[a]University G. D'annunzio, "SS Annunziata" Hospital, Chieti, Italy
[b]Catholic University of the Sacred Heart, Rome, Italy

Background: Language disturbances can be usually found in various pathological acute pictures involving the dominant frontal and temporal lobes. Prolonged aphasia as the only manifestation of focal status epilepticus is rarely described and only a few cases have been documented. Several EEG patterns have been associated with Aphasic Status Epilepticus (ASE) including Lateralized Periodic Discharges (LPDs). LPDs pattern is usually correlated with structural lesions of cortical or subcortical areas due to some pathological conditions such as acute stroke, brain tumours, infections, traumas and metabolic diseases. The origin of LPDs is a controversial issue and only a few existing neurophysiological hypotheses address causes and circumstances of LPDs onset and if they represent an ictal or inter-ictal pattern.

Methods: We report two cases of ASE associated with LPDs. Aphasic Status Epilepticus was defined according to Rosenbaum's criteria modified by Grimes & Guberman. All these patients underwent a 21 derivations EEG recording according to the 10-20 international system, 3T Magnetic Resonance Imaging (MRI) of the brain and were tested with Aphasia Rapid Test (ART) to better define aphasia's severity. In addition, a review of the past literature was performed by the search terms "Aphasic Status Epilepticus" and "Lateralized Periodic Discharges" on PubMed. A total of 6 articles were available for further analysis.

Results: We stress the electro-clinical correlation between ASE and Lateralized Periodic Discharges. It has been recently reported that the association between LPDs and seizure is more consistent in the presence of particular LPDs features with an increased seizure risk with higher periodic discharges frequency and "Plus modifier" such as superimposed fast activity. In the previous literature, LPDs have been sometimes associated with ASE but they have not always been marked as ictal pattern even though, in some cases, a clear electro-clinical correlation was described with patient's good clinical response to the anti-seizure therapy.

Conclusions: We highlight the importance of considering focal SE in the differential diagnosis of patients presenting aphasia and how LPDs can represent an ictal EEG pattern with regard to ASE.

doi:10.1016/j.yebeh.2019.08.029

Epilepsy & Behavior 101 (2019) 106755

Aphasic Status Epilepticus Revisited

Sonia Jaraba Armas[a,b], Jacint Sala[a], Misericordia Veciana[c], Jordi Pedro[c], Àngels Camins[d], Laura Rodríguez Bel[e], Jaume Mora[f], Cristina Gámez[e], Mercè Falip[a]
[a]Epilepsy Unit, Neurology Department, Hospital Universitari de Bellvitge, L'Hospitalet De Llobregat, Barcelona, Spain
[b]Neurology Department, Hospital De Viladecans, Viladecans, Barcelona, Spain
[c]Neurophysiology Unit, Neurology Department, Hospital Universitari de Bellvitge, L'Hospitalet De Llobregat, Barcelona, Spain
[d]Radiology Department, Institut de Diagnòstic per la Imatge (IDI), Centre Bellvitge, Hospital Universitari de Bellvitge, L'Hospitalet De Llobregat, Barcelona, Spain
[e]PET Unit, Institut de Diagnòstic per la Imatge (IDI), Hospital Universitari de Bellvitge-IDIBELL, L'Hospitalet de Llobregat, Barcelona, Spain
[f]Nuclear Medicine Department, Hospital Universitari de Bellvitge - IDIBELL, L'Hospitalet de Llobregat, Barcelona, Spain

Background: Prolonged aphasic status epilepticus (ASE) in patients without previous seizures and unknown cerebral lesions is rare, and in many occasions an acute stroke is suspected. Some of these patients may be thrombolised and admitted into stroke units. The aim of the study is to describe electroclinical and neuroimaging characteristics, aetiologies and outcome of patients presenting as de novo ASE.

Methods: We designed an unicentric study including consecutive patients presenting to the Neurology Service with new onset status epilepticus of unknown origin (NORSE) between 2011 to 2018. Final diagnosis was obtained after an acute phase complete work-up and considering the follow-up as an outpatient (minimum one year). Patients with ASE (considering aphasia as the main seizure type) were selected. Aetiology and diagnostic procedures included: video-EEG monitoring, serum and CSF biochemistry, serologies and PCR for neurotropic agents, nonspecific immunological analysis and antineuronal antibodies and onconeuronal antibodies. Necropsic studies were performed in some cases. Neuroimaging studies included ictal SPECT, MRI with a protocol for status epilepticus and FDG-PET.

