Steroid-Responsive Myeloneuropathy Associated With Antithyroid Antibodies

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Abstract

Background/Objective: To present information about 2 steroid-responsive, antithyroid antibody-positive patients with myeloneuropathy and myelopathy.

Methods: Case reports.

Results: A 48-year-old woman and a 42-year-old man presented with acute onset tetraparesis and magnetic resonance imaging studies showing cervical spinal lesions. Nerve conduction and biopsy studies of the woman were suggestive of a demyelinating polyradiculoneuropathy. Detailed diagnostic workup turned out to be negative for both patients, except for highly elevated antithyroid antibodies with normal thyroid functions and imaging. Both patients responded remarkably well to high-dose steroid treatment, and their symptoms disappeared in a few months. Both patients' antithyroid antibody levels were reduced shortly after steroid treatment and in parallel with the amelioration of symptoms.

Conclusions: Antithyroid antibodies might be associated with acute demyelinating myeloneuropathy or myelopathy pathogenesis and might indicate a good response to steroid treatment in these syndromes.

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Key Words: Tetraparesis; Myeloneuropathy; Thyroid; Hashimoto's encephalopathy; Guillain-Barré syndrome; Myelitis; Antithyroid antibodies; Steroid therapy

INTRODUCTION

Simultaneous involvement of the spinal cord and the peripheral nerves (myeloneuropathy) is a rare entity most often associated with vitamin B₁₂ deficiency, copper deficiency, and various infectious agents (1). Noninfectious inflammatory myeloneuropathy is even more infrequent, and there are only a few reported cases with transverse myelitis and chronic inflammatory demyelinating polyneuropathy, or Guillain-Barré syndrome (2). We report the clinical courses of 2 antithyroid antibody-positive patients: 1 with myeloneuropathy and 1 with isolated transverse myelitis.

CASE REPORTS

Patient 1

A 48-year-old woman presented with progressive generalized muscle weakness and sensory loss and reached maximum disability in 7 days. She had no history of

antecedent infections, immunizations, premonitory symptoms, or fever. Neurologic examination revealed predominantly distal symmetrical flaccid tetraparesis (2/5 on the Medical Research Council scale), stocking-and-glove sensory loss, impaired position and vibration senses, absent deep tendon reflexes, and unresponsive plantar reflexes. Nerve conduction study findings were suggestive of Guillain-Barré syndrome on the basis of lost F-wave responses, markedly reduced motor/sensory nerve conduction velocities, and normal action potential amplitudes. A sural nerve biopsy revealed segmental demyelination and remyelination with no inflammatory infiltrates.

Five days after her admission, the patient developed neck pain and urinary urgency and frequency with an inability to control urinary elimination. A repeat neurologic examination showed a cervical sensory level. Magnetic resonance imaging (MRI) revealed a hyperintense lesion approximately at the C3 level on T2/FLAIR-weighted images, enhancing contrast on T1-weighted sequences (Figure 1).

Patient 2

A 42-year-old man presented with difficulty walking in the last 2 days with no history of preceding events or

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Figure 1. Patient 1. (A) Sagittal T2-weighted magnetic resonance image of the cervical spinal cord shows a hyperintense lesion approximately at the C3 level. (B) The lesion enhances contrast on the T1-weighted sequence.

fever. Neurologic examination revealed symmetrical tetraparesis (4/5 on the Medical Research Council scale), upper thoracic sensory level, impaired vibration sense, increased deep tendon reflexes, and extensor plantar responses. Magnetic resonance imaging showed a hyperintense lesion at the C4-C5 level on T2/FLAIR-weighted images, enhancing contrast on T1-weighted sections.

Both patients' cranial/dorsal MRIs, complete blood count, serum biochemistry tests (including serum B₁₂, folate levels, and thyroid function tests), sedimentation rate, and serum C-reactive protein levels were normal. Cerebrospinal fluid examination showed no cells, increased protein levels (case 1, 80 mg/dL; case 2, 65 mg/dL), and normal glucose levels. Patient 1 had no oligoclonal bands, and patient 2 had only cerebrospinal fluid oligoclonal bands. Serologic screening for vasculitic/autoimmune, paraneoplastic, and infectious disorders (*Treponema pallidum* subp *pallidum*, herpes simplex virus, cytomegalovirus, Epstein-Barr virus, human immunodeficiency virus, human T-lymphotropic virus type 1, *Borrelia burgdorferi*, and *Mycoplasma pneumoniae* infections) yielded negative results. Although both serum thyroid peroxidase (441 IU/mL; normal, <5.6 IU) and thyroglobulin (1,060 IU/mL; normal, <4.1 IU) antibodies of Case 1 were markedly elevated, Case 2 showed only increased thyroid peroxidase-antibody levels (246 IU/ml). The thyroid scintigraphy/ultrasound and whole-body computed tomography screening were unremarkable for both patients.

Both patients remarkably responded to high-dose methylprednisolone (1,000 mg intravenously for 7 days). In 1 week, patient 1 significantly improved; in 2 months, neurologic examination, MRI, and nerve conduction studies were normal. Patient 2 started improving in 2 weeks, and his neurologic examination and MRI returned to normal in 3 months. No neurologic signs or clinical thyroiditis have been noted during 8- and 2-year follow-up periods for patients 1 and 2, respectively. Although patient 1 continues to have elevated thyroglobulin-antibody levels (225 IU/mL), serum thyroid peroxidase antibody levels of both patients declined after steroid treatment and were reduced to normal limits within months.

DISCUSSION

Hashimoto's encephalopathy typically presents with central nervous system findings, normal thyroid functions, and elevated antithyroid antibody levels in a manner identical to our patients (3). Antithyroid antibody-positive patients with myelopathy or Guillain-Barré syndrome (but not myeloneuropathy) have also been reported (4). Moreover, biopsy-proven central nervous system demyelination has been detected in Hashimoto's encephalopathy (5).

The absence of antecedent infections, fever, an increased sedimentation rate, and systemic findings indicates that our patients most likely do not represent a vasculitic/rheumatologic syndrome or viral infection. Antithyroid antibody seropositivity was the only abnormal parameter, suggesting Hashimoto's myeloneuropathy as a possible diagnostic option. Reduction of antithyroid antibody levels of our patients shortly after steroid treatment and symptom cessation particularly supports the notion that antithyroid antibodies might have a pathogenic significance rather than simply being epiphenomena.

Then again, as is often argued, the association between neurologic symptoms and antithyroid antibodies may be purely coincidental, because these antibodies and clinical thyroiditis are found in 10% to 20% and 1% to 3% of the general population, respectively (3). Antithyroid antibodies might also simply be indicating the patient's general tendency to develop autoimmune disease.

Nevertheless, one characteristic feature of antithyroid antibody-positive neurologic involvement is a remarkable response to steroid treatment, as also observed in our patients (3).

CONCLUSIONS

Antithyroid antibody testing might be a valuable tool in treatment decisions and could be added to the work-up panel for nontraumatic myelopathy, especially myeloneuropathy, and provide support for trials of steroid treatment in those patients.

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