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.[3] 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.[10] 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, thalmai, basal ganglia and hippocampi.[6] The important point is that patients typically recover following corticosteroids and in some cases immunogloblins. 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

1. Jansen HJ, Doebe SR, Louwerse ES, van der Linden JC, Netten PM. Status epilepticus caused by a myxoedema coma. Neth J Med 2006;64:202-5.
2. Dugbartey AT. Neurocognitive aspects of hypothyroidism. Arch Intern Med 1998;158:1413-8.
3. Brain L, Jellinek EH, Ball K. Hashimoto's disease and encephalopathy. Lancet 1966;2:512-4.
4. McGinley J, McCabe DJ, Fraser A, Casey E, Ryan T, Murphy R. Hashimoto's encephalopathy: an unusual cause of status epilepticus. Ir Med J 2000;93:118.
5. Chong JY, Rowland LP, Utiger RD. Hashimoto encephalopathy: syndrome or myth? Arch Neurol 2003;60:164-71.
6. Duffey P, Yee S, Reid IN, Bridges LR. Hashimoto's encephalopathy: Postmortem findings after fatal status epilepticus. Neurology 2003;61:1124-6.
7. Chaudhuri A, Behan P. The clinical spectrum, diagnosis, pathogenesis and treatment of hashimoto's encephalopathy (recurrent acute disseminated encephalomyelitis). Curr Med Chem 2003;10:1945-53.
8. Ferracci F, Bertiato G, Moretto G. Hashimoto's encephalopathy: epidemiologic data and pathogenetic considerations. J Neurol Sci 2004;15;217:165-8.
9. Fujii A, Yoneda M, Ito T. Autoantibodies against the amino terminal of alpha-enolase are a useful diagnostic marker of Hashimoto's encephalopathy. J Neuroimmunol 2005;162(1-2):130-6.
10. Sheng B, Lau KK, Li HL, Cheng LF. A case of Hashimoto's encephalopathy with demyelinating peripheral neuropathy. Eur Neurol 2005;53:84-5.
11. Marshall GA, Doyle JJ. Long-term treatment of Hashimoto's encephalopathy. J Neuropsychiatry Clin Neurosci 2006;18:14-20.
12. Creutzfeldt CJ, Haberl RL. Hashimoto encephalopathy: a do-not-miss in the differential diagnosis of dementia. J Neurol 2005;252:1285-7.

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 hypothyroid[1,2] and respond well to corticosteroid therapy.[2,3] 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.

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

1. Henderson LM, Behan PO, Aarli J, et al. Hashimoto's encephalopathy: A new neuroimmunological syndrome. Ann Neurol 1987;22:140-1.
2. Kothbauer-Margreiter I, Sturzenegger M, Komor J, et al. Encephalopathy associated with Hashimoto's thyroiditis. Diagnosis and treatment. J Neurol 1996;243:585-93.
3. Hartmann M, Schaner B, Schelgmann K, Bucking A, et al. Hashimoto's-Enzephalopathie. Steroidsensitive Enzephalopathie bei Hashimoto-Thyroiditis. Nervenartz 2000;71:489-94.