International Journal of Contemporary Pediatrics
Siddiqui A et al. Int J Contemp Pediatr. 2026 May;13(5):770-773
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291
DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20261144

Case Report

Fahr's syndrome secondary to idiopathic hypoparathyroidism: a report of two adolescent cases

Anam Siddiqui*, Harveen Kaur, Jasmine Singh, Chandrika Azad

Department of Pediatrics, Government Medical College and Hospital, Chandigarh, Punjab, India

Received: 25 January 2026
Revised: 05 March 2026
Accepted: 06 April 2026

***Correspondence:**
Dr. Anam Siddiqui,
E-mail: doll.anamsiddiqui786@gmail.com

ABSTRACT

Fahr's syndrome is a rare neurological disease characterized by calcification of basal ganglia and/or other areas of brain controlling motor activity. It occurs secondary to an identifiable condition, most commonly hypoparathyroidism. Hypoparathyroidism is an uncommon endocrinopathy with a wide range of presentations. We describe two adolescent cases presenting with seizures, neuropsychiatric manifestations and extra-pyramidal symptoms with bilateral basal ganglia calcification on neuroimaging. Both had hypo-calcemia, hyper-phosphatemia and low parathyroid levels. Vitamin D and calcium supplementation with or without phosphate binders is the mainstay of treatment with limited role of antiepileptics. Despite being rare, clinicians need to consider Fahr's syndrome as a differential in such cases. Good response is seen after treatment of underlying hypo-parathyroidism.

Keywords: Fahr's, Hypo-parathyroidism, Basal ganglia, Calcification, Seizures

INTRODUCTION

Hypoparathyroidism is an uncommon endocrinopathy characterized by a parathyroid hormone (PTH) deficient state resulting in hypocalcemia, hyperphosphatemia, and hypercalciuria. The clinical presentation is mainly attributable to these biochemical derangements leading to increased neuromuscular excitability and heterotopic calcification of soft tissues.[1] Although rare, basal ganglia calcification can occur in patients with hypo-parathyroidism defined as Fahr's syndrome. Fahr's disease is distinguished by familial idiopathic calcification of basal ganglia while Fahr's syndrome refers to cases with an underlying etiology for the basal ganglia calcification. Hypoparathyroidism is the most common etiology of Fahr's syndrome.[2-4]

In this case series, we describe two adolescents who presented with seizures, neuropsychiatric manifestations and extra-pyramidal symptoms with basal ganglia calcification secondary to hypoparathyroidism and improved remarkably after calcium and vitamin D therapy.

CASE REPORTS

Case 1

A twelve years old developmentally normal adolescent female, born out of non-consanguineous marriage with smooth perinatal transition, presented to our center with the chief complaint of multiple episodes of agitation with jerky movements of bilateral upper and lower limbs followed by mutism lasting for 3-4 hours for the last 15 days. These episodes were not associated with up rolling of eyeballs, frothing from the mouth, bowel/bladder incontinence, or any altered consciousness. The patient was diagnosed with epilepsy 2 years back and was seizure free for the last 2 years on oral phenytoin (5 mg/kg/day) and valproate (20 mg/kg/day). There was no history of involuntary movements, tingling sensation in limbs, jaundice, neck surgery, or radiation exposure. There was no history of seizures or similar complaints in family. At admission, vitals were stable and there were no cutaneous lesions, dysmorphism or short stature. Gum hypertrophy was present. Chvostek's sign was negative. Fundus

