Graves Disease with Hashimoto’s Encephalopathy

P Baburaj a, B V Shankara a, Edwin J George a, B L Harikrishnan b

a. Department of General Medicine, Jubilee Mission Medical College & Research Institute, East Fort, Thrissur, Kerala
b. Department of Emergency Medicine, Jubilee Mission Medical College & Research Institute, East Fort, Thrissur, Kerala*

ABSTRACT

The authors report the case of an elderly diabetic lady who presented with generalized fatigability, loss of weight, memory disturbances, irrelevant talk and disorientation. She had goiter and proptosis. Her thyroid function tests revealed hyperthyroidism and markedly elevated anti TPO antibody levels. Imaging modalities and uptake study were suggestive of Graves’ disease. The clinical findings, investigations of Graves’ disease with Hashimoto’s encephalopathy and treatment strategies are discussed here.

Keywords: Graves’ disease, Hashimoto’s encephalopathy, TSH receptor antibody

INTRODUCTION

Hashimoto’s encephalopathy is first described by Brain et al in 1966 and it is also known as Steroid -responsive encephalopathy associated with autoimmune thyroiditis (SREAT) and Nonvasculitic autoimmune meningoencephalitis (NAIM). Majority of patients are perimenopausal females with goiter and a family history of thyroid dysfunction. They can have two types of presentation; Acute stroke-like that accounts for 25% and diffuse progressive pattern of slow cognitive decline seen in 75% of the patients.

Two clinical patterns may overlap over the course of the disease. Exact pathogenesis is unknown but considered to be an autoimmune encephalopathy where precise role of antithyroid antibodies is unclear. Till date, no shared antigen has been identified between thyroid gland and brain, except alpha enolase.

CASE REPORT

Sixty-five years old Khairunnissa, who is a diabetic on irregular treatment came with complaints of generalized weakness, loss of weight, memory disturbances, irrelevant talk and disorientation. She had goiter and proptosis. Her thyroid function tests revealed hyperthyroidism and markedly elevated anti TPO antibody levels. Imaging modalities and uptake study were suggestive of Graves’ disease. The clinical findings, investigations of Graves’ disease with Hashimoto’s encephalopathy and treatment strategies are discussed here.

Keywords: Graves’ disease, Hashimoto’s encephalopathy, TSH receptor antibody

*See End Note for complete author details
Two-thirds of patients may experience focal or generalized tonic-clonic seizures, and 12% may present with status epilepticus. Also, myoclonus or tremor is seen in up to 38% of patients. The mechanism of Hashimotos encephalopathy does not appear to be related to the thyroid status, which can vary greatly from hypothyroid to hyperthyroid states. The presence of elevated antithyroid antibodies is an essential part of Hashimotos encephalopathy diagnosis, and suggests the presence of thyroid autoimmunity.

The pathogenic role of thyroid antibodies remains unknown, there is no evidence that they react with brain tissue or affects nerve function, and also no clear correlation between the severity of the neurologic symptoms and the concentration of these antibodies. Infrequently, the titers of antithyroid antibodies (TPOAb and TgAb) are measured in the CSF. A systematic review found that 13% of published cases of HE reported antithyroid antibodies in the CSF. However, the titers of antithyroid antibodies in the CSF do not correlate with the clinical stage of the disease, and the sensitivity and specificity of this finding remain unclear.

An autoantibody against the amino terminal end of the enzyme α-enolase, an antigen of the thyroid and the brain, has been identified as a potential biomarker of Hashimotos encephalopathy. A study found serum autoantibody reactivity in five of six patients with Hashimotos encephalopathy compared with two of 17 patients with Hashimoto's thyroiditis but no Hashimotos encephalopathy and in none of 25 healthy control subjects. This antigen is also found in endothelial cells, suggesting an autoimmune vasculitic mechanism; however, this has not been confirmed by neuroimaging techniques. An elevated protein concentration in CSF can be seen in 78% of patients, and in 20% of patients, it may be greater than 100 mg/dL.

The differential diagnoses of Hashimotos encephalopathy include stroke or TIA, cerebral vasculitis, carcinomatous meningitis, toxic metabolic encephalopathies, paraneoplastic syndromes, CJD, degenerative dementia and psychiatric diseases.

The long-term prognosis is variable, although a high percentage of patients respond to treatment; others could have a progressive or a relapsing course. The symptoms usually improve with glucocorticoid therapy; however, it is not necessary because of treatment. A systematic review of 85 cases published of Hashimotos encephalopathy found clinical response in 98% of patients treated with glucocorticoids, 92% of patients treated with glucocorticoids and levothyroxine.
and 67% of patients treated with levothyroxine only. Other measures include plasmapheresis, intravenous Immunoglobulin and steroid sparing drugs.

Although Hashimotos encephalopathy is usually associated with Hashimoto thyroiditis which predominantly present as hypothyroidism, this patient had features of Graves’ disease with raised anti-TPO antibodies which is found to be rare. Our patient had a dramatic improvement with steroid, beta blocker and carbimazole.

Clinical clues for diagnosis include presence of metabolic encephalopathy, goitre with variable thyroid function, significant elevation of anti-thyroid antibody and no other cause for encephalopathy.

END NOTE

Author Information

1. Dr. P Baburaj, MD
   Professor of Medicine & Unit Head, Jubilee Mission Medical College & Research Institute, East Fort, Thrissur, Kerala, PIN 680005, Tel No: 0487 2335236

2. Dr. BV Shankara, MBBS DNB
   Assistant Professor, Department of Medicine Jubilee Mission Medical College & Research Institute, East Fort, Thrissur, Kerala, PIN 680005, Tel No: 09895089602

3. Dr. Edwin J George, MBBS
   Postgraduate, Department of Medicine Jubilee Mission Medical College & Research Institute, East Fort, Thrissur, Kerala, PIN 680005, Tel No: 09895419147

4. Dr. B L Harikrishnan, MBBS
   Assistant professor, Department of Emergency Medicine, Jubilee Mission Medical College & Research Institute, East Fort, Thrissur, Kerala, PIN 680005, Tel No: 09846569012

Conflict of Interest: None declared

Cite this article as:

REFERENCES

10. Rodríguez A, Jicha G, Steves T, Benarroch E, and Westmoreland B. EEG changes in a patient with steroid responsive encephalopathy associated with antibodies to thyroperoxidase