Results

A 62-year-old man with poorly differentiated papillary thyroid carcinoma with extensive brain, lung, lymphatic metastases post thyroidectomy, radioiodine therapy, chemotherapy with dabrafenib and trametinib, and stereotactic radiation surgery presented with recurrence of brain metastases one year after the diagnosis of a 2.1 cm metastatic lesion in the left parieto-occipital region causing right homonymous hemianopsia. The metastases initially improved in size and number with radiation and serial brain MRIs were stable. Biopsy of the mets revealed a poorly differentiated carcinoma. He was subsequently treated with pembrolizumab for six months. Two months after treatment initiation, he reported episodic behavioral arrest, confusion, and expressive aphasia concerning for seizures. Continuous EEG monitoring revealed a left-sided focus without seizures, and the episodes persisted despite levetiracetam and clobazam. Four lumbar punctures revealed lymphocytic pleocytosis (10-14 cells), elevated protein (48–68 mg/dl), negative cytology, flow cytometry and viral studies including JC virus. Paraneoplastic panels in serum and cerebrospinal fluid were negative. Repeat MRIs findings were most consistent with radionecrosis and noted improvement of the metastatic lesions. Suspicion was raised for pembrolizumab-induced encephalitis and he received high-dose steroids with minimal response however, clinical improvement noted with reduced episode frequency after intravenous immunoglobulin induction therapy and rituximab maintenance therapy.

Conclusions

PD-1 ICI-related encephalitis is a diagnosis of exclusion that should be considered in patients with encephalopathy or other neurological deficits following 3 months of treatment initiation and response to immunosuppressive therapy. Higher incidences are reported in males. Early recognition is crucial to prevent long-term neurologic damage. Outcomes are depend on patient characteristics and clinical presentation.

Disclosure: Dr. Zahid has nothing to disclose. Dr. Poursheykhi has nothing to disclose. Dr. Saeed has nothing to disclose. Dr. Tremont has nothing to disclose.

Neuroblastoma Presentation With Multiple Cranial Nerve Involvement

Aysha Arshad, Janetta Arellano, Anastasia Chumakova, Sharief Taraman

Objective

NA.

Background

We report a case of neuroblastoma, a pediatric neuroendocrine tumor of the sympathetic nervous system, in a 3-year-old female with multiple cranial nerve involvement.

Design/Methods

A 3-year-old afebrile, lethargic female presented with bilateral eyelid droop, right head tilt, slurred speech, gagging, abnormal walking and no bowel movement. Neurological examination noted bilateral ptosis, dysarthria, left tongue deviation, proximal weakness in upper and lower extremities, areflexia in biceps and patellar tendons, dysmetria, and wide-based gait. MRI of the brain showed heterogeneous appearance of the clivus and MRI of the spine showed right adrenal mass and heterogeneous enhancement of multiple vertebrae, suggesting possible metastatic disease. Serum and cerebrospinal studies were unremarkable. Patient was treated with intravenous methylprednisolone and plasmapheresis for suspected paraneoplastic syndrome; however, she continued to clinically progress. Adrenal mass biopsy results and elevated urine VMA and HVA levels were consistent with the diagnosis of neuroblastoma. Nuclear imaging and meta-iodobenzylguanidine scan were negative. Paraneoplastic panels and Lambert Eaton panel were negative for autoantibodies. Total resection of the abdominal mass and right adrenal gland with continued steroid taper, resulted in reported near total symptom resolution.

Results

Our patient presented with antibody negative paraneoplastic polyneuropathy due to neuroblastoma. Cranial nerve involvement in neuroblastoma can result from tumor involvement of the sympathetic chain, paraneoplastic syndromes, or metastasis to the skull. Our patient's initial imaging suggested potential metastatic or inflammatory involvement of the clivus, suspected of causing her cranial nerve symptoms. However, nuclear studies were negative for metastatic disease and patient's symptoms resolved after resection of the main mass suggesting a paraneoplastic etiology. Although her paraneoplastic panel and Lambert-Eaton panel were negative for common associated antibodies, it is likely an unmeasured autoantibody, cytokine, hormones, or peptide was at play.

Conclusions

Neuroblastoma should be considered as a differential for a neurological presentation involving multiple cranial nerves in a child.

Disclosure: Miss Arshad has nothing to disclose. Dr. Arellano has nothing to disclose. Dr. Chumakova has nothing to disclose. Dr. Taraman has received personal compensation for serving as an employee of Cognoa. Dr. Taraman has received personal compensation for serving as an employee of Pediatric Subspecialty Faculty. Dr. Taraman has received personal compensation in the range of $10,000-$49,999 for serving as a Consultant for Cognito Therapeutics. Dr. Taraman has stock in Handzin. Dr. Taraman has stock in Cognoa. The institution of Dr. Taraman has received research support from Innovative Health Solutions. Dr. Taraman has received intellectual property interests from a discovery or technology relating to health care. Dr. Taraman has received intellectual property interests from a discovery or technology relating to health care. Dr. Taraman has received intellectual property interests from a discovery or technology relating to health care.

Co-Occurring NMDA-Receptor and Anti-GAD65 Antibodies in the CSF of a Patient With Encephalitis: Case Report

Caleb McEntire, Giovanna Manzano, Jenny Linnoila

Objective

We describe a 58-year-old woman who presented with rapid cognitive changes and was found to have concurrent CSF NMDA and GAD65 receptor antibodies.

Background

Antibodies against NMDA and GAD65 receptors are associated with highly morbid autoimmune encephalitides. One case of co-occurring NMDA-R and anti-GAD65 antibodies in a patient with progressive cognitive changes and type I diabetes has previously been described. Herein, we describe a case of a previously high-functioning woman who experienced rapidly progressive cognitive changes secondary to autoimmune encephalitis (AE) with co-existent NMDAR and GAD65 antibodies.

Design/Methods

NA.

Results

A 58-year-old woman with adult-onset insulin-dependent diabetes mellitus, hypertension, and prior left-sided Bell's palsy presented to medical care for subacute cognitive decline characterized initially by inattention and difficulty with activities of daily living, progressing to profound global aphasia and seizures. Diagnostic testing revealed GAD65 antibody positivity in serum (33 nmol/L) and CSF (17.3 nmol/L) and negative serum but positive CSF NMDAR antibody. CSF showed pleocytosis (37 nucleated cells, 97% lymphocytes), elevated glucose (109 mg/dL), and normal protein (41 mg/dL). EEG showed right temporal epileptiform discharges. MRI was unrevealing. She was treated with IV steroids, IVIg, and rituximab, and has slowly improved on follow-up.

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

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Neurology 2022;99;S73-S74
DOI 10.1212/01.wnl.0000903592.90530.d1