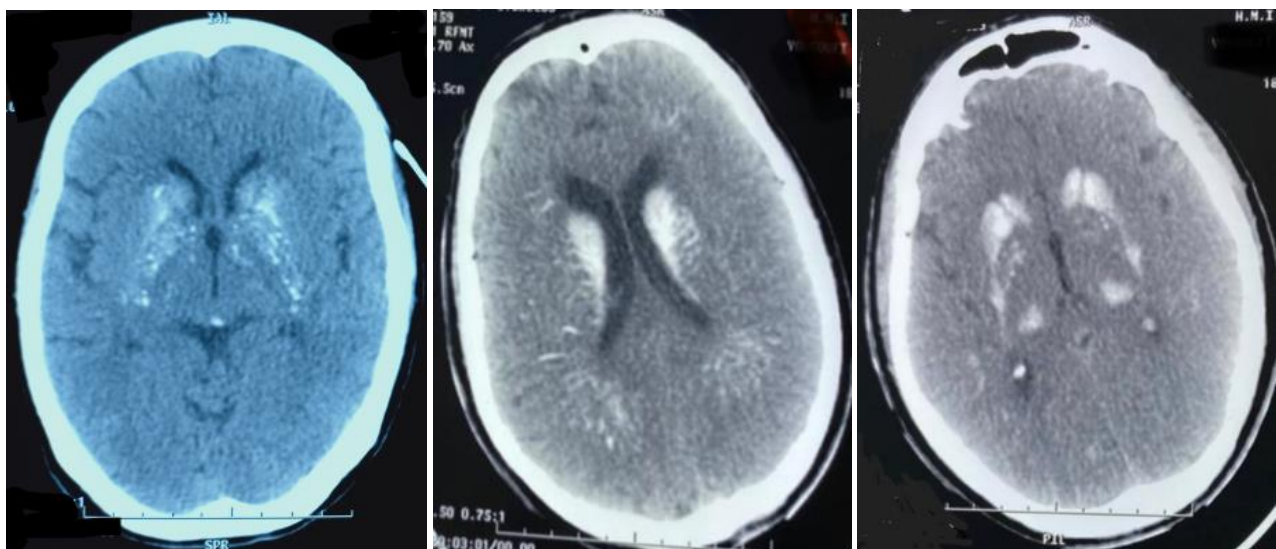


Figure 1: Non-contrast brain CT showing bilateral, symmetrical basal ganglia calcifications

Table 1: Clinical, radiological, biochemical and outcome characteristics of the patients

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7	Case 8
Age (years)	54	51	55	53	57	48	60	39
Sex	F	M	F	M	F	F	F	F
Medical history	Dementia; receiving Lexomil® and Zepam®	Epilepsy treated with sodium valproate (>10 years)	Thyroidectomy	Dementia	None reported	None reported	Thyroidectomy	None reported
Clinical presentation	Postictal coma; anxiety; confusion	Cognitive impairment; GTCS; right hemiparesis; facial palsy; positive Chvostek and Trousseau signs	GTCS; cognitive impairment	Tremor; abnormal movements; parkinsonism	Irritability; anxiety; GTCS	Altered consciousness; right hemiplegia	Altered consciousness; tetany	Shock associated with intrauterine fetal demise
Brain CT	BGC	BGC; left-sided hypodensity (IS)	BGC	BGC	BGC	BGC; left thalamic and capsulolenticular ICH	BGC; cerebellar calcifications	BGC
Calcium and phosphate metabolism	Hypoparathyroidism	PHP	Postoperative hypoparathyroidism	Hypoparathyroidism	PHP	PHP	Postoperative hypoparathyroidism	Normal
Treatment	Calcium and vitamin D supplementation	Calcium and vitamin D supplementation; anticoagulant; antiplatelet agent	Calcium and vitamin D supplementation	Calcium and vitamin D supplementation; sodium valproate; BZD	Calcium and vitamin D supplementation	Calcium and vitamin D supplementation; management of ICH	Calcium and vitamin D supplementation	Fetal evacuation
Outcome	Neurological improvement	Resolution of seizures	Resolution of seizures	Resolution of seizures; return to baseline GCS	Resolution of seizures	Death (septic shock secondary to VAP)	Neurological improvement	Death (MODS)

Abbreviations: BGC, basal ganglia calcifications; BZD, benzodiazepine; CT, computed tomography; GCS, Glasgow Coma Scale; GTCS, generalised tonic-clonic seizure; ICH, intracerebral haemorrhage; IS, ischaemic stroke; MODS, multiple organ dysfunction syndrome; PHP, pseudohypoparathyroidism; PTH, parathyroid hormone; VAP, ventilator-associated pneumonia. F, female; M, male. "None reported" indicates no relevant medical history reported in the source manuscript.

