# A Rare but Treatable Cause of Paroxysmal Nonkinesigenic Choreoathetosis

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Paroxysmal dyskinesias are a heterogeneous group of movement disorders characterized by episodic recurrent abnormal involuntary movements. Phenomenology of the movements can range from chorea, athetosis, ballism, dystonia in isolation or in combination.[1] They are classified as paroxysmal kinesigenic choreoathetosis (PKC), paroxysmal nonkinesigenic choreoathetosis (PNKD), and paroxysmal exercise-induced dyskinesia (PED) depending on the trigger and can be inherited or secondary. Secondary causes include trauma, stroke, meningovascular syphilis, encephalitis, multiple sclerosis, and metabolic disorders such as diabetes, hypoparathyroidism, hyperparathyroidism, and kernicterus.[2] We describe a young patient with secondary PNKD with some peculiarities.

A 19-year-old male with normal birth and developmental history presented with involuntary movements of the body since 1½ year of age. The movements were episodic lasting 30 s to 5 min and were described as jerky, nonrhythmic, choreoathetoid in character with the intermittent dystonic posturing of neck, face, upper and lower limbs [Video 1]. The frequency of the episodes was variable ranging from 10 to 100 attacks per day. There was no loss of consciousness, tongue bite or urinary incontinence in any of the episodes. The patient denied any premonitory urge before the movements, and he could not suppress them. There was no history of gait disturbances, cognitive decline, or cranial nerve involvement. Family history was negative. He was treated with multiple drugs (carbamazepine, valproate, and haloperidol) in the past with no benefit. The patient also underwent magnetic resonance imaging (MRI) brain (T1-weighted, T2-weighted, proton density images) during childhood which was reported to be normal. On examination, mini-mental state examination was 30/30. Speech, cranial nerves, power, and reflexes were normal. He underwent video electroencephalography (EEG) recording with the possibility of seizures versus movement disorder. Both ictal and interictal EEG did not show any epileptiform discharges. Routine investigations including hemogram, renal and liver function tests were normal. His calcium was 8.7 mg/dl (8.6–10.2 mg/dl) and phosphorus was 3.7 mg/dl (2.7–4.5 mg/dl). Ceruloplasmin, peripheral blood film for acanthocytes and thyroid profile were normal. Computed tomography (CT) head showed bilateral globus pallidus calcification [Figure 1]. His parathyroid hormone came out to be 10.17 pg/ml (normal 15–65 pg/ml) and Vitamin D 8.78 ng/ml (11–42 ng/ml). He was managed with acetazolamide, levetiracetam, calcium and Vitamin D supplementation and clonazepam following which the attacks diminished in duration as well as frequency.

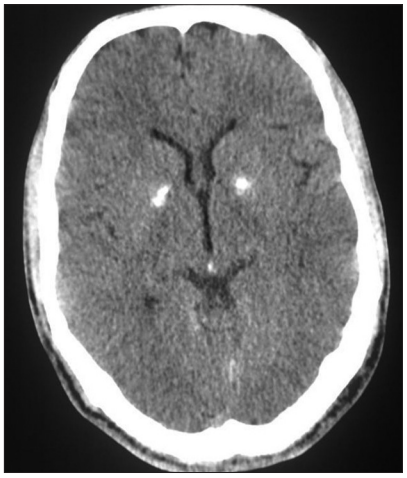

**Figure 1:** Computed tomography head showing bilateral globus pallidus calcification

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This case is interesting as it remained a diagnostic dilemma for almost 17 long years. A simple investigation like CT head helped clinched the diagnosis which was initially missed on MRI brain done previously in childhood. An important caveat is that susceptibility weighted or gradient echo images which are good at picking up calcification were not done at that time.

A constellation of neurological symptoms can occur in hypoparathyroidism, namely, seizures, tetany, and limb and perioral paresthesias, psychiatric manifestations like psychosis and depression and extrapyramidal features.[3] PNKD secondary to hypoparathyroidism is very rarely reported; though reports of PKC secondary to hypoparathyroidism and pseudohypoparathyroidism do exist.[4-6]

Pathogenesis of involuntary movements in hypoparathyroidism involves hypocalcemia which usually responds to calcium supplementation.[7] However, paroxysmal dystonic movements can occur with extensive deposits of calcium in basal ganglia with normocalcemia.[8] Other mechanisms described in literature include seizures arising from the subcortical structures, gliosis, and degeneration with long-standing calcium deposition and some form of channelopathy owing to its intermittent nature.[9] Perfusion abnormalities in the caudate nucleus in cases of PKC on Single-photon emission CT imaging have also been described in the literature.

Treatment includes the use of benzodiazepines, acetazolamide, and levetiracetam and there are few case reports of deep brain stimulation in refractory cases.[10] Our patient had only mildly decreased calcium, and he responded to a combination of acetazolamide, clonazepam in addition to calcium and Vitamin D supplementation.

To conclude, we describe a rare but treatable secondary cause of PNKD. A simple investigation like CT scan can help in clinching the diagnosis and is more sensitive than MRI brain in such a scenario. MRI brain with susceptibility weighted sequences to look at calcification or hemorrhage should be ordered routinely in patients presenting with a movement disorder.

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## Conflicts of interest

There are no conflicts of interest.

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