Serology panel revealed pan hypovitaminoses in Vitamins A, B1, B6, B12, and D.

Conclusions

Traditional GCA workup initially resulted inconclusive for the patient, whose condition deteriorated as the patient's altered mental status and dizziness spells continued unremittingly. This case highlights the link between large vessel vasculitis and malabsorption syndromes, with the involvement of the superior mesenteric artery, a medium sized vessel, in GCA previously unrecognized. Furthermore, this case is a superb example of multiple etiologies of treatable causes of reversible dementia.

Disclosure: Miss Shapiro has nothing to disclose. Dr. Renner has received personal compensation for serving as an employee of United States Medical Licensing Examination. Dr. Renner has received personal compensation for serving as an employee of London School of Hygiene and Tropical Medicine. Dr. Renner has received personal compensation for serving as an employee of University of Nagasaki. Miss Farese has nothing to disclose.

Stiff Person Syndrome Misdiagnosis: Clinical and Ancillary Testing Characteristics

Nicholas Chia, Andrew McKeon, Eoin Flanagan, Divyanshu Dubey, Nicholas Zalewski, Sean Pittock, Anastasia Zekeridou

Objective

To assess stiff person syndrome (SPS) misdiagnosis and identify factors differentiating SPS from non-SPS.

Background

SPS is a heterogeneous immune-mediated central hyperexcitability disorder that is challenging to differentiate from alternative diagnoses.

Design/Methods

Patients referred to the Mayo Autoimmune Neurology Clinic for SPS (01-Jul-2016 to 30-Jun-2021) were included. SPS diagnosis was defined as compatible clinical syndrome confirmed by an autoimmune neurologist and either serum positivity for high-titer GAD65-IgG (>20.0 nmol/L), glycine-receptor-IgG or amphiphysin-IgG (seropositive cases), or confirmatory electrodiagnostic studies (seronegative cases). Seven patients were excluded (diagnostic uncertainty). Patients were compared for clinical presentation, examination findings, laboratory and electrodiagnostic testing, and treatment responses.

Results

Of 173 cases, 48 (28%) were diagnosed with SPS and 125 (72%) with non-SPS. Age and sex did not significantly differ in the two groups. Most SPS patients were seropositive (41/48 total: GAD65-IgG 27/41, glycine-receptor-IgG 12/41 and amphiphysin-IgG 2/41). Fibromyalgia/chronic pain syndrome or functional neurological disorder were the most common non-SPS diagnoses (81/125, 65%). True SPS patients more commonly had a history of exaggerated startle (81% vs 56%, $p = 0.02$), unexplained falls (76% vs 46%, $p = 0.001$) and prior autoimmunity (50% vs 27%, $p = 0.005$). On examination, SPS patients more often had hypertonia (60% vs 24%, $p < 0.001$), hyperreflexia (71% vs 43%, $p = 0.001$) and exaggerated lumbar lordosis (67% vs 9%, $p < 0.001$) but less likely had functional signs (6% vs 33%, $p = 0.001$). SPS patients more often had abnormal electrodiagnostic studies (74% vs 17%, $p < 0.001$), and at least moderate symptomatic improvement was more likely with benzodiazepines (51% vs 16%, $p < 0.001$) or immunotherapy (45% vs 13% $p < 0.001$). Seventy-one non-SPS patients received immunotherapy; only 4 had an autoimmune neurological condition.

Conclusions

SPS misdiagnosis is common and most alternative diagnoses were non-neurologic. Misdiagnosis may be reduced by considering clinical and paraclinical factors; improved diagnostic accuracy will reduce exposure to unnecessary treatments and health care costs.

