Eye-of-the-tiger Sign

7 years old male child presented with history of falls and progressive gait difficulty. Speech and swallowing difficulties were also reported by the parents. Family history was significant for a similar symptom complex in his elder brother who was now bedridden. Physical examination revealed signs consistent with extrapyramidal and corticospinal dysfunction. Levels of copper, ceruloplasmin, lipids and amino acids in the blood were all within normal limits. No acanthocytes were seen in the peripheral smear. MRI showed symmetric hyperintense signal changes in the anterior medial globus pallidus with surrounding hypointensity in the globus pallidus on T2 weighted images—the ‘eye-of-the-tiger’ sign characteristic of Hallervorden-Spatz disease (HSD). Genetic studies were positive for a mutation in the pantothenate kinase (PANK2) gene on band 20p13 thus confirming the diagnosis. Familial HSD which is inherited recessively is characterized by iron deposition in the globus pallidus and the substantia nigra pars reticulata. Other disease presenting with extrapyramidal-pyramidal-dementia complex like Wilson disease and neuroacanthocytosis should be considered in the differential diagnosis of HSD. The disease is relentlessly progressive with extrapyramidal dysfunction and death in the second to third decade of life.

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