examination showed grade one papilledema. The higher mental functions were intact with no cranial nerve deficits. Motor and sensory systems examination was normal. The rest of the systemic examination was non-contributory. Laboratory investigations were suggestive of hypo-calcemia, hyper-phosphatemia, hypo-parathyroidism and vitamin D insufficiency [serum calcium: 1.52 mmol/l (2.0-2.59 mmol/l), ionized calcium: 0.77 mmol/l (1.15-1.3 mmol/l), serum phosphate: 2.52 mmol/l, (0.81-1.45 mmol/l), 24-hour urinary calcium: 5.0 mmol/d (2.7-7.5 mmol/d), and intact PTH levels in serum: 11.0 ng/l (12- 80 ng/l), 25(OH)-D levels: 18 ng/ml (30-100 ng/ml)]. Renal and liver function tests, thyroid function tests (TFT), HbA1c, serum cortisol, serum ferritin, serum magnesium, serum ceruloplasmin, and 24-hour urinary copper were normal. Electrocardiogram (ECG) showed a QTc interval of 0.48 sec. The electroencephalogram (EEG) came out to be normal. Non-contrast computed tomography (NCCT) head revealed bilateral symmetrical hyperintensities in basal ganglia suggestive of calcifications (Figure 1A). Hence, the patient was finally diagnosed as a case of Fahr's syndrome secondary to hypo-parathyroidism. He was given intravenous (IV) calcium gluconate therapy followed by oral supplementation with calcium carbonate 500 mg BD and calcitriol 0.25 microgram per day in two divided doses. There was no recurrence of the symptoms thereafter and serum levels of calcium and phosphate normalized. Phenytoin was replaced with levetiracetam in view of toxicity in the form of gum hypertrophy. At six months follow up, child has not experienced any recurrence of seizures and tapering of anti-epileptics (AEDs) has been initiated.

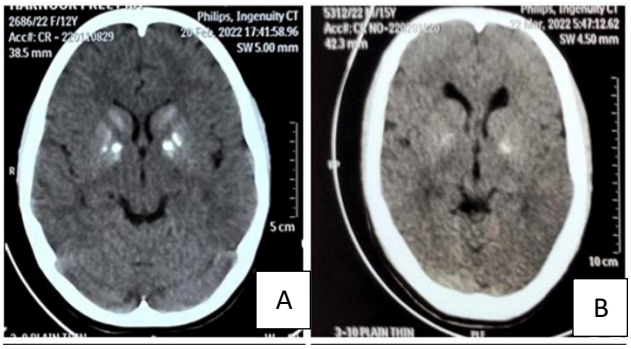

Figure 1 (A and B): NCCT head showing hyperdensities suggestive of calcifications in bilateral basal ganglia involving head of caudate nucleus and lentiform nucleus.

Case 2

A 15 years old developmentally normal boy, with no parental consanguinity with uneventful birth history, presented to our center with the chief complaint of two episodes of abnormal body movements in the form of jerky movements of bilateral upper and lower limbs with up rolling of eyeballs with each episode lasting for around 2-3 minutes followed by regaining of consciousness with no residual neurological deficits. There was no preceding history of fever, vomiting or loose stools. There was no history of excessive twitching, tingling sensation in limbs, jaundice, neck surgery, or radiation exposure. Family history was insignificant. At admission, vitals were stable without any cutaneous lesions, dysmorphism or short stature. Chvostek's sign was negative. Higher mental functions were intact with no cranial nerve deficits. Motor and sensory systems examination was normal. The rest of the systemic examination was non-contributory. Laboratory investigations were suggestive of hypo-calcemia, hyper-phosphatemia, hypo-parathyroidism and vitamin D insufficiency [serum calcium: 1.50 mmol/l (2.0-2.9 mmol/l), ionized calcium: 0.53 mmol/l (1.15-1.3 mmol/l), serum phosphate: 2.29 mmol/l (0.81-1.45 mmol/l), 24-hour urinary calcium: 5.6 mmol/d (2.7-7.5 mmol/d), and intact PTH levels in serum: 0.0 ng/l (12-80 ng/l), 25(OH)-D levels: 16.3 ng/ml (30-100 ng/ml)]. Investigations revealed normal renal, liver and thyroid function tests. Serum levels of electrolytes, cortisol, ferritin, magnesium, ceruloplasmin and calcium were within normal limits. 24-hour urinary copper was normal. ECG showed a QTc interval of 460 msec. EEG was normal. NCCT head revealed hyperintensities in bilateral basal ganglia(left>right) suggestive of calcifications (Figure 1B). Hence, diagnosed with Fahr's syndrome secondary to hypo-parathyroidism was made. She was also treated with given IV calcium gluconate therapy followed by oral supplementation with calcium carbonate 500 mg BD and calcitriol 0.25 microgram per day in two divided doses and sevelamer carbonate 1000 mg TDS. For seizures, IV phenytoin was started and hiked up to 6.5 mg/kg/day followed by IV valproate @20 mg/kg/day. During the hospital stay, the patient was noted to have dystonia for which oral trihexyphenidyl was started. She was shifted to oral AEDs and discharged with normal serum calcium levels with no fresh seizures. AEDs are being tapered on follow-up visits with no recurrence of seizures.

