Chiari Malformation Presenting as Cerebellar Degeneration

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Introduction

Computed tomography (CT) is the primary imaging modality in the investigation of suspected cerebellar degeneration. A case is presented in which an inaccurate clinical and radiological diagnosis of cerebellar degeneration resulted from CT imaging demonstrating loss of cerebellar substance. Subsequent clinical recognition of down beat nystagmus led to MR examination and a diagnosis of Chiari malformation was made. Visualization of the loss of cerebellar substance should not be considered to exclude Chiari malformation.

Case Report

A 34 year old right handed male, not on any antiepileptics, presented with feeling of imbalance and tendency to fall since last three years. Onset was insidious with slowly progressive course. There was no history of alcohol abuse or any other substance, seizure, trauma, toxic exposure or surgery. There was no family history of similar complaints.

General examination was unremarkable. Fundus examination was normal. Initial neurological examination revealed horizontal and rotatory nystagmus on lateral gaze

with mild dysarthria. Gait was ataxic and wide based with inability to do tandem walking. Motor and sensory systems were normal. Routine laboratory investigations were within normal limits. CT head revealed generalized atrophy of cerebellum with prominent folia. Left hemisphere was affected more than the right with vermis also showing atrophic changes. There was prominence of pericerebellar cisterns more so on the left side (Fig 1). Further testing showed subtle down beat nystagmus. Down beat nystagmus, however, a type of central vestibular nystagmus frequently found in lesions near cervicomedullary junction [1], prompted a search for a Chiari malformation. MR examination confirmed cerebellar degeneration (Fig 2) and also revealed tonsillar descent below the foramen magnum (Fig 3). There was no syringohydromyelia or any other brain anomaly confirming the diagnosis of Chiari type I malformation with cerebellar degeneration.

Discussion

The prevalence of the Chiari type I lesion in the normal population was formerly grossly overestimated but is probably under 1% [2]. It is also becoming clear that it is an acquired deformation, the mechanism often seeming to be the posterior fossa growing at a slower

Fig. 1 : CT showing cerebellar degeneration with prominent pericerebellar cisterns

Fig. 2 : MRI T1W axial section confirms the findings seen in Fig. 1.

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