Turkish Journal of Neurology

Turk J Neurol 2025;31(3):380-382
Doi: 10.55697/tnd.2025.317

LETTER TO THE EDITOR

Paroxysmal kinesigenic dyskinesia arising from isolated thalamic infarction: A rare convergence

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Paroxysmal kinesigenic dyskinesia (PKD) is characterized by recurrent, brief episodes of involuntary movements triggered by sudden voluntary actions. These movements often include a combination of dystonia, chorea, athetosis, and ballism, typically lasting less than a minute and occurring multiple times per day.[1] Unlike epilepsy, PKD does not present with postictal confusion or unawareness during episodes.[1] Most PKD cases are idiopathic or familial, but secondary PKD has also been reported. Treatment with phenytoin or carbamazepine can significantly reduce the frequency or prevent PKD attacks.[1] Here, we presented two cases of PKD associated with isolated thalamic infarction, which, to our knowledge, is the first such report.

Case 1- A 19-year-old left-handed female patient was admitted with episodic abnormal posturing of the left wrist, hand, fingers, and leg for three days, without loss of consciousness. Episodes were preceded by a sensation of tightness in the left wrist, hand, and leg. Transient paresthesias in the left upper limb would end before the onset of dyskinesia. The frequency of episodes was more than 10 per day, triggered by movements such as walking or getting out of bed. The patient had no prior history of systemic or neurological disorders. Neurological examination revealed no abnormalities apart from left-sided hemihypoesthesia. Choreoathetosis and dystonia were triggered by actions as mild as standing or walking (Video 1). Blood tests showed

Video 1. 0-56. sec: (Case 1) Choreoathetosis and dystonia were elicited by actions such as standing up or walking. Dyskinesia disappeared after treatment. 57-105. sec: (Case 2) Dyskinesia on the left wrist, fingers and toe, disappeared after treatment.

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E-mail: ismailkoc.tip@gmail.com
Received: October 16, 2024 **Accepted:** February 03, 2025 **Published online:** February 26, 2025
Cite this article as: Koç İ, Kökoğlu S, Kayalı Şendur N. Paroxysmal kinesigenic dyskinesia arising from isolated thalamic infarction: A rare convergence. Turk J Neurol 2025;31(3):380-382. doi: 10.55697/tnd.2025.317.

elevated lupus anticoagulant levels (first evaluation: 2.34; control evaluation: 2.66; normal <1.2) and genetic testing revealed a homozygous A1298C MTHFR mutation. Magnetic resonance imaging (MRI) revealed increased signal intensity in the right thalamus, specifically in the ventral posterolateral and ventral lateral nuclei, indicative of subacute infarction (Figure 1). Electroencephalography was normal between and during episodes. Other tests were unremarkable. The patient was treated with acetylsalicylic acid, folic acid, and carbamazepine, resulting in complete resolution of the symptoms. Written informed consent was obtained from the patient.

Case 2- A 39-year-old right-handed female patient was admitted with 10 days of intermittent, episodic involuntary movements in the left wrist, fingers, toes, and ankle, with left foot inversion. Episodes lasted less than a minute and occurred more than five times per day. The symptoms began suddenly, involving both left upper and lower extremities. There was no pain or loss of consciousness during episodes, and sudden movements, such as getting up from a chair, triggered the attacks. The patient also experienced occasional transient paresthesia and hypoesthesia in the left upper limb, which ended before the dyskinesia (Video 1). The patient had no prior medical history, and the initial neurological examination and laboratory tests were normal. Brain MRI revealed subacute infarction in the right thalamus, involving the ventral posterolateral nucleus (Figure 2). Written informed consent was obtained from the patient.

Prolonged electroencephalography monitoring, including exercise provocation, showed nonspecific findings. The patient was diagnosed with PKD associated with thalamic infarction and was treated with carbamazepine and acetylsalicylic acid, resulting in complete resolution of PKD episodes. Furthermore, laboratory and genetic studies were unremarkable.

Neither patient experienced recurrence of symptoms while on carbamazepine treatment. After one year of attack-free follow-up, the medication was gradually tapered off, and the patients were attack-free at four years of follow-up.

Paroxysmal kinesigenic dyskinesia can manifest up to 100 times per day, often preceded by a sensory aura. The paroxysmal and stereotyped nature of PKD, along with its dramatic response to phenytoin or carbamazepine, are hallmark features of this disorder.[1]

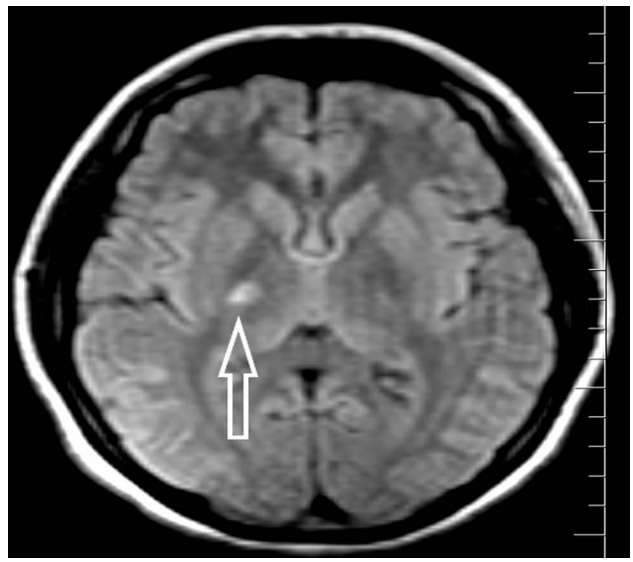

Figure 1. Increased signal in the area of the ventral posterolateral and ventral lateral nuclei of right thalamus in fluid attenuating reversal recovery (FLAIR) sequence compatible with subacute infarction.

