Spastic Paraparesis, Abnormal Muscle Biopsy and Positive Antithyroid Antibodies

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Abstract

A 35 year old lady presented with progressive spastic paraparesis and hyperintense signals in the spinal cord and brain. She was noted to have high titres of antithyroid antibodies and primary hypothyroidism. Her muscle biopsy showed perivascular lymphocytes around endomysial vessels. We highlight the association of spinal cord involvement and abnormal muscle biopsy in a case of Hashimoto’s encephalopathy.

INTRODUCTION

The association between neurological illness and antithyroid antibodies is well established. Most of the previous articles on the topic have highlighted the encephalopathic presentation of the illness. But there are rare reports of an association between myelopathy and antithyroid antibodies. The exact role of antibody in the genesis of encephalopathy or myelopathy is not clear. Some autopsy studies have revealed perivascular lymphocytic infiltration in the brain of patients with Hashimoto’s encephalopathy; on the basis of which a vasculitic basis for the disease has been proposed.

CASE REPORT

A 35 year old lady was admitted to our department with two months history of difficulty in walking. On evaluation she was found to have spasticity of the lower extremities, diminished position sense and vibration sense in her lower extremities, positive Romberg test, spastic ataxic gait, preserved deep tendon reflexes and bilateral extensor plantar response. There was no sphincter involvement or sensory level. She also had alopecia and hyperpigmentation of the lower limbs. She had myoclonic jerks in her lower extremities. She had a history of thyroid enlargement in her first pregnancy.

Her routine blood examination, peripheral smear and serum B 12 levels were normal. ANA, ANCA, rheumatoid factor, VDRL, HBsAg, Anti-HCV, HIV, Anti-Ro and Anti-La Antibody were negative. APLA was negative and angiotensin converting enzyme (ACE) levels were normal (18; range 3-52). VEP and BERA were normal. SSEP showed prolonged N13. A thyroid function test showed primary hypothyroidism. A test for antithyroid peroxidase and antithyroglobulin antibodies were positive in high titer (1:256, 1:300 respectively). An imaging of the spine and brain showed brain atrophy as well as patchy asymmetric T2W hyperintensities of the periventricular regions and similar hyperintensities in the spinal cord (Fig. 1). A lumbar puncture showed normal pressure, 40 cells, all lymphocytes, with protein 66 mg/dl and sugar 48 mg/dl. No oligoclonal bands were detected in the CSF. A muscle biopsy was done as part of the vasculitic work up. Muscle biopsy showed perivascular inflammation around the endomysial vessels. An electroencephalogram was done which showed generalised polyspike discharges at a frequency of 3-4 Hz. A diagnosis of Hashimoto’s encephalopathy was made and patient initiated on steroids. She showed a significant improvement in her symptoms over a one month period.

DISCUSSION

Fig. 1: T2W MRI showing brain atrophy and hyperintense signals in the cord.
This patient had progressive spastic paraparesis. A compressive etiology is ruled out by the negative MRI. Among the non-compressive etiologies which can present with similar imaging abnormalities, the important ones are primary progressive multiple sclerosis (PPMS), vasculitic, nutritional and toxic etiologies, hereditary diseases like hereditary spastic paraplegia, HTLV associated myelopathy, sarcoidosis etc. PPMS is unlikely in view of the absent brain lesions, lack of oligoclonal bands in CSF, normal VEP and BERA. Investigation for a vasculitic etiology was negative. Nutritional diseases like B12 deficiency can mimic this clinical picture. But her serum B12 and peripheral smear were normal. There is no history of exposure to any toxins. Hereditary disorders are unlikely in view of the absent family history. Sarcoidosis is excluded by the lack of other associated features, normal chest X-ray and normal ACE levels. Hence a diagnosis of Hashimoto’s encephalopathy was entertained in this patient with spastic paraparesis, primary hypothyroidism, positive antithyroid antibodies and cortical myoclonus.

Hashimoto’s encephalopathy is a rare disease characterized by subacute or acute encephalopathy, myoclonic jerks, seizures, tremulousness, focal neurological deficits mimicking stroke, amnesia, dementia and positive antithyroid antibodies. The first description of an association between Hashimoto’s thyroiditis and encephalopathy was in 1966 by Brain et al.1 Our patient had evidence of a brain involvement in the form of myoclonus, abnormal EEG, and abnormal MRI. She had evidence of spinal cord involvement in the form of spasticity, loss of position sense, imaging abnormalities on MRI in spinal cord. Previous reports have also emphasized imaging abnormalities in the brain2-3 of patients with hashimotos encephalopathy as well as the raised CSF protein and pleocytosis in such patients. Imaging abnormalities in the spinal cord and initial presentation as a posterolateral syndrome as in our case have not been highlighted in the literature. A review of literature revealed only two cases where antithyroid antibodies where associated with myelopathy.4,5

The exact pathogenesis of the syndrome has not been clearly defined. Various postulations as to the etiology includes an abnormality of the immune system, cerebral vasculitis, recurrent demyelination or a toxic effect of TRH on the nervous system. Whether the antibody is directly pathogenic or is it simply a surrogate marker of for other, as yet unidentified, antibodies that cross the blood-brain barrier and initiate an autoimmune encephalopathy is still a matter of debate. Autoimmune mechanism is highly probable in view of the higher incidence in females, fluctuating course and response to steroids. A biopsy from the muscle in this patient revealed perivascular lymphocytes around the endomysial vessels. This suggests that the pathology in hashimotos encephalopathy may not be restricted to brain and thyroid but, it may affect other organs as well. A brain biopsy from a patient with Hashimoto’s encephalopathy showed perivascular mononuclear cells suggesting a possible vasculitic process as the basis of this syndrome.6 The basic pathologic process may be causing similar histological changes in the various organs as we have found in the muscle and above mentioned study in the brain. Most of the studies highlight encephalopathy as the major manifestation of Hashimoto’s encephalopathy. Encephalopathy may be minimal as in our case were the only manifestations may be myoclonus and MRI changes. Our patient showed a significant improvement with steroid therapy and a course of plasma exchange.

REFERENCES


Announcement

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