Conclusions

Co-existent CSF anti-NMDAR and anti-GAD65 AE is not well-described in the literature. While this patient's serum GAD65 titer was in a range that correlates with neurologic autoimmunity, it was not as high as is typically reported in AE, and could simply be a marker of her insulin-dependent diabetes. However, it was also definitively positive from the CSF, which usually correlates well with GAD65-associated AE. Clinically, the patient had AE features that can be seen associated with either NMDAR and/or GAD65 antibodies. It is interesting to consider whether the GAD65 positivity is most likely to correlate with the patient's adult-onset diabetes, her AE, or both.

Disclosure: Dr. McEntire has nothing to disclose. Dr. Manzano has nothing to disclose. Dr. Linnoila has received research support from NIH/NINDS. Dr. Linnoila has received personal compensation in the range of $10,000-$49,999 for serving as a expert respondent on autoimmune encephalitis with U.S. government/DHHS/vaccine injury compensation program.

Evidence for and Against Subclinical Disease Activity and Progressive Disease in MOG Antibody Disease and Neuromyelitis Optica Spectrum Disorder

Negar Molazadeh, Angeliki G. Filippatou, Eleni S. Vasileiou, Michael Levy, Elias S. Sotirchos

Objective

To investigate the evidence for and against relapse-independent clinical progression and/or subclinical disease activity in patients with Myelin Oligodendrocyte Glycoprotein Antibody Disease (MOGAD) and Aquaporin-4 IgG Seropositive Neuromyelitis Optica Spectrum Disorder (AQP4-IgG+ NMOSD).

Background

MOGAD and AQP4-IgG+ NMOSD are generally relapsing disorders, without clinical progression or subclinical disease activity outside of relapses. With advances in the diagnosis and treatment of these conditions, prolonged periods of remission without relapses can be achieved, and the question of whether progressive disease courses can occur has re-emerged.

Design/Methods

We conducted a narrative literature review in Ovid MEDLINE and Embase, exploring the evidence for and against relapse-independent progression in MOGAD and AQP4-IgG+ NMOSD. We classified the results in four categories : 1)Clinical observations, 2)MRI findings, 3) Retinal imaging, and 4)Fluid-based biomarkers.

Results

As the optic nerve is the major site of involvement in MOGAD and AQP4-IgG+ NMOSD, much of the data comes from the visual pathway studies. Possible pathophysiologic mechanisms of the OCT abnormalities in the absence of symptomatic optic neuritis are: 1) primary neurodegenerative process in retina, 2) subclinical inflammation of the optic nerve, 3) chiasmal involvement leading to abnormal findings in the unaffected eye, and 4)trans-synaptic retrograde degeneration originating from inflammatory lesions in the posterior visual pathway. Except for chiasmal involvement, these mechanisms are not specific to the visual pathway, and are considered as potential explanations for subclinical disease activity outside of the retina. Outside of the visual pathway, there is a lack of sufficient evidence to support the existence of subclinical disease activity and/or relapse-independent clinical progression in MOGAD and AQP4-IgG+ NMOSD, although recent fluid-based biomarker data (serum NfL and GFAP) support that neuro-axonal and/or astrocytic damage may be ongoing between attacks in these diseases.

Conclusions

This review highlights the many unknowns that remain in our understanding of the pathophysiology and clinical course of MOGAD and AQP4-IgG+ NMOSD.

Disclosure: The institution of Dr. Molazadeh has received research support from Genentech, Inc. Dr. Filippatou has nothing to disclose. Dr. Vasileiou has nothing to disclose. Dr. Levy has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Mitsubishi Pharma. Dr. Levy has received personal compensation in the range of $500-$4,999 for serving as a Consultant for UCB Pharma. Dr. Levy has received personal compensation in the range of $5,000-$9,999 for serving as a Consultant for Sanofi. Dr. Levy has received personal compensation in the range of $5,000-$9,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Alexion. Dr. Levy has received personal compensation in the range of $5,000-$9,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Horizon. Dr. Levy has received personal compensation in the range of $5,000-$9,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Genentech. Dr. Levy has received personal compensation in the range of $500-$4,999 for serving as an Editor, Associate Editor, or Editorial Advisory Board Member for Elsevier. Dr. Levy has received personal compensation in the range of $10,000-$49,999 for serving as an Expert Witness for Various law firms. The institution of Dr. Levy has received research support from National Institutes Health. Dr. Sotirchos has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Viela Bio. Dr. Sotirchos has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Genentech. Dr. Sotirchos has received personal compensation in the range of $5,000-$9,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Alexion. The institution of Dr. Sotirchos has received research support from National Institutes of Health. The institution of Dr. Sotirchos has received research support from National Multiple Sclerosis Society. Dr. Sotirchos has received personal compensation in the range of $500-$4,999 for serving as a Speaker with Biogen. Dr. Sotirchos has received personal compensation in the range of $500-$4,999 for serving as a Speaker with Viela Bio.