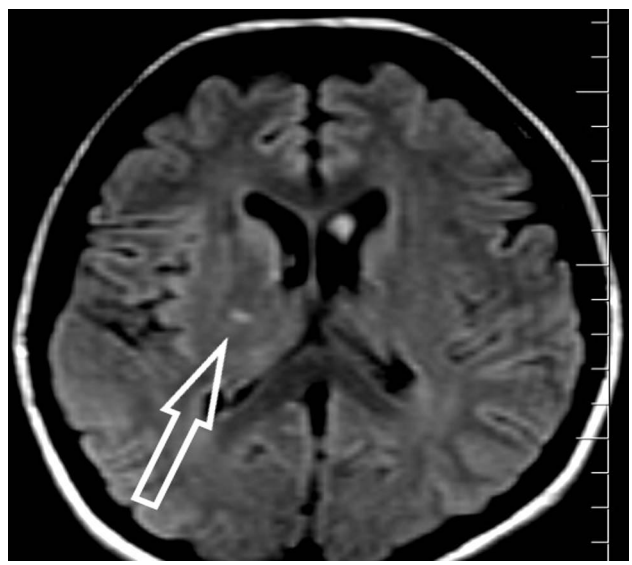

Figure 2. FLAIR sequence compatible with subacute infarction involving the ventral posterolateral nucleus area of the right thalamus.

The pathophysiology of PKD remains uncertain. The basal ganglia have been implicated as the primary site of dysfunction, with decreased inhibition of thalamic reticular nuclei by the medial globus pallidus and substantia nigra proposed as a potential mechanism.[2,3] Cortical structures, the cerebellum, and the thalamus were also implicated in the pathophysiology of PKD.[4] Our cases provided further

evidence of a link between thalamic damage and PKD development. Most PKD cases associated with thalamic infarction involve other brain structures,[3] but our cases illustrated PKD resulting from isolated thalamic infarction, supporting the structural and functional relationship between the thalamus and PKD.

Peripheral somatosensory information is relayed through ventral anterior/lateral nuclei and ventral posterior nucleus.[5] Based on this, we speculated that the lateral thalamic nuclei, which connects to the motor cortex, may play a significant role in the pathophysiology of PKD, as suggested by the involvement of the lateral thalamus observed in our patients. This is supported by PKD's paroxysmal manifestations, indicating network instability in the basal ganglia-thalamocortical pathway.

Kim et al.[2] showed that reduced bilateral thalamic volume and regional shape deformations were primarily localized to the anterior and medial aspects of the thalami in PKD patients. Although the lesion locations differed from those observed in our patients, these discrepancies may be influenced by the smaller sample size and exclusive inclusion of primary PKD patients in their study. Furthermore, prior research supports our findings, demonstrating that while other brain regions may be affected depending on the neuroimaging modality used, only the thalamus consistently exhibits involvement across all imaging techniques, including MRI, positron emission tomography, single-photon emission computed tomography, and diffusion tensor imaging.[4]

In conclusion, while it is not clear which specific thalamic nuclei are responsible, the thalamus appears to play a crucial role in PKD pathophysiology. Given that PKD can manifest years after a stroke,[4] long-term follow-up of patients with thalamic infarctions may help identify additional cases and further elucidate the mechanisms underlying PKD.

Acknowledgment: We would like to thank Assoc. Prof. Dr. Rezzak Yılmaz for his valuable insights and guidance during the development of this work.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

Author Contributions: Preparation of the project, analyzing, interpreting data of the study: İ.K., S.K., N.K.Ş.; Obtained data: İ.K., S.K.

Conflict of Interest: The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding: The authors received no financial support for the research and/or authorship of this article.

REFERENCES

1. Uysal Tan F, Balaban H, Ulkatan S, Akbostancı C. Paroxysmal dyskinesia: Report of four cases. Turk J Neurol 2001;7:101-108.
2. Kim JH, Kim DW, Kim JB, Suh SI, Koh SB. Thalamic involvement in paroxysmal kinesigenic dyskinesia: A combined structural and diffusion tensor MRI analysis. Hum Brain Mapp 2015;36:1429-41. doi: 10.1002/hbm.22713.
3. Camac A, Greene P, Khandji A. Paroxysmal kinesigenic dystonic choreoathetosis associated with a thalamic infarct. Mov Disord 1990;5:235-8. doi: 10.1002/mds.870050309.
4. Liu W, Xiao Y, Zheng T, Chen G. Neural mechanisms of paroxysmal kinesigenic dyskinesia: Insights from neuroimaging. J Neuroimaging 2021;31:272-6. doi: 10.1111/jon.12811.
5. Long Z, Xu Q, Miao HH, Yu Y, Ding MP, Chen H, et al. Thalamocortical dysconnectivity in paroxysmal kinesigenic dyskinesia: Combining functional magnetic resonance imaging and diffusion tensor imaging. Mov Disord 2017;32:592-600. doi: 10.1002/mds.26905.