Results: From 35 patients with NORSE, 16 patients (43%) with ASE were selected. 13 (81%) were women, mean age 70.4 (SD14.5), mean age at ASE onset 66 (SD 15.9), 9 (56%) patients had died. TC scan, done in the first 24 hours, was normal in all patients. MRI done during the first week was normal only in 3 patients (17.5%), in 4 (25%) periictal changes were found. First available EEG was normal or showed minor abnormalities (focal slowing or generalized slowing) in 6 (40%), in 5 patients (31%) seizures were recorded and the rest showed a lateralized periodic pattern. SPECT and/or PET were available in 12 patients and showed focal hypermetabolism or hyperperfusion in 8 (66%). Final aetiologies were symptomatic epilepsy (6), toxic/metabolic (2), amyloid angeitis (2), SMART syndrome (1), infectious encephalitis (1), unknown (2), neurodegenerative disorder (1), autoimmune systemic disease (1). Only 4/16 (25%) responded to corticotherapy. No patient with limbic encephalitis debuted with ASE.

Conclusions: Aphasic status epilepticus is a severe entity in which high suspicion is needed. PET or SPECT studies may be specially helpful in diagnosing this entity.

doi:10.1016/j.yebeh.2019.08.030

Epilepsy & Behavior 101 (2019) 106756

The features of status epilepticus in children with progressive myoclonus epilepsy - a single center experience

Ruzica Kravljanac, Biljana Vucetic Tadic
Institute For Mother and Child Healthcare of Serbia, Faculty of Medicine University of Belgrade, Beograd, Serbia

Background: Progressive myoclonus epilepsy (PME) is characterized by various epileptic phenotype since PME has heterogeneous etiology. The main feature of PME is neurological devastating and resistant epilepsy. The aim of the study is evaluation of status epilepticus (SE) in children with progressive myoclonus epilepsy (PME).

Methods: The retrospective study included children with PME and SE with prominent motor symptoms, treated in Institute in period from 1998 to 2018. PME was diagnosed by enzyme, genetic and/or histopathology investigations. SE was defined as clinical seizure duration >30 min, and classified according the new classification (Trinka et al. 2015). Evaluated features were: age, type, duration, SE recurrence and response to the treatment.

Results: During the course of disease, 21 of 46 children with PME (45.65%) experienced total of 50 episodes of SE. The etiology was: neuronal-ceroid lipofuscinosis (7), Gaucher disease 2 (2), Nieman-Pick C (4), mitochondrial disorders (4), Lafora disease (2), Krabbe disease (1), KCNC1 (1). The age was 0.2-18 (mean 8.4) years. Nine patients experienced SE during the first year of disease, and in four cases, SE was the first epileptic event. All episodes had prominent motor symptoms: convulsive (25), myoclonic (13), focal including epilepsia partialis continua (12). Response to the treatment was variable, with common side effects. Most effective drug was midazolam in intravenous infusion with mean dosage of 0.35 mg/kg/h. The artificial ventilation was necessary in 7 episodes, in 4 together with circulatory support. Refractory SE was in 62% episodes, including nine SRSE. Recurrence rate was nearly 50%.

Conclusion: Children with PME frequently experience SE. Episodes are mostly convulsive, refractory to AEDs with high recurrence rate. SE appearance in later phase of disease contribute to prominent drug adverse effects. Managing SE in children with PME is challenge and requires rational approach in order to stop the seizure, and, on the other hand, to prevent side effects and worsening of general and neurological patient`s condition.

doi:10.1016/j.yebeh.2019.08.031

Epilepsy & Behavior 101 (2019) 106757

Refractory and super-refractory Status epilepticus- analysis of etiological factors

Cristina Munteanu, Stanislav Groppa
State University of Medicine and Pharmacy "Nicolae Testemiţanu", Chisinau, Moldova (the Republic of)
Institute of Emergency Medicine, Chisinau, Moldova (the Republic of)

Background: Status epilepticus (SE) can be a life-threatening condition associated with multiple complications, including death, and can progress to refractory and super-refractory SE. Treatment guidelines recently published partially addresses to the treatment of refractory and super-refractory SE.

Materials and Methods: We conducted a retrospective study of patients diagnosed as having SE- for a period of 2 years (01/01/2017-12/31/2018), according to the clinical presentation and EEG findings, also who had appropriate instrumental diagnosis included cerebral neuroimaging with CT and MRI to identify which condition are leading to refractoriness. The following data were analysed: age, sex, SE clinical type (convulsive and non-convulsive (NCSE)), neurological presentation, EEG features, etiology according to instrumental findings, neuroimaging study (brain CT and MRI), and/or blood examinations, and response to staged applied treatment protocol.

