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MULTIPLE CRANIAL NERVE PALSIES ASSOCIATED WITH THYROID AUTOIMMUNITY: HASHIMOTO'S THYROIDITIS- A CASE REPORT

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ABSTRACT Hashimotos's encephalopathy is an uncommon neurologic syndrome associated with Hashimoto's thyroiditis. Sometimes it may manifest years before onset of thyroid disease. The patients are usually euthyroid or midly hypothyroid. Antibodies against thyroid peroxidase are useful diagnostic marker of Hashimoto's thyroiditis. High levels of this antibody are associated with high prevalence of hashimoto's encephalopathy[1]. Clinical features are variable includes behavioral and cognitive changes, myoclonus, seizures, hemiparesis, involuntary movements, cerebellar signs, psychosis and coma, with relapsing and progressive course. Diagnosis is often overlooked but it is important as it is treatable cause of encephalopathy. Multiple cranial nerves involvement in Hashimoto's thyroiditis was reported in a very few patients, a rare association. Here we present a case of multiple cranial nerve involvement in a case of hashimoto's thyroiditis.

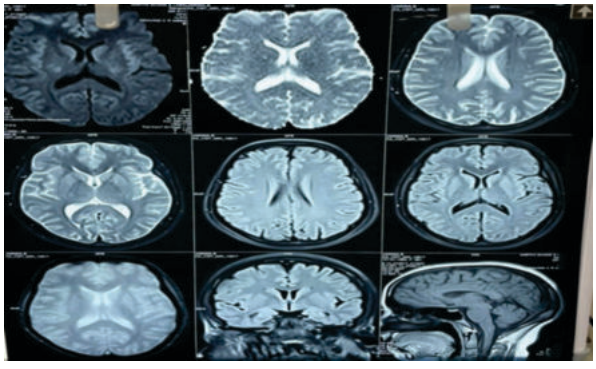
KEYWORDS : Hashimoto's Encephalopathy, Thyroid Peroxidase Antibodies, Cranial Nerve Palsies, Hashimoto's Thyroiditis.

INTRODUCTION:

Hashimoto's encephalopathy is an atypical manifestation of hashimoto's thyroiditis. It is an steroid responsive encephalopathy with relapsing and progressive course often treatable. The cause of Hashimoto's encephalopathy is proposed to be autoimmune because of its association with other immunologic disorders (myasthenia gravis, glomerulonephritis, primary biliary cirrhosis, pernicious anemia and rheumatoid arthritis), female to male ratio of 4:1,female predominance, occur more frequently in ages 30-46 years, inflammatory findings in cerebrospinal fluid (CSF) and response to treatment with steroids[2]. Other authors suggest that Hashimoto's encephalopathy represent an autoimmune cerebral vasculitis resulting from either endothelial inflammation or immune complex deposition[3]. Clinical findings are variable[4,5]. The varied range of clinical presentations often leads to a delay in diagnosis and initiation of treatment. Although neurological presentations like confusion, dizziness, hemiparesis, par aesthesia and involuntary movements have been documented, the involvement of multiple cranial nerves was reported very rarely in literature.

CASE REPORT:

A 50 years old female , not known to be hypertensive, diabetic, hypothyroidism, presented to us with sudden onset of left sided facial weakness, unable to close eyes tightly, deviation of angle of mouth to right side for a one day. This episode was preceded by pain & heaviness behind the jaw. No H/O fever, headache, neck rigidity, LOC,weakness of limbs. Facial nerve conduction and blink reflex studies showing Lt facial neuropathy.The patient was diagnosed as left LMN facial paralysis, Bell's palsy and was treated with steroids and improved. Later MRI Brain scan was done and is normal. After 6 months patient developed double vision on seeing distant objects and unable to move right eye laterally and diagnosed as isolated right lateral rectus paralysis. She had left facial synkinesis due to old LMN facial paresis sequelae. At this time there is no H/O headache, blurring of vision, redness of eye, vomiting, other cranial nerve abnormalities ,weakness of limbs. MRI brain could not be done due to dentures. CT scan was done normal. All routine blood investigatioins are normal except hb%,ESRand CRP elevated. She was further investigated for anemia. ELISA for HIV was nonreactive. Further investigations revealed that ANA,Anti ds DNA, HBsAg, antiHepC virus, lupus anticoagulant and VDRL results were negative. Also, the anticardiolipin antibody IgG level was normal. Her thyroid profile was done normal, as there is a strong family history of hypothyroidism was present, the possibility of an autoimmune etiology was suspected and we went for complete thyroid profile investigations including Free T3,Free T4,TSH,ultrasound thyroid, TPO Ab's. Free T3 ,Free T4,TSH were normal but TPO Ab's were high(112IU/ml).Ultrasound showing thyroiditis. FNAC thyroid nodule S/O Lymphocytic autothyroiditis. Considering the clinical and laboratory findings, a diagnosis of "multiple cranial nerve palsies due to Hashimoto's encephalopathy" was made. The patient was started on 1 gm methylprednisolone, administered intravenously daily over 5 days. There was a rapid clinical response. She was switched over to oral prednisone starting at 1 mg/kg daily, with a slow taper. She came back for his routine check up after 2weeks and had experienced significant resolution of symptoms.



