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Case Report

Hashimoto's Encephalopathy Presenting with Seizure and Neurocognitive Symptoms: A Case Report

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Introduction

Hashimoto's encephalopathy (HE) is a rare disorder associated with Hashimoto's thyroiditis. It was initially described in 1966, and it remains a controversial disorder [1]. Clinical findings are variable and nonspecific. In this case report, we present the case of a patient with subacute onset of declining upper brain functions associated with Hashimoto's encephalopathy.

Case presentation

The patient is a 30 year-old female who is known to have history of right sided Bell's palsy diagnosed in 2015, history of sleeve gastrectomy in July 2017, H. Pylori infection, ventricular bigeminy, iron deficiency anemia, vitamin B12 and vitamin D deficiencies. She is on multivitamins (compliant to them) and she lost follow up with general surgery after 2 months from her surgery. Over few months ago, patient's family noticed that she has episodes of transient amnesia, confusion, increased desire to sleep, social withdrawal and reduced verbal output. Additionally, her family observed episodes of disorientation and inappropriate behavior. Initially the patient was admitted in other health care facility in May 2019, where she presented with generalized tonic colonic seizures and started on levetiracetam 500mg BID. EEG was normal. MRI Head and MRA Brain were both unremarkable. MRI Brain showed bilateral frontal white matter hype- intense signal intensities (unidentified bright objects). Extensive workup were done and were normal including: ferritin, vitamin B12, and vitamin D, TSH & T4. Autoimmune workup were within normal limits including: (C-ANCA, P-ANCA, Cardiolipin IgG, Cardiolipin IgM, DNA AB, ENA, B2 Glycoprotein IgG, and B2 Glycoprotein IgM). Cooper and zinc levels were also within normal limits. CT abdomen/pelvis showed mild hepatosplenomegaly and small liver cyst. Levetiracetam discontinued as there was no clear indication and the patient was discharged with following differential diagnosis: encephalitis, metabolic encephalopathy, possible autoimmune related encephalopathy vs psychiatric element.

After eight days, she went to other health care facility as her family was not happy with her condition. In that facility, again multiple investigations were done including neuroimaging, EEG, lumbar puncture and labs workup. EEG showed focal epileptiform discharges. Examination of the CSF was normal. The laboratory tests showed no significant change compared with the patient's previous laboratory results except the presence of anti-thyroid peroxidase antibodies (319.9 high, reference range <34KU/L) as well as high serum thyroglobulin concentration (177.40 high, reference range 1.4-78.0micro/L). T3 and T4 were normal. US thyroid was unremarkable. Paraneoplastic and antinueornal antibodies were sent and came back negative. A diagnosis of Hashimoto encephalopathy was made and she was treated with pulse methylprednisolone for five days. Her condition dramatically improved after first dose of methylprednisolone, and she was discharged home on lacosamide 50mg BID for seizures along with tapering dosage of oral prednisolone. Five days later, her sister brought her to our facility as she had another episode of abnormal behavior. On examination, patient was awake, alert, oriented to person & place, not oriented to time & date. Having word finding

difficulties for complex things. Able to repeats the names. Poor attention and poor short memory. No focal neurological deficits. The laboratory showed ESR of 2mm/hr, CRP 0.2mg/L, TPO AB 305 IU/ml (reference range <34 IU/ml), thyroglobulin AB 19.5IU/ml (reference range <115 IU/ml). EEG done and showed moderate intermittent to almost continuous left temporal non-epileptiform disturbance of cerebral activity. In addition, there was minimal intermittent diffuse non-specific disturbance of cerebral activity. Previous investigations were reviewed. US pelvis done to rule out teratoma (NMDA encephalitis) and it was unremarkable. Patient re-started on pulse methylprednisolone for 3 days (then changed to oral prednisone at doses of 1 mg/kg body weight) and started on immunoglobulin 0.4g/kg/day for 5 days. Lacosamide dose increased to 100mg BID. Thiamine level sent and she was started on intravenous thiamine 100mg TID. Thiamine level came back normal 141nmol/L (reference range 70-180). Considering the clinical deterioration after tapering down steroids and laboratory findings, a diagnosis of Hashimoto encephalopathy was confirmed. At discharge, the patient was treated with prednisone at doses of 1 mg/kg body weight, with tapering dosage. The patient came back to neurology clinic after two weeks for follow-up, was still on prednisolone. Repeated TPO AB was decreased from 305 to 59 IU/ml, thyroglobulin AB decreased from 19.5 to <10 IU/ml. She developed acne vulgaris due to steroids use, for which she was seen by dermatology, and treated with topical medications. In addition, she developed itchy eyes, seen by ophthalmology and diagnosed with steroid induced glaucoma. She was back to her normal life with continuous medications.

