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Gluten Ataxia: Cerebellar Ataxia Complicating Celiac Disease: A Case Report in Pediatrics

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Summary

Celiac disease is an autoimmune enteropathy related to gluten intolerance that can have several neurological complications, including cerebellar ataxia. The mechanism involved remains poorly understood. The gluten-free diet is the mainstay of therapy, and allows a dramatic improvement in these symptoms.

***Observation:** We report the observation of a 10-year-old patient, followed in our training for celiac disease since 2014 confirmed by a positive IgA anti transglutaminase rate and a jejunal biopsy in favor of villous atrophy, initially put on a gluten-free diet with poor compliance, she presented with a picture of cerebellar ataxia evolving in a context of apyrexia.*

The clinical examination on admission showed a child with cerebellar ataxia. A cerebral CT scan and a lumbar puncture were done in first intention in order to eliminate all diagnostic and therapeutic emergencies. During her hospitalization in the department, the patient benefited from an MRI showing cerebral atrophy, as well as an anti-transglutaminase and anti-gliadin antibody assay. Therapeutically, the introduction of a strict gluten-free diet allowed an improvement of the symptomatology, which allowed us to retain the celiac origin of this ataxia.

***Conclusion:** Celiac ataxia remains a neurological complication very rarely observed in children. Reducing the duration of exposure to gluten with the Gluten Free Diet will improve the prognosis of these neurological disorders.*

Introduction

Celiac disease is an autoimmune enteropathy related to gluten intolerance that regresses after dietary exclusion of wheat gluten and equivalent prolamins from other cereals considered toxic (rye, barley and to a lesser extent, oats). Various extra digestive manifestations may occur in unusual forms of the disease. Cerebellar ataxia is one of these rare manifestations, present in 9% of cases [1,2], the mechanism involved remaining poorly understood. We report a pediatric case of cerebellar ataxia complicating celiac disease.

Observation

This is a 10-year-old girl from a non-consanguineous marriage with no particular pathological history, in particular no previous similar episode or similar case in the family, no flu-like syndrome preceding the symptomatology and no notion of dysimmune terrain or neoplastic pathology followed in our training for celiac disease since 2014 confirmed by a positive IgA anti transuglutaminase and antigliadin rate, a jejunal biopsy showing villous atrophy as well as a positive HLA DQ2 genotyping. Initially put on a gluten-free diet with poor compliance,

She presented with a cerebellar ataxia evolving in a context of apyrexia. The clinical examination on admission showed a child with cerebellar ataxia with walking and balance disorders that had been progressively developing for 4 months. There were no associated neurological or extra neurological signs. The patient was apyretic without alteration of the general state.

The clinical examination revealed a pale skin and mucous membranes, and a static cerebellar syndrome, consisting of a "shaky walk", an enlargement of the sustentation polygon, and a dance of the anterior leg tendons, without any deep sensitivity disorders. A cerebral CT scan and a lumbar puncture were performed as a first line of treatment in order to eliminate all diagnostic and therapeutic emergencies. Cerebellar damage was suspected, requiring a cerebral magnetic resonance imaging, which showed cerebellar atrophy. The biological workup showed a microcytic hypochromic anemia at 9 g/dl, with a low level

of serum iron and vitamin B12, and normal VitE. Anti-endomysium and anti-transglutaminase antibodies were clearly positive. The diagnosis of a cerebellar ataxia complicating a celiac disease was retained. Thus, a strict gluten-free diet with correction of deficiencies was introduced. The evolution was marked by clinical improvement (weight gain, disappearance of the ataxia).

Discussion

Celiac disease is an autoimmune enteropathy related to gluten intolerance. Its prevalence is high if one takes into account not only the symptomatic forms, but also the frustrated and latent forms (1/300) [3,4]. The two peaks of frequency are early childhood, most often between six months and two years of age, and young adulthood between 20 and 40 years of age, with a female predominance.

Cerebellar ataxia represents a formidable complication of this disease, it was described in 1966. Representing 40% of ataxias of unknown origin, 15% can appear in people of all ages, including children [7].

Cerebellar damage in CD is related to destruction of cerebellar Purkinje cells, triggered by gluten intake [7]. The presence of lymphocytic infiltrates in the cerebellum, the posterior cords of the medulla and the peripheral nerves pleads for an immunological mechanism at the origin of neuronal destruction; the pathogenic role of anti-neuronal antibodies has been evoked. Antibodies directed against Purkinje cells have been detected in the serum of patients suffering from Coeliac Disease with ataxia, with a cross-reactivity between antigliadin antibodies and certain epitopes of Purkinje cells [13]. There seems to be a relationship between the duration of exposure to gluten and the severity of ataxia and cerebellar atrophy [7]; this may explain the reversible or non-reversible character of cerebellar ataxia, depending on the duration of its evolution, after strict GFD [9].

Neuropsychiatric manifestations remain one of the most debated issues in celiac disease complications, accounting for 10% of cases [6].

In 1966, Cooke and Thomas-Smith published an article on neurological disorders associated with celiac disease [8]. Several observations were subsequently reported concerning patients who presented with ataxia complicating celiac disease, however, neurological manifestations may not only complicate the disease but may also be the only manifestation [9].

However, neurological manifestations may not only complicate the disease but also be the only manifestation [9]. antigliadin antibodies have a high sensitivity not only for patients with celiac disease but also for patients with minimal or no lesions of the gut and where the primary target organ is the cerebellum or peripheral nervous system. The evidence supporting this claim comes from HLA genotyping of patients with neurological disorders associated with gluten sensitivity. Since celiac disease has one of the strongest HLA associations of all autoimmune diseases (HLA-DQ2 present in more than 90% of celiacs) one would expect that anti-gliadin antibody positive patients would have a similar HLA genotype if they are truly gluten sensitive.

In fact, 85% of patients with neurological disorders associated with gluten sensitivity have an HLA genotype compatible with celiac disease compared to 25% in the normal population [11]. Therefore, ataxias can be reversible with a gluten-free diet [12].

Conclusion

Cerebellar ataxia is one of the rare extradigestive complications of celiac disease and is very rarely observed in children. The gluten-free diet is the mainstay of therapy, so reducing the duration of exposure to gluten will improve the prognosis of these neurological disorders.

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