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Role of Immunotherapy in Down Syndrome Disintegrative Disorder (DSDD)

Nidhiben Anadani, Deepti Chrusciel

Objective

To describe case series of patients with DSDD, successfully treated with immunotherapy including Intravenous Immunoglobulin (IVIG) at a single academic center.

Background

Down syndrome is the most common chromosomal disorder, and in most cases, is due to trisomy of chromosome 21. DSDD is under-recognized, rapidly progressive neuropsychiatric syndrome with various postulated etiology including psychological stress, primary psychiatric disorder and autoimmunity.

Design/Methods

Case-1: A 20-year-old fun loving female with trisomy-21 and infantile spasms started having complex partial seizures, hallucinations, speech regression, tics, abnormal head movement and obsessive-compulsive behavior. Case-2: A 20-year-old female cheerleader with trisomy-21, started having rapid regression in language, cognition, social skills and agitation over one year. Case-3: A 22-year-old female dancer with trisomy-21, started having subacute onset depression, hallucinations, sleep changes, anorexia and speech regression over one year.

Results

Case-1: MRI brain and cerebrospinal fluid (CSF) studies were normal including negative autoimmune encephalitis panel. Serum thyroglobulin and thyroid peroxidase antibody were high. Prolonged oral steroid therapy helped but caused adverse effects. She was able to return to her premorbid baseline with chronic IVIG therapy every 10 weeks. Case-2: MRI brain and CSF were normal. Serum autoimmune encephalitis panel, thyroglobulin antibody and thyroid peroxidase antibody were negative. Pulse IV steroids improved symptoms, however she regressed after stopping steroids. IVIG every 6 weeks along with electroconvulsive therapy improved neurological symptoms. Case-3: MRI brain and EEG were normal. CSF showed elevated white blood cell count. Serum Thyroid antimicrosomal and thyroglobulin antibody were high. One dose of IVIG caused significant improvement in neurological symptoms for 6 weeks.

Conclusions

DSDD should be considered in patients with down syndrome with rapid regression. It is often associated with positive thyroid peroxidase antibody suggesting immune mediated etiology. Various immunotherapy treatments have been reported in literature including steroid, IVIG, mycophenolate and rituximab with significant improvement in selected patient with autoimmunity.

Disclosure: Dr. Anadani has nothing to disclose. Dr. Chrusciel has nothing to disclose.

EEG Characteristics in Hospitalized Patients With Acute COVID-19 Symptoms

Ganesh Murthy, Daniel Fayard, Ryan Chung, Steve Chung

Objective

Our objective was to evaluate the incidence of seizures, pattern of EEG abnormalities, and localization of abnormal discharges in hospitalized patients with COVID-19.

Background

The COVID-19 epidemic has revealed significant neurological manifestations including de novo seizures in patients who do not have a prior history of epilepsy or clear epilepsy risk factors. Our center is located in Arizona, which in the early part of January 2021 had more cases per capita than any other place in the world.

Design/Methods

We performed a retrospective review to observe the electroencephalogram (EEG) patterns of hospitalized adult patients with COVID-19 between March 2020 and February 2021.

Results

We identified 99 patients who were COVID-19 positive and had EEG testing during the same hospitalization. The most common EEG abnormality was diffuse background slowing, which was seen in 63.6% of patients (n = 63/99), compare to 15.1% of focal background slowing. Epileptiform discharges were seen in 11.1% of patients and seizures were found in 5.1% of patients, as newly diagnosed seizures. When combining all focal abnormalities, the most common location for these abnormalities was in the frontal regions 36.4% (n = 8/22). Even though 21 patients had acute focal neuroradiologic findings, only 5 had correlated EEG abnormalities within the same region. When EEG was obtained with suspected seizures (n = 33), 4 cases (12.1%, n = 4/33) indeed showed ictal pattern compared to 1.6% when seizures was not suspected (p = 0.087).

Conclusions

Abnormal EEG findings are most commonly found in the frontal lobe among hospitalized patients with acute COVID-19 symptoms. De novo seizures may be seen with COVID-19 infection. Suspicion of seizures should be raised in patients with COVID-19 encephalopathy. The utility of an EEG may help allow us better insight into how and where the COVID infection affects our central nervous system.

