Sibi Gopinath has nothing to disclose. Dr. Saraf has nothing to disclose. Dr. Mathai has nothing to disclose. Mrs. Antony has nothing to disclose.

Rapidly Progressive Dementia With Recurrent Seizures and Hyponatremia; A Case of LGI1 Limbic Encephalitis

Joshua Luster, Ashley Barasa, William Hoffman

Objective

N/A.

Background

Leucine-Rich Glioma Inactivated Protein-1 (LGI1) autoimmune encephalitis was first described in 2001 as one of the syndromes caused antibodies against the voltage-gated potassium channels (VGKC) until it was discovered in 2010 that antibodies were instead being directed towards the protein LGI1. This often presents in males in their 60's and is often associated with faciobrachial dystonic seizures, which have become path pneumonic for this disease process.

Design/Methods

N/A.

Results

77-year-old female with history of hyponatremia, anxiety, hypertension, and lacunar infarct presented for a concern for seizures. She presented for multiple episodes of reported generalized tonic seizures and was eventually found to have right frontotemporal seizures with impaired awareness. Magnetic Resonance Imaging (MRI) was repeated multiple times but were significantly degraded due to motion artifact and read as limited. Further discussion with husband was concerning for memory loss over the past 4 months, but patients children disputed this with several years of memory loss. After neuropsychological testing which demonstrated significant decline across multiple domains. MRI was revisited which was concerning for bilateral mesial temporal hyperintensities on Fluid-Attenuated Inversion Recovery (FLAIR). Patient underwent lumbar puncture given unremarkable workup thus far. CSF and serum both demonstrated LGI1 autoantibodies for which the patient received a 5 days course of IV methylprednisolone, IV immunoglobulins, and was eventually transitioned to rituximab with complete recovery of long term memory.

Conclusions

This case demonstrates the complexity of evaluating a patient for reported rapidly progressive dementia and some of the pitfalls involved in the workup. This case demonstrates that when the initial workup is unremarkable, the patient should be evaluated for uncommon causes, such as autoimmune encephalitis. We diagnosed an atypical presentation of autoimmune encephalitis and documented the initial treatment and response to both first line and second line treatment with future plans to titrate the anti-epileptic drugs.

Disclosure: Dr. Luster has nothing to disclose. Miss Barasa has nothing to disclose. The institution of Dr. Hoffman has received research support from United States Air Force.

Co-Occurrence of Sj/ITPR1 and NMDA Antibodies: A Case Report

William Chapman, Allison Jordan, Joseph Broderick, Simona Ferioli

Objective

To highlight a case of concurrent anti-SJ/ITPR1 and anti-NMDA encephalitis.

Background

The anti-Sj/inositol 1,4,5-trisphosphate receptor (ITPR1) has been associated with autoimmune cerebellar ataxia and malignancy. Reports of patients with anti-Sj/ITPR1 describe isolated cerebellar ataxia as well as various manifestations throughout the central and peripheral nervous system. Anti-NMDA encephalitis presents with subacute decline, seizures, movement disorder, alterations in behavior and cognition, autonomic dysfunction, and central hypoventilation but is rarely associated with cerebellar ataxia in adults.

Design/Methods

NA.

Results

A 28-year-old female with no relevant medical history presented to an outside hospital with acute onset headache, diplopia, nystagmus, and vertigo. MRI and MRV were unremarkable. CSF analysis showed a lymphocytic pleocytosis. She was empirically treated with acyclovir, although viral serologies were negative. On initial assessment in our clinic, neurologic exam showed square wave jerks, ataxic eye movements, resting tremor, appendicular and gait ataxia. She progressively declined with gait instability, autonomic dysfunction, neuropsychiatric symptoms, and significant weight gain from compulsive hyperphagia. Her course was complicated by respiratory failure and tracheostomy was placed for mechanical ventilation. Malignancy screening with mammogram, CT, and full body PET was negative. Transvaginal ultrasound was nondiagnostic. Serum paraneoplastic autoantibody panel was negative. EEG showed severe generalized slowing. Repeat CSF studies were positive for anti-SJ/ITPR1 and anti-NMDA. She was treated with high-dose IV methylprednisolone, plasmapheresis, and rituximab. She has residual moderate/severe ataxia, but is now conversant, without trach dependence, and ambulates with assistance.

Conclusions

There is no definite current evidence for the pathogenicity of the ITPR1 antibody. Given the rarity of cerebellar ataxia in anti-NMDA encephalitis in adults, one could argue for a pathogenic role of ITPR1 in our case. No underlying malignancy was identified in our patient. We will continue surveillance since the clinical syndrome may precede tumor identification by several years.

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Expanding Frontiers in Autoimmune Encephalitis

Habib Moutran Barroso, Saúl Reyes, Jaime Rodríguez Orozco, Hellen Kreinter Rosembaun, Claudio Alejandro Jiménez Monsalve, Juan Esteban Cote, Jaime Toro

Objective

To characterize a case series of Colombian patients with autoimmune encephalitis (AE).

Background

AE is often an under-recognized entity and antibody testing is not widely available in many developing countries. There is a lack of population-based data on AE in Colombia.

Design/Methods

We made a comprehensive review of the literature on AE in Colombia. Additionally, we contacted researchers in other tertiary care institutions in Bogotá, Colombia to obtain information on additional unpublished cases.

Results

45 individuals were included and antibodies were identified in 73.3% of them. The most prevalent antibody was NMDA followed by LGI-1. Clinical characteristics according to the specific antibody were similar to

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