HASHIMOTO'S ENCEPHALOPATHY: AN UNUSUAL CAUSE OF SEIZURES

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ABSTRACT

Hashimoto's encephalopathy is a rare, chronic relapsing and remitting encephalopathy associated with auto-antibodies against thyroid components. Clinically it is characterized by tremor, myoclonus, stroke-like episodes, seizures, impairment of consciousness, and dementia. We described here a 13-year-old boy who presented with generalized tonic clonic seizures associated with Hashimoto's encephalopathy. After corticosteroid treatment, his neurological features improved dramatically. As a result, Hashimoto's encephalopathy should be considered in cases of unexplained encephalopathy and seizures in pediatric patients.

Key Words: *Hashimoto's encephalopathy, seizure, child*
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EPİLEPTİK NÖBETİN NADİR BİR NEDENİ OLAN HASHİMOTO ENSEFALOPATİSİ OLGUSU

ÖZET

Hashimoto ensefalopatisi, nadir görülen, tiroid antikorları ile ilişkili relaps ve remisyonlarla seyreden bir ensefalopati tablosudur. Klinik olarak, tremor, miyoklonus, nöbetler, inme benzeri ataklar, bilinç bozukluğu ve demansla karekterizedir. Jeneralize tonik klonik nöbetle başvuran 13 yaşındaki bir erkek Hashimoto ensefalopatisi olgusu sunulmaktadır. Kortikosteroid tedavisinden sonra hastanın nörolojik bulguları dramatik olarak düzeldi. Sonuç olarak, Hashimoto ensefalopatisi çocuk hastalar arasında nedeni açıklanamayan ensefalopati ve nöbet olgularında akla gelmelidir.

Anahtar Kelimeler: Hashimoto ensefalopatisi, nöbet, çocuk. **Nobel Med 2012; 8(2): 121-122**

INTRODUCTION

Hashimoto encephalopathy (HE) is a rare, steroid-responsive disorder associated with Hashimoto thyroiditis resulting in variable clinical manifestations such as seizures, behavioral and psychiatric manifestations, movement disorders, and coma.[1] The underlying pathology is not completely understood, but evidence of autoimmune mechanisms has been thought. HE is mainly diagnosed in the adult population, but is rare in children. Differential diagnosis is important in especially paediatric patients with encephalopathy. We observed a patient with HE who presented with generalized seizures.

CASE REPORT

A 13-year-old boy presented to the hospital with generalized tonic-clonic seizures. He complained progressive muscle wasting, fatigue, being withdrawn, deteriotation in his school performance and intermittent generalized headache for he last 12 months. His previous medical history and family history were unremarkable.

On examination, pulse rate was 70/min and blood pressure was normal. There was no palpable goiter. Neurologic examination revealed decrease mental alertness. His face appeared hypomimic, and there was minimal dystonia in upper extremities. Other examinations were within normal limits.

In laboratory, full blood count, urea and electrolytes, liver function tests, lactate and ammonia were normal. Thyroid function tests revealed a hypothyroid state with increased serum levels of thyroid-stimulating hormone (TSH 11.2 mIU/mL; normal, 0.40-4.0 mIU/mL) and reduced free T4 (0.83 pmol/L; normal,→

10.3-24.4 pmol/L) levels. High levels of antithyroid antibodies were present, with antimicrosomal antibodies was 560 IU/mL (normal, 0-60 IU/mL), and antithyroglobulin antibodies was 111.1 IU/mL (normal, <40 IU/mL). Thyroid ultrasound was suggestive of thyroiditis. Electroencephalography (EEG) showed slowing of the background and epileptic activity on the left occipital region. Cranial magnetic resonance imaging was normal. He was started levothyroxine and oral methylprednisolone 1mg/kg/day. Phenytoin was began first, and changed to oxcarbazepine for seizures. His neurological features improved dramatically, and he was seizure-free within 15 days.

DISCUSSION

Hashimoto encephalopathy is a rare, life-threatening but treatable condition associated with antithyroid antibodies. Thyroid status may be either euthyroid, hypothyroid and rarely even hyperthyroid. Generally, accepted diagnostic criteria are based on the presence of neurological or psychiatric symptoms, elevated anti-thyroid antibodies and corticosteroid responsiveness.[1] Our patient met all of these three criteria.

The clinical features of HE are heterogeneous, and a high degree of suspicion is necessary for diagnosis. The clinical presentation may be stupor, coma, tremor, myoclonus, seizures, neuropsychiatric changes (impairment of cognitive functions, behavioral and mood disturbances, hallucinations) or focal neurological deficits.[1] Seizures are reported in 66% of HE patients, especially in children. Generalized tonic clonic followed by complex partial seizures, are the most common types of seizures.[2] Focal motor seizures and status epilepticus have also been reported.[1] Alink et al., reported 25 (80%) HE patients who presented with seizures.[3] Hoffmann et al., also reported a 6 year-old girl with progressive epilepsy resistant to anticonvulsive treatment and unclear encephalopathy related to HE. Berger et al., described a patient with HE, who suffered from HE causing refractory epilepsy with progressive cognitive decline.[4,5] Our patient had generalized tonic-clonic seizures. Electroencephalogram changes are nonspecific in 90% of cases, usually manifest as intermittent slow wave activity.[6] The EEG in our patient, has shown generalised slowing and epileptic discharges on the left occipital region. Neuroimaging is usually normal, as seen in our patient. However, MRI may occasionally demonstrate bilateral subcortical high signal lesions on T2-weighted images.[1,2]

The pathophysiology of HE is much debated but remains speculative. Postulated mechanisms include an autoimmune cerebral vasculitis, an antineuronal antibody mediated reaction or toxic effect of thyroid stimulating hormone on the central nervous system.[2,7] From the therapeutic experience of the previously published cases of HE, steroids seem to be the most effective agent. Both high doses of oral prednisone and short courses of intravenous methylprednisolone have been shown effective, but pulse intravenous therapy is the preferred choice for acute encephalopathy of HE. Rapid improvement may be seen in a few days, and the mean time to significant clinical improvement is 3-4 weeks with the steroid theraphy. This case responded succesfully to therapy within 15 days, and at the end of the first month, he recovered dramatically.

CONCLUSION

In conclusion, HE is a rare, but likely underdiagnosed disease in children. It should be considered in the differential diagnosis with seizures, encephalopathies, and behavioral changes especially in children with unexplained and acquired clinical features to seizures.

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