

Title: Anti-Ganglioside Antibodies and Acute Motor Axonal Neuropathy: A Case Report

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Introduction

Guillain-Barré syndrome (GBS) is an acute, inflammatory autoimmune polyradiculoneuropathy characterized by abrupt onset progressive limb weakness and diminished reflexes. Heterogeneous subtypes of GBS are further differentiated by clinical, pathological, and electrophysiological criteria.[1] The predominant trigger is antecedent infection that induces aberrant immune-mediated responses targeting peripheral nerves and spinal roots.[2] *Campylobacter jejuni* (*C. jejuni*) is the pathogen most frequently associated with acute motor axonal neuropathy (AMAN), a sub-group of GBS characterized by axonal degeneration of motor nerves. There is a strong association between AMAN and GM1, GD1a, GD1b, and Ga1NAc-GD1a antibodies.[3] Here we describe a patient who developed AMAN with elevated anti-GM1 and anti-GD1a antibody titers but negative *C. jejuni* serology.

Case Description

A 59-year-old male presented with rapidly progressing bilateral upper and lower extremity weakness. He was previously admitted 12 days prior for suspected viral gastroenteritis and treated with intravenous fluids and discharged home at baseline. Two days prior to this admission, he developed right hand and foot weakness, which quickly progressed to the contralateral side. Initial exam revealed rapidly progressing flaccid quadriparesis without sensory loss. Bilateral biceps, triceps, brachioradialis, patellar, and Achilles muscle tendon reflexes were absent. Cranial nerves were intact. Nerve conduction study (NCS) performed three days following symptom onset revealed reduced compound muscle action potential amplitudes/velocities in right-sided median, ulnar, peroneal, and tibial nerves with preserved sensory responses. Electrophysiologic findings were suggestive of axonal degeneration with evidence of demyelinating features. Six weeks later, repeat NCS demonstrated worsening motor response and evidence supporting a generalized axonal motor polyneuropathy consistent with AMAN. Cerebrospinal fluid analysis did not show albuminocytologic dissociation. Serology tested negative for *Salmonella*, Shiga toxin 1/2, and *Campylobacter*. Anti-GM1 and anti-GD1a antibody titers were high. Patient received two 5-day courses of intravenous immunoglobulin (IVIG) following symptom onset which halted, but did not reverse, progression of weakness. After 3 months of inpatient rehabilitation, he was discharged home at supervision level for ambulation.

Discussion

The role of anti-ganglioside antibodies in the immunopathogenesis of GBS is not clearly understood. It has been proposed that antibody formation is triggered by molecular mimicry between *C. jejuni* lipopolysaccharides and neural tissue, which induces a cross-reactive immune response producing axonal degeneration.[4] This explains the association between anti-GM1 antibody and antecedent *C. jejuni* infection; however, there have been several studies which have found only

a 50% concordance between serologically positive *C. jejuni* and anti-GM1 antibody.[1] The low sensitivity of anti-*C. jejuni* assay and the presence of other microorganism(s) that triggers anti-ganglioside responses and AMAN could explain this discordance.

Conclusions

Despite negative *C. jejuni* serology, patient presented with high anti-ganglioside antibody titers and AMAN. Electrodiagnostic studies showed evidence of axonal degeneration, which is thought to be an indicator of poor prognosis. However, after intensive inpatient rehabilitation, patient demonstrated significant functional recovery at 3-months following symptom onset.

References

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Biography

Daniel Kuo received his Doctor of Osteopathic Medicine degree from Touro University Nevada College of Medicine in Las Vegas, NV in 2016. He completed his internship at Nassau University Medical Center in East Meadow, NY. He is currently a Physical Medicine and Rehabilitation resident at Tufts Medical Center in Boston, MA and is expected to complete his residency in 2020.

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