Acute Axonal Polyneuropathy Combined with Acute Disseminated Encephalomyelitis: Case Report

Roberto Bomprezzi, MD, PhD
Michael Snyder, MD
Timothy Lee Vollmer, MD†

Axonal variants of the acute inflammatory polyneuropathies known as Guillain-Barré syndromes are more rare than the demyelinating forms. The occurrence of disseminated involvement of the CNS in the same patient, also in an acute stage, is even more unusual. We present the case of a young woman who developed a rapidly evolving quadriplegia consistent with acute axonopathy and radiological evidence of inflammatory acute encephalomyelitis while recovering from an infectious illness. This case supports the view that para-infectious inflammatory diseases of the nervous system have an autoreactive etiology and that both the CNS and peripheral nervous system can be affected simultaneously.

Key Words: acute disseminated encephalomyelitis, Guillain-Barré syndrome, polyneuropathy

Abbreviations Used: ADEM, acute disseminated encephalomyelitis; CNS, central nervous system; CSF, cerebrospinal fluid; MR, magnetic resonance

Division of Neurology, Barrow Neurological Institute, St. Joseph's Hospital and Medical Center, Phoenix, Arizona

†*Division of Neurology, University of Colorado Health Science Center, Denver, Colorado*

An autoreactive immune process directed against self-components of the peripheral nervous system after an acute infectious illness are believed to cause the acute inflammatory polyneuropathy known as Guillain-Barré syndrome. Cross-reactivity between antigens of infectious agents and constituents of the self have been shown to be the basis of the nerve damage.[3] Similarly, the pathogenic mechanisms of ADEM have been linked to a self-limited autoreactive process that follows an infectious illness or a vaccination contaminated with neural tissue. Based on the hypothesis of a widespread autoreactive immune response triggered by a microorganism, the simultaneous presentation of Guillain-Barré syndrome and ADEM may seem a likely event that nonetheless is seldom encountered in clinical settings. We present the rare case of a woman who developed severe acute axonal sensorimotor polyneuropathy and showed radiological evidence of multifocal CNS involvement.

Case Report

A 30-year-old woman sought treatment for a sore throat. She was given oral antibiotics at an urgent care center. Two weeks later she developed nausea, vomiting, and abdominal pain that motivated a second visit to the urgent care where she was prescribed anti-emetics. Soon thereafter, she complained of chest pain, shortness of breath, and palpitations and was hospitalized at another facility. Within 24 hours of admission, she developed anisocoria with poorly reactive pupils (left greater than right) followed by rapidly progressive external

Table 1. Summary of Nerve Conduction Study (NCS) and Electromyography (EMG) in Patient with Guillain-Barré Syndrome and ADEM

Right ulnar motor NCS showed small CMAP amplitudes without temporal dispersion; distal latency moderately prolonged despite normal conduction velocities. Right median motor NCS showed very small, temporally dispersed CMAP amplitudes. Although distal latency was significantly prolonged and conduction velocity was moderately slowed, they are probably commensurate with the CMAP amplitudes.
Right peroneal NCS recordings from extensor digitorum brevis and tibialis anterior showed either extremely small or absent CAMP responses. Right tibial motor NCS showed absent CMAP response.
Right median and ulnar antidromic sensory NCS recordings from digits 2 and 5, respectively, showed absent SNAPs. Right superficial radial sensory NCS showed a slightly small SNAP amplitude with normal conduction velocity.
Right sural and superficial sensory NCS recordings showed absent SNAPs.
Right ulnar F-waves were absent.
EMG of right tibialis anterior, medial gastrocnemius, vastus medialis, first dorsal interosseus, and deltoid showed various degrees of Fibs/PSWs and fasciculations. No MUPs generated in any muscle. Most severe denervation changes noted in deltoid and tibialis anterior.

CMAP, compound motor action potential; SNAP, sensory nerve action potential; Fibs, fibrillations; MUP, motor unit potential; PSW, positive sharp waves.

