

to have less high risk disease. However, more studies are needed to determine whether IM medications play a role in neuro-autoimmune disease progression and mortality. A limitation of this study is that data was collected from a single institution and does not represent the general population.

Disclosure: Mr. Ahmed has nothing to disclose. Mr. Ahmed has nothing to disclose. Mr. Trivedi has nothing to disclose. Mr. Ors has nothing to disclose. Mr. Ahmed has nothing to disclose. Mr. Jaffry has nothing to disclose. Dr. Souayah has received publishing royalties from a publication relating to health care.

Do Those With Neuro-Autoimmune Disease Carry a Higher Burden of Disease?

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Objective

To investigate the burden of disease and their prognosis amongst patients with neuro-autoimmune disease (NAD).

Background

NAD has been shown to increase overall mortality and early death among patients. However, the overall burden of disease in NAD patients has not yet been fully characterized.

Design/Methods

A retrospective analysis on 34464 patients hospitalized at a tertiary care center in a major metropolitan area was conducted. The outcomes compared included the prevalence of comorbidities, high risk comorbidities, and rheumatoid arthritis and/or lupus (RAL) among patients with and without neuro-autoimmune disease (NAD and nNAD). The outcomes of the initial disposition after discharge, length of hospital stay, ICU admission, and death among patients with comorbidities and with or without neuro-autoimmune disease (cNAD and ncNAD) was also determined.

Results

There is no significant difference in the level of comorbidity (53% vs 54%) or high risk comorbidities (19% vs 24%) between patients in NAD and nNAD, respectively ($p > 0.05$). 4.7% of NAD and 2.2% of nNAD patients had RAL ($p < 0.02$). The mortality was 5% in cNAD and 4.3% in ncNAD ($p > 0.05$). ICU admissions was 16% in cNAD and 20% in ncNAD ($p > 0.05$). 42% of patients in cNAD and 72% in ncNAD were discharged home ($p < 0.0001$). The average length of stay was 10 and 6.7 days for patients in cNAD and ncNAD, respectively ($p < 0.01$).

Conclusions

These results suggest that NAD may not affect the overall burden of disease in patients but may increase the prevalence of RAL. Furthermore, comorbidity status may correlate with length of stay and disposition in patients with NAD.

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“Obvious” Indications for Neural Antibody Testing in Epilepsy or Seizures: The ONES Checklist

Yiu-Chia Chang, Maryam Nouri, Seyed Mirsattari, Jorge Burneo, Adrian Budhram

Objective

To develop a checklist that identifies patients who have “obvious” indications for neural antibody testing, and compare its diagnostic performance to predictive scores.

Background

Numerous predictive scores have been developed to help determine which patients with epilepsy or seizures of unknown etiology should undergo neural antibody testing. However, their diagnostic advantage compared to only performing testing in patients with “obvious” indications (e.g. broader features of autoimmune encephalitis) requires further study.

Design/Methods

We developed the “Obvious” indications for Neural antibody testing in Epilepsy or Seizures (ONES) checklist through literature review. We then retrospectively reviewed patients who underwent neural antibody testing for epilepsy or seizures at our center between March 2019 and January 2021, to determine and compare the sensitivity and specificity of the ONES checklist to the recently-proposed Antibody Prevalence in Epilepsy and Encephalopathy (APE2)/Antibodies Contributing to Focal Epilepsy Signs and Symptoms (ACES) reflex score.

Results

One-hundred-seventy patients who underwent neural antibody testing for epilepsy or seizures were identified. Seventy-four of 170 (43.5%) with a known etiology were excluded from sensitivity/specificity analyses; none had a true-positive neural antibody. Of the 96 patients with an unknown etiology, fourteen (15%) had a true-positive neural antibody. The proportion of false-positives was significantly higher among patients with a known etiology (3/3, 100%) compared to an unknown etiology (2/16, 13%) ($P = .01$). There was no significant difference of the APE2/ACES reflex score compared to the ONES checklist with regard to sensitivity (93% for both, $P > .99$) or specificity (71% versus 78%, $P = .18$) for true-positive neural antibodies.

Conclusions

Compared to only performing neural antibody testing in patients with epilepsy or seizures of unknown etiology who have “obvious” indications, predictive scores confer no clear diagnostic advantage. Pre-specified definitions of what constitutes a true-positive neural antibody is required in future studies to avoid false-positives that can confound results.

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Application of APE2 and RITE2 Scores in a Stanford Cohort of Autoimmune Encephalitis Patients

Trevor Rafferty, Anna Koeppen-Babcock, Srikanth Muppidi, Scheherazade Le

Objective

The goal of our study was to apply the APE2 and RITE2 scores in a cohort of autoimmune encephalitis (AE) patients at Stanford with immune-mediated seizures.

Background

Early identification and immunotherapy in those with immune-mediated seizures are associated with better neurologic outcomes and reduction of seizures. There have been previously published scoring systems to identify antibodies (Ab) and responsiveness to immunotherapy that were applied to our cohort.

Design/Methods

This was a retrospective study at Stanford University Hospital with chart review of the electronic medical record between 2008-2021. Patients

were included if they had acute symptomatic seizures secondary to autoimmune encephalitis or autoimmune-associated epilepsy as defined by the International League Against Epilepsy (ILAE) and possible, auto-Ab negative but probable, or definite AE using diagnostic criteria from Graus. Patients were excluded if no Ab panel was drawn or the patient was lost to follow-up. Chart review was used to calculate scores.

