

Steroid Un-Responsive Encephalopathy Associated with Autoimmune Thyroiditis (SUEAT) in Pediatric Patients.

Geetanjali Rathore

Objective

We report a series of children with encephalopathy associated with thyroid antibodies who are refractory to steroid monotherapy.

Background

Steroid Responsive Encephalopathy associated with autoimmune thyroiditis (SREAT) is a rare condition, with only a few isolated cases reported in children. Marked clinical improvement following treatment with steroids, is a hallmark of SREAT.

Design/Methods

An IRB approved chart review was conducted on patients <18 years diagnosed with autoimmune encephalitis. A retrospective analysis of clinical features, diagnostic tests, response to therapy and long term follow up was conducted on patients positive for Thyroperoxidase (TPO) antibodies.

Results

52 patients <18 years were diagnosed with autoimmune encephalitis, 10 (19.2%) of these were positive for TPO antibodies. Median age at disease onset was 14.5 years (range 6-18 years) with only 1 male patient being. Mental status (90%) and behavior changes (100%) were most common presentations, seizures were detected in only 1 patient. MRI (20%) and EEG (30%) abnormalities were uncommon, and only 1 patient had evidence of inflammation in cerebrospinal fluid (CSF). Autoimmune encephalitis and paraneoplastic antibody panels were negative besides 2 (20%) patients having concomitant Thyroglobulin (TG) antibodies. All patients needed additional IVIG after steroids treatment, 7 (70%) patients received Rituximab and 3 (30%) patients needed Plasmapheresis. All patients recovered at an average of 4.4 years follow up.

Conclusions

Encephalopathy associated with thyroid antibodies can be steroid unresponsive in the pediatric population. Further immune therapy, including plasmapheresis, should be considered in these patients, even in the absence of other para-clinical evidence of inflammation.

Disclosure: Dr. Rathore has nothing to disclose.

The Role of Plasmapheresis In Pediatric Antibody- Negative Autoimmune Encephalitis

Geetanjali Rathore

Objective

We show the efficacy and relative safety of plasmapheresis as a treatment option for antibody negative AIE in children.

Background

Plasmapheresis is well established therapy for antibody mediated autoimmune encephalitis (AIE). In patients with no identified antibody, the role of plasmapheresis is unclear. Starting plasmapheresis becomes even more controversial in children with antibody negative AIE

Design/Methods

An IRB approved chart review was conducted on patients <18 years diagnosed with autoimmune encephalitis. A retrospective analysis of response to plasmapheresis and long term follow up was conducted on patients that did not have an identified antibody.

Results

52 patients <18 years were diagnosed with autoimmune encephalitis, 14 (26.9%) of these tested negative for antibodies. 2 (14%) patients

received only steroids, while all others received Steroids plus IVIG. 7 (58%) patients received rituximab for poor response/relapse following Steroids plus IVIG. Of these, 3 (43%) patients further underwent plasmapheresis for presumed refractory AIE. All patients had improvement after plasmapheresis and remained symptom free, including seizure freedom, at 2 year follow up. One patient needed repeat plasmapheresis for presumed relapse. No adverse effects reported.

Conclusions

Several studies have shown that timely delivery of immunotherapies is crucial and delay in treatment due to negative autoantibodies can lead to poorer outcomes. Plasmapheresis is safe and should be considered for refractory/relapsing AIE in children, even in the absence of an identified antibody. Larger studies in future can help solidify the findings from our cohort.

Disclosure: Dr. Rathore has nothing to disclose.

First Case Report of AMPA Receptor Encephalitis Presenting With Features of Parkinsonism

Chirag Lalwani, I.M Thushara, Sudheeran Kannothe, Anand Kumar, Vivek Nambiar, Sibi Gopinath, Udit Saraf, Annamma Mathai, Dayana Antony

Objective

NA.

Background

Autoimmune Encephalitis is an inflammatory condition of the brain due to antibodies against onconeural proteins. Paraneoplastic Parkinsonism is very rare. We report the first case of AMPA receptor encephalitis presenting with symptoms of parkinsonism, an atypical presentation of a rare entity.

