Abstract

Objective: Paranodal antibodies against neurofascin155 (NF155) have been well-described in chronic and acute demyelinating diseases of both the central and peripheral nervous system, but have not yet been associated with acute axonal pathology¹. Here we characterize the demographic, clinical, laboratory, and electrodiagnostic features in a cohort of adult women of Native American ethnicity who presented with rapidly progressive, severe, subacute painful paresthesias and were found to have axonal polyneuropathy associated with positive serum NF155 antibody.

Methods: Five patients presented with severe, rapidly progressive, symmetric, painful paresthesias, and were found to have sensory greater than motor axonal polyneuropathy. All patients were tested with the Washington University sensory and motor neuropathy panel which includes NF155 and contactin-1. All five patients underwent EMG/NCS testing. Demographic and clinical data were collected.

Results: All cases presented with rapidly progressive distal neuropathic pain, sensory ataxia, areflexia, and mild symmetric distal weakness. All were women of self-reported full or partial Native American ethnicity. All had evidence of severe axonal polyneuronopathy on EMG/NCS. Four of the five cases were seropositive for NF155. Seropositive patients were trialed on a course of IVIg with two of these patients subsequently receiving plasmapheresis. This treatment led to a mild subjective and clinical improvement in just one of the patients. Clinical follow up is ongoing in three of the five patients. Two patients have since died from unrelated causes.

Conclusions: This series highlights rapidly progressive axonal polyneuronopathy associated

with NF155 in a small cohort of women of Native American ethnicity. To our knowledge, this is

the first time these antibodies have been described in association with acute axonal pathology.

References

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