LETTER TO THE EDITOR

An Overview of Hashimoto's Encephalopathy

Dear Editor,

Hashimoto's encephalopathy is a rare complication of autoimmune thyroid disease with an estimated prevalence of 2.1/100,000.[1] It is also known as steroid responsive encephalopathy associated with autoimmune thyroiditis, SREAT.[2] The pathogenesis is still unknown, but the possible mechanism could be due to cerebral vasculitis with or without immune complex deposition.[3] It is an inflammatory condition proposed by the presence of elevated TPO,[4] elevated CSF protein,[4] histological feature of vasculitis of venules, and lymphocytic perivascular cuff.[1]

Patients may present with encephalopathy (100%) which usually develop over 1 to 7 days, tremor (84%), transient aphasia (73%), seizure (66%), gait ataxia (63%), hypersomnolence (63%), myoclonus (38%), neuropsychiatric symptoms (36%), and stroke-like symptoms (27%).[1,5] Variable thyroid function tests could be seen in patients with Hashimoto's encephalopathy despite similar neurological findings. Goiter was found in 63% of the reported cases, subclinical hypothyroidism in 35%, normal thyroid function in 30%, overt hypothyroidism in 20%, and hyperthyroidism in 7% of cases.[1,6] Lab tests in patients with Hashimoto's encephalopathy usually show elevated TPO antibodies (100% of reported cases) and elevated thyroglobulin antibodies in 73% of the reported cases.[1,7] Upon CSF examination, 78% of the reported cases had elevated protein, and normal leukocyte count in 76% of cases.[1,7] The minority of patients, about 15%, had elevated ESR and CRP. Most of the patients (about 98%) with Hashimoto's encephalopathy had abnormal EEG.[7] EEG abnormalities could be generalized slowing, focal slowing, prominent triphasic waves, epileptiform abnormalities, and frontal intermittent rhythmic delta activity.[7] 50% of the patients with Hashimoto's encephalopathy had abnormal imaging in CT and MRI, in the form of cerebral atrophy, abnormalities in the white matter and diffused subcortical or focal cortical abnormalities.[7,8] Single photon emission computed tomography (SPECT) showed focal hypoperfusion in 73% of cases, global hypoperfusion in 9%, and 18% of cases had normal SPECT. Treatment options include steroids.[6] Steroids could be given as oral prednisone (50-100 mg/day), or intravenous methylprednisolone (1 g/day). Thyroid hormone replacement therapy with steroid was administered for patients presenting with hypothyroidism and Hashimoto's encephalopathy, and there was 92% improvement of reported cases using this combination therapy, and 67% improvement in patients taking levothyroxine alone. There has been only one case having been reported to recover from Hashimoto's encephalopathy after thyroidectomy.[8]

Hashimot's encephalopathy is a rare complication of autoimmune thyroid disease, which is most of the time underdiagnosed. The pathogenesis is still unknown. Steroid is considered the mainstay of treatment.

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Keywords: Hashimoto's Encephalopathy; Steroid; SREAT

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