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.

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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.

Neuroblastoma Presentation With Multiple Cranial Nerve Involvement

Aysha Arshad, Janetta Arellano, Anastasia Chumakova, et al.

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