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Eye of the Tiger Sign in a 8-Year-Old Child with Epilepsy NBIA of the PKAN Type

Kaoutar Maslouhi*, Asma Cherif, Rania Bouanane, Kaouthar Sfar, Nazik Allali, Latifa Chat and Siham E Haddad Department of Pediatric radiology Pediatric Teaching Hospital, Mohammed V University, Rabat-Morocco

A 8-years-old girl frome a consanguines mariage, Suffren frome a intellectuel déficient, Epilepsy and psychosomatique Delay frome Barth. Clinical examination revealed dysarthria, stiffening and dystonic movements mainly of the upper member with an EEG compatible with epilepsy. An initial head CT showed bilateral calcifications of the globus pallidus (Figure 1). A brain MRI was performed, revealing diffuse bilateral and symmetric hypointensity of both the globus pallidus with relatively central hyperintensity in T2-weighted images (Figure 2). These areas showed susceptibility artifacts (low signal) in T2* sequences (Figure 3). No other signal alterations were observed in other regions of the basal ganglia. These findings were consistent with the eye of the tiger sign that is characteristic of pantothenate kinaseassociated neurodegeneration but not pathognomic.

Discussion

PKAN is characterized by the distinctive eye of the tiger sign, featuring a circular hyperintense center surrounded by hypointensity in the globus pallidus on T2-weighted images (T2WI). This classic sign sets PKAN apart from other NBIA disorders, including COASY protein-



Figure 1: Axial CT scan showed bilateral calcifications of the globus pallidus.



Figure 2: Brain MRI in axial T2-weighted sequence revealing diffuse bilateral and symmetric hypointensity of both the globus pallidus with relatively central hyperintensity.



Figure 3: Brain MRI in axial $T2^*$ sequence revealing Globus pallidus with low signal.

associated neurodegeneration (CoPAN), which shares similarities in coenzyme A metabolism with PKAN [1,2]. In infancy, the initial change appears as a linear hyperintense streak along the medial border of the globus pallidus, evolving into an isolated hyperintense center in the anteromedial region during early childhood. The surrounding T2 hypointensity tends to enlarge with age. The eye-of-the-tiger sign may exhibit variations in shape in adults, and its appearance can be influenced by factors like diamagnetic calcium deposits. While this sign can mimic patterns seen in other conditions, its PKAN specificity is best defined by the region-specific pattern of iron deposition on SWI [3].

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*Corresponding author: Kaoutar Maslouhi, Department of Pediatric radiology Pediatric Teaching Hospital, Mohammed V University, Rabat-Morocco Email: dr.maslouhikaoutar@gmail.com

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