examination was unremarkable. MRI of the brain revealed multifocal enhancing lesions and edema as well as multifocal signal abnormality throughout the supratentorial and infratentorial parenchyma. Her cerebrospinal fluid showed elevated opening pressure, elevated nucleated cells, elevated protein and normal glucose. CSF cytology, gram stains and cultures were unremarkable. Possibility of sarcoidosis, mycobacterium and fungal etiologies were ruled out. Other work up included but not limited to MRA head, MRV head, diagnostic cerebral angiogram, extensive serum and CSF autoimmune labs were also unremarkable. She underwent a brain biopsy which showed necrotizing granulomatous inflammation with associated dystrophic calcification. Patient was started on high dose methylprednisolone followed by a 6-week prednisone taper. There was a complete resolution of her symptoms, as well as improvement in her follow up MRI brain over a six month period.

Design/Methods
N/A.

Results
N/A.

Conclusions
We describe what we believe would be the third reported case of necrotizing granulomatous meningitis of an unknown cause in humans. This case demonstrates the response of necrotizing granulomatous meningoencephalitis in humans to steroid therapy. Yet, further studies are necessary to determine symptomatology and pathogenesis, as well as the treatment in humans.

Disclosure: Dr. Lateef has nothing to disclose. Dr. Gharaibeh has nothing to disclose. Ms. Zamir has nothing to disclose. Dr. Sheikh has nothing to disclose.

Ischemic Stroke in Neurosarcoidosis: A Retrospective Cohort Analysis

Spencer Hutto, Kevin Kyle, Denis Balaban, Nagagopal Venna

Objective
To provide a detailed analysis of the clinical features and course of ischemic stroke secondary to sarcoidosis of the CNS and to observe the effects of immunosuppression in the prevention of recurrent cerebrovascular disease.

Background
Cerebrovascular disease is rarely reported in neurosarcoidosis and constitutes one of its least well-described forms, though recognition for it has grown in the last decade with recent studies estimating a higher frequency of occurrence than previously known.

Design/Methods
Patients with ischemic stroke were included if the mechanism was directly attributable to sarcoidosis of the CNS. Patients were excluded if an alternative stroke etiology was of equal or higher likelihood than CNS sarcoidosis.

Results
Neurologic disease was the initial presenting manifestation of sarcoidosis in 8/11 (72.7%), and ischemic stroke was an inaugural manifestation of sarcoidosis in 4/11 (36.4%). Small vessel disease was the predominant ischemia subtype (10/11, 90.9%) with pontine perforating vessels (6/11, 54.5%) and lenticulostriate arteries (3/11, 27.3%) being the vessels most often affected. Vessels with a more rostral supratentorial distribution were uncommonly affected. Common neuroinflammatory accompaniments included leptomeningitis (10/11, 90.9%) and cranial nerve disease (4/11, 36.4%). Recurrent strokes occurred in 8/11 (72.7%), and recurrent neuroinflammation occurred in 7/11 (63.6%). Antiplatelet drugs were used in 6/11 (54.5%). Most patients (10/11, 90.9%) required at least two lines of immunosuppression to achieve inflammatory disease remission in this context; infliximab was the most successfully employed immunosuppressant (7/8 treatment courses, 87.5%). The presenting median modified Rankin Scale score of 4.0 improved to 2.0 over a median period of follow-up of 52.0 months.

Conclusions
Ischemic strokes in neurosarcoidosis occur in a caudal-to-rostral distribution, tend to affect small caliber blood vessels that lack collateral blood flow, and typically associate with inflammatory leptomeningeal disease.

Disclosure: Dr. Hutto has nothing to disclose. Dr. Kyle has nothing to disclose. The institution of Dr. Balaban has received research support from Biogen. Dr. Venna has nothing to disclose.

Pachymeningitis in Biopsy-Proven Sarcoidosis: Clinical Course, Radiographic Findings, Response to Treatment, and Long-Term Outcomes

Pressley Chakales, Max Herman, Ling Chen Chien, Spencer Hutto

Objective
To study the clinicoradiographic features of pachymeningeal involvement in neurosarcoidosis and its evolution over time in response to treatment.