Picture : MRI Images of Brain Normal study

DISCUSSION:

Hashimoto's encephalopathy is an unusual neurologic disorder whose etiology, pathogenesis and histologic characteristics are unclear. A systematic review published in 2003 reported only 85 well documented cases in the literature; however, this syndrome may be under recognized. Two major patterns of presentation are described:

1) 25% of patients follow a stroke like pattern of multiple recurrent episodes of focal neurologic deficits with a variable degree of cognitive dysfunction and consciousness impairment[2].
2) The remaining 75% present with a diffuse progressive pattern of slow cognitive decline with dementia, confusion and hallucinations[2].

These two clinical patterns may overlap over the course of the disease. 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% patients, hyper reflexia and other pyramidal tract signs in 85% patients and psychosis, visual hallucinations and paranoid delusions have been reported in 25–36%

patients [6]. However, the presence of multiple cranial nerve palsies is a unique and unreported presentation of this disease.

Mechanism of Hashimoto's encephalopathy does not appear to be related to the thyroid status, which can vary greatly in patients with this disease. In two recent reviews, 23–35% of patients had subclinical hypothyroidism, 17–20% had hypothyroidism, 7% had hyperthyroidism and 18–45% were euthyroid[1,5]. The development of neurologic symptoms may occur up to three years before the onset of autoimmune thyroiditis[7,8]. The presence of elevated serums levels of antithyroid antibodies remains an essential characteristic of HE diagnosis, and suggests the presence of thyroid autoimmunity[5]. Hashimoto thyroiditis have antibodies to various thyroid antigens, the most frequently detected include anti-thyroid peroxidase (anti-TPO), anti thyroglobulin (anti-Tg), and to a lesser extent, TSH receptor-blocking antibodies (TBII)[9,10],a small percentage of patients with Hashimoto thyroiditis (approximately 10-15%) may be serum antibody negative .Other antithyroid antibodies found in Hashimoto thyroiditis include thyroid-stimulating antibody and cytotoxic antibody [11].

The differential diagnosis of Hashimoto's encephalopathy includes delirium, rapidly progressive dementia, seizures or focal neurologic deficits[5],stroke or TIA, cerebral vasculitis, carcinomatous meningitis, toxic ,metabolic encephalopathies, paraneoplastic syndromes, CreutzfeldtJakob disease, degenerative dementia and psychiatric diseases[12]. The long term prognosis is variable, although a high percentage of patients respond to treatment; others may have a progressive or a relapsing course[1,5]. The symptoms usually improve with glucocorticoid therapy; however, it is not necessary. A systematic review of 85 published cases of Hashimoto's encephalopathy found clinical response in 98% patients treated with glucocorticoids, 92% patients treated with glucocorticoids and levothyroxine and 67% of patients treated with levothyroxine only[2].

CONCLUSIONS:

This syndrome may go unrecognized for a long time and the patient may be subjected to numerous investigations without definite benefit. Our case report uncovers a very unusual presentation of Hashimoto's thyroiditis in the form of multiple cranial nerve palsies. Henceforth, while evaluating patients with a diagnostic dilemma,autoimmune etiology of thyroid should be thought of and might help in instituting early treatment especially in middle aged women.

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