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Case Report Open Access

Progressive Ataxia Revealing Gluten Sensitivity

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Background

Ataxia is the commonest neurological manifestation of coeliac disease. Some individuals with genetic susceptibility to the disease have serological evidence of gluten sensitivity without gastrointestinal symptoms or evidence of small-bowel inflammation. The inaugural manifestation of disease in such patients may be ataxia. We describe the clinical, radiological, and neurophysiological features of this disorder [1].

Case Report

A 67 year- old-man, without any medical history, presented with progressive late onset gait disorder since 6 years. Neurological examination revealed a horizontal gaze-evoked nystagmus, severe truncal ataxia and scanning speech. His achileen reflexes were absent and tactile sensibility in lower extemities decreased bilaterally with distal predominance. His scale for the assessment and rating of ataxia (SARA) score reachs 22/40 (Schmitz-Hubsch 2004). Brain MRI showed important cerebellar atrophy (Figures 1-3). The electrophysical findings showed a sensorimotor axonal neuropathy. Laboratory tests for Alpha-fetoprotein, Vitamin E, serum protein electrophoresis, plasma cholesterol, creatine-kinase, acanthocyte were normal. Analyses of cerebrospinal fluid with dosage of anti Yo and anti-Hu were also normal. His anti-gliadine antibodies (AGA) IgA titer was positive and his AGA IgG titer was negative. The anti-transglutaminoses antibodies (ATG) titer was positive and anti-endomisium antibodies titer was positive. Duodenal biopsy was normal. The diagnosis of gluten ataxia was established based on clinical and biological data. For secure diagnosis a gluten free diet was initiated. The patient was treated by intraveinous corticotherapy during 5 days with high doses (1 gram/day) relayed by oral corticotherapy using 60 milligrams per day. His neurological symptoms showed marked improvement and his SARA score decreased to 13/40 points after 2 months.

Discussion

The term gluten sensitivity refers to a state of immunological responsiveness to ingested gluten in genetically susceptible individuals (HLA DQ2 Genotype) [1]. In several studies, patients with sporadic ataxia had a high frequency occurrence of AGA antibodies compared with healthy controls. All patients have gait ataxia and most have limb ataxia. Gluten ataxia usually presents with pure cerebellar ataxia or in combination with myoclonus, Palatal tremor [2,3], Opsoclonus [4], or Chorea [5]. Gluten ataxia usually has an insidious onset with a mean age at onset of 53 years like our case report. Rarely, the ataxia can be rapidly progressive, mimicking paraneoplastic cerebellar degeneration. Gaze-evoked nystagmus and other ocular signs of cerebellar dysfunction are seen in up to 80% of cases [2]. Less than 10% of patients with gluten ataxia will have any gastrointestinal symptoms but a third will have evidence of enteropathy on biopsy and sensorimotor lengthdependent axonal neuropathy in up to 60% of patients [2]. Antiendomysium antibodies are detectable in only 22% of patients [2], anti-TG2 IgA antibodies are present in up to 38% of patients with gluten ataxia, but often at lower titres than those seen in patients with coeliac disease [2]. This finding is in line with data that have provided evidence for intrathecal antibody production against TG in patients with neurological diseases [6]. Upto 60% of patients with gluten ataxia have evidence of cerebellar atrophy on MRI [7]. The response to treatment with a gluten-free diet depends on the duration of the disease that's why prompt treatment is more likely to improve or stabilize the ataxia [8]. The use of intravenous immunoglobulins can be helpful but it is still in trial [9]. A treatment with corticoides offer good results on neurological manifestations especially for ataxia which usually did not respond to diet [10]. Neuropathologic findings found in patients with gluten ataxia when autopsied showed perivascular cuffing with inflammatory cells, predominantly affecting the cerebellum, and resulting in loss of Purkinje cells implying that the neurologic insult may be immune mediated. It is yet unknown whether such immune-mediated damage is primarily cellular or antibody driven. Despite it was complex, gluten sensitivity [11] can be considered as a potential curable etiology of ataxia and should be treated by association of gluten diet free and an anti-inflammatory therapy such as corticotherapy (or) immunoglubuline which can offer in some cases a considerable improvement [12].

