RESEARCH PAPER

Eye of the Tiger beyond Childhood: A Rare Case of Pantothenate Kinase-Associated Neurodegeneration in Late Adulthood

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ABSTRACT
Pantothenate kinase-associated neurodegeneration (PKAN) is an uncommon inherited disorder marked by iron deposition within the basal ganglia, particularly the globus pallidus. Although typically identified in childhood, atypical late-onset presentations may occur. We describe a 59-year-old male presenting with progressive motor impairment. MRI demonstrated the classical "eye of the tiger" appearance within the globus pallidus. This report underscores the diagnostic value of characteristic imaging findings and highlights the need to consider Neurodegeneration with Brain Iron Accumulation (NBIA) disorders even in older patients with movement abnormalities.

Informed consent for participation and publication
Informed consent was obtained from individual participant included in the study.

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INTRODUCTION

Pantothenate kinase-associated neurodegeneration represents a major subtype of disorders grouped under neurodegeneration with brain iron accumulation (1,2). It arises from mutations affecting the (Pantothenate kinase 2) PANK2 gene, leading to disordered iron metabolism within deep gray matter structures (1,3,4).

While the classical form manifests early in life with rapidly progressive extrapyramidal symptoms, atypical variants may present later with a more indolent clinical course. Such presentations can mimic other neurodegenerative conditions, making diagnosis challenging (2,6,7).

MRI plays a central role in evaluation, with the "eye of the tiger" pattern—T2 hypointensity with central hyperintensity in the globus pallidus—serving as a key radiological indicator guiding the diagnosis (3,5).

CASE REPORT

A 59-year-old male presented with gradually progressive difficulty in walking, slowness of movements, and intermittent involuntary movements over the past two years. There was no significant past medical or family history. Neurological examination revealed rigidity, bradykinesia, and mild dystonia. Cognitive functions were relatively preserved.

MRI of the brain was performed for further evaluation. T2-weighted and FLAIR sequences (Figure a) demonstrated bilateral symmetrical hypointensity in the globus pallidus with a central region of hyperintensity in the anteromedial globus pallidus, producing the characteristic "eye of the tiger" appearance. T2*-weighted gradient-echo (GRE) images (Figure b) demonstrated blooming in the globus pallidus, consistent with iron deposition.

No significant abnormalities were noted in other regions of the brain. Based on clinical presentation and imaging findings, a diagnosis of pantothenate kinase-associated neurodegeneration was considered.

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DISCUSSION

Pantothenate kinase-associated neurodegeneration (PKAN), also known as Hallervorden–Spatz syndrome, is a common form of neurodegeneration with brain iron accumulation due to autosomal recessive mutation of PANK2 gene of chromosome 20p13 (1,2). PKAN has an estimated prevalence rate of 1-3/ 1,000,000 population (3). This gene is necessary for production of PANK 2 enzyme which is needed for synthesis of coenzyme A (CoA) (1,3). Therefore, PKAN results in pathological iron accumulation, particularly within the globus pallidus (1,3).

Clinically, PKAN is categorized into early-onset (classic) and late-onset (atypical) forms. The former has onset before 6 years of age with rapid progression while the latter demonstrates slower progression and may present with parkinsonian features, movement disorder progressing to speech disturbances, or neuropsychiatric manifestations, often leading to delayed recognition (2,6).

The “eye of the tiger” sign on MRI reflects underlying pathological changes. The hypointense rim is attributed to iron deposition, whereas the central hyperintense region in the anteromedial globus pallidus corresponds to neuropil destruction and gliosis (5). Even though the classical location is globus pallidus, it can progress to involve substantia nigra in advanced stages (5). Although highly suggestive, this sign is not entirely exclusive so should be interpreted alongside clinical findings and the definitive diagnosis is by genetic testing for PANK2 gene mutation (5).

In older individuals, PKAN may be mistaken for more common conditions such as Parkinson’s disease or other extrapyramidal disorders based on the clinical presentation with progressive extrapyramidal syndrome. Hence, awareness of this imaging signature is essential for accurate diagnosis (7).