Design/Methods

NA.

Results

A 71-year-old female with multiple comorbidities presented in a state of stupor with complaints of insidious onset slowly progressive recent memory impairment, progressive slowness in performing daily activities, inability to communicate, and acute onset urinary incontinence. CNS examination initially showed a GCS of E2V1M4 with Grade 2 rigidity in all extremities. MRI Brain showed subtle T2 FLAIR hyperintensities in the peri-ventricular and subcortical white matter. Serum and CSF studies showed AMPA 1 antibody positivity. The FDG PET showed an avid speculated soft tissue density lesion in the upper inner quadrant of the right breast with active right axillary lymph nodes (Histopathology- Infiltrating duct carcinoma Grade 2 NST T2N2aMx). The patient was managed using IVIg and steroids following which her sensorium improved to a GCS of E4M6V5. UPDRS at this point was 29. She subsequently underwent therapy for her tumor following which there was a significant decrease in parkinsonian symptoms and an improvement in memory without the use of any antiparkinsonian medications (UPDRS score-6). During the four years of follow-up, she has remained independent and can perform all her activities of daily living. Hence this autoimmune encephalitis case can be classified as a definite paraneoplastic neurological syndrome (PNS Care score-9).

Conclusions

We propose that Parkinsonism, in our case, is probably a paraneoplastic neurological syndrome associated with antibodies against the AMPA receptor, as the symptoms and signs recovered with cancer treatment.

Disclosure: Mr. Lalwani has nothing to disclose. Miss Thushara has nothing to disclose. The institution of Dr. Kannothe has received research support from Novartis. Dr. Kumar has nothing to disclose. Dr. Nambiar has nothing to disclose.

Sibi Gopinath has nothing to disclose. Dr. Saraf has nothing to disclose. Dr. Mathai has nothing to disclose. Mrs. Antony has nothing to disclose.

Rapidly Progressive Dementia With Recurrent Seizures and Hyponatremia; A Case of LGI1 Limbic Encephalitis

Joshua Luster, Ashley Barasa, William Hoffman

Objective

N/A.

Background

Leucine-Rich Glioma Inactivated Protein-1 (LGI1) autoimmune encephalitis was first described in 2001 as one of the syndromes caused by antibodies against the voltage-gated potassium channels (VGKC) until it was discovered in 2010 that antibodies were instead being directed towards the protein LGI1. This often presents in males in their 60's and is often associated with faciobrachial dystonic seizures, which have become pathognomonic for this disease process.

Design/Methods

N/A.

Results

77-year-old female with history of hyponatremia, anxiety, hypertension, and lacunar infarct presented for a concern for seizures. She presented for multiple episodes of reported generalized tonic seizures and was eventually found to have right frontotemporal seizures with impaired awareness. Magnetic Resonance Imaging (MRI) was repeated multiple times but were significantly degraded due to motion artifact and read as limited. Further discussion with husband was concerning for memory loss over the past 4 months, but patient's children disputed this with several years of memory loss. After neuropsychological testing which demonstrated significant decline across multiple domains. MRI was revisited which was concerning for bilateral mesial temporal hyperintensities on Fluid-Attenuated Inversion Recovery (FLAIR). Patient underwent lumbar puncture given unremarkable workup thus far. CSF and serum both demonstrated LGI1 autoantibodies for which the patient received a 5 day course of IV methylprednisolone, IV immunoglobulins, and was eventually transitioned to rituximab with complete recovery of long term memory.

Conclusions

This case demonstrates the complexity of evaluating a patient for reported rapidly progressive dementia and some of the pitfalls involved in the workup. This case demonstrates that when the initial workup is unremarkable, the patient should be evaluated for uncommon causes, such as autoimmune encephalitis. We diagnosed an atypical presentation of autoimmune encephalitis and documented the initial treatment and response to both first line and second line treatment with future plans to titrate the anti-epileptic drugs.

Disclosure: Dr. Luster has nothing to disclose. Miss Barasa has nothing to disclose. The institution of Dr. Hoffman has received research support from United States Air Force.

