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Int J Case Rep Images 2024;15(1):66–68.
www.ijcasereportsandimages.com

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Pantothenate kinase-associated neurodegeneration (PKAN) with a typical "eye of the tiger": A radiology case report

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CASE REPORT

A 7-year-old boy was referred to the Radiology Department for the evaluation of progressive developmental regression, dystonia, and dysarthria.

The neurological examination revealed upper limb tremor, spastic paraparesis with rigidity, dystonic movements, and dysarthria.

His birth history was unremarkable, and developmental regression began at the age of 4.

Laboratory tests and electroencephalogram (EEG) results were normal.

Imaging

The patient underwent brain scanning using a 1.5 T magnetic resonance imaging (MRI) system, with the routine protocol involving three-dimensional fluid attenuated inversion recovery (3D FLAIR)-weighted images, 3D T1-weighted images, susceptibility-weighted imaging (SWI), and diffusion-weighted imaging (DWI).

Both T2 and SWI images revealed bilateral and symmetric high signal intensity within the globus pallidus, surrounded with a low signal intensity area (Figure 1).

No other lesions were observed, including other basal ganglia and substantia nigra.

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Received: 17 January 2024
Accepted: 03 March 2024
Published: 17 April 2024

This combination of finding is suggestive of a typical "eye of the tiger," characteristic of pantothenate kinase-associated neurodegeneration (PKAN), though not pathognomonic.

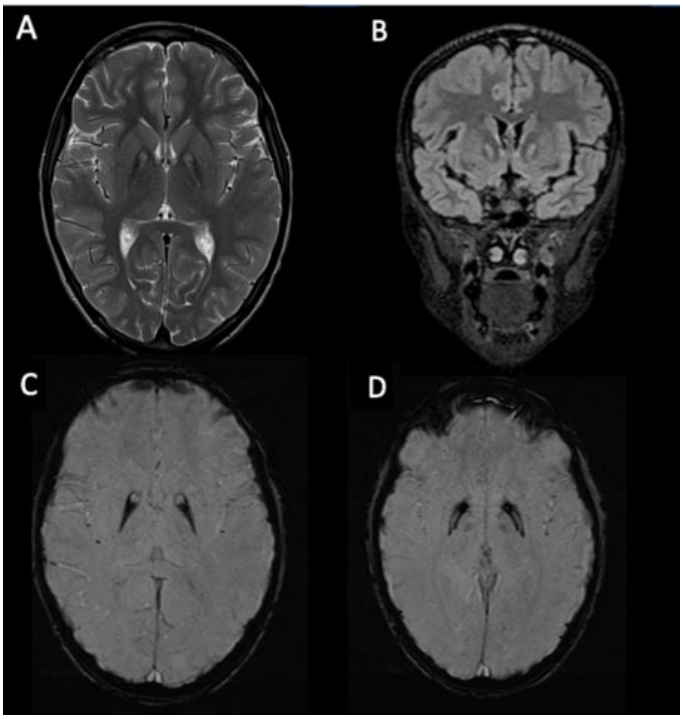

Figure 1: (A) Brain MRI axial sequence in T2-weighted, (B) coronal sequence in FLAIR, (C and D) and axial sequences in SWI, showing bilateral aspect of "eye of the tiger" sign, with central area of hyperintensity surrounding a region of hypointensity in the globus pallidus in T2-weighted and FLAIR, and hypointensity in SWI.

DISCUSSION

Pantothenate kinase-associated neurodegeneration (PKAN), formerly called Hallervorden–Spatz syndrome, is the most usual type of neurodegeneration with brain iron accumulation (NBIA), accounting for half of the NBIA cases and has an estimated prevalence of 1–3/100,000 [1, 2].

It is an autosomal recessive disorder resulting from a mutation in the pantothenate kinase 2 gene (PANK2). A PANK2 is essential for the production of the "pantothenate kinase 2 enzyme," which regulates coenzyme A (CoA) synthesis. Insufficiency of this enzyme leads to the destruction of the phospholipid membrane, primarily in the basal nuclei and retina, resulting in secondary iron accumulation [2].

Pantothenate kinase-associated neurodegeneration is subdivided into two main types, based on clinical presentation [1–3]:

- Classic PKAN: (75%) characterized by an early onset (usually between 3 and 4) and a faster disease progression. Symptoms include dystonia, dysarthria, rigidity, choreoathetosis, and retinopathy pigmentosa.
- Atypical PKAN: (25%) characterized by a later onset (usually at the age of 14) and slower progression, often revealed by dysarthria, mild gait abnormalities, subtle dystonia, and neuropsychiatric features.

Neuroimaging in PKAN reveals distinctive iron accumulation in the globus pallidus. As the disease progresses, it may also affect the substantia nigra.

Computed tomography (CT) scans, while not particularly useful or specific in diagnosing PKAN, may show hypodensity or calcifications in the globi pallidi.

The typical MRI finding, known as "the eye of the tiger," is identified on T2 imaging as a round medial area of high signal intensity surrounded with a low signal intensity globus pallidus [3, 4].

The low signal intensity is best appreciated on T2*/SWI-weighted images due to susceptibility effects, correlating pathologically with areas of abnormal iron deposition, while the center area of high signal intensity "the eye" corresponds to neuronal loss with gliosis [3, 4].

Over time, the low signal intensity may dominate the radiological presentation, and the central high signal intensity can disappear [3, 4].

This sign has been documented in various other conditions, including carbon monoxide poisoning, cortical basal ganglionic degeneration, multiple-system atrophy, and other forms of NBIA, especially neuroferritinopathy [3, 4].

While "the eye of the tiger" is a characteristic finding, it is not specific or sensitive for PKAN and should be correlated with clinical context, confirmed by genetic testing demonstrating PKAN2 gene mutation.

CONCLUSION

In conclusion, our case report highlights the characteristic neuroimaging finding, "the eye of the tiger," indicative of pantothenate kinase-associated neurodegeneration (PKAN) in a 7-year-old boy presenting with dystonia and dysarthria. Although this radiological sign is highly suggestive of PKAN, it is not pathognomonic and can be observed in other conditions. Therefore, a comprehensive evaluation integrating clinical presentation, neuroimaging, and genetic testing when available is imperative for accurate diagnosis and appropriate management.

Keywords: Computed tomography, Eye of the tiger, Magnetic resonance imaging, PKAN

How to cite this article

Horache K, Messaoud O, Elkettani N, Fikri M, Jiddane M, Touarsa F. Pantothenate kinase-associated neurodegeneration (PKAN) with a typical "eye of the tiger": A radiology case report. Int J Case Rep Images 2024;15(1):66–68.

Article ID: 101447Z01KH2024

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doi: 10.5348/101447Z01KH2024CI

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Author Contributions

Kenza Horache – Conception of the work, Design of the work, Analysis of data, Drafting the work, Final approval

of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Ola Messaoud – Conception of the work, Design of the work, Analysis of data, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Guarantor of Submission
The corresponding author is the guarantor of submission.

Source of Support
None.

Consent Statement
Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.