and underwent bilateral optic nerve sheath fenestration. Overall, his encephalopathy resolved but vision remained poor.

Conclusions

This case illustrates an unique manifestation of GFAP astrocytopathy including rapid visual loss, persistent intracranial hypertension and absence of characteristic MRI findings.

Disclosure: Dr. Jean has nothing to disclose. Dr. Canissario has nothing to disclose. Dr. Diep has nothing to disclose. Dr. Williams has nothing to disclose. Dr. Samkoff has nothing to disclose.

Autoimmune Encephalitis With Ganglionic a3-AChR Antibodies

Ghaida Khalaf Zaid, Chetan Saini, Leila Gachechiladzi, Mahmoud Salhab

Objective

To describe a case of autoimmune encephalitis presented as new onset refractory nonconvulsive status epilepticus and autonomic failure caused by alpha-3 ganglionic acetylcholine receptor autoantibodies (a3-AChR Ab).

Background

Autoimmune autonomic ganglionopathy (AAG) is rare acquired dysautonomia in adults due to a3-AChR Ab. A patient typically presents with subacute autonomic dysfunction, and encephalitis has not been reported yet.

Design/Methods

NA.

Results

A 60 years old female presented with progressive worsening of mental status for two weeks. Vital signs were normal. The patient was somnolent, nonverbal, with tonic non-reactive pupils on the exam. CT head showed multiple subcortical hypodensities. MRI brain demonstrated T2-FLAIR hyperintense lesions in bifrontal lobes sparing the U-fibers, with abnormal leptomeningeal/pial perivenular type enhancement. MRI C/T spine, CT chest/Abd/pelvis were negative. Although serum and CSF testing showed inflammatory markers, meningoencephalitis and autoimmune encephalitis panel were negative, except for the serum Paraneoplastic panel was positive with high titer a3-AChR Abs. EEG showed nonconvulsive status epilepticus, which required management with three antiseizure medications. Initially, the patient had little response to empirical pulse methylprednisolone therapy. Later showed good clinical response with plasmapheresis. Later on she presented with nonobstructive small bowel obstruction, resolved with another pulse steroid therapy, discharged on dexamethasone taper. On a three-month follow-up, the patient was in near clinical remission, supported with resolved lesions on a repeat MRI. She never relapsed after two years of follow-up.

Conclusions

Studies have shown that a3-AChR Abs can result in diverse neurological manifestations, mainly dysautonomia. Our patient presented with encephalitis, dysautonomia, and refractory nonconvulsive status epilepticus. Scans suggested diffuse leukoencephalopathy, patient was found to have high 3-ACHR antibody seropositivity. Initially, the patient was steroid-resistant but showed clinical improvement with plasmapheresis. We have described an interesting case of encephalitis likely associated with a3-AChR Ab that has not been previously described. More studies are required to confirm this association.

Disclosure: Dr. Zaid has nothing to disclose. Dr. Saini has nothing to disclose. Dr. Gachechiladzi has nothing to disclose. Dr. Salhab has nothing to disclose.

Autistic Regression Sequelae or Relapse for NMDA Receptor Antibody Encephalitis

Ghaida Khalaf Zaid, Chetan Saini, Khadija Awais, Namrata S Shah

Objective

To highlight a pediatric presentation of autistic regression secondary to relapsing NMDA receptor antibody encephalitis (NMDARAE) post-HSV infection.

Background

It is reported that 30% of patients develop NMDA receptor antibodies (NMDARA) after HSV Encephalitis. Previous studies have demonstrated a significant association between prior HSV-1 infection and NMDARAE, a diagnosis often overlooked due to diverse neurological manifestations.

Design/Methods

NA.

