Hashimoto's encephalopathy

By Prof Sarosh Irani, Mayo Clinic Florida, USA and reviewed by Assist Prof Omar Siddiqi, Harvard Medical School and Beth Israel Deaconess Medical Center, Boston, USA

Background Hashimoto's encephalopathy (HE) is a rare condition, which is probably of autoimmune origin. Autoimmunity describes disorders in which the immune system mistakenly attacks the body's own cells. HE can affect all age groups but typically affects females of around 50 years of age.

Recent insights into other forms of autoimmune encephalitis have taught us that HE may not represent a single diagnosis, but a syndrome which includes a several more specific conditions. Therefore, the symptoms and signs of HE are very varied and – as medicine advances – patients with HE may receive more accurate, alternative diagnoses which better describe their condition. Nevertheless, HE may a useful term to help guide treatments in a small number of patients, as described below.

What are the symptoms? The clinical presentation of HE (the symptoms) typically reaches its worst within a few weeks and often includes imbalance, episodes which look like strokes, psychiatric disturbances, jerky movements (some of which may be seizures) and sometimes coma. These are a vague list of symptoms – and so it is their co-occurrence which is key to considering HE.

What are the causes? Currently, the exact cause is unknown, but it is probably an autoimmune condition. The thyroid antibodies are thought likely to be a marker of the illness rather than the cause of the problem, but are very common in the general population. Hence, thyroid antibodies have very limited diagnostic value. Thyroid function is usually normal.

How is it diagnosed? HE is a diagnosis of exclusion. To diagnose HE it is important to rule out a wide range of other conditions including Creutzfeldt-Jacob disease, viral or other antibody-associated forms of encephalitis and dementia such as Alzheimer's. Useful tests to look for alternative diagnoses include magnetic resonance imaging (MRI) of the brain, the electroencephalogram (EEG), cerebrospinal fluid (CSF) analysis, hormonal and metabolic blood screens, and viral studies.

How is it treated? Most patients with HE improve with high dose steroids (most often prednisolone). Improvement may be rapid, taking days to weeks, but sometimes can take many months. Because of the improvement with steroids, some experts refer to the condition as “steroid responsive encephalopathy associated with autoimmune thyroiditis (SREAT)”.

In some patients who show a limited response to steroids doctors may use:

- plasma exchange (when some of a person’s blood is taken out from a vein, and the plasma part of the blood which contains antibodies is separated and replaced with new plasma and then put back into the vein in a drip)
- intravenous immunoglobulin (IVIG) (a blood product given into the vein in a drip)

The prognosis with treatment is generally good. Steroids are often continued for a few months.

Prognosis & outcomes of Hashimoto's encephalopathy Most people with HE recover well, but response to treatment can vary from person to person.

Research shows most HE patients respond to corticosteroids and many see significant improvement in a few months. In a study of 251 cases 91% of patients responded completely. While many do well, some may relapse and need longer or repeated treatment. If steroids don’t work, other options like azathioprine, IVIG or plasma exchange may be used.

Outcome for HE depends on how severe the condition is and how quickly treatment is started. Many make a full recovery but some may have ongoing cognitive or neurological issues. In rare cases if treatment is delayed or not given HE can be life threatening so early diagnosis and treatment is key.

How has the understanding of HE changed in the last decade? A number of autoantibodies, other than thyroid antibodies, have been described in patients with many similar features to typical cases of HE over the last decade. Therefore, the concept of HE is becoming fragmented into a number of other types of autoimmune encephalitis which appear to have their own autoantibodies, prognosis and associated features.

In addition, for those patients without autoantibodies, a long list of alternative diagnoses should be considered to ensure the right patients are being treated with the appropriate medications.

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Encephalitis International, 32 Castlegate, Malton, North Yorkshire, YO17 7DT, UK

Administration: +44 (0) 1653 692583 **Support**: +44 (0) 1653 699599

Email: mail@encephalitis.info **Website**: www.encephalitis.info