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Differential Diagnosis in the Management of CPI Immunotoxicity: Case Series of Etiologies not to Miss

Timothy Gregory, Sudhakar Tummala

Objective

To present on treatable conditions arising with exposure to checkpoint inhibiting immunotherapy for malignancy. Each case was diagnostically obscured by presumed immunotoxicity.

Background

Neurological immune-related adverse events (n-irAEs) are rising in incidence with adoption of checkpoint inhibitors (CPIs) for many cancers. 1-3% of patients treated with CPIs experience severe n-irAEs with potential for persistent functional disability or mortality. Diagnosis can be challenging for immunologically vulnerable patients with frequently multifactorial problems from their cancer and potential infectious, metabolic, and iatrogenic complications.

Design/Methods

Three informative cases from a single institution were analyzed.

Results

1. An 80-year old woman with metastatic melanoma and recent treatment with ipilimumab+pembrolizumab developed acute leg weakness. 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 minor 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.

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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 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.

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