Disclosure: Dr. Chia has nothing to disclose. The institution of Dr. McKeon has received research support from Euroimmun AG. The institution of Dr. McKeon has received research support from National Institutes of Health. Dr. McKeon has received intellectual property interests from a discovery or technology relating to health care. Dr. McKeon has received intellectual property interests from a discovery or technology relating to health care. Dr. McKeon has received publishing royalties from a publication relating to health care. The institution of Dr. Flanagan has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Alexion. Dr. Flanagan 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. Flanagan has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Horizon Therapeutics. Dr. Flanagan has received personal compensation in the range of $500-$4,999 for serving on a Speakers Bureau for Pharmacy times. The institution of Dr. Flanagan has received research support from Viela Bio. Dr. Flanagan has a non-compensated relationship as a Member of medical Advisory Board with The MOG Project that is relevant to AAN interests or activities. Dr. Flanagan has a non-compensated relationship as a Editorial board member with Journal of The Neurologic Sciences that is relevant to AAN interests or activities. Dr. Flanagan has a non-compensated relationship as a Editorial board member with Neuroimmunology Reports that is relevant to AAN interests or activities. The institution of Dr. Dubey has received personal compensation in the range of $500-$4,999 for serving as a Consultant for UCB. The institution of Dr. Dubey has received personal compensation in the range of $5,000-$9,999 for serving as a Consultant for Astellas. Dr. Dubey has received personal compensation in the range of $0-$499 for serving on a Speakers Bureau for AGRIMS. Dr. Dubey has received personal compensation in the range of $0-$499 for serving on a Speakers Bureau for Advances in Neurology. Dr. Dubey has received personal compensation in the range of $0-$499 for serving on a Speakers Bureau for Moffit Cancer Center. Dr. Dubey has received research support from Department of Defense. Dr. Dubey has received intellectual property interests from a discovery or technology relating to health care. Dr. Dubey has received intellectual property interests from a discovery or technology relating to health care. Dr. Dubey has received intellectual property interests from a discovery or technology relating to health care. Dr. Zalewski has nothing to disclose. Dr. Pittock has received personal compensation in the range of $5,000-$9,999 for serving as a Consultant for Genentech, Inc. Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Sage Therapeutics, Inc. The institution of Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Astellas. Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Prime Therapeutics. Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving as a Consultant for UCB. Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Roche/Genentech. The institution of Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Alexion. The institution of Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving as a Consultant for MedImmune/Viela Bio. Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for UCB, Inc. Dr. Pittock has received personal compensation in the range of $5,000-$9,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Hoffman/LaRoche AG. Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Genetech. Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for F. Hofman/LaRoche. The institution of Dr. Pittock 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. Pittock has received research support from Grifols. The institution of Dr. Pittock has received research support from NIH. The institution of Dr. Pittock has received research support from Viela Bio/MedImmune/Horizon. The institution of Dr. Pittock has received research support from Alexion Pharmaceuticals. The institution of Dr. Pittock has received research support from F. Hoffman/LaRoche/Genentech. Dr. Pittock has received intellectual property interests from a discovery or technology relating to health care. Dr. Pittock has received intellectual property interests from a discovery or technology relating to health care. The institution of Dr. Zekeridou has received research support from Roche/Genentech. Dr.

Zekeridou has received intellectual property interests from a discovery or technology relating to health care. Dr. Zekeridou has received intellectual property interests from a discovery or technology relating to health care.

Characterization of Cardiac Bradyarrhythmia Associated With LGI1-IgG Autoimmune Encephalitis

Hannah Zhao-Fleming, Anza Zahid, Tong Lu, Xiaojing Sun, Sean Pittock, Hon-Chi Lee, Divyanshu Dubey

Objective

To evaluate and characterize cardiac arrhythmias associated with LGI1-IgG (Leucine-rich glioma inactivated 1–IgG) autoimmune encephalitis (AE).

Background

AE is increasingly identified as a potentially treatable cause of encephalitis. LGI1-IgG is one of the most common pathogenic neural specific autoantibodies associated with AE in adults. Prior cases of bradyarrhythmias and sudden death have been reported in LGI1-IgG AE, however, larger cohort studies are lacking.

Design/Methods

In this retrospective descriptive study, we identified Mayo Clinic patients (May 1, 2008–December 31, 2020) with LGI1-IgG AE who had electrocardiogram proven bradyarrhythmias during the initial presentation. Inclusion criteria were 1) LGI1-IgG positivity with a consistent clinical syndrome; 2) electrocardiographic evidence of bradyarrhythmia; and 3) sufficient clinical details. We excluded patients with alternate reason for bradyarrhythmias. We collected demographic/clinical data including details of bradyarrhythmia (severity, duration, treatments), and neurologic and cardiac outcomes.

Results

We found that patients with LGI1-IgG AE had bradyarrhythmia at a frequency of 8% during the initial presentation. The bradyarrhythmia was often asymptomatic (6/11, 55%); however, the episode was severe with one patient requiring a pacemaker. Outcome was also generally favorable with the majority (8/11, 73%) having full resolution without further cardiac intervention. Lastly, we found that mouse and human cardiac tissues express LGI1 (mRNA and protein), suggesting that LGI1-IgG may influence cardiac tissue itself.

