has received personal compensation in the range of $500-$4,999 for serving as a Consultant for TG Therapeutics. The institution of Dr. Paz Soldan has received research support from National Institutes of Health. The institution of Dr. Paz Soldan has received research support from Western Institute for Biomedical Research. The institution of Dr. Paz Soldan has received research support from Biogen. The institution of Dr. Paz Soldan has received research support from Novartis. The institution of Dr. Paz Soldan has received research support from Clene Nanomedicine. Dr. Clardy has received personal compensation for serving as an employee of Veterans Health Administration (VHA). Dr. Clardy has received personal compensation for serving as an employee of University of Utah Health. Dr. Clardy has received personal compensation in the range of $0-$499 for serving as a Consultant for Clarion. Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a Consultant for ExpertConnect. The institution of Dr. Clardy has received personal compensation in the range of $0-$499 for serving as a Consultant for VidaBio. The institution of Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Genentech. The institution of Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Alexion. The institution of Dr. Clardy has received personal compensation in the range of $0-$499 for serving as a Consultant for GuidePoint. Dr. Clardy has received personal compensation in the range of $10,000-$49,999 for serving as an Editor, Associate Editor, or Editorial Advisory Board Member for Neurology/AAN Publications. The institution of Dr. Clardy has received research support from Alexion Pharma. The institution of Dr. Clardy has received research support from Sumaira Foundation for NMO. The institution of Dr. Clardy has received research support from Immune Deficiency Foundation. The institution of Dr. Clardy has received research support from National Institute of Health, National Institute of Neurological Disorders and Stroke (NIH/NINDS). Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a Grand Rounds Travel and Lodging with AAN. Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a Speaker Honoraria for Grand Rounds Travel with U of Iowa. Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Biogen. Dr. Clardy has received research support from National Multiple Sclerosis Society. The institution of Dr. Rose has received research support from Guthy Jackson Charitable Foundation. The institution of Dr. Rose has received research support from NIH. The institution of Dr. Rose has received research support from NIH. The institution of Dr. Rose has received research support for REVHEALTH LLC.

Given her EMG and CSF findings, she began treatment for suspected CPI-induced atypical GBS and myositis. Concomitantly she was found to have B12 and folate deficiencies, then gradually improved to baseline with vitamin repletion, steroids, and plasma exchange. 2. A 27-year-old woman with metastatic melanoma and recent treatment with ipilimumab+nivolumab developed autoimmune hepatitis and intractable vomiting. Three weeks after she began dabrafenib and trametinib, she developed confusion, diplopia, and ataxia along with weakness and areflexia. She was treated for possible GBS, but was concurrently found to have thiamine deficiency with sequela of Wer­nicke’s encephalopathy on MRI Brain. Her confusion improved with thiamine supplementation but had persistent weakness. 3. A 57-year-old woman with lung adenocarcinoma who had progressed on durvalumab began pembrolizumab. Two weeks later, she developed fevers, rash, and lethargy. She was treated supportively but continued to worsen until neurological workup revealed limbic hyperintensities on MRI Brain and CSF pleocytosis with +HSV1. She had minimal clinical improvement with acyclovir but remained cognitively debilitated.

Conclusions

Given frequently complex clinical circumstances when working up n-irAEs, a systematic approach and a broad differential must be utilized for this important intersection of cancer neurology and immunology.

Disclosure: Dr. Gregory has nothing to disclose. Dr. Tummala has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Genentech. Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a Consultant for REVHEALTH LLC.

Giant Cell Arteritis of the Superior Mesenteric Artery Presenting With Wernicke Encephalopathy From Thiamine Deficiency

Sarah Shapiro, David Renner, Ludovica Farese

Objective

N/A.

Background

Giant cell arteritis (GCA) is one of the most common systemic vasculitides in adults over the age of 50 with incidence ranging from 15 to 35 per 100,000 individuals. The disorder is often included in the differential diagnosis of maladies producing atypical facial pain, headache, visual loss, amaurosis fugax, jaw pain, elevated inflammatory markers, and anemia. GCA is typically known to affect cranial arteries with physical exam findings of tenderness to palpation of the temporal arteries and cranial neuropathies. Clinical diagnosis is further supported by new headache, temporal artery abnormality, elevated ESR (= 50 mm/h), and abnormal artery biopsy.

Design/Methods

N/A.

Results

A 68-year-old female with history of primary generalized seizures presented to clinic with a 6-week history of paroxysms of acute confusional episodes, the inability to arise from a seated position due to lower extremity weakness bilaterally, alterations of consciousness without loss of consciousness, severe anorexia, and weight loss. MRI with contrast including Axial FLAIR/T2/Diffusion revealed bilateral pan-lobar cortical and subcortical atrophy with ex-vacuo ventriculomegaly and mild leukoaraiosis in the subcortical white matter tracts. PET-CT body revealed linear uptake involving the aortic root, extending into subclavian arteries bilaterally with segmental involvement of proximal common carotids, and extending inferiorly to the level of the common iliac arteries and the mesenteric arteries. Temporal artery biopsy revealed presence of granulomas with multinucleated giant cells.
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 amphiophysin-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 electrodagnostic 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 amphiophysin-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 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 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 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 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 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 Scientific Advisory or Data Safety Monitoring board for Horizon Therapeutics.
Giant Cell Arteritis of the Superior Mesenteric Artery Presenting With Wernicke Encephalopathy From Thiamine Deficiency
Sarah Shapiro, David Renner and Ludovica Farese
Neurology 2022;99;S11-S12
DOI 10.1212/01.wnl.0000903116.61112.46

This information is current as of December 5, 2022

Updated Information & Services
including high resolution figures, can be found at:
http://n.neurology.org/content/99/23_Supplement_2/S11.2.full

Subspecialty Collections
This article, along with others on similar topics, appears in the following collection(s):
Cerebrospinal Fluid
http://n.neurology.org/cgi/collection/cerebrospinal_fluid
CT
http://n.neurology.org/cgi/collection/ct
Low pressure syndrome
http://n.neurology.org/cgi/collection/low_pressure Syndrome

Permissions & Licensing
Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:
http://www.neurology.org/about/about_the_journal#permissions

Reprints
Information about ordering reprints can be found online:
http://n.neurology.org/subscribers/advertise