

Conclusion: GRIN2A (16p13.2) codes for a subunit of the NMDA receptor, and is known to be associated with variant phenotypes of focal epilepsy and Landau Kleffner syndrome. Several candidate genes in the interval of 16p11.2 gain (SEZ62, DOC2A, and others) expressed in the developing brain may provide insights into a gene dosage effect resulting in SES.

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Encephalopathy with Super Refractory Status Epilepticus Related to Chemotherapy in a Young Patient with Osteosarcoma

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Background: Neurotoxic side effects (SEs) of chemotherapy occur frequently. Chemotherapeutic agents may cause both peripheral and central neurotoxicity. Incidence of neurologic syndromes with Methotrexate (MTX) covers a range from 2.3% to 15% and are frequently central. Cisplatin (CDDP) mostly induces peripheral neurological damage, albeit in adults there have been several reports on central neurotoxicity, induced seizures have been estimated at 10% and occur from 6h to 3 months after treatment onset. Only very few cases of severe neurologic central dysfunction following chemotherapy have been reported in children.

Methods: We describe a case of a young patient affected by osteosarcoma treated with chemotherapy and complicated by an acute encephalopathy characterized by super refractory epileptic status and altered mental status with aggressive behaviour and hallucinations.

Results: 13-year-old male with primary high-grade osteosarcoma of tibia received MTX and CDDP containing polychemotherapy. He developed fever, confusion, psychomotor agitation and non-convulsive epileptic seizures after the first course of drugs administration (MTX 12 g/sm; CDDP 120 mg/sm). Imaging, lumbar puncture and laboratory values were within normal limits, EEG revealed frontal status epilepticus that persisted despite lorazepam IV, phenytoin IV and oral oxcarbazepine administered at increasing dose; only after high dose of continuous IV midazolam there was a good clinical and electrical improvement; SE recurred on weaning of midazolam. At this point, to switch from IV to oral therapy, high oral lorazepam dose every 4 h/day was started. After a week EEGs were without paroxysmal discharges. His mental status improved after risperidone although it is an off label use. After two months, his osteosarcoma was treated with surgical resection. As well as a very good response was achieved (post-chemotherapy necrosis grade: 99%), he received further courses of low-dose cisplatin (80 mg/sm) and methotrexate (8 and 10 g/sm), with no further seizures. He currently is on antiepileptic and anti-psychiatric therapy.

Conclusions: Health providers should be aware of the potential central neurotoxicity associated with chemotherapy in children, after excluding other causes (metastasis, cerebrovascular accident, venous thrombosis, paraneoplastic syndromes, infective complications). Understanding the mechanism and predictors neurotoxicity is important to improve treatment outcomes in paediatric patients.

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Prolonged repeated episodes of non convulsive status epilepticus with slight cognitive impairment in a 71 yo man

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Background: "Non-convulsive status epilepticus (NCSE) is one of the great diagnostic and therapeutic challenges of modern neurology. Because the clinical features of this disorder may be very discrete and sometimes hard to differentiate from normal behaviour, NCSE is usually overlooked and consequently not treated properly".

Methods and Results : We report the case of a 71 years old health man (only hypertension) that, during last 5 years, presented at least 2-3 episodes/year of slight confusional state. The wife referred that the husband showed events – lasting until 2 days - characterized by mild confusional state. During these events he had difficult to: a) find objects of common daily use, b) maintain goals of ordinary decisions and projects and c) assume usual daily therapy. He was admitted in our Neurology Dept. only when, during last episode (jan.13,2019), he presented a tonic-clonic seizure. EEG done when admitted in our dept. showed subcontinuous polyspike and wave bilateral discharges (2-3Hz) interrupted by normal alpha activity; bolus of 1000 mg Levetiracetam i.v. infusion in 5' reduced gradually activity frequency with progressive prolonging of normal pattern intervals (from 2" until 20" and more).

Brain TC, routine haematochemical examination, EKG were normal. No fever or use of psychotropic drugs/substances assumption. Cognitive and brief term memory were normal before and after LEV. Brain MRI will be done next week.

Conclusions: we report the case to reflect about opportunity to recur to aggressive treatment during NCSE and how long extend aed therapy in an apparent non symptomatic . Finally, may we consider our case a recurrent absence status with in consideration of slight compromise of daily performances?

References

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Correlation between initial clinical and electroencephalographic findings and follow-up of elderly with nonconvulsive status epilepticus

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Background: There are controversies concerning ictal EEG patterns and therapy procedures for the treatment of nonconvulsive status epilepticus (NCSE). Objective: To correlate clinical and ictal EEG data after administration of benzodiazepine (BZD) and/or antiepileptic

drugs (AEDs) in elderly individuals with NCSE diagnosis, in accordance with ILAE, at a significance level of $p < 0.05$.

