Sir,

Paroxysmal dyskinesias are rare movement disorders, which include paroxysmal choreoathetosis and paroxysmal dystonic choreoathetosis. They are mostly idiopathic in nature but have been reported secondary to certain metabolic disturbances. Among secondary causes of paroxysmal dyskinesias, hypocalcemia and hypoparathyroidism are well described. Here, we report a case of hypocalcemia with hypothyroidism and pseudohyoparathyroidism (PHP) presenting with paroxysmal dyskinesia.

A 17-year-old south Indian male patient presented to the Department of Neurology at Kannur medical college with a history of paroxysmal events for the past 2 months occurring daily, characterized by episodic, sudden, involuntary movements of limbs, which were unprovoked and paroxysmal, irregular, and lasting for few seconds to several minutes. His past medical history revealed global developmental delay and infantile seizures, treated with antiepileptics. The patient complained of typical carpo-pedal spasm at age of four, investigations of which indicated hypocalcemia and hypothyroidism. He was then started on levothyroxine and calcium supplements, which rendered him asymptomatic till recent times.

Patient’s mother described the evolving episodes as involuntary movements involving bilateral upper/lower limbs in a nonrepetitive/nonrhythmic pattern, appearing a few times per day initially, which later progressed to multiple, uncontrollable vigorous movements with greater frequency. However, the patient was totally conscious and oriented with an apparent discomfort during the episodes; these symptoms developed following the withdrawal of oral calcium and vitamin D3 supplements.

On examination, the patient appeared much younger than the stated age, with a height of 140 cm and weight of 36.5 kg. He had dysmorphic facial features, mal-formed and mal-occluded teeth, and short and stout hands and feet with shortening of metacarpals and metatarsals [Figure 1]. Examination of the mental status showed severe retardation. Cranial nerves, motor, sensory, and cerebellar systems were normal. Deep tendon reflexes were brisk, gait was normal, and cranium and spine were normal.

Laboratory investigations revealed hypocalcemia (serum calcium = 6.1 mg/dL), elevated serum phosphorus (10 mg/dL), and raised parathormone (PTH = 124.4 pg/mL) consistent with the diagnostic findings of PHP. Thyrotrophin secreting hormone (TSH) was initially high (97.63 IU) and decreased with therapy; T3 and T4 levels were normal. Radiographic examination of both hands revealed shortening of 4th and 5th metacarpals [Figure 2]. Magnetic resonance imaging of brain demonstrated bilateral symmetric dense hyperintensities (calcifications) in frontoparietal subcortical white matter, basal ganglia, and dentate nuclei. His electroencephalogram (EEG) study was unremarkable with normal findings. He was restarted on calcium and vitamin D3 supplementation for several years but there was no reduction in episodic abnormal movements. Later, he was started on oral oланzapine with which the patient had no significant improvement. On adding oxcarbazepine twice daily, he improved symptomatically. The number and duration of episodes reduced in intensity and totally disappeared within one week.

Paroxysmal dyskinesia is a hyperkinetic movement disorder causing attacks of involuntary movements; it is triggered by sudden voluntary movements/exercise and also it is triggered during sleep/even at rest. These attacks increase in number during puberty. This rare disorder affects around 1 in 1,50,000 only and has a male preponderance.[1] There are 2 types of paroxysmal dyskinesia: primary and secondary. While primary can be familial/sporadic, secondary dyskinesias can be due to many other medical conditions like hypocalcemia and PHP.[2]

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**Figure 1:** Shows short and stout feet with shortened metatarsals

**Figure 2:** Radiograph image showing short metacarpals
The movement disorder manifested in this patient was consistent with paroxysmal dyskinesia with the radiographic findings demonstrating basal ganglia calcifications. The relationship between these calcifications and extrapyramidal symptoms is not clear. But according to previous literature, extrapyramidal signs may be found in patients with calcification of basal ganglia in cases of untreated pseudohypoparathyroidism. The hypocalcemic seizures can range from syncopal episodes and movement disorders to generalized/focal seizures. Anticonvulsant therapy may delay the diagnosis by decreasing or eliminating the clinical features of tetany. Usually, in cases of PHP, the failure to diagnose from a history and physical examination is often the crux of the problem. Indeed, the most common misdiagnosis in hypocalcemic patients is an idiopathic seizure disorder. The movements in the present patient under study did not fit any seizure type and EEG episodes were normal, ruling out the possibility of hypocalcemia-induced simple seizures.

Hypocalcemia with elevated parathyroid hormone levels in the absence of renal disease is diagnostic of pseudohypoparathyroidism. PHP is in itself an uncommon disease often under diagnosed. Fuller Albright first introduced the term “pseudohypoparathyroidism” in 1942 to describe patients who presented with parathyroid-hormone-(PTH) resistant hypocalcemia and hyperphosphatemia along with an unusual constellation of developmental and skeletal defects, including short adult stature, obesity, brachydactyly mostly affecting 4th and 5th metacarpals and metatarsals, and ectopic ossifications collectively termed Albright hereditary osteodystrophy.

Intracranial calcifications are encountered accidentally in 0.3%–1.2% of routine radiological examinations. PHP as a cause of hypocalcemia can cause bilateral intracranial calcifications in the basal ganglia, thalamus, cerebral white matter, and cerebellum; this can be related to the neurological abnormalities like extrapyramidal signs and dyskinesia. Patients with PHP type 1a may develop resistance to other hormones such as TSH, gonadotrophins, and growth hormone (GH)-releasing hormone. PHP type 1a is characterized by impaired sensitivity to both TSH and TRH, with a mild-to-moderate elevation of serum TSH concentration. Hypothyroidism is generally mild and involves a slightly elevated TSH concentration with normal or slightly low concentrations of thyroid hormone. In our patient, hypothyroidism was diagnosed first and PHP later. It may be because the treatment of hypocalcemia resulted in a substantial diminution of symptoms initially. Their co-existence was identified much later in life. This co-existence of hypothyroidism with PHP along with symptoms of hypocalcemia leading to secondary paroxysmal dyskinesia has not been reported yet.

Carbamazepine and phenytoin are known treatments in managing polycystic kidney disease, but oxcarbazepine was much more effective in epileptic patients in lesser doses, without associated adverse effects. The present patient too responded well to oxcarbazepine.

This case highlights an important and a rare presentation of hypocalcemia in the form of a “Secondary Paroxysmal Dyskinesias in Pseudohypoparathyroidism and Hypothyroidism”—i.e., paroxysmal dyskinesia was secondary to PHP and hypocalcemia. It also reiterates the significance of clinical examination in patients with paroxysmal movement disorders and the importance of obtaining metabolic studies in such cases. It is essential to diagnose the underlying PHP early so that appropriate genetic counselling and prompt hormonal replacement therapy are offered to affected individuals and their first-degree relatives.

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There are no conflicts of interest.

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