Title

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CLINICAL VIGNETTE

A 68-Year-Old Female with Severe Alzheimer's Dementia and Hashimoto's Encephalopathy

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Case Presentation

A 68-year-old female was brought in by family for severe, progressing agitation for several months. Past medical history includes anxiety, depression, dementia, Hashimoto's thyroiditis, and hypothyroidism. The patient was first diagnosed with cognitive deficits seven years ago after her husband died. She was evaluated by a Neurologist and started on donepezil but was lost to follow-up. She re-established care with Neurology three years ago, with laboratory remarkable for TPO antibody > 900 u/mL and APOE 3/3. MRI brain showed significant cerebral atrophy including in the medial temporal lobe structures. She was diagnosed with dementia with paranoid fixations and mood instability and did not start treatment for presumed hypothyroidism. Significant memory impairment and functional decline prompted her children to move her closer to them last year. She was described by the family as conversational at baseline, recognizing family members, and able to perform daily activities (ADL) such as feeding, dressing, and showering.

On presentation, the patient was alert, but inattentive, and not interacting, and with intermittent agitation. Her vital signs were within normal limits and exam noted myoclonus. Laboratory testing was notable for WBC 11.5 x10E3/uL, TSH 13.3 mcIU/mL and FT 4 0.70 ng/dL. Endocrinology was consulted and recommended continuing levothyroxine with plan to repeat thyroid tests in 4-6 weeks. Her agitation was not felt to be related to her thyroid dysfunction. Psychiatry was consulted for medical management of agitation and evaluation for transfer to a psychiatric facility. Neurology was also consulted, and EEG showed moderate diffuse slowing and an MRI brain with and without contrast showed severe bilateral posterior cerebral hemisphere volume loss and bilateral hippocampal volume loss, mildly progressed compared to outside hospital MRI from two years prior. Dementia laboratory work up for Hashimoto's encephalopathy also known as steroid responsive encephalopathy of autoimmune thyroiditis (SREAT) and a lumbar puncture was recommended to rule out any other etiologies. Lumbar puncture was unremarkable. Lab work up was significant for TPO antibody 127 u/mL and normal TG antibody. Neurology determined that the patient's overall clinical history and current presentation was most consistent with progressive Alzheimer's dementia though the patient's previously untreated Hashimoto's thyroiditis is thought to have complicated her overall clinical course.

Discussion

Hashimoto's encephalopathy (HE) is a rare auto-immune disorder associated with elevated levels of anti-thyroid antibodies (ATA: antithyroid peroxidase and anti-thyroglobulin).[1] Though no specific level of ATA has been associated with HE, case reports of those affected had anti-TPO and/or anti-TG levels ranging from 900-1000 u/mL.[2] The presence of ATAs is considered to be nonspecific as they can be elevated in patients without encephalopathy and also may be associated with other auto-immune diseases[1] but elevated ATA levels are required for the diagnosis. The pathophysiology is not known and may involve vasculitis or other inflammatory changes of the brain leading to encephalopathy.[2,3] The clinical presentation can vary from "stroke like signs, seizures, amnestic syndrome, ataxia, myoclonus, cognitive impairment, and dementia, to psychiatric manifestations".[1] HE is a diagnosis of exclusion that is made clinically[1,2] supported by negative or nonspecific lumbar puncture and MRI findings and, usually, response to steroid therapy.[2] Though not all cases have been found to be steroid responsive, HE is also known as "steroid responsive encephalopathy associated with autoimmune thyroiditis" because an almost instant response is seen, with reversal of some or most symptoms, once high dose steroids are initiated.[2,3] Not only can the response to steroids be robust but it can occur years after the diagnosis is made making the prognosis of HE good.[2] Untreated HE however leads to cognitive decline that is progressive.

Conclusion

HE is a largely reversible disease process that should be evaluated for in patients with elevated ATA and clinical signs or symptoms of cognitive, neurological, and/or psychiatric disturbance. Once diagnosis is made, treatment should not be delayed.

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