

Unusual presentations of Hashimoto's encephalopathy: trigeminal neuralgiaform headache, skew deviation, hypomania

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Abstract Hashimoto's encephalopathy (HE) is a term used to describe an encephalopathy of presumed autoimmune origin characterized by high titers of antithyroid peroxidase antibodies. We describe three patients showing unusual clinical presentations like trigeminal-neuralgia, skew deviation, hypomania associated with HE. The purpose of this article is to describe the patients with unusual clinical pictures of HE, something that has not been reported in elsewhere in medical literature.

Keywords Hashimoto's thyroiditis · Hashimoto's encephalopathy · Antithyroid antibody

Case 1

A 67-year-old woman with a complaint of memory loss and inappropriate behaviors for 20 days admitted. She had no medical history or drug use. On admission, she showed confusion. The results of blood chemistries and a drug screen were within normal limits. Cranial magnetic resonance imaging (MRI) identified multiple hyperintense lesions on frontal lobes and caudate nuclei. Electroencephalography (EEG) was diffusely slow without epileptiform activity. Cerebrospinal fluid (CSF) analysis showed a mild increase in protein content (48 mg/dL). Thyroid function tests revealed a free-triiodothyronine (f-T3) level

of 1.51 pg/ml, free-thyroxine (f-T4) level of 0.81 pg/ml, and thyroid stimulating hormone (TSH) level of 7.26. Antithyroid-peroxidase antibodies (anti-TPO-Ab: 78.6 IU/ml) and antithyroglobulin antibodies (anti-Tg-Ab: 397 IU/ml) were increased. Hypoechoic thyroid was detected on ultrasonography. Hypothyroidism and HE were considered, and methylprednisolon (1,000 mg/d) was administered intravenously for 5 days, then followed by oral prednisolone and levothyroxine. The clinical symptoms remained stable for 3 months although her EEG, MRI findings disappeared and thyroid-antibody concentrations were slightly decreased. The patient remained symptom-free and had thyroid-antibody titers within normal limits after 6 months of treatment with prednisolone and levothyroxine.

Case 2

Beginning in March 2006, a 43-year-old woman developed intractable headache, vertigo, and unsteady gait. In May 2007, she experienced fluctuating memory disorder and depressive mood. During this period, she had been seen by several neurologists and psychiatrists and treated by vestibulosuppressant/antidepressants. On admission, she complained severe, short-lasting, lancinating pain shooting from the corner of the mouth to the angle of the jaw on the left side mimicking trigeminal neuralgia. Neurological examinations disclosed truncal ataxia. A cranial MRI was unremarkable. An EEG exhibited diffuse slow wave activity and absence of normal background rhythms. The analysis of CSF disclosed high protein level (68 mg/dL). TSH, f-T3, and f-T4 were within the normal range. Anti-TPO-Ab (132 IU/ml) and anti-Tg-Ab (97 IU/ml) were increased. Thyroid ultrasonography showed typical pattern of Hashimoto's thyroiditis. Depression was diagnosed

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based on her reported symptoms and signs on psychiatric interviews. A diagnosis of HE was suspected. After 4 weeks of treatment with prednisolone, patient was symptom-free with normal electroencephalographic and immunologic changes. The steroid therapy was tapered, and the patient remained symptom-free despite the fact that the patient was receiving no steroid therapy for 1 year.

Case 3

A 51-year-old man with no medical history presented confusion, diplopia, vomiting, and headache. Neurological examination showed stupor and right medial rectus muscle palsy. A MRI revealed hyperintense lesions on the right midbrain and bilateral thalamus. CSF protein was 86.5 mg/dL. EEG showed diffuse slowing. The patient recovered spontaneously over 5 days and he was symptom-free at discharge. Three months later, he was hospitalized again with the new neurologic and psychiatric signs. On examination, he had left hemiparesis and skew deviation. Hypomania was diagnosed based on psychiatric interviews. Brain MRI, CSF, and EEG revealed the previous findings and all laboratory results were normal. Despite TSH, f-T3, and f-T4 were within the normal range, thyroid antibodies were checked in both serum and CSF at his second admission and found positive (anti-TPO-Ab: 1,300 IU/ml, anti-Tg-Ab: 94.6 IU/ml). Thyroid ultrasonography was consistent with Hashimoto's thyroiditis. The patient was

diagnosed as HE because of the recurrent encephalopathy, positive antithyroid antibodies, and extensive negative workup. He was treated with oral prednisone which resulted in dramatic improvement on motor-sensorial and hypomanic symptoms. Skew deviation was decreased. He was then discharged on tapering doses of oral prednisone. He has been symptom-free except minimal vertical gaze palsy.

All patients' CSF parameters including CSF antibodies against virus and bacteria, serum vasculitis and tumor markers were negative.

The mostly reported clinical picture of HE is a relapsing-remitting encephalopathy characterized by seizures, headache, stroke-like episodes, and psychosis [1–3]. We recommend that the antithyroid-antibodies should be detected in cases of unexplained neurological symptoms and presentations not responding to conventional therapies.

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