Short Communication

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Eye of Tiger Sign in Paediatrics

Khurram Khaliq Bhinder[1*], Mahnoor Sukaina[2], Zenab Farooq[1], Humaira Bashir[1], Laiba Khan[3] and Ameena[4]

[1]Department of Radiology, Shifa International Hospital, Islamabad, Pakistan
[2]Department of Psychiatry, Karachi Medical and Dental College, Karachi, Pakistan
[3]Department of Medicinal Science, Women Medical and Dental College, Abbottabad, Pakistan
[4]Department of Medicinal Science, Lahore General Hospital, Lahore, Pakistan

Abstract

Hallervorden-Spatz disease is a rare autosomal recessive disorder that can be present in a paediatric age group. The disease is classified based on the onset of symptoms, such and classic and atypical. Early symptoms onset is generally appearing in the first decade of life manifesting cognitive impairment and neurodegeneration. However, a child can be present with psychiatric disorders and speech disorders after the first decade of life. The mainstay diagnosis is made based on T2 weighted MRI, exhibit an eye-of-tiger sign, typically due to the accumulation of iron in globus pallidus, basal ganglia, or substantia nigra. A typical classified Hallervorden-Spatz presented at our hospital, the child was diagnosed based on disease traits and confirmed by radio imaging.

Keywords: Hallervorden spatz; Depressive symptoms; Cognitive impairment; Psychotic; Parkinsonism

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Corresponding author: Khurram Khaliq Bhinder, Department of Radiology, Shifa International Hospital, Islamabad, Pakistan, E-mail: kkbhinder@yahoo.com

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Description

Hallervorden-Spatz syndrome is a rare autosomal recessive familial disease, due to mutation of pantothenate kinase 2 (PANK2) genes involving band 20p13- p12.3 typically causing neurodegeneration. It has an early onset usually in infancy or the first decade of life starting from 7 years to 15 years of age [1]. While having progressive outcome overtime at the second decade of life and eventually most fatalities are reported at the second or third decade of life. The common symptoms are psychotic explicating signs of impulsiveness, depressive symptoms, and cognitive impairment [2]. These signs neuro-degenerative to accumulation of iron in the brain, furthermore other common signs manifest in the disease are, dysarthria and extrapyramidal signs including parkinsonism, increase in muscle tone causing dystonia and rigidity while leading to an unprogressive milestone for children [3].

Pediatric age group child presented in the outpatient facility of our hospital with presenting complaints of pursing off lower lip off and on with slurring of speech, decreased vision, and frequent history of falls for 2-3 years. Physical examination especially the neurological examination was unremarkable. Such history was seen in his elders' brother also. An urgent MRI Brain was ordered. Multiplanar, multi-sequential MRI of the brain without contrast was performed according to departmental protocols. Symmetrical T2 hyperintense signal areas with surrounding low-intensity areas giving an eye-of-tiger sign in bilateral medial globus pallidi. These areas showed signal drop out on T2* images signifying iron deposition. No other significant abnormality was reported in the brain. Diagnosis of Hallervorden-Spatz disease was made on radiological findings. Findings were not confirmed pathologically because of the financial status limitation of the patient.

A major contributor in the pathogenesis of Hallervorden-Spatz syndrome is a mutation in gene encoding PKAN-2 which alters the normal phosphorylation of pantothenic acid, required to synthesize coenzyme A. Deficit amount of pantothenate kinase procced with the retention of cysteine in the basal ganglia [4,5]. The disruption in the normal mechanism further involves the formation of free radicles due to iron chelation in globus pallidus accompanied by immediate cysteine auto-oxidation physiologically there is a normal level of iron in globus pallidus and pars reticularis, however, excess iron may escape and leak into the axoplasm, leads to axonal disorders [6] .Moreover, iron and zinc interaction promotes cytokine-mediated damage to the neurons which enable oxidative stress by microglia. The globus pallidus and substantia nigra involves in the excessive iron accumulation and pigmentation in the blood vessels, it can also be characterized by the cerebral and basal ganglia atrophy, with an accumulation of axonal spheroids especially involving globus

pallidus and basal ganglia [7].

