Immune-mediate cerebellar ataxias in children: a case report

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Objective

To describe a case of a child with cerebellar ataxia of immune origin.

Patient and methods

A 2 years old white female was admitted to our hospital for an acute symptomatology characterized by irritability, slowed speech, difficulties in walking, limb incoordination; over a period of a week she suffered from vomitimg and diarrhea. On the neurological examination the child was unable to walk and scanning speech, intention tremor and ataxia were reported. Serum biochemical parameters analyse, electroencephalogram (EEG), cerebrospinal fluid analysis, brain computed tomography (CT) scan and magnetic resonance imaging (MRI) of the brain and spinal cord were performed.

Results

Laboratory data (complete blood count, electrolytes, renal and liver function tests, thyroid function, erythrocyte sedimentation rate, C-reactive protein), EEG, brain CT and MRI were negative, so a diagnosis of acute cerebellitis was made; therefore, treatment was started with methylprednisolone, without any benefit.
Given the dramatic and rapid progression of symptoms and lack of response to treatment, an immune/paraneoplastic origin of the syndrome was suspected and chest radiography, abdominal ultrasound, antibody screening (Abs to RI, YO, CV2 ,Hu, GAD, amphiphysin) and tumor markers were performed.
Antibody screening showed increased levels of Abs anti CV2. The patient was treated with immunoglobulin and the dose of methylprednisolone was increased. After a few weeks, a clinical improvement with complete remission of symptoms was observed. Long-term cortisone therapy and prolonged follow-up to exclude the onset of neoplasm have been programmed. After three months Abs anti CV2 values have become normal.

Discussion

Immune mediate cerebellar ataxias include cerebellar ataxia associated with anti GAD antibodies, Haschimoto's encephalopathy, primary autoimmune cerebellar ataxia, gluten ataxia, Miller Fischer syndrome, ataxia associated with systemic lupus erytematosus and paraneoplastic cerebellar degeneration **(Tab.1)**. Humoral mechanism, cell mediated immunity, inflammation and vascular injuries contribute to cerebellar deficits. Antibodies to CV2 are often associated with an underlying neoplasm such has small cell lung cancer or thymoma and neurological syndromes include limbic encephalitis, chorea, encephalomyelitis and cerebellar involvement, characterized by symmetrically distributed static and kinetic symptoms. Treatment consists of intravenous followed by oral high dose corticosteroids and intravenous immunoglobulins. No underlying cancer was found in our patient, so we made a diagnosis of immune mediated cerebellar ataxia.

Table 1 The classification of immune-mediated cerebellar ataxias

1. Autoimmunity that mainly targets the cerebellum[a] or its related structures[b]:	
Cerebellar autoimmunity not triggered by another disease:	
Anti-GAD Abs associated cerebellar ataxia	
Cerebellar type of Hashimoto's encephalopathy	
Primary autoimmune cerebellar ataxia	
Others	
Cerebellar autoimmunity triggered by another disease or condition:	
Gluten ataxia	(gluten sensitivity)
Acute cerebellitis	(infection)
Miller Fisher syndrome	(infection)
Paraneoplastic cerebellar degenerations	(neoplasm)
2. Autoimmunity that simultaneously targets various parts of the CNS:	
Multiple sclerosis	
Ataxia in the context of connective tissue diseases such as SLE	

Table 1. modified from Zuliani L. et al JNNP 2012

Conclusion

Immune mediated cerebellar ataxia is a condition which must be included in the differential diagnosis of cerebellar dysfunction in children. Early diagnosis is important for the detection and the successful treatment not only of the autoimmune syndrome, but also of the underlying disease.