Coexistance of Probable Neurosarcoidosis and Generalized Myasthenia Gravis: Case Report

Itziar Abad, Mariona Hervas Pujol, Jordi Estela, Maria Luisa Viguera, Oriol Barrachina-Esteve, Gisela Ribera, Carlos Feijoo, Olalla Vazquez, Carme Lozano, Ana Paula Caresia

Objective

NA.

Background

Sarcoidosis is an immune mediated disorder characterized by the formation of non-necrotizing granulomas in multiple organs. The broad range of manifestations of sarcoidosis in the nervous system includes cranial neuropathy, meningitis or myelopathy. Generalized Myasthenia Gravis is a neuromuscular disorder characterized by the formation of antibodies against the acetylcholine receptor, and typically presents with muscle weakness and fatigue. Association of Myasthenia Gravis and Neurosarcoidosis is infrequent.

Design/Methods

NA.

Results

Case report: A thirty-five year old woman, natural from Senegal was admitted to the hospital with a two month history of left peripheral facial nerve palsy, progressive dysphonia and dysphagia, associated with fever, headache and asthenia. Examination revealed, left facial numbness and weakness (V and VII nerve) and right vocal cord paresis (X nerve). Chest ray showed bilateral hilar lymphadenopathy and cranial MRI revealed enhancement of both Meckel cavum, bilateral trigeminal nerves and canalicular segment of the left facial nerve. CSF examination detected elevated protein and lymphocytic pleocytosis. For further study a PET/TC was done which revealed supradiaphragmatic and

infradiaphragmatic hypermetabolic adenopathies. Biopsy of an adenopathy was performed objecting non-caseating granulomas consistent with the diagnosis of probable neurosarcoidosis. Moreover, the patient explained weakness and muscle fatigue since she was 18 years old, which was observed at proximal limb muscles. Acetylcholine receptor antibodies were detected and electromyographic study showed a decremental response to repeated nerve stimulation.

Conclusions

It is well known that autoimmune disorders may coexist in some patients. Neurosarcoidosis and Myasthenia Gravis are two rare diseases with different pathogenesis. Although their coexistance could be coincidental it may also suggest immunologic mechanisms triggering the occurrence of these diagnoses together.

Disclosure: Miss Abad has nothing to disclose. Mariona Hervas Pujol has received personal compensation in the range of $0-$499 for serving on a Scientific Advisory or Data Safety Monitoring board for Sanofi, Merk, Bayer, Biogen, Almirall. Jordi Estela has nothing to disclose. Miss Viguera has nothing to disclose. Dr. Barrachina-Esteve has nothing to disclose. Ms. Ribera has nothing to disclose. Dr. Feijoo has nothing to disclose. Mrs. Vazquez has nothing to disclose. Miss Lozano has nothing to disclose. Dr. Caresia has nothing to disclose.

Autoimmune Encephalitis Misdiagnosis: A Review of Reported Cases

Maria Daniela Orellana Zambrano, Gregory Day, Elia Sechi, Alfonso Lopez

Objective

To identify autoimmune encephalitis (AE) mimics and clinical features reported in the literature.

Background

Recent evidence suggesting that AE is as frequent as infectious encephalitis has increased awareness and testing for immune-mediated causes of neurological impairment. Consistent with this theme, several publications have focused on patients in whom a diagnosis of AE was initially overlooked. On the contrary, AE remains a rare diagnosis in clinical practice, opening up the possibility for symptoms, signs, and test findings associated with other etiologies to be misattributed to AE.

Design/Methods

Case reports published in PubMed in English language before 04/2022 were reviewed. Cases in whom AE was clearly suspected during the diagnostic work-up or misdiagnosed were included.

Results

A total of forty-five patients with a final diagnosis different from AE were included from 40 reports. Median age was 52 (range 5-86) years; 30/45 (67%) were male. Twenty-eight patients fulfilled the criteria for possible AE (62%), five for definite AE (11%), and twelve neither (27%). Features suggestive of AE were acute/subacute altered mental status (ranging from abnormal behavior to coma), (82%); new-onset refractory status epilepticus, (7%); CSF pleocytosis (42%) or oligoclonal bands (9%), and apparent response to immunotherapy (38%). In 26 cases, imaging corresponded to the anatomical classification of limbic encephalitis, 15 had one or more cortical/subcortical T2-abnormalities, one meningeal involvement, one brainstem involvement, and two had normal MRI. In 12 patients, clinically not relevant neural autoantibodies were detected in serum and/or CSF, including GAD, Anti-Zic4, CASPR2, VGKC, anti-N-type calcium channel antibody, anti-LGI1, and GQ1B. We identified four common AE mimic categories: neoplasms (15 patients), infectious diseases (9 patients), genetic diseases (9 patients), and neurodegenerative diseases (7 patients). Five patients had other etiologies.

