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Extensive leukoencephalopathy as a differential diagnosis of motor neuron disease: case report and literature review

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Background: Weakness is frequently a reason to attend neurology consultation which entails a challenge due to its wide range of diagnostic possibilities. We present the case of an elderly woman with long-standing right upper limb weakness. Methods: Patient data was obtained from medical records and an extensive neuropathological evaluation was performed. Results: Weakness started off in her right hand, which progressed insidiously to her right hemibody. with hyperreflexia, atrophy, fasciculations and foot drop. Subsequently, bulbar and left hemibody deterioration began. She died due to ventilatory failure four years after the first symptom appeared. A tigroid pattern in the neuroimaging studies suggest white matter involvement, while the neuropathological studies showed loss of motor neurons in the spinal cord. Conclusions: The available evidence does not allow us to dismiss the hypothesis of a motor neuron disease or a leukodystrophy. Cases like this one establish a diagnostic challenge due to their complexity and unusual etiology.

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Implications of Gold Coast Criteria in diagnosis of amyotrophic lateral sclerosis in a large subspecialty clinic

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Background: Criteria to formalize the diagnosis of amyotrophic lateral sclerosis (ALS) and refine clinical trial populations have evolved. The recently proposed Gold Coast criteria (GCC) are intended to simplify use and increase sensitivity. We evaluated sensitivity of GCC and potential impacts on therapeutic trial enrollment. Methods: We performed a single center retrospective study including patients diagnosed with ALS between 2016 – 2021. We determined criteria met at diagnosis according to revised El Escorial (rEEC), Awaji (AC) and GCC. We compared sensitivity and examined impacts GCC would have on enrollment in landmark ALS trials. Results: We included 203 people with ALS. Sensitivity of GCC (96.1%, 95% confidence interval [CI] = 92.2-98.2%) was significantly higher than rEEC (89.8%, 95% CI 84.6-93.4%, $\chi^2 = 5.3$, $p = 0.01$) and AC (89.3%, 95% CI 84.1-93.0%, $\chi^2 = 6.1$, $p = 0.006$). GCC was more sensitive than clinically definite or probable rEEC (47.6%, 95% CI 40.6-54.6%, $\chi^2 = 117.6$, $p = < 0.001$) and use would result in increased eligibility in landmark therapeutic trials. Conclusions: GCC are more sensitive than rEEC and AC at time of diagnosis in ALS. Use of GCC in our population would expand clinical trial participation and make results more widely generalizable.

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Objective measures of balance deficits in sensory ganglionopathy

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Background: Sensory ganglionopathy (SG) is a rare form of neuropathy affecting the dorsal root ganglia and leading to non-length-dependent sensory abnormalities. Although balance problems are frequently reported by patients, a comprehensive balance assessment in SG has not been conducted. This study quantifies balance deficits in SG and examines their relation to patient-reported outcome measures (PROMs). Methods: Prospective data was collected from five participants with SG. Balance assessments included Fullerton Advanced Balance scale, Berg Balance scale, and 360 degree turn. Participants completed PROMs assessing balance confidence (ABC scale), pain, fatigue, quality of life (QoL), and daily activity and participation. Assessment also included neurological exam, nerve conduction studies (NCS) and posturography. Results: All participants had severe SG on NCS with normal strength and significant sensory abnormalities. Balance scores indicated severe balance deficits in all participants and aligned with posturography and truncal sway measures. PROMs revealed low confidence in balance, high levels of pain and fatigue, difficulties with daily activities, and reduced QoL. Conclusions: Although balance testing is not part of routine clinical practice, PROMs and targeted assessment may help monitor patients with SG and their response to treatment. Larger sample sizes are needed to understand the impact of balance on PROMs and optimize bedside balance testing.

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Determining individual substantial response in amyotrophic lateral sclerosis: utilizing a new method on CENTAUR trial results

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Background: In ALS, determining whether individuals have a substantial response to therapy is a challenge for the field. ALS naturally progresses at variable rates and a personalized approach is required to determine if individuals have a substantial response. A new method to evaluate individual response is proposed and applied to data from the CENTAUR trial of sodium phenylbutyrate/ursodoxicoltaurine (PB/TURSO). Methods: In a post hoc analysis, CENTAUR participants whose actual rate of change from baseline in the ALSFRS-R at week 18 was ≤ their own trial baseline progression rate (ΔFS) were defined as having a substantial individual response in slowing ALS progression. Results: Substantial individual response was observed in a greater proportion of participants receiving PB/TURSO (41%, n=87) vs placebo (19%, n=48; *P*=0.0076). Conclusions: Response versus