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Correspondence

Focal Status Epilepticus With Unilateral Brain Edema: An Expanding Spectrum

Dear Sir,

We read with great interest the study by Zhang and Ma[1] on neuroimaging findings in focal status epilepticus-related unilateral brain edema (FSE-UBE). They have aptly distinguished their cohort from infantile hemiconvulsion-hemiplegia-epilepsy (IHHE) syndrome, citing different age groups, pre-existing epilepsy, developmental delay, and the absence of fever as possible reasons. The International League Against Epilepsy defined IHHE as a specific syndrome with new-onset refractory status epilepticus in patients aged less than two years. They classically have unilateral seizures with fever and unilaterally abnormal neuroimaging followed by transient or permanent hemiparesis.[2]

We would like to share our experience and propose an umbrella diagnosis for "acute encephalopathy with brain swelling" for entities with cytotoxic brain edema, one of them being FSE-UBE.[3] FSE-UBE is not uncommon and we came across several patients with FSE-UBE with pre-existing epilepsy, developmental delay, or cerebral palsy. These children behaved like IHHE, had acute hemiparesis (which later showed gradual improvement) and consequent focal epilepsy (which was drug-resistant in some). We are eager to know the outcome of Zhang and Ma's cohort. This entity is rarely reported from the Western world probably because of better domiciliary and emergency care of at-risk patients with status epilepticus.

Recently, Nukui et al.[4] reported another entity, acute encephalopathy with acute brain swelling with febrile illness. We envisage that all these entities, IHHE, FSE-UBE, and acute encephalopathy with diffuse brain swelling with febrile infection, need to be clubbed together as acute encephalopathy with brain swelling considering the common underlying pathophysiology as cytotoxic brain edema secondary to status epilepticus, febrile illness, ongoing cytokine storm, and so forth. Channelopathies like *CACNA1A*, *SCN1A*, and so forth may also be contributory.

Nukui et al. have precisely highlighted the importance of recognizing this entity. Recognition of such entities will avoid an unyielding long battery of metabolic investigations (as done in Zhang and Ma's cohort), especially in a resource-limited setting. Infectious, immune, and genetic evaluation may be warranted in an appropriate clinical setting.

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