Conclusions

Despite well-defined clinical diagnostic criteria, the misdiagnosis of AE encompasses atypical presentation of common disorders and less likely rare diagnoses.

Disclosure: Miss Orellana Zambrano has nothing to disclose. Dr. Day has received personal compensation in the range of $5,000-$9,999 for serving as a Consultant for Parabon Nanolabs. The institution of Dr. Day has received personal compensation in the range of $5,000-$9,999 for serving as a Consultant for Eli Lilly. Dr. Day has received personal compensation in the range of $500-$4,999 for serving as an Editor, Associate Editor, or Editorial Advisory Board Member for DynaMed (EBSCO Health). Dr. Day has received personal compensation in the range of $10,000-$49,999 for serving as an Expert Witness for Barrow Law. Dr. Day has stock in ANI Pharmaceuticals. The institution of Dr. Day has received research support from National Institutes of Health / NIA. The institution of Dr. Day has received research support from Chan Zuckerberg Initiative. The institution of Dr. Day has received research support from Alzheimer's Association. The institution of Dr. Day has received research support from National Institutes of Health / NINDS. The institution of Dr. Day has received research support from Horizon Therapeutics. Dr. Day has received personal compensation in the range of $500-$4,999 for serving as a Presenter at Annual Meeting (CME) with American Academy of Neurology. Dr. Day has received personal compensation in the range of $500-$4,999 for serving as a Content Development (CME) with PeerView, Inc. Dr. Day has received personal compensation in the range of $5,000-$9,999 for serving as a Content Development (CME) with Continuing Education, Inc. Dr. Day has a non-compensated relationship as a Clinical Director with AntiNMDA Receptor Encephalitis Foundation that is relevant to AAN interests or activities. Dr. Sechi has nothing to disclose. Dr. Lopez has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Horizon. Dr. Lopez has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Genentech.

The Complicated Course of a Patient With Faciobrachial Dystonic Seizures Associated With LGI1-Antibody Limbic Encephalitis

Shirin Sadeghpour, Dilasha Neupane, Mariam Mouti, Jeffrey Clark

Objective

Highlighting diagnostic and treatment challenges of Faciobrachial Dystonic Seizures (FBDS) associated with LGI1-antibody limbic encephalitis (LE)

Background

Anti-LGI1 LE presents with FDBS as its hallmark: brief, recurrent, contractions of facial and upper limb muscles. Patients have associated cognitive decline and psychiatric disturbance. Temporal lobe involvement is often found on MRI.

Design/Methods

NA.

Results

This 85-year-old female presented with a 2-week history of involuntary "twitching" in the face and arms.Family reported that she hadalsobeen uncharacteristically quiet.Neurological exam and a CT headwere normal.rEEG showed no epileptiform activity.One monthafter onset, episodes became longer and more frequent.Observation during outpatient evaluation led to consideration ofFDBS based on semiology. MRI revealedT2/flairhyperintensity in medial temporal lobes consistent with LE.InpatientcEEG was obtained: 26 episodes were marked, with no ictal EEGcorrelate seen.IVIG and methylprednisolone were started.InitialCSF studies were unremarkable andencephalitis/meningitis panelwas negative. Autoimmuneand paraneoplastic encephalopathy panel later revealed LGI-1antibodiesin the CSF. Chest and abdominopelvic CT were unrevealing of underlying malignancy. Whilereceiving methylprednisolone and IVIG, she developed impaired orientation, hallucinations, and agitation. A 5-day course was completed but with worsening mentation and limited improvement in FBDS.Thus,a decision was made to initiate PLEX (now 1.5 months fromsymptomonset). FBDS episodes resolved withPLEX and mentation improved, butshe development bleeding and retroperitoneal hematoma requiring transfusion, delaying completion of PLEX. Her course was further

Coexistance of Probable Neurosarcoidosis and Generalized Myasthenia Gravis: Case Report

Itziar Abad, Mariona Hervas Pujol, Jordi Estela, et al.

Neurology 2022;99;S74-S75
DOI 10.1212/01.wnl.0000903600.28886.23