DISCUSSION

Hypoparathyroidism is known to have varied clinical presentation attributable to hypo-calcemia, hyperphosphatemia, hypercalciuria leading to increased neuromuscular excitability and heterotopic calcification. Basal ganglia calcification occurs in hypoparathyroid patients with hyperphosphatemia as seen in both the cases described.[1] Fahr's syndrome is rare neurological condition characterized by progressive neuropsychiatric symptoms and/or extrapyramidal features in the presence of calcifications in areas of brain controlling motor movement especially basal ganglia and/or dentate nucleus of cerebellum, sub-cortical white matter , corona radiata or thalamus. It can be a result of conditions such as endocrinopathies e.g., hypoparathyroidism (idiopathic or secondary), pseudohypoparathyroidism, intrauterine or perinatal infections, brucella infection, Kenny-Caffey syndrome type 1, neuroferritinopathies, mitochondrial myopathy or tuberous sclerosis complex.[2] Hypoparathyroidism is the most common etiology of

Fahr's syndrome. In our case series, both cases had underlying idiopathic hypoparathyroidism. Causes for secondary hypoparathyroidism were ruled out.[1]

Patients with Fahr's syndrome can present with headache, seizures, stroke-like events, confusion, changes in personality, reduced memory and movement disorders such as ataxia, chorea, or dystonia, anxiety, and psychosis.[2,5] Lopez-Vilegas et al reported a variety of neurological symptoms, depending on the specific regions of the basal ganglia affected by calcification, in 18 patients with Fahr's syndrome.[6] Our first case had personality changes with mutism and chorea while our second case had seizures and dystonias. Basal ganglia calcification has been reported in 73.8% in a cohort of patients with idiopathic hypo-parathyroidism by Goswami et al.[7] In this cohort, progression of basal ganglia calcification was related to the calcium: phosphorus ratio during follow-up, hence highlighting the importance of phosphorus control in such patients. Also, clinicians must remember that despite seemingly akin clinical and radiological presentation, Fahr's disease is a separate entity with bilateral symmetrical idiopathic basal ganglia calcification usually with autosomal dominant or rarely sporadic inheritance. In the absence of any identifiable cause, Fahr's disease lacks an effective treatment, whereas patients with Fahr's syndrome show significant improvement after correcting the primary etiology.[2]

Oral calcium and vitamin D supplementation remain the mainstay of treatment for hypoparathyroidism. Patients presenting with signs and symptoms of acute hypocalcemia or with total serum calcium below 1.9 mmol/l require treatment with IV 10% calcium gluconate 1-2 ml per kg as slow IV infusion under cardiac monitoring. If hypocalcemia persists, IV calcium gluconate boluses or continuous infusion might be needed. After resolution of acute symptoms, 30-75 mg of elemental calcium/kg/day in divided doses is given orally in children and adolescents. Vitamin D is given in its active form i.e., calcitriol, in a dose of 20 to 60 ng/kg/day along with parent vitamin D. Importantly, calcium levels must be maintained at the lower limit of normal to prevent hypercalciuria and nephrocalcinosis. Also, serum calcium-phosphate product must remain <4.4 $mmol^2/L^2$.[1,8] High phosphate levels may require phosphate binders also as seen in the second case. Symptomatic treatment has to be individualized and includes antipsychotics and antidepressants for psychiatric manifestations, AEDs for seizures and trihexyphenidyl for dystonias.[7]

