

refractory status epilepticus (NORSE [n = 23]) or rapidly progressive dementia (n = 14). Fifty-three of 72 patients (74%) with available data had associated malignancy, primarily small cell lung cancer (SCLC). Two P/Q type VGCC-IgG positive patients had isolated Lambert-Eaton Myasthenic Syndrome. Additionally, 27 patients presented with non-specific symptoms or alternate diagnoses (atypical presentation); none had underlying SCLC (13 with malignancy data). Most patients with anti-GABABR encephalitis/seizure presentations had GABABR-IgG detected in CSF (95%, 52/55; of 3 who were negative, 1 had NMDA-IgG and another AMPA-IgG and CRMP5-IgG) and/or GABABR-IgG detected by CBA in serum at a dilution of 1:100 (76%, 31/41). Five encephalitis/seizure patients with GABABR-IgG detected in CSF were CBA positive only at 1:10 dilution in serum. Forty-eight of 76 patients had samples positive on tissue IFA (serum = 17/42, CSF = 40/49). Among 27 patients with atypical presentations, most did not have GABABR-IgG CSF CBA positivity (91%, 21/23) and/or GABABR-IgG 1:100 serum titer by CBA (0/17). None of 27 cases were tissue IFA positive (serum = 0/27, CSF = 0/21).

Conclusions

GABABR-IgG is associated with autoimmune encephalitis/seizure presentations. Our study highlights the importance of evaluating CSF and testing serum at both 1:10 and 1:100 dilutions, as seropositivity at only the 1:10 dilution by CBA without CSF GABABR-IgG positivity has poor clinical specificity.

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A Case of Paraneoplastic NMO With Sarcoma and Unusual Cauda Equina Enhancement

Mohammed Hussein, Danny Samkutty, Sarah Durica, Nidhiben Anadani

Objective

To describe a case of sarcoma associated with NMO presenting with myeloradiculitis.

Background

Neuromyelitis optica is a demyelinating disease of the CNS that predominantly affects the spinal cord and optic nerves. Reports of NMO occurring in the setting of cancer suggest that aquaporin-4 autoimmunity may have a paraneoplastic basis. Here, we describe a patient with metastatic sarcoma who tested positive for aquaporin-4 IgG consistent with NMO.

Design/Methods

66-year-old man with left thigh soft tissue sarcoma with metastasis to the lungs admitted for 2-day history of urinary retention. Neurological exam showed bilateral lower extremity weakness, with decreased patellar reflexes. MRI spine showed cord swelling with enhancement from C6 downwards into the thoracic spine with associated syringomyelia and diffuse enhancement of the cauda equina. MRI brain was normal without abnormal enhancement in the optic nerves. CSF analysis demonstrated elevated protein and 0 oligoclonal bands. Was started on methylprednisolone 1 gram daily for 5 days empirically for transverse myelitis. Extensive serum and CSF workup for other causes of myelopathy including rheumatologic, infectious, and nutritional etiologies were unremarkable until NMO IgG antibody came back positive at 1:10000 titer. He had minimal clinical improvement after methylprednisolone course and so was started on plasmapheresis for five days. His weakness improved slightly but continued to have urinary retention. Discharged to a long-term acute care facility on prednisone 60 mg daily with intermittent bladder catheterization and plan for follow-up with Neuroimmunology.

Results

NA.

Conclusions

Myeloradicitis has been reported with anti-MOG disease but is not a typical finding for NMO. We report this case to highlight this unusual finding. In addition, sarcoma is an uncommon cause of paraneoplastic syndromes and to our knowledge, this is the first report of sarcoma being associated with paraneoplastic NMOSD.

Disclosure: Dr. Hussein has nothing to disclose. Dr. Samkutty has nothing to disclose. Dr. Durica has nothing to disclose. Dr. Anadani has nothing to disclose.