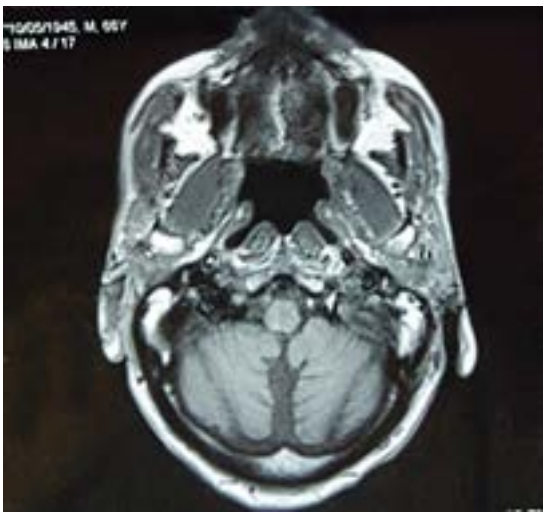

Figure 1: Axial cerebral MRI of the Patient (T_1 weighted): cerebellar atrophy.

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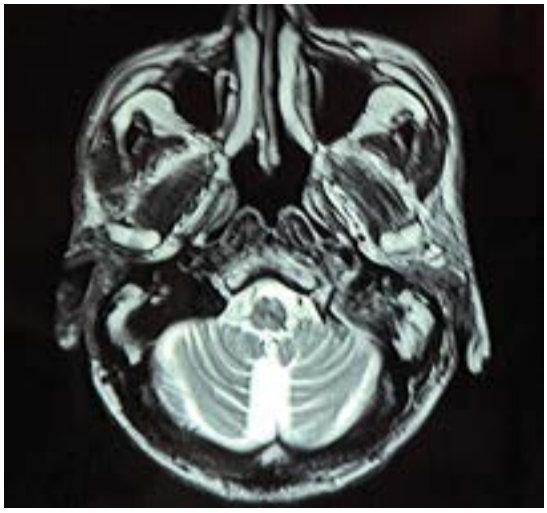

Figure 2: Axial Cerebral MRI of the Patient (T_2 weighted): Cerebellar Atrophy.

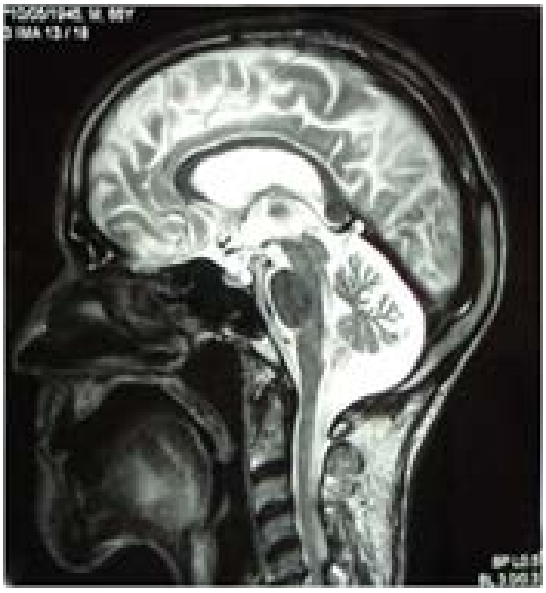

Figure 3: Sagittal Cerebral MRI (T_2 weighted): Cerebellar Atrophy.

Conclusion

Gluten sensitivity seems to be a major factor in some neurological disorders including auto-immune disorder. The timing of the diagnosis and treatment of these patients appears to be crucial because of the loss of Purkinje cells which is irreversible. Thus, antigliadin antibodies should be an essential part of the investigation of patients with sporadic idiopathic ataxia at first presentation. If diagnosis was performed, there is no time to loss and immediate gluten diet free with corticotherapy should be started to avoid irreversible sequellae.

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