In their study including children who presented with seizures as the first manifestation of hypoparathyroidism, Liu et al did not see any improvement with addition of AEDs to calcium supplementation.[9] This finding was consistent even in patients with sub-cortical calcifications. However, seizure suppression could be significantly correlated with normalization of serum calcium levels. In addition, most patients in this study were taking anticonvulsants like phenytoin sodium, valproic acid, or carbamazepine which are detrimental to serum calcium levels.[8] Modi et al studied a cohort of patients with seizures due to idiopathic hypoparathyroidism. In their study, 71% of the patients were successfully withdrawn from anti-epileptic therapy with no seizure recurrence and withdrawal of AEDs led to significant increase in serum calcium concentration.[10] Again stressing the importance of normalising calcium levels rather than chasing anticonvulsants for seizures in hypo-parathyroid patients. Therefore, authors suggest careful use of AEDs in parathyroid disorders as they might be ineffective or even detrimental.

CONCLUSION

The above two cases highlight the importance of screening for hypo-calcemia as the cause of seizure in children. Any child with hypo-calcemia and hyperphosphatemia must undergo PTH levels. Despite its rarity, Fahr's syndrome must be kept as a differential in cases presenting with seizures, neuropsychiatric manifestations and/or extrapyramidal features with basal ganglia calcification on neuroimaging. Hypoparathyroidism can have varied presentation and it is the most common cause of Fahr's syndrome. Cases of hypoparathyroidism must be treated efficiently with calcium and vitamin D and followed up. Anticonvulsants have a limited role in these cases.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Al-Azem H, Khan AA. Hypo-parathyroidism. Best Pract Res Clin Endocrinol Metabolism. 2012;26(4):517-22.
2. Zhou YY, Yang Y, Qiu HM. Hypo-parathyroidism with Fahr's syndrome: A case report and review of the literature. World J Clin Cases. 2019;7(21):3662-70.
3. Saleem S, Aslam HM, Anwar M, Anwar S, Saleem M, Saleem A, et al. Fahr's syndrome: literature review of current evidence. Orphanet J Rare Dis. 2013;8:156.
4. Brandi ML, Bilezikian JP, Shoback D, Bouillon R, Clarke BL, Thakker RV, et al. Management of Hypoparathyroidism: Summary Statement and Guidelines. J Clin Endocrinol Metab. 2016;101:2273-83.
5. El Aoud S, Elleuch M, Charfi N, Hadj Kacem F, Mnif M, Rekike N, et al. Fahr's syndrome in Southern Tunisia: A broad spectrum of clinical and etiological features. Tunis Med. 2018;96:490-4.
6. López-Villegas D, Kulisevsky J, Deus J, Junqué C, Pujol J, Guardia E, et al. Neuropsychological alterations in patients with computed tomography-detected basal ganglia calcification. Arch Neurol. 1996;53(3):251-6.

7. Goswami R, Sharma R, Sreenivas V, Gupta N, Ganapathy A, Das S. Prevalence and progression of basal ganglia calcification and its pathogenic mechanism in patients with idiopathic hypo-parathyroidism. Clin Endocrinol (Oxf). 2012;77:200-6.
8. Maio SD, Soliman AT, Sanctis VD, Kattamis C. Current treatment of hypoparathyroidism: theory versus reality waiting guidelines for children and adolescents. Acta Biomed. 2018;89(1):122-31.
9. Liu MJ, Li JW, Shi XY, Hu LY, Zou LP. Epileptic seizure, as the first symptom of hypo-parathyroidism in children, does not require antiepileptic drugs. Childs Nerv Syst. 2017;33:297-305.
10. Modi S, Tripathi M, Saha S, Goswami R. Seizures in patients with idiopathic hypoparathyroidism: effect of antiepileptic drug withdrawal on recurrence of seizures and serum calcium control. Eur J Endocrinol. 2014;170:777-83.

Cite this article as: Siddiqui A, Kaur H, Singh J, Azad C. Fahr's syndrome secondary to idiopathic hypoparathyroidism: a report of two adolescent cases. Int J Contemp Pediatr 2026;13:770-3.