Symptoms are progressive as the disease due to association PANK2 mutation and initiate during the first decade of life, hence it worsens during the second decade and eventually leads to fatality. Classic familial pantothenate kinase genetic (PANK2) mutation affects the children before the first decade, usually four to five years of age, with characteristic features of corticospinal tract involvement, abnormal gait, extensor toe sign, over-responsive reflexes, spasticity [8]. Moreover, symptoms manifest rigidity, abnormal muscle tone leading to dystonia, following with milestone delay; children lose the ability to move after the first decade of life [9]. Atypical forms of disease influence the patient's speech and psychiatric abilities, the common symptoms are an impulsive explosion of emotions, violent attacks, depressive symptoms [10,11]. These atypical symptoms are slowly progressive and can manifest classic symptoms after the first decade till the third decade of life [12]. Further complications are dementia due to neuro-degenerative PANK2 mutation, abnormal muscle tone, retinal pigment degeneration due to iron accumulation in the brain, limb stiffness, later causing dysarthria.

The MR radiological diagnostic feature of HSS is the eye-of-tiger-sign a hallmark feature that can be visualized through the imaging, the hypo-intensive image on T2 weighted MRI due to excessive iron deposition in globus pallidus. The hyper-intensive region on MRI indicates the necrotic pattern, gliosis, and edema followed by neutrophil cavitation, loss of neurons, vacuolization, and disintegration of the region on T2 weighted MRI. Antioxidant treatments have shown positive responses for patients with PKAN2 mutation, including coenzyme Q, pantothenate [13]. Furthermore, there are controversies regarding iron chelation therapy for excessive iron deposition. However supportive treatments are beneficial for complications associated with HSS. Dopamine agonists are indicated for the treatment of tremor, anticholinergics with dopamine agonists have shown better clinical outcomes for rigidity and tremor, moreover, baclofen and trihexyphenidyl for dystonia can be prescribed in **Figures 1-3**.

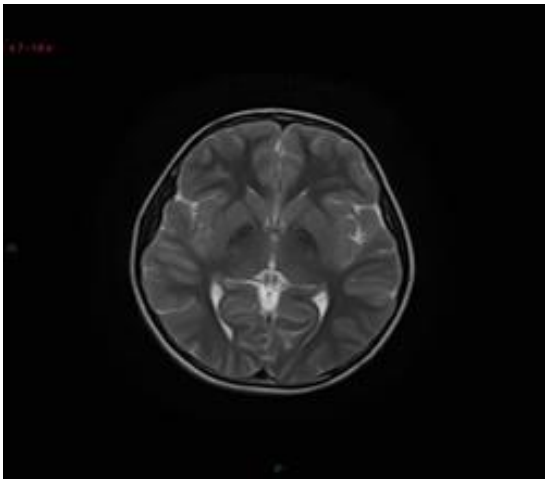

Figure 1: Axial T2 MRI brain.

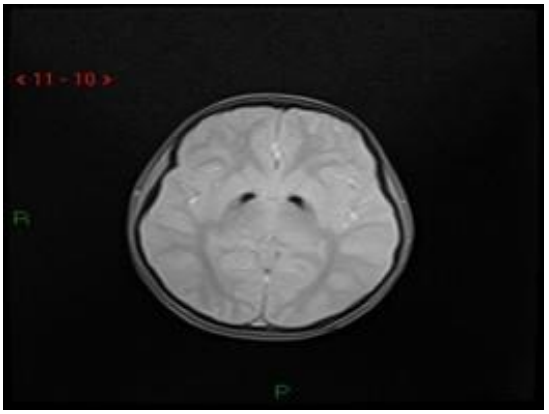

Figure 2: Axial T2 axial view.

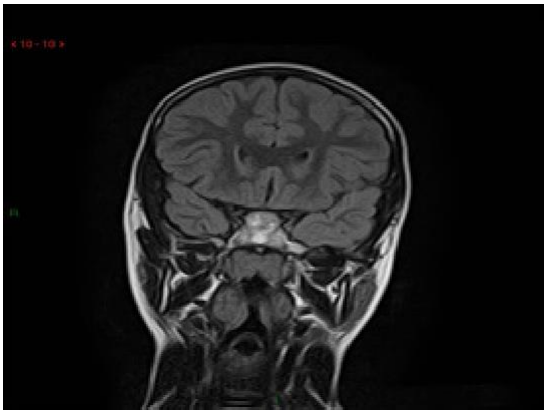

Figure 3: Coronal T2 MRI brain.

Conclusion

Hallervorden Spatz disease should be kept in differentials in paediatric population with cognitive impairment and dystonia. MRI is the gold standard modality is diagnosing such rare diseases. "Eye of Tiger" sign is nearly pathognomonic for hallervorden spatz disease. Timely action taken by the physicians and surgeons on the proper radiological reports can save the life of patient. Thus, imaging can play a momentous role in making precise diagnosis on time.

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