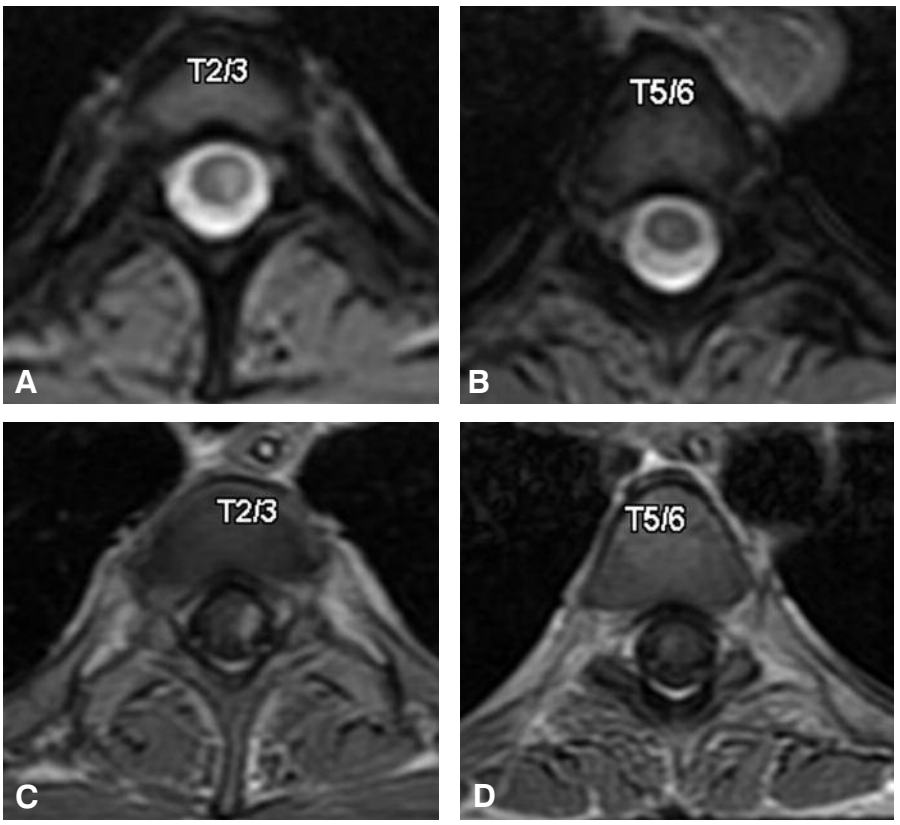


Figure 1. (*A* and *B*) Axial T2-gradient echo MR images of lesions at T2-3 and T5-6 and (*C* and *D*) axial T1-gradient echo MR images with contrast of the same lesions show contrast-enhancing demyelinating lesions.

ophthalmoplegia, flaccid quadriplegia, and respiratory failure requiring ventilator assistance. CSF testing showed no white blood cells. Her protein level was 59 mg/dL, and her glucose level was 58 mg/dL. MR imaging of the brain, obtained to clarify the anisocoria, was normal. She was diagnosed with Guillain-Barré syndrome and underwent six cycles of plasmapheresis before being transferred to our hospital.

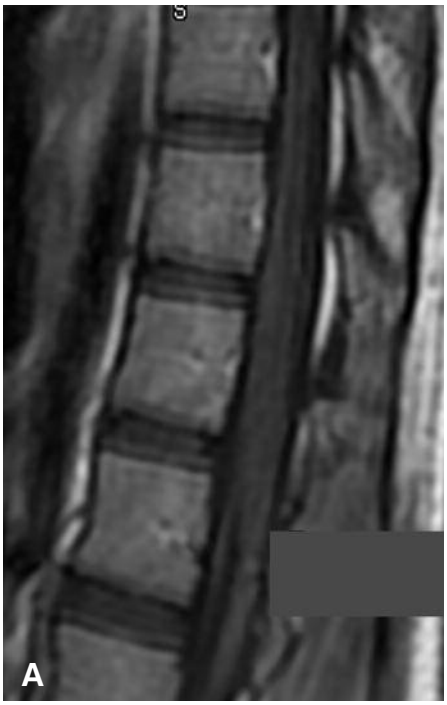
At that time the anisocoria had resolved. She could move her eyes in all directions but exhibited hypertropia of the left gaze. She remained quadriplegic with minimal movements in all limbs. Her sensation perception was impaired all over but was worse in her lower body with an upper sensory level of around T8.

