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 Kannoth, 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 onconeuronal 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.

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