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Pantothenate Kinase-Associated Neurodegeneration (PKAN) With a Typical "Eye of the Tiger": A Radiology Case Report

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Ca e Repor

A 7-ear-old bo as referred to the radiolog department for the evaluation of progressive developmental regression, d stonia and d sarthria. , e neurological e amination revealed upper limb tremor, spastic paraparesis ith rigidit , d stonic movements and d sarthria. His birth histor as unremarkable, and developmental regression began at the age of 4. Laborator tests and EEG results ere normal.

, e patient under ent scanning using a 1.5T MRI s stem, ith the protocol involving 3D FLAIR images, a 3D T1- eighted images, susceptibilit - eighted imaging (SWI), and di usion- eighted imaging (DWI). Both T2 and SWI images revealed bilateral and s mmetric h perintensit ithin the globus pallidus, surrounded ith a h pointensit area. No other lesions ere observed, including other basal ganglia and substantia nigra.

, is combination of , nding is suggestive of a t pical e e of the tiger. , characteristic of pantothenate kinase-associated neurodegeneration (PKAN), though not pathognomonic.

Di c ion

PKAN, former l called Hallervorden-Spat s ndrome, is the most usual t pe of neurodegeneration ith brain iron accumulation (NBIA), accounting for half of the NBIA cases and has an estimated prevalence of 1-3/100000 [1].

It is an autosomal recessive disorder resulting from a mutation in the pantothenate kinase 2-gene (PANK2). PANK 2 is essential for the production of the pantothenate kinase 2 en me , hich regulates coen me A (CoA) s nthesis. Insu cienc of this en me leads to the destruction of the phospholipid membrane, primaril in the basal

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Informed con en

Written informed consent as obtained from a legall authori ed representatives for anon mi ed patient information to be published in this article.

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