Discussion

Hashimoto's encephalopathy (HE) is a rare neurological disorder associated with Hashimoto's thyroiditis, originally described in 1966 [1]. The disorder is estimated to affect 2.1 per 100,000 individuals in the general population, and it is more common in females than male with a ratio of 4:1 [1]. The pathogenesis of HE remains unknown, and no correlation exists between the concentration of thyroid antibodies and severity of symptoms. The reaction between anti-thyroid antibodies with the brain tissues remains unclear. The formation of autoantibodies against the N-terminal of α-enolase (NAE) may cause infarcts in the bilateral gangliocapsular region and left frontal periventricular deep white matter lesion as in our patient [1]. Clinical findings are variable and nonspecific including seizures, stroke-like episodes, cognitive decline, neuropsychiatric symptoms, and myoclonus. There are two major clinical patterns of the disorder. The first pattern account for 25% of patients and present with sudden vasculitis [1]. The second pattern account for 75% and present with a diffuse progressive pattern of slow cognitive decline with dementia, confusion and hallucinations [1]. These two clinical patterns may overlap over the course of the disease [1]. Two-thirds of patients experience focal or generalized tonic-colonic seizures. Status epilepticus has been reported in 12% [1]. The diagnosis of Hashimoto's encephalopathy is a clinical supported by laboratory evidence of auto-thyroid antibodies. Anti-thyroid antibodies will be elevated but can be associated with other autoimmune disease such as myopathy, dementia, bipolar disease, and they may present in normal population in about 2%-20% [1]. There is no specific diagnostic test for HE. EEG findings seen in 90-98% of patients including slow background activity, focal spikes or sharp waves and transient epileptic activity [1]. Imaging findings are nonspecific as in our patient including cerebral atrophy, focal cortical abnormality, diffuse subcortical abnormality and nonspecific subcortical focal white matter abnormality. The CSF analysis may show high protein concentrations, mild rise in lymphocytes count, high IgG, oligoclonal bands and anti-thyroid antibodies may be seen. Hashimoto's encephalopathy is a diagnosis of exclusion, differential must include any condition that cause delirium such as stroke or transient ischemic attack, cerebral vasculitis, carcinomatous meningitis, toxic metabolic encephalopathies, paraneoplastic syndromes, Creutzfeldt-Jakob disease, degenerative dementia and psychiatric diseases. Diagnosis of HE should meet the following three points: cognitive disorder; cerebrospinal fluid analysis that can exclude infectious diseases; and high titers of anti-thyroid antibodies in serum [1]. These findings and a response to corticosteroids generally define this disorder which were achieved in our patient. Regarding treatment for HE, high dose corticosteroids 1-2mg/kg is considered first line of choice. As in our patient symptoms improved significantly and rapidly after initiation of corticosteroid treatment. Clinical improvement with corticosteroid therapy is usually observed in the first 4-6 weeks and it has been considered to be part of the criteria for diagnosis of HE [2]. Other therapies such as plasmapheresis and immunosuppressant medications such as cyclophosphamide and methotrexate have been successfully used in patients non-responsive to corticosteroids. For patient who presenting with seizures, the use of corticosteroids prior to antiepileptic therapy may be effective. For patients with hypothyroidism, it's believed that thyroxine therapy should be provided for those patients with thyroid dysfunction although there is no correlation has been found between thyroxine therapy and outcomes of the disease [1]. The long-term prognosis is variable, although a high percentage of patients respond to treatment; others could have a progressive or a relapsing course [1-4].

Conclusion

Hashimoto's encephalopathy (HE) is an unusual autoimmune disorder frequently presents with neurocognitive symptoms and normal findings in several different examinations. This disorder can be misdiagnosed, therefore it should be kept in mind when evaluating a patient with neurocognitive dysfunction and high

titers of anti-thyroid antibodies. Additionally, it is high responsive to steroids so other causes of neurocognitive dysfunction should be excluded before initiating treatment.

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