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RESEARCH ARTICLE

A CHALLENGING CASE OF CEREBELLAR ATAXIA IN A CHILD

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Abstract

Gluten ataxia is one of the extra-intestinal manifestations of celiac disease. It is a rare condition in children, mainly seen in genetically predisposed persons. We report the case of an acute onset of ataxia in a three years-old child treated for celiac disease with excellent compliance to free-gluten diet. The diagnosis and management were challenging.

Introduction:-

Gluten ataxia (GA) is an immune-mediated disease triggered by dietary intake of gluten, associated with cerebellar symptoms, often related to genetic susceptibility. To date, there are no guidelines on how to manage it. We report the case of a 3 years old child diagnosed with celiac disease (CD), who had an acute onset of cerebellar ataxia (CA).

Case report:

A three years-old boy presented to our center for diarrhea evolving for 18 months. Past medical history found a dietary intake of gluten starting at twelve-months old, and a failure to thrive and loss of gait at two-years old. Physical examination showed severe malnutrition and failure to thrive (Height=-3SD, weight=-4SD). He had generalized edema, clinical signs of rickets, and generalized cutaneous itchy blisters. Laboratory workup found features of malabsorption: iron deficiency anemia, low blood albumin levels, hypocholesterolemia, and low total vitamin D level. The anti-gliadin antibodies level was very high (more than eight times the normal range for IgA, and more than forty times for IgG). He had positive HLA DQ8 and negative HLA DQ2. Cutaneous biopsy found a dermatitis herpetiformis. We started gluten-free diet (GFD). Compliance was excellent. Diarrhea was persistent, so oral metronidazole was given for seven days. After what, zinc, iron and vitamin D supplementations were started. After two weeks of GFD, the child had no more diarrhea or edema. He begun gaining weight. Meanwhile there was no improvement of gait. After four weeks of GFD, the child had an acute onset of generalized tremor, myoclonus, and facial central paralysis. Neurological evaluation found CA. Calcium, magnesium, zinc, vitamin B12 and B9 blood levels were normal. Brain magnetic resonance imaging (MRI) found multiple periventricular and cerebellar hypersignals (figure 1). Screening for autoimmune diseases was negative (antinuclear antibodies, anti NMO antibodies, anti MOG antibodies). Oral sodium valproate was started. GA was highly suspected, so we started methyl prednisolone pulses (1 g/1.73m^2/day for three days in a raw). Afterwards, we started prednisone (2mg/kg/day). After Two weeks of steroid therapy, there was a mild improvement of gait, no more facial paralysis, subtle myoclonus and persistent tremor. A unique pulse of 2 g/kg of IV immunoglobulins (Igs) was than given. After four weeks of Igs, the child had no more neurological symptoms and a normal gait. Tapering of oral steroids by 0.2 mg/kg/month was then started. At six months follow up, he was still asymptomatic. His vitamin D and hemoglobin levels normalized.

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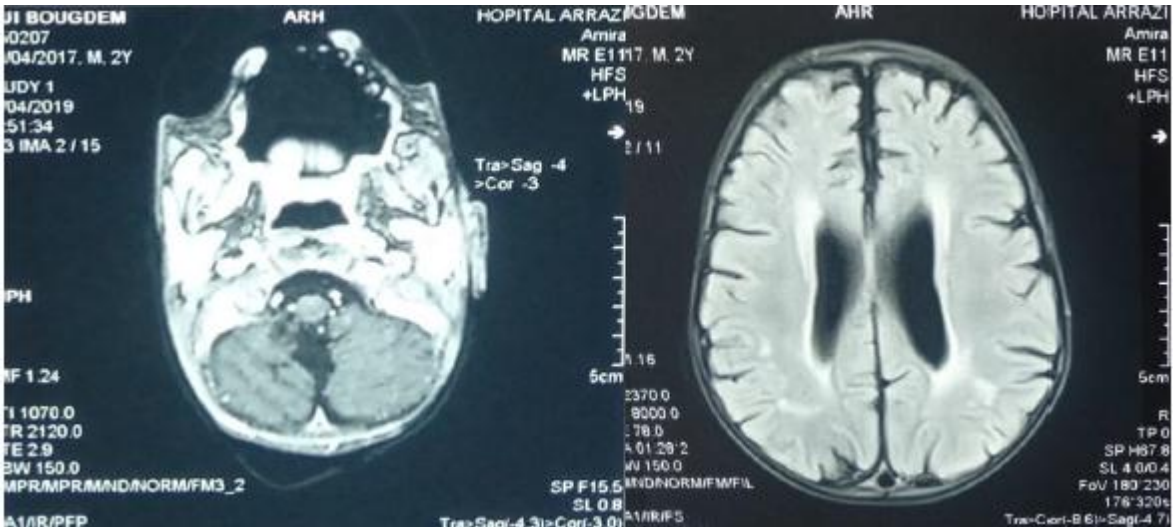

Figure 1:- Multiple periventricular and cerebellar hypersignals on MRI.

Discussion:-

GA is one of the extraintestinal manifestations of gluten sensitivity (GS) that have only been recognized in the last decade [1]. It is currently defined as CA associated with the presence of serological evidence of GS [2]. It is due to a patchy loss of Purkinje cells throughout the cerebellar cortex. There is marked perivascular cuffing with inflammatory cells, mainly T lymphocytes within the cerebellar white matter. These findings are in favor of an immune-mediated pathogenesis [3]. GA is characterized by insidious onset of predominantly gait ataxia, often associated with symptoms and signs suggestive of peripheral neuropathy (60%) [4]. By definition, all patients will have positive IgG and/or IgA antigliadin antibodies. The HLA type DQ2 is seen in 70% of these patients [5]. A few pediatric cases were described in literature. Their management was based on the results of adult's case reports and trials. The benefit of a strict gluten-free diet in the treatment of GA is controversial, and partial remissions or relapses are frequent. In this case, the drugs used are oral and/or IV steroids and intravenous Igs [1]. The risk with GA is permanent disability, that is why early diagnosis and treatment are imperative.

Conclusion:-

The diagnosis of GA should be raised in all pediatric cases of cerebellar ataxia, and anti-gliadin antibodies should be dosed. In our case, the strict compliance to gluten free diet did not improve the ataxia. However, the combination of steroids and IV Igs was effective, and total remission was reached after only one month of the treatment. More studies are necessary to determine the best way to manage this rare condition.

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