Missed Opportunities to Prevent N-methyl-D-Aspartate Receptor (NMDAR) Encephalitis in a DREAMer

Paul Crane, Matthew Jensen, Suzanne Liu, Justin Abbatemarco, Jana Wold, Holly Leydard, Umang Swami, Michelle Miranda, Stacey Clardy

Objective

Describe a case of NMDAR encephalitis in a young Latino male patient, additionally the factors resulting in delayed preventative and diagnostic medical care, which contributed to the development of a preventable case of NMDAR encephalitis.

Background

Adolescent undocumented immigrants in the United States face a history of prejudice and bias that perpetuates disparities and stigmas related to their healthcare. The lack of culturally informed practices among healthcare workers can create multiple lost opportunities to deliver standard of care practices, including routine testicular exams. The treatment of NMDAR encephalitis with immunotherapy, and resection of culpable tumors when present, can be lifesaving. Recognition of the germ cell tumor association has also renewed awareness of the importance of screening for such tumors.

Design/Methods

N/A.

Results

Case: A 25-year-old male who immigrated from Mexico to the U.S. at age 13 presented to the hospital for concern of status epilepticus. His past medical history included atypical developmental delay beginning in late teenage years. A large abdominal mass was identified on imaging as a stage IIIC (pT1bN0M1bS2) NSGCT (70% teratoma/30% seminoma) tumor arising from an unresected, undescended left testicle. Autonomic instability in the setting of this malignancy prompted an evaluation for, and diagnosis of, NMDAR encephalitis. His course was complicated by altered mental status, seizures, sympathetic storming, and orofacial dystonia. After tumor resection, and initiation of immune therapy, the patient showed a remarkable recovery.

Conclusions

This patient's preventive healthcare was impacted at multiple timepoints by changing political policies and a lack of culturally informed practices that unpredictably disrupted reliable access to medical care. Recognition of care gaps allows us to expand our differential diagnoses, and enact a comprehensive approach to fill in gaps. Effective communication, incorporating focused discussions within culturally sensitive frameworks, requires ongoing education for clinicians regarding the populations they serve to prevent disease and minimize health care disparities.

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Neuronal Uptake of Paraneoplastic and Other IgGs is Mediated by the Fc Portion of the IgG Molecule and Involves Previously Uncharacterized Neuronal FcγRI Receptors: Implications for Antibody-Mediated Neuronal Injury

Tammy Smith, Suzanne Liu, Noel Carlson, Stacey Clardy, John Greenlee

Objective

To investigate the mechanisms by which neurons take up paraneoplastic and other antibodies.

Background

Our laboratory has previously demonstrated that neurons can take up both normal and paraneoplastic IgGs and that paraneoplastic autoantibodies such as anti-Yo and anti-Hu can bind to their intracellular target antigens to produce neuronal death. In this study we investigated how neuronal antibody uptake occurs.

Design/Methods

We first compared neuronal uptake of normal and paraneoplastic Fab fragments with that of normal IgG Fc fragments or whole paraneoplastic IgGs. To determine whether neurons expressed receptors capable of binding the Fc portion of the IgG molecule, paraformaldehyde-fixed mouse and rat brains sections were probed with antibodies for the three major types of Fc receptors: FcγRI (CD64), FcγRII, (CD32) and FcγRIII (CD16). Neuronal uptake of antineuronal IgGs was compared between wild type mice and knockout mice lacking the FcγRI receptor. We also investigated whether neuronal IgG uptake could be blocked by normal IgG.

Results

Neurons incorporated the Fc fragment of normal IgG but not the Fab fragment. Intact paraneoplastic IgGs were taken up by neurons, but immunospecific Fab fragments were excluded. Neurons throughout cerebrum, cerebellum, and brainstem showed immunolabeling for FcγRI, but only rare neurons expressed FcγRII or FcγRIII. Uptake of paraneoplastic IgG and neuronal death were not observed in cultures from FcγRI knockout mice but were extensive in cultures from wild type controls. Paraneoplastic antibody uptake could be inhibited by normal IgG.

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Mohammed Hussein, Danny Samkutty, Sarah Durica, et al.

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