those reported in the literature. Interestingly 1 patient had Zika virus (ZIKV) encephalitis 4 months before developing LGI-1 AE, and another patient developed an immunodeficiency following a NMDA related AE. A total of 5 patients had a negative antibody screen. There was no data on antibodies in 7 patients.

Conclusions

The clinical phenotypes of patients with AE in our case series were similar to those previously published. It is important to highlight a novel infectious trigger (ZIKV) and a postencephalitic immune mediated complication that, to the best of our knowledge, have not been previously reported in the literature. The initial diagnostic approach of AE can be based on clinical findings and conventional tests, especially in countries with limited resources where antibody testing is not accessible to all clinicians.

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Concurrent Autoimmune Encephalitis, Diabetes, and Thyroiditis After a Single Dose of Pembrolizumab

Ali AlMoamen, Maria del Pilar Guillermo Prieto Eibl, Olimpia Carbunar, Luis Tornes, Kamil Detyniecki

Objective

We describe the case of a patient with an extensive autoimmune response after one dose of pembrolizumab, emphasizing the importance of early recognition of the diverse presentation of autoimmune complications from checkpoint inhibitors.

Background

A 55-year-old woman with a myxoid chondrosarcoma of the right hand, previously treated with chemotherapy that received one dose of pembrolizumab, with an excellent tumor response. One month later she developed progressive memory impairment and new onset of severe hyperglycemia (glucose > 700 mg/dL) and profound hypothyroidism (TSH of 90 mcIU/mL), attributed to pembrolizumab. She was treated with hormone replacement for autoimmune diabetes and hypothyroidism. Shortly after, she had her first generalized convulsive seizure. Initial MRI brain was unremarkable. Formal neurological evaluation two weeks later was concerning for fluctuating cognitive impairment and staring spells, for which she was admitted. EEG demonstrated focal status epilepticus of the left posterior quadrant. She required multiple agents to control her refractory seizures. MRI brain showed FLAIR hyperintensities in the bilateral hippocampi and cerebral hemispheres. CSF: WBC 3 (76% lymph), normal protein and glucose, negative infectious workup, 7 CSF specific oligoclonal bands, and positive anti-Hu (1:8 CSF and 1:400 serum) and positive serum anti-GAD-65 antibodies (>1:4800). Anti-TPO antibodies were 103 IU/ml. She received 1 gram of intravenous solumedrol for 5 days. Due to partial response, she received five sessions of plasma exchange. Follow up MRI brain showed worsening FLAIR hyperintensities in the bilateral hippocampi, therefore, she was started on IVIG. Patient's mental status continued to improve, and she was discharged to acute rehabilitation on a slow prednisone taper.

Design/Methods

N/A.

Results

N/A.

Conclusions

Checkpoint inhibitor therapy is associated with a variety of systemic and neurological autoimmune complications. A high level of suspicion is needed for early identification of these syndromes and prompt management. Monitoring of treatment response is crucial as it may require treatment escalation.

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An Atypical Case of GFAP Astrocytopathy

Maxime Junior Jean, Ryan Canissario, Judy Diep, Zoe Williams, Lawrence Samkoff

Objective

To describe a case of anti-GFAP astrocytopathy with atypical features.

Background

Glial fibrillary acidic protein (GFAP) astrocytopathy is a steroid-responsive autoimmune meningoencephalitis that is commonly characterized by preceding viral illness followed by encephalopathy and papillitis without significant effect on visual acuity. We describe an atypical case of GFAP astrocytopathy presenting with profound vision loss and intracranial hypertension.

Design/Methods

Case Report

Results

31 yo male with a history of hypertension developed flu-like symptoms for one week. Subsequently, he experienced blurry vision and presented to the hospital. He was found to be hypertensive with acute kidney injury. He was treated for hypertensive urgency and discharged. However, patient's vision continued to deteriorate and he developed non-threatening visual hallucinations. He was readmitted to the hospital. His eye exam revealed bilateral loss of visual acuity, retinal hemorrhages and severe papilledema. His laboratory work-up was notable for LP findings of lymphocytic pleocytosis, elevated protein and opening pressure of 54. He was started on acetazolamide and transferred to tertiary medical center. There, he developed encephalopathy with psychosis. An extensive infectious/autoimmune/malignancy workup was completed. This included three repeat LPs showing persistent intracranial hypertension, lymphocytic pleocytosis, high protein and oligoclonal bands. He underwent imaging with CT chest/abdomen/pelvis, MRV and MRI head/orbits, which showed no evidence of malignancy, patent vasculature and pachymeningeal enhancement, respectively. His work-up was notable for positive IgG GFAP in CSF leading to diagnosis of GFAP astrocytopathy. He was treated with pulse dose IV steroids followed by slow steroid taper and concurrent PLEX therapy. Due to severity of case, he was later started on cyclophosphamide. For papilledema, he was continued on acetazolamide

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.

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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.

An Atypical Case of GFAP Astrocytopathy

Maxime Junior Jean, Ryan Canissario, Judy Diep, et al.

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