Case Report

Hashimoto's Encephalitis: Unusual Cause of Reversible Dementia

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Abstract

Hashimoto's encephalopathy (HE) is a poorly understood and often misdiagnosed rare autoimmune disease with varied neurological and psychiatric features. The low prevalence and varied clinical features coupled with unclear pathogenesis and histopathologic characteristics have caused still doubts in any particular diagnostic criteria. Therefore, more case studies are needed to characterize the clinical, laboratory and imaging features and outcomes of HE patients. We describe a case of such a patient with HE presenting with dementia and focus on its early recognition as the cognitive changes are reversible.

Keywords: Cognitive, dementia, encephalitis, encephalopathy, Hashimoto

Introduction

Hashimoto's encephalitis or encephalopathy (HE) is a rare autoimmune disease often under diagnosed. It can present as rapidly progressive dementia (RPD) or dementia of unknown origin which is treatable with high dose steroids. We present a case of HE to signify the importance of its early recognition and management.

Case Report

The present case report is about a 58-year-old female patient who was brought by her family members with a 4 month history of progressive memory impairment; both recent and remote memory loss, inability to carry out activities of daily living namely dressing, toileting, bathing and cooking. She also had urinary and fecal incontinence and was mute. She demonstrated features of compulsive behavior in the form of repeated folding of hands. On examination, she was conscious but was not oriented to time, place or person with gross psychomotor retardation. Mini-mental state examination (MMSE) could not be performed as she was not speaking and would follow few commands. Her voice was hoarse and would speak only if provoked. There was no cranial nerve involvement, motor weakness or sensory deficits. Her deep tendon reflexes were normal with flexor plantar response. There were no signs of cerebellar incoordination or of meningeal irritation.

Her routine laboratory work-up was normal (hemoglobin = 13.2 g%, total white blood cell = 12900 cumm, platelet count = 425000/cumm, packed cell volume = 42.3%, mean corpuscular volume = 92.8 fl, random blood glucose = 101 mg/dl, serum amylase = 119 U/lS, serum sodium = 137 mg/l, serum potassium = 5 mg/l, serum calcium = 8.7 mg/dl, total bilirubin = 0.7 mg/dl, direct bilirubin = 0.3 mg/dl, indirect bilirubin = 0.4 mg/dl, serum glutamic oxaloacetic transaminase = 28 U/l, serum glutamic pyruvic transaminase = 36 U/l, alkaline phosphatase = 184 U/l, serum urea = 39 mg/dl, serum creatinine = 1.2 mg/dl, serum uric acid = 6.2 mg/dl, rheumatoid factor: 4.6 IU/ml, anti-nuclear antibodies and double-stranded deoxyribonucleic acid: Negative, erythrocyte sedimentation rate: 61 mm in 1st h, serum B12 level: 192 pg/dl, test for venereal disease research laboratory, human immunodeficiency virus I and II serology were non-reactive). Chest X-ray, urine/stool examination, electrocardiogram and cerebrospinal fluid analysis yielded normal results. Examination of fundus did not show any sign of raised intracranial pressure or abnormal deposits. Thyroid profile revealed; T_3: 2.34 pg/ml, T_4: 1.21 ng/dl, thyroid stimulating hormone (TSH): 5.4, anti-thyroid peroxidase antibodies (anti-TPO): 1300 U/ml (N < 65 U/ml)

Access this article online

Quick Response Code:

Website: www.jfmpc.com

DOI: 10.4103/2249-4863.141650

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and anti-thyroglobulin antibodies (anti-TGA): 2049.70 U/ml. Magnetic resonance imaging (MRI) brain scan showed mild diffuse cerebral atrophy and electroencephalogram (EEG) revealed diffuse slowing with generalized theta/delta activity. A diagnosis of HE with dementia was made and she was treated with intravenous methylprednisolone 1 g/daily for 5 days and later shifted to oral prednisone 60 mg/day. There was a drastic improvement in her symptoms within 1 week. She also received oral thyroxin 50 µg/day. She was regularly evaluated every 2-3 weeks showing gradual improvement in symptoms and by 4 months MMSE score was 30/30 and she was performing all her routine activities of daily living. Her anti-TPO and anti-TGA levels markedly reduced to normal limits and EEG became normal. Her oral steroid therapy was tapered over a period of 4 months and the patient remained symptom free on subsequent follow-up.

