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Case Report

Hashimoto's encephalopathy revealed by behavioral disorder: A Case Report

Behavioral disorder in Hashimoto's encephalopathy

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Abstract

Introduction: Hashimoto's encephalopathy (HE) is defined by the association of encephalopathy and increased thyroid antibodies, with various modes of presentation. However, behavioral disturbances are rare and exceptionally revealing in pediatrics.

Case Presentation: We report the case of an 11-year-old girl with no significant medical history, admitted to the pediatric neurology unit for behavioral disturbances, preceded by a convulsive episode resembling an absence seizure. The biological workup was normal, and the radiological assessment revealed no abnormalities. Immunological tests showed elevated anti-thyroperoxidase antibody levels. The diagnosis of Hashimoto's encephalopathy was made, and the patient was treated with a corticosteroid bolus, with favorable progression.

Conclusion: Given the wide variety of symptoms, clinical suspicion is essential. Although it may seem excessive, this diagnosis should be considered in the absence of other causes of encephalopathy, as it justifies corticosteroid treatment, and a favorable response can serve as a recurrent diagnostic criterion.

Keywords

Hashimoto's encephalopathy, behavioral disturbances, child, anti-thyroperoxidase antibodies, corticosteroids

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Introduction

Hashimoto's encephalopathy, or encephalopathy associated with anti-thyroperoxidase (anti-TPO) antibodies, is a rare autoimmune condition, well described in adults, but it can also affect children.[1] Its pathophysiology remains poorly understood, and it can present in various forms. It is characterized by nonspecific neurological and/or psychiatric symptoms. It is associated with increased thyroid antibody levels in the serum and, sometimes, in the cerebrospinal fluid (CSF) of affected patients. Electroencephalographic (EEG) abnormalities are nonspecific, as are those found on cerebral magnetic resonance imaging (MRI). It often responds to corticosteroids.[2] In this work, we will detail the clinical, epidemiological, pathophysiological, and therapeutic aspects of Hashimoto's encephalopathy in children, in light of the available medical literature and our clinical observation.

The aim of this case report is to present a pediatric case of Hashimoto's encephalopathy presenting with behavioral disturbances and to highlight the importance of early recognition and corticosteroid therapy.

Case Presentation

This concerns an 11-year-old girl with no significant medical history, admitted to the pediatric neurology department for behavioral disturbances, which were preceded by a 20-day history of a seizure episode, resembling an absence seizure. Upon admission, her Glasgow Coma Scale (GCS) was 15. She presented with behavioral disturbances, including hetero-aggressiveness and incoherent speech. The rest of the neurological examination was unremarkable. Her vital signs were stable; she was hemodynamically stable and afebrile. There were no meningeal signs or focal deficits. The initial biological tests were normal, including renal and liver function tests, C-reactive protein, sedimentation rate, toxicology screens for blood and urine, viral serologies (hepatitis, cytomegalovirus, Epstein-Barr virus, HIV, herpes, rabies), and metabolic testing. Brain MRI and CSF examination were normal. EEG showed normal background activity with diffuse slow-wave bursts predominantly in the anterior regions. Further investigations, including a serum and CSF profile for autoimmune encephalitis (anti-NMDA antibodies), were normal. Ophthalmological examination was normal. The patient was treated with intravenous immunoglobulins (2g/kg) without improvement. Testing revealed elevated anti-thyroperoxidase (anti-TPO) antibodies (54.5 UI/ml, normal: < 5.6 UI/ml). The thyroid profile was normal (TSH: 3.2 µUI/ml, range: 0.7-6.4 µUI/ml), and the cervical ultrasound was normal. A diagnosis of Hashimoto's encephalopathy (HE) was made, and treatment with intravenous methylprednisolone bolus (30mg/kg/day) was initiated, with a spectacular response to steroid treatment after 5 days. The patient was discharged from the hospital and had a favorable outcome without relapse after 3 months.