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Progressive Multifocal Leukoencephalopathy Associated With Sarcoidosis: A Multi-Center Case Series

Caleb R.S. McEntire, MD, Anita Fletcher, MD, Michel Toledano, MD, Samantha Epstein, MD, Sabrina Tan, MD, Yang Mao-Draayer, MD, PhD, Samantha Banks, MD, Allen Aksamit, MD, Jeffrey M. Gelfand, MD, MAS, Kiran Thakur, MD, Irene Cortese, MD, Shamik Bhattacharyya, MD

Objective

We aim to describe the clinical, laboratory, and radiographic features that characterize patients with progressive multifocal leukoencephalopathy (PML) in the context of sarcoidosis (S-PML).

Background

Sarcoidosis has been associated with CD4+, CD8+, and CD19+ lymphopenia, T-cell anergy, and increased infection risk. S-PML has been reported in approximately 60 cases. PML is often mistaken for neurosarcoidosis, leading to harmful administration of high-dose steroids. Preliminary evidence suggests that experimental therapies such as interleukins 2 and 7, checkpoint inhibitors, polyomavirus-specific T-cell therapy, and infliximab may offer promise for treatment. To ensure optimal outcome, it is crucial to identify S-PML accurately and with minimal delay.

Design/Methods

Data and imaging for patients were collected retrospectively from the electronic medical record from Mass General-Brigham network hospitals, National Institutes of Health, Mayo Clinic, Columbia University Irving Medical Center, University of California San Francisco, University of Michigan, and Beth Israel Deaconess Medical Center.

Results

Twenty-five patients with definite S-PML were identified. Median age at diagnosis of sarcoidosis was 54 years, and median time between sarcoidosis and PML diagnosis was 12 months. Sarcoidosis was isolated to lung in 14/25 patients; 10/25 had multisystem involvement; one patient had isolated dermatologic sarcoidosis. Of all patients, 16/25 patients had never received immunosuppressive medications prior to neurological symptom onset. Median serum lymphocyte count at time of PML diagnosis was 430 cells/uL (range: 50-1490). On MRI, 8/25 patients had infratentorial lesions only, while 8/25 had both infratentorial and supratentorial lesions, and 6/25 showed contrast enhancement. Seven/twenty-five patients progressed to death.

Conclusions

This study characterizes the clinical, laboratory, and imaging features of S-PML patients from seven major US medical centers. These data will be used to identify risk factors for development of PML in the context of sarcoidosis and to investigate any biomarkers that might aid in accurate and timely diagnosis.

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"Trigeminal Tract Sign" in Patients With Herpes Zoster Ophthalmicus: A Case Series of a Novel Imaging Finding

Heather Yong, Carla Wallace, Ronak Kapadia

Objective

Herein, we report three patients presenting with zoster ophthalmicus caused by varicella zoster virus (VZV) with unexpected and novel cervico-medullary findings on magnetic resonance imaging (MRI).

Background

Zoster opthalmicus involves reactivation of latent VZV in the ophthalmic division of the trigeminal nerve producing a dermatomal rash. In a minority of patients this leads to ophthalmic findings and damage to surrounding peripheral nerves through perineural/intraneural inflammation. MRI findings in zoster opthalmicus are often nondescript or normal leading to misdiagnosis and delays in treatment.

Design/Methods

This was a case-series describing patients in Calgary, Alberta who presented with zoster opthalmicus, and who were noted incidentally to have similar appearing lesions in the ipsilateral cervico-medullary region on brain MRI. Standard MRI imaging of the brain and selected high-resolution images of the orbits and globes were reviewed by neuroradiology (CW).

Results

Three patients, with varying phenotypes of zoster opthalmicus, are described: a 70-yo male with ipsilateral decreased visual acuity and oculomotor/abducens palsy; a 75-yo female with ipsilateral oculomotor/facial nerve palsies; and a 35-yo immunocompromised female with ipsilateral blepharoconjunctivitis and optic neuritis. In all 3 cases, MRI revealed profound T2 and fluid-attenuated inversion recovery (FLAIR) hyperintensity along the spinotrigeminal tract of the clinically affected side, extending from the posterolateral pons and medulla into the cervical region.

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