Results

This case highlights a 5-year-old female presenting with fever and refractory status epilepticus, was diagnosed with HSV Encephalitis requiring prolonged hospitalization. At discharge she had insomnia, mutism, dyskinetic movements, atonic seizures, and developmental regression. At 3 months post-discharge EEG exhibited multiple generalized myoclonic, myoclonic tonic, and atonic seizures, MRI brain demonstrated right temporal lobe encephalomalacia with immunologic workup demonstrating positive serum and CSF NMDARA. She underwent treatment with intravenous steroids followed by plasmapheresis and then rituximab with an improved clinical response and seizure control at 6 months. At 9 months, she displayed behavioral changes with diagnosis of ADHD with autistic regression. Workup was positive for NMDARA in the CSF (18 months post-presentation) and EEG showed diffuse epileptiform discharges activated during sleep. She was treated with steroids followed by rituximab. At follow-up, she showed improved social interaction, sleep, and seizure control with persistence of some autistic and ADHD features.

Conclusions

About 90% of patients with NMDARE present with prominent behavioral manifestations, with challenges in differentiating from psychiatric diseases. Relapse is reported in 12-24% of cases and is associated with delayed treatment and the female gender. NMDARAE relapse post-HSV Encephalitis is underreported, especially cases demonstrating autistic regression, as in our case. In conclusion, given overlapping and subtle symptoms, it is crucial to recognize the varying presentations and early diagnosis of NMDARAE relapse for effective treatment and better outcome.

Disclosure: Dr. Zaid has nothing to disclose. Dr. Saini has nothing to disclose. Dr. Awais has nothing to disclose. Dr. Shah has nothing to disclose.

CSF Indices in Autoimmune Encephalitis: Promising Predictors of Treatment Response

Ekaterina Popova, Pranav Nair, Annamma Mathai, Sruthi Sasikumar, Siby Gopinath, Vivek Nambiar, Anand Kumar, Saraf Udit Umesh, Jyothi Leelamaniamma, Sudheeran Kannoth

Objective

To study CSF Indices and their association with prognosis of autoimmune encephalitis.

Background

Till date, there is no published data on use of CSF indices in diagnosing or prognosticating of autoimmune encephalitis.

Design/Methods

This retrospective, single-center (AIMS, Kochi, India) cohort study included 60 adult patients with autoimmune encephalitis who were first admitted from August 2016 till November 2021. We used a univariate binary logistic regression for the entire cohort (n = 60) or the cohort of seronegative cases (n = 54) and a two-tailed Fisher's exact test for a small group of seropositive cases (n = 6). A Chi-square test was performed to describe the results in rare cases if logistic regression failed to work.

Results

In the entire cohort (n = 60) a statistically significant association was found between a good fast treatment response and a total count of cells in the CSF more than 4 cells/mm^3 (OR 4.571, 95% CI 1.31–15.956, p = 0.017), IgG Local Synthesis (OR 7.273, 95% CI 1.562–33.863, p = 0.011), and Integrative Parameter of Local IgG Synthesis proposed by Ziadie M. et al. (OR 5.318, 95% CI 1.271–22.250, p = 0.022). Good fast response was defined as an improvement with single agent from the first line therapy by mRS-9Q of at least 3 points in case of severe disease and at least 2 points in case of moderately severe disease at the time of discharge. Higher Albumin Index values associated with higher odds of having poor GCS Score (OR 1.165, 95% CI 1.011–1.343, p = 0.035). In the cohort of seronegative cases (n = 54) we obtained similar results. In the cohort of seropositive cases (n = 6), none of the patients had a good fast response.

Conclusions

In our research, evidence of Local IgG Synthesis in CNS and CSF total cell count more than 4 cells/mm^3 showed association with a good and fast treatment response in patients with autoimmune encephalitis.

Disclosure: Miss Popova has nothing to disclose. Mr. Nair has nothing to disclose. Dr. Mathai has nothing to disclose. Ms. Sasikumar has nothing to disclose. Siby Gopinath has nothing to disclose. Dr. Nambiar has nothing to disclose. Dr. Kumar has nothing to disclose. Dr. Saraf has nothing to disclose. Mrs. Leelamaniamma has nothing to disclose. The institution of Dr. Kannoth has received research support from Novartiz.

Recurrent Acute Necrotizing Encephalopathy with underlying RANBP2 mutation

Dhanalakshmi Angappan, Christopher Hollen

Objective

N/A.

