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RESEARCH ARTICLE

HALLERVORDEN-SPATZ DISEASE – MRI EVALUATION

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ABSTRACT

Hallervorden-Spatz Disease is a rare inherited CNS disorder that is characterized by abnormalities in the globus pallidus. 32 year old man presented with unsteady gait and progressive generalized motor difficulty associated with dysarthric speech since two years. MR studies demonstrate iron storage in the basal ganglia in patients

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INTRODUCTION

Hallervorden-Spatz Disease is a rare inherited CNS disorder that is characterized by abnormalities in the globus pallidus with motor and speech difficulties. Presence of decreased signal intensity in T2-weighted images, compatible with iron deposits, and of a small area of hyperintensity in its internal segment ("eye of the tiger" sign) is referred to as Hallervorden-spatz disease (Hayflick *et al*., 2003) Though the disease can present in childhood or adolescence. The usual age of presentation is 2nd–5th decade with basal ganglia and globus pallidus being the most common sites of involvement[5] About 40% of patients with "eye of the tiger" sign present initially unsteady gait and progressive generalized motor difficulty associated with dysarthric speech.

MATERIALS AND METHODS

The scan was performed using 1.5 Tesla ACHIEVA (Philips Healthcare). Sagittal and Axial T1 and T2 weighted sequences and coronal STIR sequence were taken for cranium. Axial T1, T2 Weighted and FLAIR sequences. Coronal and sagittal T2 Weighted sequences and axial DWI sequences are taken for cranium.

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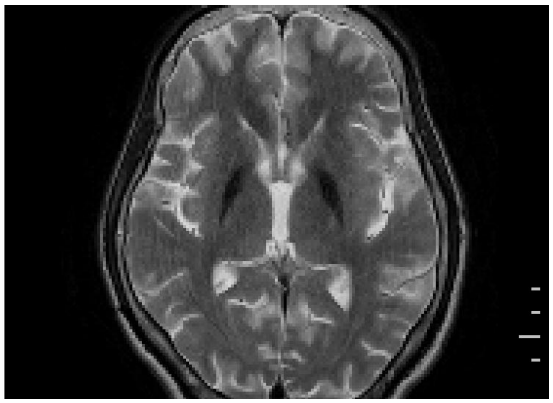

Figure 1. Hypointensity with central hyperintensity in globus pallidus on T2WI

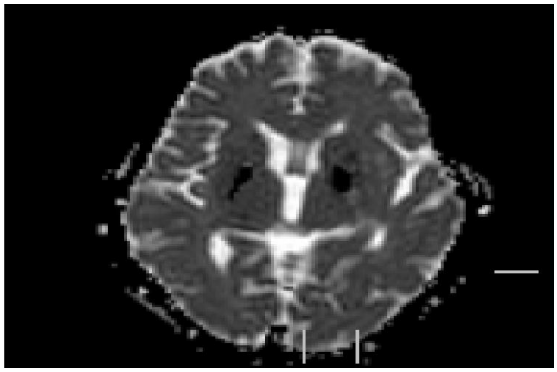

Figure 2. Low signal on ADC sequences

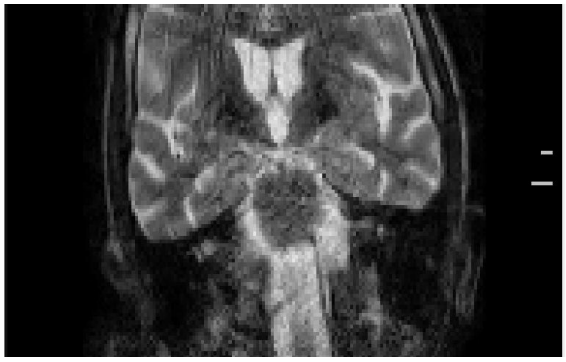

Figure 3. T2WI coronal shows globus pallidus hypointensity with central hyperintensity

DISCUSSION

Hallervorden-spatz disease is a condition characterized by decreased signal intensity in T2-weighted in globus pallidus with small area of hyperintensity in its internal segment ("eye of the tiger" sign) (Hayflick *et al*., 2003). Genetic studies have revealed an autosomal recessive hereditary disorder, presumably of metabolic origin. Symptoms can include unsteady gait, progressive generalized motor difficulty associated with dysarthric speech. (Dooling *et al*., 2005) Features often present between the ages of 2nd–5th decade. Clinico-imaging features are fairly diagnostic of HSD even in the absence of the histopathological confirmation of the disease in the globus pallid (Jhou *et al*., 2001 and Hartig *et al*., 2006) When a young patient presents with progressive extra-pyramidal symptoms and mental deterioration with typical MR appearances of bilaterally symmetrical hyperintense changes (gliosis) with peripheral hypointensity (iron deposition) in the globus pallidi on T2WI a possibility of HSD needs to be strongly considered (Castelnau *et al*., 2005). CT scan usually shows hypodensity in both basal ganglionic regions, along with mild to moderate atrophy of the brain and basal ganglionic regions, due to gliosis but some time it may be normal (Hartig *et al*., 2006 and Castelnau *et al*., 2005) To summarize, HSD should be suspected in patients with progressive generalized motor difficulty Routine biochemical investigations should always be performed to rule out metabolic causes ,knowledge of the associated conditions will not only help to rectify the treatable cause but will also prevent unnecessary treatment in others.

REFFERENCES

Castelnau, P, Cif, L, Valente, EM, Vayssiere, N, Hemm, S, Gannau, A, Digiorgio, A, Coubes, P. 2005. Pallidal stimulation improves pantothenate kinase-associated neurodegeneration. *Ann Neurol*., 57:738–741.

Dooling, E.C., Schoene, W.C., Richardson, E.P., 2005. Jr. Hallervorden-Spatz syndrome. Arch Neurol.1974; 30: 70–83 4. Gregory A, Hayflick SJ. Folia Neurodegenerationwith brain iron accumulation. *Neuropathology,* 43(4):286-96.

Hartig, M.B, Hortnagel, K., Garavaglia, B., Zorzi, G., Kmiec, T., Klopstock, T., Rostasy, K., Svetel, M., Kostic, V.S., Schuelke, M., Botz, E., Weindl, A., Novakovic, I., Nardocci, N, Prokisch, H, Meitinger, T. Genotypic, 2006. phenotypic spectrum of PANK2 mutations in patients with neurodegenerationwith brain iron accumulation. *Ann Neurol*., 59(2):248-56.

Hayflick, S.J, Westaway, S.K, Levinson, B, Zhou, B, Johnson, M.A, Ching, K.H, Gitschier, J. Genetic, 2003. clinical, and radiographic delineation of Hallervorden-Spatz syndrome. *N Engl J Med.,* 348:33-40.

Jhou, B, Westaway, S.K, Levinson, B, Johnson, M.A, Gitschier J, Hayflic, K 2001. A novel pantothenate Kinase gene (PANK2) is defective in Hallervorden-Spatz syndrome. Nature Genetics, 28:345-9.

Sethi, K.D, Adams, R.J, Loring, D.W., elGammal, T. 1988. Hallervordenspatz syndrome: Clinical and magnetic resonance imaging correlations: *Ann Neurol*., 24:692-4.

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