Hypertrophic nerves producing myelopathy in fulminant CIDP

Figure Hypertrophic nerves in chronic inflammatory demyelinating polyradiculoneuropathy

(A) Sagittal and (B) parasagittal T2-weighted MRI of the cervical spine demonstrating hypertrophic nerve roots (arrows). (C) Cervical spine at autopsy showing compression from intradural hypertrophic nerve roots (arrowheads). Cervical nerve root histopathology with onion bulb formation on (D) methylene blue semithin epoxy sections and (E) Schwann cell (S-100) immunostaining.

A 59-year-old man died following a 20-year history of fulminant chronic inflammatory demyelinating polyradiculoneuropathy (CIDP). His symptoms began with gait unsteadiness and ascending severe weakness and paresthesias, followed by papilledema and compressive cervical myelopathy from hypertrophic nerve roots (figure). Initial aggressive immunotherapy resulted in a return to ambulation and employment (previously reported as case 1).¹ Years later, he became quadriplegic due to a combination of immunotherapy-resistant CIDP (including cyclophosphamide) and consequent worsening cervical myelopathy. Varied extent and type of immune mechanisms in CIDP are inferred by such treatment-refractory patients.²

Nathan P. Staff, MD, PhD, Juan J. Figueroa, MD, Joseph E. Parisi, MD, Christopher J. Klein, MD, Rochester, MN

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Address correspondence and reprint requests to Dr. Christopher Klein, Department of Neurology, Mayo Clinic, 200 1st Street SW, Rochester, MN 55905; klein.christopher@mayo.edu

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