DISCUSSION

FS is a rare clinicopathological entity characterised by bilateral, symmetrical, non-arteriosclerotic basal ganglia calcifications, most often occurring in patients with parathyroid disorders [3].

The pathophysiology of striatopallidodentate calcinosis remains incompletely understood. Studies have demonstrated basophilic mineral deposits within the arterial, capillary and venous walls of the basal ganglia. These deposits contain calcium, iron, zinc, copper, magnesium, aluminium and potassium, together with an organic mucopolysaccharide matrix. These abnormalities are accompanied by a minimal glial reaction and variable degrees of neuronal loss [4,5]. Most authors attribute these deposits to a metabolic disturbance of oligodendroglial cells, followed by

vascular and perivascular injury and calcium deposition [6].

Such deposit within small-vessel and capillary walls may reduce tissue perfusion and contribute to cerebral ischaemia [7,8], or weaken vessel walls and predispose to haemorrhagic stroke. In our series, FS was identified during the evaluation of stroke in two patients: one with ischaemic stroke and the other with haemorrhagic stroke.

Long-term use of antiseizure medications may interfere with calcium and phosphate metabolism by inducing vitamin D deficiency; phenytoin may also impair intestinal calcium absorption. These effects could potentially contribute to basal ganglia calcification [7,9]. This mechanism, described mainly with enzyme-inducing antiseizure medications, remains hypothetical

in our patient receiving sodium valproate, particularly given the concomitant pseudohypoparathyroidism.

FS is difficult to suspect clinically because its manifestations do not conform to a specific clinical pattern. It may remain asymptomatic or be detected incidentally on imaging performed for another indication [10], as occurred in three of our patients presenting with ischaemic stroke, haemorrhagic stroke and shock, respectively.

Neuropsychiatric manifestations predominate and include personality and behavioural changes, cognitive impairment, intellectual disability and seizures, which may occasionally be mistaken for episodes of tetany. Extrapyramidal manifestations and, less commonly, pyramidal signs may also occur [4,6]. QT interval prolongation, observed in three of our patients, is consistent with hypocalcaemia and supports routine ECG assessment because of the risk of cardiac arrhythmias.

The diagnosis is established by brain CT demonstrating bilateral, symmetrical basal ganglia calcifications.

Parathyroid disorders are the most common underlying causes, particularly primary or postoperative hypoparathyroidism [6], characterised by hypocalcaemia, hyperphosphataemia and reduced parathyroid hormone (PTH) levels. In our series, hypoparathyroidism (four patients, including two following thyroidectomy) and pseudohypoparathyroidism (three patients) accounted for all identified aetiologies. No calcium or phosphate metabolism abnormality was detected in one patient.

The differential diagnosis includes other causes of intracerebral calcification, such as endocrine, systemic, degenerative and neoplastic disorders. In these conditions, calcifications are generally neither bilateral nor symmetrical, in contrast to the findings in our patients [4,7].

FS may present with diverse and occasionally severe manifestations, despite a relatively straightforward and effective treatment. Correction of calcium and phosphate metabolism abnormalities often leads to substantial symptomatic improvement, accounting for the generally favourable prognosis reported in FS [4,5]. In our series, both deaths were attributed to intercurrent conditions, namely sepsis and obstetric shock, rather than FS itself.

The limitations of this study include its retrospective, single-centre design, small sample size and the absence of EEG assessment in half of the patients.

CONCLUSION

FS is a rare clinicopathological entity that should be considered in patients presenting to the emergency department with seizures, unexplained neuropsychiatric symptoms or signs of hypocalcaemia, particularly following thyroidectomy. Its identification should prompt investigation for an underlying parathyroid disorder, allowing appropriate treatment, which usually leads to a favourable clinical outcome.

Declaration of competing interests: The authors declare no competing interests related to this article.

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Consent and ethics: Data were collected retrospectively in anonymised form, with due regard for patient confidentiality.

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