Nerve conduction testing and electromyography were performed, and a severe acute axonal sensorimotor neuropathy was confirmed (Table 1).

A second test of her CSF demonstrated worsening of the albuminocytological dissociation, along with increased intrathecal IgG synthesis, although no oligoclonal bands were present. A comprehensive list of viral and bacterial causes was tested by serology and, where available, by polymerase chain reaction. It was concluded that the patient had no ongoing active infection. Anti-ganglioside GM1 antibodies were negative.

MR imaging of the brain was repeated and showed a few scattered lesions in the white matter and juxtacortical regions, some of which showed contrast enhancement. Multiple areas of contrast-enhancing signal abnormalities were noted in the thoracic spinal cord, the most prominent at T2-3 and T5-6 (Fig. 1). The postcontrast scans also showed a marked increase in signal intensity at all thoracic nerve roots consistent with acute polyradiculoneuropathy (Fig. 2).

The patient was treated with a course of intravenous immunoglobulins at standard dosing levels. On hospital Day 7 her status began to improve, and she slowly and gradually recovered motor function. After a stay at a rehabilitation facility, she continued to improve. Ap-



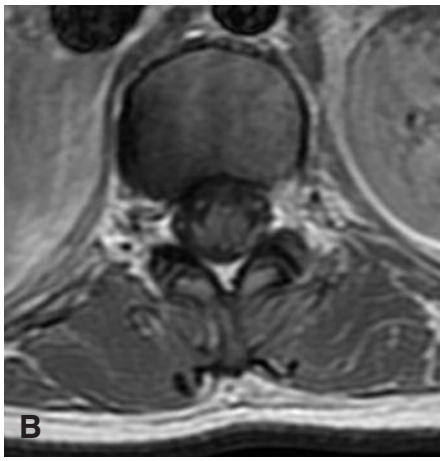


Figure 2. (*A*) Sagittal and (*B*) axial MR images of the lumbar spine show contrast enhancement of the nerve roots consistent with acute inflammatory polyradiculoneuropathy.

proximately 4 weeks later she had regained the ability to ambulate with bilateral support. The sensory level related to the spinal cord lesion had resolved. Considering the rate of her initial recovery, resumption of normal neurological function with perhaps some residual sensory disturbances is expected for this patient.

Discussion

There is sufficient evidence that cross-reactivity between antigens of infectious agents and constituents of self is at the basis of the nerve damage in Guillain-Barré syndrome.[3] The antigenic target for the more common variant of Guillain-Barré syndrome, the demyelinating form, is still unknown. However, the more rare axonal variants have been associated with the detection of anti-ganglioside auto-antibodies in the serum.[2]

There are few reported cases of parainfectious inflammatory conditions involving the CNS and peripheral nervous system, for which the term acute combined demyelination was introduced.[1] Both Guillain-Barré syndrome and ADEM manifest with various degrees of severity, and they tend to be monophasic and self-limited. Their clinical presentations are characterized by a prodromic phase, followed by neurological deficits that peak early and recover gradually. Although high doses of corticosteroids have been shown to speed recovery from ADEM, they are not indicated in Guillain-Barré syndrome. Plasmapheresis and intravenous immunoglobins have been successfully used for both Guillain-Barré syndrome and ADEM, and they should be the treatments of choice in cases of acute combined demyelination syndrome.

That the serology for the anti-ganglioside antibodies was obtained after our patient had undergone plasmapheresis may account for her negative results. Interestingly, the severity of the neuropathy that affected the sensory and motor nerves did not mask the sensory findings related to the demyelinating lesions of the spinal cord. Considering the negative initial MR imaging study of the brain and the timing of the manifestations of the neurological deficits, the CNS involvement was obviously part of a delayed diffuse auto-immune response likely triggered by the same infectious agent that caused the initial upper respiratory illness and polyneuropathy. The early treatment with plasmapheresis and IV immunoglobulins may have contributed significantly to the patient's recovery from the inflammatory changes in both the CNS and peripheral nervous system.

References

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