Autoimmune encephalitis: modifiable and non-modifiable predictors of relapse

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Background: Approximately 25% of encephalitis cases in North America are autoimmune (AIE). For most forms of AIE, it is unclear which patients have the highest relapse risk and whether standard treatments reduce this risk. Our objective was to determine the overall risk of relapse and whether chronic immunosuppressive therapy modifies that risk. Methods: We performed a chart review consisting of all patients with AIE presenting to the Calgary Neurology Clinic and Tom Baker Cancer Centre between 2015 and 2020. Predictors of relapse were determined with use of t-test. Results: Outcome data was assessable in 39 patients, 17/39 (44%) patients relapsed, and most relapses (76%) occurred within 3 years. Patients not on any immunosuppression at the time of relapse had a greater increase in CASE score, a proxy for presentation severity, at relapse compared to those on immunosuppression (p=0.0035). Conclusions: The risk of relapse in AIE is high (44%). Immunosuppression at the time of relapse, which may occur up to 3 years after initial presentation, lessens the relapse severity, although it remains unclear if it can reliably prevent relapses. Our data enforces the importance of long-term follow up and that ongoing immunosuppression may be helpful, particularly in the first 3 years after initial presentation.

Is there utility in duplicating antibody testing for autoimmune encephalitis? A comparison of results obtained from Mayo and Mitogen Dx

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Background: Autoantibody testing for suspected autoimmune encephalitis (AIE) in Alberta is commonly performed by Mitogen Dx (MDx) using cell-based assays (CBAs) for cell surface antibodies and line immunoassay (LA) for intracellular antibodies without confirmatory tissue immunofluorescence/immunohistochemistry (TIFF/IHC). Duplicate testing is often sent to Mayo Clinic (MC) in case of disagreement. Methods: Antibody panel results were obtained for all patients who had testing sent to both MC and MDx from adult hospitals in Alberta between 2018 and 2020. Positive antibodies were evaluated to be pathogenic/non-pathogenic by chart review and expert consensus. Results: Thirty-four individuals had antibody panels completed at both labs. Overall agreement (positive/negative panel) was fair (κ = 0.24, p = .08), even after excluding low-titre GAD65 antibodies through MC (n=9, 26.5%). MDx reported more non-pathogenic serum results, including: anti-SOX1 (n=3), anti-NMDAR (n=2) and anti-GABA(B)R (n=1). All pathogenic antibodies (n=3) were positive in both laboratories. Conclusions: No new pathogenic antibodies were identified by sending duplicate testing to MC; however, a larger number of non-pathogenic antibodies were reported by MDx, likely due to lack of confirmatory TIFF/IHC. Antibody testing for AIE should be done in labs performing confirmatory TIFF/IHC on all CBA/LA results to avoid unnecessary investigations and/or treatments.

Relevance and home completion rate of patient reported outcome measures set in chronic inflammatory neuropathies

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Background: Patient-centred care is important in the management of chronic inflammatory neuropathies (CIN) given the heterogeneity in disease course and treatment response. Patient Reported Outcome Measures (PROMs) support value-based healthcare by aligning treatment goals with what matters most to patients. This study evaluated the relevance of PROMs to patients and the feasibility of using them in clinical management. Methods: PROMs assessing quality of life, pain, fatigue, and overall disability were collected prospectively from 32 patients with CIN every three months over a 12-month period. Results: Completion rate was 92%. Home vs. in-clinic completion increased from 56% to 85% over the course of the study. There was an association between completion of the panel and perceived relevance. The PROMs were consistently rated as highly relevant, with disability and fatigue measures rated highest. Conclusions: PROMs are appraised as highly relevant among patients with CIDP and MMN. Patients require support initially but adapt to electronically delivered home completion of questionnaires. We recommend inclusion of PROMs into routine clinical practice as a means of capturing aspects of health that are not easily assessed in a clinic visit.

Antibody testing for autoimmune encephalitis: a multisite study examining clinical practices in a large Canadian city

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Background: Antibody panels are one diagnostic tool within the comprehensive evaluation of suspected autoimmune encephalitis (AIE). Over-reliance on antibody panels contributes to misdiagnosis and inflated healthcare costs. Methods: Inpatients or outpatients who had AIE antibody testing ordered from one of four adult hospitals in Calgary between January 2018 – January 2020 were included. Results: Antibody panels were sent for 469 individuals during the 2-year period; 42 were positive (9.0%) of which 10 were pathogenic. Of 150 individuals included in chart review, 27 (18.0%) met criteria for possible AIE at presentation and 16 (10.8%) met criteria for definite AIE at final diagnosis. Overall, antibody testing was ordered in both serum and CSF in 36.3% (versus 69.2% meeting possible AIE criteria); MRI brain was performed in 92.7% (possible AIE 92.6%), EEG in 78.7% (possible AIE 100.0%), and lumbar spine tap in 56.5% (possible AIE 58.1%).