Research of autoimmunity clues in electrical status epilepticus during sleep (ESES)

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INTRODUCTION

Electrical status epilepticus in sleep (ESES) is an epileptic encephalopathy, as defined by the International League Against Epilepsy (ILAE). It is a spectrum disorder consisting of different seizure types, and combinations of cognitive, motor and behavioral disturbances associated with a particular electroencephalogram (EEG) pattern activated during slow sleep. The pathophysiological mechanisms underlying this condition are still not understood. Although the majority of the cases have unknown etiology, it has been sometimes associated with brain pathologies such as migrational disorders, shunted hydrocephalus, polymicrogyria, porencephaly, and thalamic lesions. While seizures can be successfully treated with conventional and newer anticonvulsive drugs, they do not have the same effect on cognitive functions and particular EEG pattern. Currently, immunomodulatory treatments such as corticosteroids or intravenous immunoglobulins (IVIGs) are considered to be more effective than conventional antiepileptic drugs. Therefore, the effectiveness of these drugs in the treatment of ESES suggests a relationship between immune system activation and ESES.

OBJECTIVES

In this study, we aimed to investigate hints of peripheral immune system abnormalities in cases with ESES who did not receive immunological treatment before.

MATERIALS & METHODS

- It is a retrospective study with a multicenter and prospective design,
- 19 patients diagnosed with ESES were included in the study,
- Demographic features: mean age and gender ratio,
- Predominant seizure types: is it generalized or focal?
- EEG findings: those with spike wave index (SWI) ≥85% are typical ESES, those with <85% atypical ESES,
- Neuroimaging findings: magnetic resonance (MR) or computed tomography (CT) images, whichever is available),
- Serum: immunoglobulin (IgA and IgG), albumin and ferritin levels,
- Serum autoantibodies: celiac and antinuclear antibody (ANA)
- Peripheral blood elements: leukocytes and monocytes counts,
- The Wechsler Intelligence Scale for Children-Revised (WISC-R): verbal -general scores were recorded.

RESULTS

19 patients in total

- The mean age of the patients was 9.0±2.2 years (5.4-13.4), 73.7% male and 26.3% female,
- Seizure type; 47.4% generalized, 52.6% focal (typical rolandic seizures in 26.3%),
- According to SWI index: 50% typical, 50% atypical ESES,
- According to neuroimaging findings: those with positive MRI findings, 37.5% symptomatic epilepsy,
- IgA and IgG levels were low in 12.5% of cases, borderline low in 6.3%.
- Albumin and ferritin levels were normal,
- Celiac and antinuclear antibody (ANA) positivity could not be detected,
- The mean leukocytes and monocytes counts were normal,
- The WISC-R test was performed in only 7 of the cases, the mean verbal score was 78.5 and overall score was 83.8.

CONCLUSIONS

However, the qualitative decrease in serum IgA could not be decisive for the differentiation of typical and atypical ESES. In addition, no significant correlation was found in favor of autoimmunity over ANA, celiac autoantibodies, albumin, leukocyte and monocyte counts.

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