CONCLUSION

This case emphasizes that PKAN may present in late adulthood with atypical late-onset. Awareness of the characteristic MRI findings is essential for early suspicion. In such cases, imaging plays a pivotal role in guiding further evaluation, including genetic testing for confirmation. Including NBIA disorders in the differential diagnosis of elderly patients with basal ganglia abnormalities can facilitate appropriate evaluation and management. Early recognition also helps avoid misdiagnosis and enables better patient counselling and prognosis.

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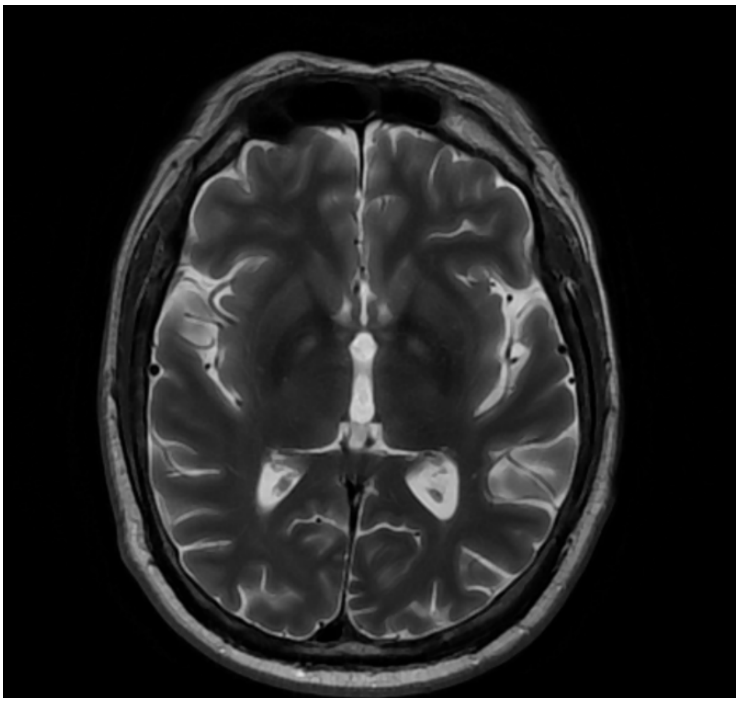
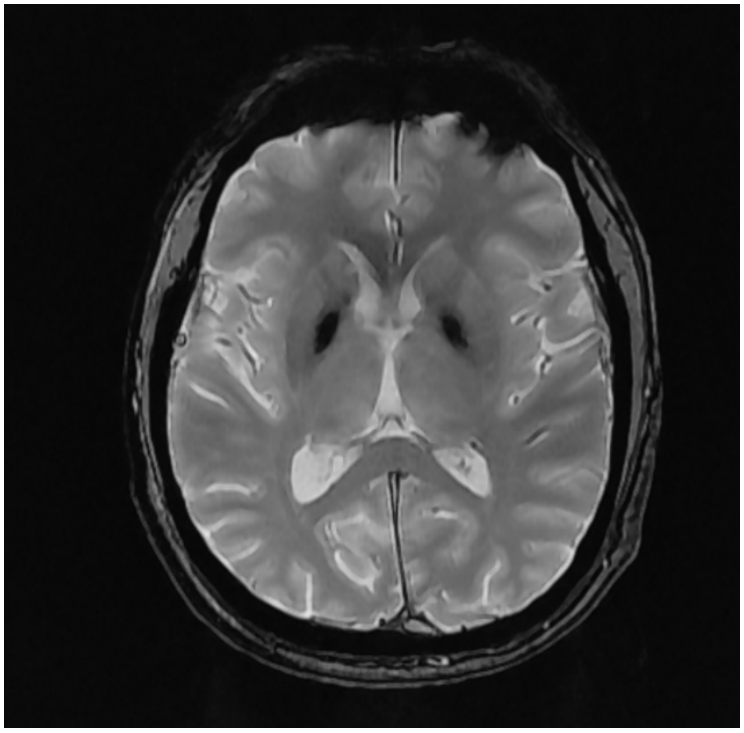
FIGURES



a



b

FIGURE LEGENDS

Figure a: T2 weighted image demonstrating classical 'eye of the tiger sign' – Bilateral symmetrical T2 hypointensity in the globus pallidus with central hyperintensity.

Figure b: T2* Gradient Echo (GRE) image showing blooming in bilateral globus pallidus.