

# Never Before Reported Acute Autonomic Sensory Motor Axonal Neuropathy Associated with Neuromyelitis Optica: A Case Report

Bruce Zhang, MD<sup>1</sup>, Jessica Calandra, DO<sup>2</sup>, Marlyn S Ramos Lamboy, MD<sup>3</sup>, Channarayapatna R Sridhara, MD<sup>3</sup>

<sup>1</sup>Department of Physical Medicine & Rehabilitation, Temple University Hospital, Philadelphia, PA

<sup>2</sup>Department of Rehabilitation Medicine, Thomas Jefferson University, Philadelphia, PA

<sup>3</sup>MossRehab, Elkins Park, PA

## CASE DIAGNOSIS

**Case Diagnosis:** 29 year old woman with neuromyelitis optica associated acute autonomic sensory motor neuropathy (AASMN), as found on electrodiagnostic studies.

## CASE DESCRIPTION

### History

Patient is a 29-year-old woman who developed acute on chronic decreased vision with coincident progressive bilateral lower extremity weakness and paresthesias and weight loss.

A comprehensive work up was obtained:

- Complete spine MRI revealing T2 hyperintensity extending from the cervical medullary junction through the entire cervical cord involving the dorsal central aspect of the cord, involvement of the thoracic cord extending down to the level of T11/T12, and mild enhancement of the roots of the cauda equina (Figure 1).
- CSF studies: colorless, elevated protein including alpha 1 globulin, elevated albumin, IgG.
- Serum Labs: low folic acid, elevated vitamin B12, low copper levels.
- Neuromyelitis optica antibodies negative
- Regardless, Neurology with strong clinical suspicion for Neuromyelitis Optica

With severe loss of vision, extremity weakness causing considerable functional decline, she was admitted to inpatient rehabilitation. Due to concern for superimposed peripheral neuropathy, she was referred for electrodiagnostic studies.

### Physical Examination

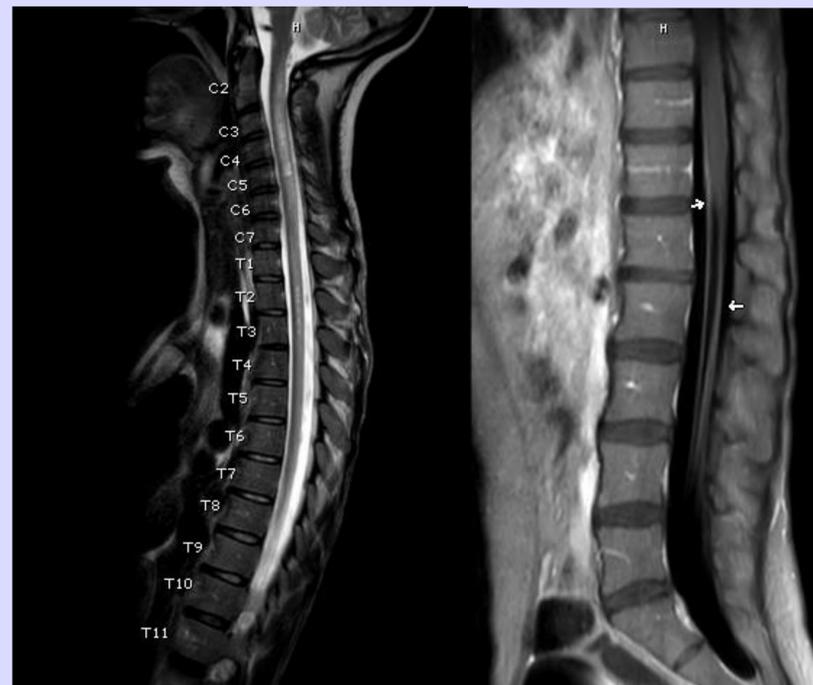
- Cachexia, diffuse atrophy, and trophic changes on her feet and hands (Figure 2).
- Diminished sensation in a stocking-glove distribution.
- Symmetric weakness more pronounced distally than proximally, with antigravity strength in her wrist extensors, but only trace ankle dorsiflexors.
- Hyperreflexia in her upper limbs but mute in her Achilles tendon bilaterally.

