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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

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EEG Characteristics in Hospitalized Patients With Acute COVID-19 Symptoms

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