Background
Meningeal inflammation is one of the most common forms of neurosarcoidosis, occurring in 16-69% of affected patients. While the clinical and radiographic features of leptomeningitis in neurosarcoidosis are well known, those of pachymeningitis are far less clear.

Design/Methods
Patients with a diagnosis of neurosarcoidosis seen at Emory University [01/2011-8/2021] were included if pachymeningeal involvement was evident by MRI and the patient's sarcoidosis was pathologically confirmed (from a neural or extraneural site).

Results
26/215 (12.1%) patients with neurosarcoidosis qualified for inclusion. Pathological confirmation came from neural tissue in 50%. Median age of onset was 43.5 years; most were male (16/26, 61.5%). Symptoms were primarily related to pachymeningitis in 20/26 (76.9%). Headache (19/26, 73.1%), visual dysfunction (12/26, 46.2%), and seizures (7/26, 26.9%) were the most common symptoms. All patients had cranial pachymeningitis; only a single patient undergoing spinal imaging (1/11, 9.1%) had spinal pachymeningitis. The falx cerebri (16/26, 61.5%) was the most commonly affected dural structure, but the anterior and middle cranial fossae and tentorium were frequently involved (12/26 each, 46.2%). The pachymeningeal lesions were unifocal (11/26, 42.3%) or multifocal (15/26, 57.7%) in distribution, nodular morphologically (23/25, 92.0%), and homogeneously enhancing (24/25, 96.0%). Symptomatic improvement occurred with steroids initially in 22/25 (88.0%). Ultimately, 23/26 (88.5%) required initiation of steroid-sparing immunosuppressants, including 8/26 (30.8%) eventually undergoing TNF inhibition. Pachymeningeal relapses occurred in 7/26 (26.9%). Median clinical follow-up was 48 months. Median mRS at last follow-up improved to 1.0 from 2.0 at presentation.

Conclusions
Sarcoid pachymeningitis often presents with headaches and visual dysfunction, usually affects the falx cerebri and anterior and middle cranial fossae, and tends to require steroid-sparing immunosuppressants. It has the potential to relapse, but the prospect for recovery is excellent.

Disclosure: Dr. Chakales has nothing to disclose. Dr. Herman has nothing to disclose. Dr. Chien has nothing to disclose. Dr. Hutto has nothing to disclose.

Case Review of Alternative Treatment in Patient With Immunotherapy Induced Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP)

Melissa Huberman, Oluwagbemiga Larinde

Objective

To emphasize recognition of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) due to Immune checkpoint inhibitor (ICI) therapy Atezolizumab and efficacy of steroids in treatment.

Background

Atezolizumab is an anti-programmed death-ligand 1 (PD-L1) immunemodulating drug that induces tumor-specific cytotoxic T-cell immunity against the PD-L1 overexpression of many aggressive lung cancers. IVIG was initially thought to have the same benefit in immunotherapy mediated CIDP however recent cases suggest better benefit with plasmapheresis and steroids.

Design/Methods

We report a case of CIDP in a 61-year-old female with high grade neuroendocrine carcinoma of the lung with extensive metastasis. After confirmation of diagnosis, patient began treatment with whole brain radiation therapy and 6 cycles of carboplatin/etoposide followed by atezolizumab maintenance every 21 days for 38 months. Initial treatment reaction included self-resolving loss of balance. Last dose of atezolizumab was given 9 days prior to brain biopsy of left frontal mass. Following biopsy, symptoms of generalized weakness and bilateral lower extremity weakness presented. When symptoms progressed, patient was admitted to hospital, 2-3 weeks after last dose of atezolizumab.

Results

The patient responded to treatment with steroids and cessation of atezolizumab, carboplatin, and etoposide. She recovered to her neurological baseline after a course of IV dexamethasone.