Ethical Approval

Not applicable.

This case report was prepared in accordance with the CARE guidelines.

Discussion

Hashimoto's encephalitis, or encephalitis associated with autoimmune thyroiditis (Steroid Responsive Encephalopathy Associated with Autoimmune Thyroiditis, SREAT), is a rare condition in children, with a prevalence estimated at 2/100,000.[3] It is defined by:

-An encephalitic presentation with cognitive disturbances, behavioral problems, neuropsychiatric signs (paranoia, hallucinations, psychosis), partial or generalized seizures, focal neurological deficits, consciousness disturbances, and dystonia.

-Elevated serum levels of anti-thyroperoxidase (anti-TPO) antibodies.

-Exclusion of other infectious, toxic, metabolic, autoimmune, or neoplastic causes.

-An effective response to corticosteroid therapy, either partial or complete.

Chronic forms, such as cerebellar syndrome, have been described, as well as myelopathies. These children are often hospitalized for recurrent, drug-resistant seizures or for behavioral problems and abnormal movements with no apparent cause. CSF analysis usually shows hyperproteinorachia, and EEG is almost always abnormal, showing either generalized background slowing or focal abnormalities. In 50% of cases, brain imaging reveals nonspecific abnormalities involving both white and gray matter. Regarding thyroid status, children are often euthyroid. A systematic measurement of anti-TPO antibodies is therefore necessary in cases of unexplained encephalopathy. Anti-TPO antibody levels are elevated and significantly higher than in typical pediatric Hashimoto's thyroiditis. The pathophysiology of this disease remains unknown, but several theories have been proposed, including cerebral vasculitis with or without immune complex deposition, which can disrupt the blood-brain barrier. The majority of children with anti-TPO encephalopathy progress to Hashimoto's thyroiditis, while the reverse is extremely rare.[1] This suggests the involvement of another, possibly associated, autoantibody that is directly responsible for the neurological involvement. Treatment is based on high-dose corticosteroids, with intravenous methylprednisolone bolus (30mg/kg/day) administered for 3 to 5 days, depending on clinical evolution. During the acute phase of the disease, the response to treatment is sometimes spectacular, with an "on/off" pattern.[4] Depending on the clinical course, oral corticosteroid therapy (1 mg/kg/day for 15 days) may be considered.1 Recurrences are possible, and treatment with immunosuppressive agents, such as cyclophosphamide or azathioprine, may be discussed based on clinical evolution, particularly if steroid dependence is observed. The clinical outcome is generally favorable in the majority of cases, but disabling sequelae such as recurrent seizures and cognitive decline have been described.[4-5]

Limitations

This report describes a single case; therefore, the findings cannot be generalized, and larger studies are needed to confirm these observations.

Conclusion

Hashimoto's encephalopathy, a rare yet curable condition in children, remains a poorly understood and often overlooked disease, lacking clear diagnostic criteria. Keeping this diagnosis in mind could allow for its consideration and confirmation. Advances in fundamental immunology have improved understanding of its pathophysiology, though much remains to be clarified.

Declarations

Animal and Human Rights Statement

All procedures were performed in accordance with the ethical standards of the institutional and/or national research committee and the 1964 Declaration of Helsinki and its later amendments, or comparable ethical standards.

Informed Consent

Written informed consent was obtained from the patient's parent for publication of this case report and the accompanying images.

Data Availability

The datasets used and/or analyzed during the current study are not publicly available due to patient privacy reasons, but are available from the corresponding author on reasonable request.

Conflict of Interest
The authors declare no conflict of interest.

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The authors declare that no AI-assisted technologies were used.

Abbreviations
CSF: Cerebrospinal fluid
EEG: Electroencephalography
GCS: Glasgow Coma Scale
HE: Hashimoto's encephalopathy
MRI: Magnetic resonance imaging
TPO: Thyroperoxidase
TSH: Thyroid-stimulating hormone

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