A Longitudinal Study of Patients with Chronic Inflammatory Demyelinating Polyneuropathy (CIDP): Identifying Ultrasonographic Features for Prognosis

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Objectives Diagnosis and treatment monitoring in CIDP is primarily based on clinical parameters. High-frequency ultrasound reflects nerve pathophysiology non-invasively and painlessly, and has been demonstrated as a useful additional diagnostic tool in CIDP. However, correlations with disease state and response to treatment have been mixed. This prospective 12-month study aimed to identify potentially useful prognostic and treatment-related biomarkers utilising neuro muscular ultrasound.

Methods We recruited 35 patients with CIDP and other immune-mediated neuropathies who were currently or about to commence treatment. 32 participants completed 12 months of follow-up, with standardised clinical and ultrasonographic assessment at baseline, 3- and 12 months. Our protocol included bilateral, whole-length assessment of the median and ulnar nerves, with unilateral assessment of other nerves, measuring cross-sectional area (CSA), echogenicity, vascularity and morphological findings.

Results As with our previous retrospective and cross-sectional studies, nerve size variability was demonstrated in nearly all CIDP participants, particularly nerve enlargements in proximal upper limb nerves. However, nerve size parameters correlated poorly with clinical state or change over time. By contrast, other morphological findings, including changes in echogenicity and fascicular appearance, demonstrated suggestive correlations with remaining stable on weaning treatment, as well as in participants who demonstrated clinical improvement. The most promising parameters will be presented.

Conclusions This longitudinal study of neuromuscular ultrasound in patients with CIDP identified morphological findings – particularly based on echogenicity and fascicular appearance rather than CSA measurement – that may have potential as biomarkers in identifying treatment-responsive patients, along with those who may have ‘burnt out’ disease.

COVID-19 MRI-Negative Myelopathy – A Distinct Syndrome? Three Patients and a Review of the Literature

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Background Acute MRI-positive transverse myelitis is a well documented post-infectious phenomenon following SARS-CoV-2 infection. There are reports of MRI-negative presentations of myelitis following SARS-CoV-2 infections.

Methods Between September and December 2022, three patients presenting to hospitals in the Auckland region, New Zealand, were prospectively followed by the authors. Their clinical features are outlined. A literature search for patients with MRI-negative myelitis/myelopathy following SARS-CoV-2 infection was conducted.

Results We document three patients with initially negative MRI who presented with symptoms and signs of myelopathy – progressive paraparesis with significant gait disturbance, impaired proprioception, Lhermitte’s phenomenon, sensory level, hyperreflexia and spasticity. Despite treatment with corticosteroids, all patients had progression of symptoms. Ten further patients with initial normal MRI of the spinal cord have been described in the literature. The mean age was 58, and 7 were women. The onset of symptoms ranged from 1 to 16 weeks after SARS-CoV-2 infection. All patients had lower limb onset of symptoms, with 10 developing paraparesis. Nearly all had significant dorsal column involvement, often with sensory ataxia. Our 3 patients exhibited early Lhermitte’s phenomenon as a distinctive feature. Most patients had hyperreflexia and a sensory level. Treatment was most commonly with intravenous methylprednisolone, with rare improvement.

Conclusions Due to the novel description of these patients alongside the absence of initial MRI findings, the diagnosis can easily be overlooked. We propose these patients have a distinct syndrome of COVID-19 MRI-negative myelopathy.