

Hashimoto's encephalopathy

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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 number of specific conditions. Therefore, the symptoms and signs of HE are varied. Nevertheless, it may a useful term to help guide treatments, 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 drowsiness, imbalance, episodes which look like strokes, psychiatric disturbances, jerky movements (some of which may be seizures) and sometimes coma.

What are the causes?

Currently, the exact cause is unknown although, but it is probably an autoimmune condition (please see above). The thyroid antibodies are thought likely to be a marker of the illness rather than the cause of the problem, and are very common in the general population. 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 whole range of other conditions including Creutzfeldt-Jacob disease, viral or other antibody-associated forms of encephalitis and dementia such as Alzheimer's. Useful tests 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 many months.

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.

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