Results: Data are presented in raw numbers, data were analysed using Excel, student's t-test, we identified 78 patients, 29 of whom were female and 49 male, with a mean age of 42.2 $\pm$ 18.4 years. Regarding clinical SE type, convulsive SE was observed in 68 patients and non-convulsive in the remaining 10/78. As regards the SE type according to age patients with convulsive SE were older than patients with NCSE. According to EEG – focal SE was in 49 cases and generalised in 29, no difference for gender, for age focal SE more characteristic for older. Lesional SE was in 52 cases, toxic-dysmetabolic in 11 patients, anoxic SE in 3 and AED non- adherence in 12 from 78 patients. The oldest patients were those with lesional SE due to posttaumatic injuries, poststroke, brain metastasis, neurosurgical interventions followed by patients with non-adherence and toxico-dysmetabolic. Also it was observed among young patients the predominance of infectious and autoimmune underlying etiology (autoimmune/ viral encephalitis), and this cases proven to be refractory to first and second- line AEDs treatment (6/10 patients with refractory SE). 22 of patients have complete SE regression after being treated with benzodiazepines, 46 needed administration of second-line drugs like- phenytoin and phenobarbital and 10 patients – 12,8 % required anesthetic drugs to control the epileptic activity (3 patients- 3,8 % developed super-refractory SE (2 case herpetic encephalitis and 1 case anoxic brain injury).

Conclusion: Predictors of refractory status epilepticus were new diagnosis of SE and nonconvulsive SE. The etiology of refractory status epilepticus appears to be similar overall to that of nonrefractory status epilepticus, but more likely associated with encephalitis (viral encephalitis, in particular) and hipoxico-anoxic brain injuries. We also can conclude that good adherence to staged treatment as well as treatment of the underlying etiology is the key to success in controlling refractory seizures.

doi:10.1016/j.yebeh.2019.08.032

Epilepsy & Behavior 101 (2019) 106758

Long term follow-up of recurrent Status Epilepticus and Stroke-Like Episodes in a MELAS family

Lorenzo Muccioli, Francesca Bisulli, Lidia Di Vito, Chiara La Morgia, Laura Licchetta, Lara Alvisi, Valerio Carelli, Marco Zanello, Paolo Tinuper
IRCCS Bologna Institute of Neurological Sciences, Department of Biomedical and Neuromotor Sciences, University Of Bologna, Bologna, Italy

Background: Mitochondrial encephalomyopathy, lactic acidosis and stroke-like episodes (MELAS) is a disorder commonly caused by the A3243G/tRNALeu mutation of mitochondrial DNA (mtDNA). MELAS patients are at high risk of developing status epilepticus (SE) during stroke-like episodes (SLEs). We describe the long-term follow-up of 2 affected members of a MELAS family with recurrent focal SE associated with SLEs. SE treatment is discussed.

Method: A complete clinical work-out including clinical, biochemical, neuroradiological and EEG assessment was performed over the years in both patients.

Results: The mother developed since 33 years of age focal epilepsy (auditory and visual simptoms, hemiclonic seizures), sensorineural deafness, migraine, intestinal dysmotility, severe cognitive impairment, right homonymous hemianopsia and hemiparesis. She presented recurrent parieto-occipital SE associated with SLEs leading to death at 40 years of age.

Her son suffered photosensitive epilepsy since 17 years of age. He presented 7 episodes of occipital SE (elementary visual hallucinations and oculo-clonic seizures) associated with hemianopia, lactic acidosis and parietal-occipital SLE and refractory epilepsia partialis continua (EPC) in one occasion. SE became progressively more difficult to treat and complicated by lactic acidosis and rhabdomyolysis. During one SE propofol was used and the patient suffered multiple organ failure (propofol infusion syndrome-PRIS). Iv high-dosage midazolam was the most effective treatment of SE.

Both patients carried the mtDNA A3243G/tRNALeu mutation with a similar degree of heteroplasmy (80%).

Conclusion: We report the long term follow-up of 2 members belonging to a MELAS family with recurrent SE and SLEs. SE became refractory to treatment in both patients leading to death in the mother. Based on the occurrence of PRIS and evidence of mitochondrial toxicity,