Background

Acute necrotizing encephalopathy (ANE) is a rapidly progressive encephalopathy that can occur in otherwise healthy children after common viral infections such as influenza and parainfluenza. Most ANE is sporadic and nonrecurrent (isolated ANE). We report a case of recurrent acute necrotizing encephalitis in a boy with an identified RANBP2 mutation, which is known to account for the majority of recurrent ANE cases.

Design/Methods

CASE REPORT Our patient is a 13-year-old boy with no significant medical or developmental history and no family history of neurodevelopmental disorders.He had his first episode at 15 months of age-which manifested as irritability, non-responsiveness and was diagnosed as acute disseminated encephalomyelitis (ADEM) and subsequently had 4 additional episodes of ANE at ages 4,4.5,5, and 10. After his third episode, testing for RANBP2 was performed and found to be positive. His typical presentation includes fever, staring spells, nystagmus and altered sensorium during these episodes typically within 24 hours of febrile-illness. He has had multiple triggering viral infections identified including adenovirus, influenza A and parainfluenza. Ultimately his ANE episodes were managed with iv pulse steroid therapy and IVIG.With treatment he has had a slow but nearcomplete recovery, including radiological resolution. He does have mild cognitive impairment and learning difficulties which have persisted.

Results

N/A.

Conclusions

This patient has had numerous episodes of ANE triggered by infection that have responded well to acute management without prophylactic immunomodulation. This is, to our knowledge, themost non-fatal recurrences that have been reported with this condition. Our case suggests that multiple relapses are possible in patients with ANE and early diagnosis and treatment of the episodic encephalopathy would result in improved outcomes. Our case raises the consideration of prophylactic immunotherapy but also demonstrates a relatively positive outcome with a watchful management approach. Keywords: acute necrotizing encephalopathy.

Disclosure: Dr. Angappan has nothing to disclose. Dr. Hollen has nothing to disclose.

Primary Immune Dysregulation in Subacute Sclerosing Panencephalitis: A Case-Control Study

Vijay Varman, Vinay Suresh, Hardeep Malhotra, Neeraj Kumar, Ravindra Garg

Objective

The primary objective was to study the pattern of immune dysregulation in cases with subacute sclerosing panencephalitis (SSPE). The secondary objective was to assess the correlation between the measured immunological variables and disability/death at 6 months,

Background

SSPE is a chronic progressive neurological condition caused by a defective measles virus. It is postulated that immune-dysregulation might result in persistent infection (immune evasion) as well as initiation of autoimmune phenomenon (via natural killer cells) leading to panencephalitis.

Design/Methods

This was a prospective observational study conducted at a tertiary-case referral-facility from January 2020 to September 2021. Thirty consecutive patients fulfilling the Dyken's criteria for SSPE and 30 age-and-sex-matched healthy controls were enrolled. Immunological profile constituted by lymphocyte subset analysis, immunoglobulin levels and complement levels were done in all cases and controls. Cases were staged as per Jabbour's system; disability was assessed using the modified Rankin Scale (mRS).

Results

Patients with SSPE had a mean age of 14.76 years (± 6.9 years). There were 25 males and 5 females; 6.7% cases belonged to Jabbour's first stage, 40% to second stage and 53.3% to third stage. Levels of absolute lymphocyte count, B-cells, T cells, helper T-cells and cytotoxic T-cells were significantly higher in cases. IgG, IgM and IgE levels were significantly higher while IgD levels were significantly lower in cases. At baseline, 13.3% of cases had a mRS score of 0-2 and 86.7% had a score of 3-6; at 6 months 10% had a mRS score 0-2 (favorable outcome) while 90% had a mRS score 3-6 (poor outcome). No correlation of immunological parameters with outcome was found.

Conclusions

Significant immune dysregulation in terms of lymphocyte subsets and immunoglobulin levels seem to exist in SSPE. These findings may pave way for targeted immunomodulator therapy that can be targeted in a larger cohort of patients.

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