Co-Occurrence of Sj/ITPR1 and NMDA Antibodies:

A Case Report

William Chapman, Allison Jordan, Joseph Broderick, Simona Ferioli

Objective

To highlight a case of concurrent anti-Sj/ITPR1 and anti-NMDA encephalitis.

Background

The anti-Sj/inositol 1,4,5-trisphosphate receptor (ITPR1) has been associated with autoimmune cerebellar ataxia and malignancy. Reports

of patients with anti-Sj/ITPR1 describe isolated cerebellar ataxia as well as various manifestations throughout the central and peripheral nervous system. Anti-NMDA encephalitis presents with subacute decline, seizures, movement disorder, alterations in behavior and cognition, autonomic dysfunction, and central hypoventilation but is rarely associated with cerebellar ataxia in adults.

Design/Methods

NA.

Results

A 28-year-old female with no relevant medical history presented to an outside hospital with acute onset headache, diplopia, nystagmus, and vertigo. MRI and MRV were unremarkable. CSF analysis showed a lymphocytic pleocytosis. She was empirically treated with acyclovir, although viral serologies were negative. On initial assessment in our clinic, neurologic exam showed square wave jerks, ataxic eye movements, resting tremor, appendicular and gait ataxia. She progressively declined with gait instability, autonomic dysfunction, neuropsychiatric symptoms, and significant weight gain from compulsive hyperphagia. Her course was complicated by respiratory failure and tracheostomy was placed for mechanical ventilation. Malignancy screening with mammogram, CT, and full body PET was negative. Transvaginal ultrasound was nondiagnostic. Serum paraneoplastic autoantibody panel was negative. EEG showed severe generalized slowing. Repeat CSF studies were positive for anti-Sj/ITPR1 and anti-NMDA. She was treated with high-dose IV methylprednisolone, plasmapheresis, and rituximab. She has residual moderate/severe ataxia, but is now conversant, without tracheostomy, and ambulates with assistance.

Conclusions

There is no definite current evidence for the pathogenicity of the ITPR1 antibody. Given the rarity of cerebellar ataxia in anti-NMDA encephalitis in adults, one could argue for a pathogenic role of ITPR1 in our case. No underlying malignancy was identified in our patient. We will continue surveillance since the clinical syndrome may precede tumor identification by several years.

Disclosure: Dr. Chapman has nothing to disclose. Dr. Jordan has nothing to disclose. Dr. Broderick has received publishing royalties from a publication relating to health care. Dr. Ferioli has nothing to disclose.

Expanding Frontiers in Autoimmune Encephalitis

Habib Moutran Barroso, Saúl Reyes, Jaime Rodríguez Orozco, Hellen Kreinter Rosembaun, Claudio Alejandro Jiménez Monsalve, Juan Esteban Cote, Jaime Toro

Objective

To characterize a case series of Colombian patients with autoimmune encephalitis (AE).

Background

AE is often an under-recognized entity and antibody testing is not widely available in many developing countries. There is a lack of population-based data on AE in Colombia.

Design/Methods

We made a comprehensive review of the literature on AE in Colombia. Additionally, we contacted researchers in other tertiary care institutions in Bogotá, Colombia to obtain information on additional unpublished cases.

Results

45 individuals were included and antibodies were identified in 73.3% of them. The most prevalent antibody was NMDA followed by LGI-1. Clinical characteristics according to the specific antibody were similar to

Neurology®

First Case Report of AMPA Receptor Encephalitis Presenting With Features of Parkinsonism

Chirag Lalwani, I.M Thushara, Sudheeran Kannothe, et al.

Neurology 2022;99;S44-S45

DOI 10.1212/01.wnl.0000903356.12164.0d

This information is current as of December 5, 2022

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/99/23_Supplement_2/S44.3.full
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): Cerebrospinal Fluid http://n.neurology.org/cgi/collection/cerebrospinal_fluid CT http://n.neurology.org/cgi/collection/ct Low pressure syndrome http://n.neurology.org/cgi/collection/low_pressure_syndrome
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 2022 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

