LETTER TO THE EDITOR

Status epilepticus and Hashimoto’s encephalopathy

Jansen et al. reported a patient with marked hypothyroidism who developed status epilepticus.1 Although hypothyroidism in itself can lead to ataxia and cognitive dysfunction, we would like to propose the alternative diagnosis of Hashimoto’s encephalopathy (HE).2 This was first described by Brain in 1966; affected individuals present with neurological problems and autoimmune thyroid disease.1 The lady in question had a combination of thyroid dysfunction with psychosis, ataxia, seizures progressing to status epilepticus and cognitive impairment which are well described in HE.3-11 Other features of this condition include myoclonus, dementia and demyelinating peripheral neuropathy.3 Her other clinical signs and symptoms, such as hoarseness and myopathy, can be attributed to the profound hypothyroidism. HE is an uncommon entity; based on a small number of patients, it has an estimated prevalence of 2.1/100,000.8 Computed tomography of the brain and cerebrospinal fluid findings may be normal but levels of serum antithyroid antibodies, such as antithyroid microsomal, antithyroid peroxidase and antithyroglobulin antibodies are elevated. More recently autoantibodies against the amino terminal of α-enolase and intrathecal synthesis of antithyroid autoantibodies have been reported as useful markers.8,9 Given its low incidence, there have been few pathological analyses but some autopsy cases have revealed focal inflammatory cell infiltrates within the stroma of the thyroid gland, lymphocytic infiltrates around venules and arterioles and gliosis of gray matter in the cortex, thalamus, basal ganglia and hippocampi.6 The important point is that patients typically recover following corticosteroids and in some cases immunoglobulins. It is worthwhile to determine antithyroid autoantibody levels in patients with unexplained encephalopathies or unexplained seizures with thyroid dysfunction.12

A.C.F. Hui*, B.L. Man, W.H. Leung
Division of Neurology, Department of Medicine and Therapeutics, Prince of Wales Hospital, The Chinese University of Hong Kong, Shatin, Hong Kong, *corresponding author: tel.: +852-2632 31 31/2632 31 33, fax: +852-2637 53 96/2637 38 52, e-mail: cfhui@cuhk.edu.hk

REFERENCES

Response from the authors

We thank Hui et al, for their comment. We agree that Hashimoto’s encephalopathy (HE) should be in the differential diagnosis. HE refers to a syndrome of persisting or fluctuating neurological and neuropsychological deficits associated with elevated blood concentrations of antithyroid antibodies. Affected individuals are usually euthyroid or mildly hypothyroid4 and respond well to corticosteroid therapy.5 Furthermore, Hashimoto’s thyroiditis can be associated with other autoimmune diseases, such as Addison disease, autoimmune gastritis (pernicious anaemia), rheumatoid arthritis, systemic lupus erythematosus, celiac disease, and diabetes mellitus type 1.

© 2006 Van Zuiden Communications B.V. All rights reserved.

NOVEMBER 2006, VOL. 64, NO. 10
We think that some findings argue against HE, namely: 1) Our patient presented with extreme hypothyroidism, 2) Autopsy of the brain showed no abnormalities, and 3) there was no clinical response to corticosteroid therapy. In addition, there was no associated autoimmune disease present. Cerebrospinal fluid examination revealed no abnormalities, although a normal cerebrospinal fluid may be present in up to 25% of HE cases. Unfortunately, we did not measure any thyroid autoantibodies. We agree that thyroid autoantibodies should be determined in every patient with unexplained encephalopathy or unexplained seizures with thyroid dysfunction.

H.J. Jansen*, P.M. Netten
Jeroen Bosch Hospital, 's-Hertogenbosch, the Netherlands, *corresponding author: tel.: +31 (0)73-699 30 81, e-mail: h.jansen@aig.umcn.nl

REFERENCES