Conclusions

Among ICI therapy, atezolizumab has been less frequently reported with neurologic adverse events than other medications of its class. Increased recognition of the association between ICI and CIDP may improve early detection and improve potentially fatal outcomes. This case and an assessment of the literature indicate ICI induced CIDP may not respond to IVIG and plasmapheresis treatment with consistency. Although IVIG and plasmapheresis maintained efficiency in some reports, adverse outcomes of IVIG treatment may indicate alternative first-line treatment. This case reports successful recovery with the sole use of immunosuppressive therapy which should be considered to optimize future outcomes of ICI associated CIDP.

Disclosure: Miss Huberman has nothing to disclose. Dr. Larinde has nothing to disclose.

Opsoclonus Myoclonus Syndrome and Supraventricular Tachycardia in a Pediatric Patient: A Case Report and Literature Review

Alec Giakas, Sydney Garner, John Korin, Hollie Edwards

Objective

NA.

Background

Opsoclonus myoclonus syndrome (OMS) is a rare movement disorder in children often associated with an underlying neuroblastoma. In other cases, it is believed that the tumor is occult or there is another immune-stimulating precipitating event. Diagnosis can be difficult, requiring 3 of 4 criteria: opsoclonus or ocular flutter, myoclonus or ataxia, behavioral or sleep disturbances, and neuroblastoma. Prompt treatment of OMS is crucial to preventing permanent neurologic sequelae. In order to better characterize the clinical profile of this syndrome and its associated conditions, we present a case report of a 9 month old male with OMAS without an associated neuroblastoma and with new onset supraventricular tachycardia (SVT) and review the associated literature.

Design/Methods

NA.

Results

A 9-month-old male with a past medical history of macrocephaly, hypotonia, and developmental delay presented with abnormal eye and body movements starting one month prior. Due to concern for OMS, the patient underwent an extensive initial workup including CT neck, chest, abdomen, and pelvis and VHA/HMA levels, which were all within normal limits. The patient's spinal fluid revealed no evidence of paraneoplastic, autoimmune, or infectious processes. Per neurology recommendations, the patient was started on IVIG and dexamethasone. After his first IVIG infusion, the patient's abnormal movements worsened, and his heart rate increased into the 300s. The IVIG was discontinued. However, the patient continued to have recurring bouts of SVT, which was eventually controlled with digoxin. After resuming treatment with IVIG and dexamethasone, the patient's opsoclonus myoclonus symptoms began to improve. All additional metabolic labs resulted normal and the patient was discharged.

Conclusions

Because OMS can cause permanent developmental delay, prompt recognition and treatment of this syndrome is necessary. This is a unique case of OMAS without neuroblastoma, associated with recurrent bouts of SVT. Recognizing rare complications of OMS is crucial to improving medical management of its sequelae.

Disclosure: Mr. Giakas has nothing to disclose. Ms. Garner has nothing to disclose. Mr. Korin has nothing to disclose. Dr. Edwards has nothing to disclose.

GABABR IgG Associated Encephalitis: Clinical Presentations and Measures to Improve Diagnostic Assay Specificity

Jennifer McCombe, John Mills, Abhigyan Datta, Mohamed Rezk, Nicholas Chia, Andrew Knight, Anastasia Zekeridou, Eoin Flanagan, Andrew McKeon, Sean Pittock, Divyanshu Dubey

Objective

To review the clinical/oncological presentations of gamma aminobutyric acid-B receptor (GABABR)-IgG and evaluate the clinical specificity of antibody testing methodologies.

Background

GABABR-IgG is an intermediate-risk paraneoplastic autoantibody commonly associated with encephalitis and/or seizures.

Design/Methods

GABABR-IgG positive patients tested at Mayo Clinic Neuroimmunology Laboratory were identified. Available archived sera were retested by cell-based assay (CBA) at 1:10 and 1:100 dilutions.

Results

105 GABABR-IgG seropositive patients with clinical details were identified (females, n = 56; median age 63 [range 8-82]). Most patients had one of three anti-GABABR encephalitis/seizure presentations: focal-onset seizures with altered mental status (n = 39), new-onset

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