### Electrodiagnostic Studies

Refer to Figures 3 and 4 for nerve conduction study and electromyography data.

In summary:

- Non-responsive tibial motor nerve stimulation with markedly reduced compound muscle action potential amplitude with normal distal latency and conduction velocity in the peroneal nerve from the tibialis anterior indicated motor axon loss
- Left superficial peroneal sensory nerve with reduced sensory nerve action potential amplitude corroborated with sensory axonal loss.
- Similar findings were noted in the upper limb median motor and sensory nerves tested
- Combined with needle examination findings involving the distal muscles in the upper and lower limbs and lack of sympathetic responses in the lower extremity, the constellation of findings suggest an **acute autonomic sensory motor neuropathy (AASMN)**
- Additionally, active denervation in L2-S1 myotomes may also suggest corresponding radiculopathy, which corroborate with MRI findings of increased signal in the cauda equina.



**Figure 1:** Left – T2 weighted sagittal thoracic spine. Note hyper-intensities traversing the spinal cord. Right – T1 weighted sagittal lumbar spine. Arrows label hyper-intensities highlighting the cauda equina.

Motor Summary Table												
Stim Site	NR	Onset (ms)	Norm Onset (ms)	O-P Amp (mV)	Norm O-P Amp	Neg Area (mVms)	Site1	Site2	Delta-0 (ms)	Dist (cm)	Vel (m/s)	Norm Vel (m/s)
Left Median Motor (Abd Poll Brev) 32.1°C												
Wrist		3.9	<4.2	2.5	>4	8.75	Elbow	Wrist	4.7	23.4	50	>49
Elbow		8.6		2.3	>4	9.53						
Left Peroneal TA 2Chan Motor (TA) 26.9°C												
BFib-TA		3.1	<4.2	0.4	>1.5	3.59	Knee-TA	BFib-TA	1.4	7.2	51	>40
Knee-TA		4.5		0.4		4.62						
Left Tibial Motor (Abd Hall Brev) 27.3°C												
Ankle	NR		<4.8		>2							
Left Ulnar Motor (Abd Dig Minimi) 32°C												
Wrist		2.8	<3.5	6.4	>4	31.69	B Elbow	Wrist	4.1	21.5	52	>49
B Elbow		6.9		7.3	>4	31.57						

Sensory Summary Table													
Stim Site	NR	Onset (ms)	Norm Onset (ms)	Peak (ms)	O-P Amp (µV)	P-T Amp (µV)	Norm P-T Amp	Site1	Site2	Delta-0 (ms)	Dist (cm)	Vel (m/s)	Norm Vel (m/s)
Left Median D2 Sensory (2nd Digit) 31.8°C													
Wrist		2.8	<3.5	3.8	12.3	20.1	>20	Wrist	2nd Digit	2.8	14.0	50	>40
Left Sup Peroneal Sensory (Ant Lat Mall) 27.8°C													
Low Leg		4.8	<3.3	6.3	4.1	4.1	>8	Low Leg	Ant Lat Mall	4.8	14.0	29	>42
Left Ulnar Sensory (5th Digit) 31.9°C													
Wrist		3.2	<3.1	4.0	10.3	40.8	>18	Wrist	5th Digit	3.2	14.0	44	>45

**Figure 3:** Nerve Conduction Studies.



**Figure 2:** Patient's left foot. Note distal skin trophic changes.

Sympathetic Skin Response Summary Table						
Stim Site	NR	Onset (ms)	Norm Onset (ms)	O-P Amp (µV)	Norm O-P Amp	
Left Palm, Left Sole SSR (Palm and Sole)						
Palm		1718.8		162.7		
Sole	NR					
Palm		1562.5		366.2		
Sole	NR					

