Encephalitis associated with autoimmune thyroiditis : a rare cause of encephalopathy in children

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Introduction

We report here the case of a patient presenting persistent status epilepticus with elevated antithyroid antibodies suggesting the diagnosis of Hashimoto encephalitis.

Hashimoto encephalitis is relatively rare in peadiatric population but has to be evocated regarding the association of acute encephalopathy with elevated antithyroid antibodies after exclusion of other etiologies (infection, tumor, toxic, metabolic).

Case Report

Reason for consulting

A ten year old girl was brought by the emergency mobile service for a **persistent status epilepticus** despite administration of Diazepam on site.

She had presented **headaches** for the last three days and one episode of **fever** was suspected but not measured four days earlier. She had **one episode of dizziness** five days earlier.

Her personal history was without particularity. Her mother has an auto-immune thyroiditis. The father's sister has a diabete mellitus and a crohn's disease.

Evaluation

- Inflammatory tests: normal
- Cerebrospinal fluid analysis: normal NMDAR, GABA B-R, Glutamate R and Glycine R antibodies : negative
- Bacteriological and viral cerebrospinal fluid analysis: normal

- Brain Scan: normal
- Magnetic resonance imaging: **vasogenic oedema**
- EEG: right status epilepticus, evolution to right focal epileptic discharges

- Laboratory evaluation: Moderate **hypothyroidism** with **high anti-TPO antibodies (anti-TPO : 250 UI/L; anti-thyroglobuline: 371 UI/mL)**
- Thyroid ultrasound: appearance of **thyroiditis**

Diagnosis and management

Diagnosis:
Auto-immune encephalitis with high anti-TPO antibodies

Hashimoto Encephalitis

Management: high-dose corticosteroid therapy followed by plasmapheresis which lead to clinical improvement.

Start **L-thyroxine** treatment

Discussion

- Hashimoto encephalitis is also known as Steroid responsive encephalopathy associated with autoimmune thyroiditis (SREAT)
- Association of acute encephalopathy with elevated antithyroid antibodies after exclusion of other etiologies (infection, tumor, toxic, metabolic) suggests the diagnosis
- Most pediatric patients present with slowly progressive encephalopathy with epileptic seizures as the most common symptom (60–80%) which require intensive treatment in order to limit brain damage
- It appears that thyroid antibodies are not directly responsible for brain damages, and the abnormalities in thyroid hormone levels are generally too mild to explain the brain disease. Other auto-immunue encephalopathies in children are associated with other autoantibodies, such as anti- NMDAR or GABA-B-R
- The standard treatment consists in a systemic corticosteroid therapy and/or plasma exchange, while other lines of immunotherapy are sometimes needed. Antithyroid antibody titer can be used to predict responsiveness to treatment in the acute stage, but cannot be used as a marker of relapse

Conclusion

Hashimoto encephalitis is a relatively rare cause of encephalitis in children but must be evocated when acute encephalopathy is associated with elevated antithyroid antibodies.