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.

Disclosure: Dr. Moutran Barroso has nothing to disclose. Dr. Reyes has received personal compensation in the range of $0-$499 for serving on a Scientific Advisory or Data Safety Monitoring board for Merck. Dr. Reyes has received personal compensation in the range of $0-$499 for serving on a Scientific Advisory or Data Safety Monitoring board for Roche. Dr. Reyes has received personal compensation in the range of $0-$499 for serving on a Scientific Advisory or Data Safety Monitoring board for BIIB. Dr. Reyes has received personal compensation in the range of $0-$499 for serving on a Speakers Bureau for Merck. Dr. Reyes has received personal compensation in the range of $0-$499 for serving on a Speakers Bureau for Novartis. Dr. Reyes has received personal compensation in the range of $0-$499 for serving on a Speakers Bureau for Biogen. Dr. Rodriguez Orozco has nothing to disclose. Hellen Kreinter Rosenbaun has nothing to disclose. Mr. Jiménez Monsalve has nothing to disclose. Dr. Cote has nothing to disclose. Dr. Toro has received personal compensation in the range of $500-$4,999 for serving as an Editor, Associate Editor, or Editorial Advisory Board Member for Watch Neurology.

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.

Disclosure: Dr. AlMoamen has nothing to disclose. Dr. Prieto Eibl has nothing to disclose. Dr. Carbunar has nothing to disclose. Dr. Tornes has nothing to disclose. Dr. Detyniecki has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Aquestive. Dr. Detyniecki has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Neurelis. Dr. Detyniecki has received personal compensation in the range of $500-$4,999 for serving as a Consultant for UCB. Dr. Detyniecki has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Greenwich.

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

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Neurology 2022;99;S46
DOI 10.1212/01.wnl.0000903372.20783.7d