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

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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.

Autistic Regression Sequelae or Relapse for NMDA Receptor Antibody Encephalitis

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