Results

Fifty-six patients were identified, and 3 were excluded. The APE2 score = 4 to predict positive serum Ab had a sensitivity of 92.7% and specificity of 6.7%. The RITE2 score = 7 to predict seizure responsiveness to immunotherapy had a sensitivity of 92.9% and specificity of 60%.

Conclusions

The APE2 and RITE2 scores were sensitive in our patients, which implies that these scores can likely be used to identify patients within the Stanford cohort who may have seropositive AE and may benefit from early immunotherapy. Our APE2 score and RITE2 scores were less specific than Dubey's, likely due to patient selection. We only included those with suspected AE with seizures, whereas Dubey included a broader, more heterogeneous patient population including those with Parkinsonism, stroke, memory disorders, and other neurologic disorders.

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Treatment Outcome of Autoimmune Associated Epilepsy (AAE) vs Acute Symptomatic Seizures in Autoimmune Encephalitis (ASSAE)—A Single Center Experience

Ning Zhong, Mark Waheed

Objective

To assess the seizure treatment outcome in autoimmune encephalitis (AE).

Background

Seizures due to AE etiology are increasingly recognized. Timely confirmation of autoimmune etiology may lead to improved outcomes by allowing for earlier immunotherapy, which is more effective than anti-seizure medication (ASM). Currently, seizures/epilepsy of autoimmune etiology can be subcategorized as autoimmune associated epilepsy or acute symptomatic seizures in autoimmune encephalitis with varied response to immunotherapy.

Design/Methods

This was a retrospective single center study including patients diagnosed with AE, with a minimum follow-up of 12 months after disease onset in alive cases.

Results

28 out of 31 patients were analyzed (3 without seizure were excluded). 12 were found to have autoantibodies (Abs) against cell membrane (CM) protein (3 anti-NMDAR, 4 anti-GABA_BR, 4 anti-LGI-1, 1 anti-GABA_AR); 4 with non-specific Abs (low titer of anti-GAD, VGCC); 11 with no Abs identified; 3 were with intracellular (IC) Abs (MOG, Hu, GFAP). All patients received immunotherapy and ASM treatment. Among the patients with CM-Abs, 10 of the 12 remained seizure free, 7 were successfully weaned off ASM. Among those with nonspecific/no Abs, only 4 of the 15 patients (26.7%) remained seizure free. All 3 patients with intracellular Abs did poorly, and only one patient with anti-MOG-Abs survived with

drug resistant epilepsy (DRE). Within the group of CM-Abs, 2 patients (16.7%) with anti-GABA_BR-Abs developed DRE, compared to over 70% of patients in group with nonspecific/no Abs or IC-Abs. Upon further investigation of DRE patients, several features were observed: 1) poor response to immunotherapy, 2) sustained abnormal brain MRI T2/FLAIR signal, 3) persistent focal epileptiform features and frequent ictal patterns in EEGs.

Conclusions

Patients with CM-Abs likely suffer ASSAE with favorable long-term outcome. Patients without identified auto-Abs or IC-Abs likely develop AAE and consequently develop DRE. Further research focusing on biomarkers predicting AAE and DRE is needed for treatment guidance.

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Spectrum of Brain MRI Features in CASPR2 Associated Autoimmune Encephalitis: A Case Report With Parietal Lobe Involvement

Luis Manrique-Trujillo, Tiffani Franada

Objective

Contactin-associated-protein-like 2 (CASPR2)-antibody-mediated autoimmune encephalitis (AE) is characterized by diverse clinical manifestations with involvement of both central and peripheral nerve systems.

Background

Furthermore, approximately 50-55% of the patients with this condition present with an abnormal brain MRI including bilateral/unilateral T2-hyperintensity in the mesial temporal lobes, thalami, basal ganglia, brainstem, or cerebellum and hippocampal atrophy/sclerosis. To our knowledge this is the first case of CASPR-Ab-related AE reporting concurrent parieto-occipital lobes enhancing and non-enhancing brain lesions.

Design/Methods

A 71-year-old male presented with 3-months of cognitive decline, generalized tonic-clonic seizures, and parasomnias. Initial neurological examination showed diminished attention with neuropsychology evaluation revealed episodic memory deficits and executive dysfunction, otherwise no focal abnormality.

Results

Brain MRI with and without contrast from showed subtle cortical enhancement in the anterior left parietal lobe, additionally T2 FLAIR hyperintensities in the bilateral hippocampi, left centrum semiovale and left dorsal pons. CSF analysis revealed 5 nucleated cells, protein and glucose were normal. Serum and CSF paraneoplastic panel was positive for CASPR-2 auto antibodies. His memory and executive function improved after plasma exchange and long-term steroids. One year later the patient developed ataxia and gait instability. Repeated examination revealed diminished vibration in bilateral lower extremities, prominent left upper extremity ataxia compared to the right and a wide based gait. Updated brain MRI demonstrated progression of T2 FLAIR hyperintensities in the cortical and subcortical areas of the left parieto-occipital fissure. IVIG was started with stabilization of the progression.

Conclusions

CASPR2-autoantibody is associated to a wide range of neurological manifestations and diagnosing this condition could be challenging. This case demonstrates the possible involvement of the parietal and occipital cortex as a target of the antibodies directed against this membrane protein. Recognition of this spectrum of symptoms and imaging findings

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