Conclusions

LGI1-IgG AE can be rarely associated with bradyarrhythmias. Although the disease course is mostly favorable, some cases may require pacemaker placement to avoid devastating outcomes.

Disclosure: Dr. Zhao-Fleming has nothing to disclose. Dr. Zahid has nothing to disclose. Dr. Lu has nothing to disclose. Mrs. Sun has nothing to disclose. Dr. Pittock has received personal compensation in the range of $5,000-$9,999 for serving as a Consultant for Genentech, Inc. Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Sage Therapeutics, Inc. The institution of Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Astellas. Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Prime Therapeutics. Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving as a Consultant for UCB. Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Roche/Genentech. The institution of Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Alexion. The institution of Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving as a Consultant for MedImmune/Viela Bio. Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for UCB, Inc. Dr. Pittock has received personal compensation in the range of $5,000-$9,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Hoffman/LaRoche AG. Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Genetech. Dr. Pittock has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for F. Hofman/LaRoche. The institution of Dr. Pittock 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. Pittock has received research support from Grifols. The institution of Dr. Pittock has received research support from NIH. The institution of Dr. Pittock has received research support from Viela Bio/MedImmune/Horizon. The institution of Dr. Pittock has received research support from Alexion Pharmaceuticals. The institution of Dr. Pittock has received research support from F. Hoffman/LaRoche/Genentech. Dr. Pittock has received intellectual property interests from a discovery or technology relating to health care. Dr. Pittock has received intellectual property interests from a discovery or technology relating to health care. Prof. Lee has nothing to disclose. The institution of Dr. Dubey has received personal compensation in the range of $500-$4,999 for serving as a Consultant for UCB. The institution of Dr. Dubey has received personal compensation in the range of $5,000-$9,999 for serving as a Consultant for Astellas. Dr. Dubey has received personal compensation in the range of $0-$499 for serving on a Speakers Bureau for AGRIMS. Dr. Dubey has received personal compensation in the range of $0-$499 for serving on a Speakers Bureau for Advances in Neurology. Dr. Dubey has received personal compensation in the range of $0-$499 for serving on a Speakers Bureau for Moffit Cancer Center. Dr. Dubey has received research support from Department of Defense. Dr. Dubey has received intellectual property interests from a discovery or technology relating to health care. Dr. Dubey has received intellectual property interests from a discovery or technology relating to health care. Dr. Dubey has received intellectual property interests from a discovery or technology relating to health care.

Are Insulin Mimetics Protective Against Comorbidity in Patients With Neuro-Autoimmune Disease?

Mohsen Ahmed, Afaaq Ahmed, Ronak Trivedi, Muhammed Ors, Nabeel Ahmed, Mustafa Jaffry, Nizar Souayah

Objective

To investigate the neuroprotective potential of insulin mimetics (IM) in patients with neuro autoimmune disease (NAD) and high risk comorbidities.

Background

IM are used to treat patients with diabetes mellitus (DM) and have been shown to protect against progressive neurological damage. Despite their neuroprotective benefits, the extent of their neuroprotection in patients with NAD has not been completely characterized.

Design/Methods

A retrospective analysis on 34464 patients hospitalized at a tertiary care center in a major metropolitan area was conducted. 168 patients were taking IM medications. 7848 patients were taking medicine other than IM for their DM (nIM). 7690 patients were taking insulin without IM medications. 26448 patients were not on any DM medication (nDM). The outcomes compared included the prevalence of NAD, diabetic neuropathy and/or retinopathy (DMNR), and the prevalence of high risk comorbidities defined as those with either heart failure, chronic kidney disease, stroke, or encephalopathy.

Results

The prevalence of NAD was 0.6%, 0.52%, 0.53%, 0.56% among patients in the IM, nIM, insulin, and nDM groups respectively ($p > 0.05$). 19.5% of NAD and 23.6% of those without NAD had high risk comorbidities ($p > 0.05$). Among those with autoimmune disease, 31% of those taking any diabetic medication and 16% of nDM had high risk comorbidities ($p > 0.05$). The prevalence of DMNR was 10% and 8% in IM and nIM groups respectively ($p > 0.05$).

Conclusions

These results suggest that IM medications may benefit patients with NAD against additional comorbidity as those without NAD and DM are expected

Stiff Person Syndrome Misdiagnosis: Clinical and Ancillary Testing Characteristics

Nicholas Chia, Andrew McKeon, Eoin Flanagan, et al.

Neurology 2022;99;S12-S13
DOI 10.1212/01.wnl.0000903120.29940.d6