### EMG:

Side	Muscle	Ins Act	Fibs	Psw	Fascs	HF	Amp	Dur	Poly	Recr	Int Pat	Comment
Left	AntTibialis	Incr	Large	Large	None	Nml	Nml	Nml	1+	Discrete	Incomplete	
Left	Med Gastroc	Incr	Nml	Large	None	Nml	Nml	Nml	1+	Nml	Incomplete	
Left	VastusMed	Nml	Nml	Nml	None	Nml	Nml	Nml	0	Reduced	Incomplete	
Left	TensorFasLat	Nml	Nml	Nml	None	Nml	Nml	Nml	1+	Nml	Complete	
Left	Iliopsoas	Incr	Nml	Large	None	Nml	Nml	Nml	0	Reduced	Complete	
Left	GluteusMax	Incr	Nml	Large	None	Nml	Nml	Nml	1+	Reduced	Complete	
Left	Deltoid (Mid)	Nml	Nml	Nml	None	Nml	Nml	Nml	0	Reduced	Complete	
Left	Biceps	Nml	Nml	Nml	None	Nml	Nml	Nml	0	Nml	Complete	
Left	Triceps	Nml	Nml	Nml	None	Nml	Nml	Nml	0	Nml	Complete	
Left	Ext Indicis	Incr	Lrg/Sm	Lrg/Sm	None	Nml	Nml	Nml	0	Nml	Incomplete	
Left	1stDorint	Incr	Nml	Lrg/Sm	None	Nml	Nml	Nml	0	Nml	Complete	
Left	Abd Poll Brev	Incr	Large	Large	None	Nml	Nml	Nml	0	Reduced	Incomplete	

**Figure 4:** Needle Electromyography

## DISCUSSION

Neuromyelitis Optica (NMO), otherwise known as Devic's Syndrome, is an auto-immune mediated disorder of the central nervous system (CNS) characterized by relapsing episodes of optic neuritis, myelitis, or both (1). It can cause considerable disability due to severely impaired ambulation and visual deficits. Autoantibodies to aquaporin-4, the most abundant water channel protein in the CNS, is believed to play a role in the pathogenesis (1).

NMO has very rarely been associated with axon loss neuropathy. Only a handful of case reports have previously described an association between NMO and peripheral neuropathy (2, 3). Peripheral neuropathies may be under-reported because electrodiagnostic studies are not routinely ordered as part of NMO work up (2). One case report described an auto-antibody serum negative NMO with superimposed acute axonal sensory motor neuropathy. The patient's functional status improved over the next year following consistent treatment with Rituximab (2).

Acute autonomic sensory motor neuropathy (AASMN) itself is a rare diagnosis, also confined to a handful of case reports. The pathogenesis remains unclear beyond an auto-immune generated response, possibly post-infectious (4). To our knowledge, AASMN associated with NMO has not previously been reported in the literature.

## CONCLUSION

Electrodiagnostic studies are necessary to diagnose peripheral neuropathies that may superimpose on central pathologies like NMO. Diagnosis of specifically AASMN associated NMO has not previously been reported in the literature.

## REFERENCES

- Jarius S, Wildemann B, Paul F. Neuromyelitis optica: clinical features, immunopathogenesis and treatment. *Clin Exp Immunol.* 2014;176(2):149-164. doi:10.1111/cei.12271
- Feyissa AM, Shanina E, Shah R, Smith RG. Neuromyelitis optica phenotype associated with therapy-responsive acute peripheral neuropathy. *Neurol Neuroimmunol Neuroinflamm.* 2015;2(3):e83. Published 2015 Mar 12. doi:10.1212/NXI.0000000000000083
- Chi LM, Gao Y, Nan GX. A case report of neuromyelitis optica spectrum disorder with peripheral neuropathy as the first episode. *Medicine (Baltimore).* 2018;97(10):e0059. doi:10.1097/MD.00000000000010059
- Kinoshita S, Sugie K, Kataoka H, Sugie M, Hirano M, Ueno S. Acute autonomic, sensory and motor neuropathy associated with meningoencephalitis. *Clin Med Case Rep.* 2009;2:17-20. Published 2009 Feb 12. doi:10.4137/ccrep.s2194

