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Pediatric quadriplegic CIDP: an update

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Background: Chronic inflammatory demyelinating polyradiculoneuropathies (CIDP) is a rare, acquired polyneuropathy, especially in children, affecting the peripheral nervous system. It most commonly presents in a symmetric, proximal and distal, sensorimotor fashion. Immunosuppression and immunomanipulation are treatment modalities. This is an update of the 14-year-old male presented, at the 2024 CNSF meeting, with severe progressive CIDP who became refractory to steroid and IVIg but responded to Rituximab. Methods: At the age of 16, at a stage of recovery where he had regained MRC grade 5 strength diffusely, all therapies were sequentially tapered (prednisone & IVIg) or withdrawn (rituximab). He did well for approximately 6 months at which point he relapsed severely to the point of needing a walker. Repeat electrodiagnostics confirmed latency, amplitude, and conduction velocity deteriorations as compared to previous. As a result, prednisone, IVIg, and rituximab were reinstated. Results: As per the first exposure to rituximab, the patient began to respond roughly 3 months after the retreatment with targeted anti-CD20 therapy. He has again made a complete recovery physically but not electrophysiologically. This is the same pattern of response that was originally manifested. Conclusions: Rituximab appears to be an effective treatment for severe IVIg-refractory pediatric CIDP.

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A case of refractory NF155 paranodopathy with near complete sponteneous recovery

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Background: CIDP is a rare immune-mediated demyelinating neuropathy which has significant phenotypic variability. Despite extensive efforts, a unifying immunopathologicalmechanism remains elusive and this is likely due to etiological heterogeneity among the variant presentations. This is best exemplified by the identification of nodal/paranodal antibodies, such as neurofascin 155 (NF155) in a small subgroup of CIDP patients, who present with a distinct phenotype and embody a poor response to IVIG. Methods: We present the case of a 39-year-old male who presented with a 2-year history of progressive stocking-glove sensory loss and sensory ataxia. Electrodiagnostics confirmed an acquired demyelinating neuropathy, with serum anti-NF155 IgG4. His case was refractory to standard immunomodulatory therapy, including adequate trials of IVIG, steroids, azathioprine, and rituximab. He also had a non-therapeutic trial of PLEX, methotrexate, and tacrolimus. Results: After cessation of all immunomodulatory therapy for 2 years, he had spontaneous remission of his CIDP and near-complete resolution of electrodiagnostic/clinicalabnormalities. Conclusions: This case highlights a unique supra-refractory seropositive CIDP variant which underwent spontaneous remission with near-complete resolution. Delayed effect from rituximab was posited as a contributor, however, the patient had no clinical or electrophysiological improvement 20-months after initiation of anti-CD20 therapy. Current data suggests that CIDP respond to rituximab within 6-12 months.

Epilepsy and EEG

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Investigation of clinical features, EEG findings, and brain imaging in psychiatric patients with epilepsy at Razi Psychiatric Hospital

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Background: This study aimed to assess the clinical features, Electroencephalography (EEG) findings, and brain imaging results in psychiatric patients diagnosed with epilepsy at Razi Psychiatric Hospital. Methods: This retrospective descriptive-analytical study was carried out on patients with epilepsy and psychiatric disorders admitted to Razi Psychiatric Hospital over two years. A total of 94 patient files seizure and epilepsy comorbidity, recorded in the hospital's health information system (HIS), were reviewed. Data collection involved a demographic checklist and an epilepsy scale; the latter, developed by DiIorio, Colleen, et al., encompassed personal characteristics, mental disorders, epilepsy, and seizures. The Kruskal-Wallis and MannWhitney non-parametric tests were utilized to compare the mean scores of variables, with SPSS software, version 21 facilitating the analysis Results: Out of 94 patients with seizure and epilepsy, 9.6% had focal seizure, 26.6% had generalized epilepsy, 36.1% had focal-generalized seizure, and 26.8% had unknown seizure. About 12% had a structural etiology, while the remaining 88% had an etiology that remained unidentified. Conclusions: The findings indicate that epilepsy, affecting individuals from adolescence through to old age, can lead to psychiatric disorders. For many patients, the etiology of their condition remains elusive, and EEG findings and brain imaging appear normal in the majority of cases