Discussion

HE should be considered in a case of sub-acute encephalopathy associated with high levels of anti-thyroid antibodies despite normal thyroid functions and in the absence of other brain diseases.

HE is a neurological disorder associated with high serum anti-thyroid antibody. It was first described by Brain *et al.*, in 1966.[1] It is a rare disease with no specific known cause. Since, the link between HE and Hashimoto thyroiditis is not clear, the condition is now termed as "steroid-responsive encephalopathy associated with autoimmune thyroiditis" (SREAT). It should be suspected in all cases with mental status changes in the setting of normal brain imaging and normal routine lab results. The acronym SREAT suggests that patients typically respond to corticosteroids.[2]

SREAT has two types of initial clinical presentation: An acute stroke like (vasculitic type) episode and a gradual cognitive impairment.[3] Diagnostic criteria for SREAT includes confusion, focal/generalized seizure, focal neurological deficit, cognitive decline, dementia, increase in anti-thyroid antibody titers and an excellent response to corticosteroids. SREAT is associated to non-specific EEG abnormalities, elevated cerebrospinal fluid proteins and nonspecific white matter changes.[4] Since clinical features of SREAT are unspecific, other etiologic factors such as infectious, metabolic, toxic, vascular, neoplastic and paraneoplastic causes have to be excluded. It is a controversial entity because it lacks definite diagnostic criteria. Keeping these factors in mind, SREAT still remains a diagnosis of exclusion.[3,5] All the symptoms are not seen in every patient and presentation varies significantly. Moreover, not all cases of SREAT respond to corticosteroids.[6] In such cases, other forms of immunosuppression may be tried.

It was initially believed that anti-thyroid antibodies have a role in the pathogenesis of SREAT; however, various studies have reported that there is no evidence of pathogenic role of antibodies in SREAT. High titers of anti-thyroid antibodies are not sufficient for diagnosis. The levels of antibody do not correlate with severity or type of clinical presentation and its levels are not influenced by corticosteroids.[3,7] Furthermore, the clinical status of thyroid gland is independent of neurological symptoms. However, the combination of encephalopathy, high serum anti-thyroid antibody concentrations and responsiveness to glucocorticoid therapy are seen in almost every case and therefore this association may not be due to chance alone. The antibodies are probably markers of some other autoimmune disorder affecting the brain.[7,8]

Diagnosis of SREAT requires a certain degree of suspicion. SREAT has been listed as a cause of RPD. Endocrinologists, Neurologists and Psychiatrists should be aware of this condition and consider it as one of the differential diagnosis in every case of RPD. All cases of encephalopathy of unknown origin and RPD during their initial screening must have TSH levels checked along with complete blood count, vitamin B12, chemistry panel and MRI along with evaluation of personal and familial history of thyroid disease. It is very important to diagnose SREAT early because it has a good prognosis upon treatment responding drastically to immunosuppressive drugs.

After being described in 1966 as HE, not many cases have been reported world-wide until now.[9] The small number possibly indicate it to be an under diagnosed condition may be due to lack of awareness. We must check for thyroid abnormalities in every case of RPD and rule out antithyroid antibody titers in cases with deranged thyroid function test. There have been no reports suggestive of any environmental or geographic factors playing a role in the pathogenesis of this condition. There is scope for research to find out the actual cause of SREAT and the role of antithyroid antibodies.

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How to cite this article: Anand KS, Garg J, Verma R, Chakraborty A. Hashimoto's encephalitis: Unusual cause of reversible dementia. J Fam Med Primary Care 2014;3:284-6.

Source of Support: Nil. **Conflict of Interest:** None declared.