Procedures: Thirty-six elderly patients (70.8 ± 7.8 years) with clinical manifestation and critical record of NCSE, treated at the PUC Hospital, Campinas, SP, Brazil, were included in the study.

Results: Eight patients with NCSE with coma and 28 patients with NCSE were not comatose. Change in basal activity occurred in 30 cases, rhythmic delta activity (RDA) occurred in 13 cases and periodic patterns (PD) occurred in 19 cases (lateralized in 18 cases and generalized in 1 case), and electrographic SE was observed in 13 (53.1%) cases. Initial clinical improvement after BZD and/or antiepileptic drug (AEDs) therapy was observed in 20 (52.6%) cases; improvement in EEG background activity occurred in 10 (27.8%) cases and in EEG patterns in 20 (55.6%) cases after 3.9 days. A total of 14 deaths occurred. No significant association was observed between initial clinical improvement and background activity improvement (Fisher's exact test, $p = 0.456$) and EEG patterns (RDA, PD, unequivocally clear focal electrographic SE) ($p = 0.091$), death ($n = 9$ vs $n = 5$, $p = 0.501$) and presence of acute or remote brain injury. The predictive factor for the occurrence of death was the diagnosis of NCSE in comatose patients ($p = 0.016$).

Conclusion: No specific electrographic discharge patterns were observed according to the type of NCSE. No relationship between EEG patterns and initial clinical improvement after BZD/AED therapy was observed in elderly patients with NCSE. Death rate was high and related to NCSE with coma.

Keywords: non-convulsive status epilepticus, EEG, elderly.

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Electroencephalographic status epilepticus is common in critically ill children undergoing continuous EEG monitoring

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Objectives: Electroencephalographic seizures are common in critically ill children undergoing continuous EEG monitoring. However, few data are available regarding seizure characteristics which would inform management decisions. We aimed to describe electroencephalographic seizure characteristics with management implications.

Methods: We performed a single-center prospective observational study of consecutive critically ill children undergoing continuous EEG monitoring.

Results: This interim analysis included 498 consecutive critically ill children who underwent EEG monitoring from April – November 2017. Subjects were 56% male, the median (IQR) age 5.9 years (1.5, 13.1), and categorical diagnoses included acute structural (40%), acute non-structural (30%), and epilepsy-related (30%). Electrographic seizures occurred in 137 (28%) patients. Seizure duration was less than 1 minute in 69 subjects (51%), 1-5 minutes in 39 subjects (29%), 6-30 minutes in 17 subjects (13%), and >30 minutes in 10 subjects (7%). Electrographic status epilepticus occurred in 29 subjects (21%) and consisted of continuous seizures in 9 subjects (31%) and frequent recurrent seizures in 18 subjects (62%). Seizure onset was focal in 69 subjects (51%), generalized in 63 subjects (46%) and multifocal in 4 subjects (3%).

Seizure spread was focal/unilateral in 51 subjects (37%) and bilateral in 85 subjects (62%).

Conclusions: Electroencephalographic status epilepticus is common and has been associated with unfavorable neurodevelopmental outcomes. However, brief seizures (< 1 minute) are also very common. These seizures may not induce secondary brain injury may not provide benefit while exposing patients to unnecessary anti-seizure medication adverse effects. Further studies are warranted to determine the optimal management of seizures of varying durations.

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The EEG diagnosis of NCSE: concordance between Salzburg Criteria and clinical practice

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Background: The diagnosis of Non Convulsive Status Epilepticus (NCSE) in everyday clinical practice can be challenging. To help identify NCSE, Salzburg criteria (SC) have been recently validated. Here we evaluate the concordance in NCSE diagnosis between neurologists not trained in SC use, who take care of the patient in the clinical setting, and an expert panel who retrospectively evaluated the EEG according to SC.

Methods: All consecutive urgent EEG done from January 1st to March 30th 2018 were considered.

A pool of three epileptologists trained in the use of SC, and not involved in the clinical evaluation of the incident case (and of the corresponding EEG), retrospectively classified the EEG pattern according to SC in three categories: definite NCSE, no NCSE, possible NCSE. Finally, we defined the degree of concordance between the diagnosis made by the neurologist who took care of the patient and the classification made by the expert.

Results: Among the 574 consecutive EEG done in emergency conditions in the examined period, 187 (33%) have been made to rule out a NCSE (105 male; median age of 73 yrs) and were evaluated by 15 physicians. The neurologist made a diagnosis of NCSE in 19 (10%) out of 187 cases. The expert panel classified 9 EEG (5%) as "definite NCSE", 96 (51%) as "no NCSE" and 82 (44%) as "possible NCSE". Among the 82 cases defined as "Possible NCSE" by the expert, 10 (12%) were diagnosed as NCSE, while 72 (88%) were considered without status.

Concordance was 100% evaluating the "no NCSE" and the "definite NCSE" categories.

Conclusions: our data show that there is a considerable discrepancy between diagnosis made by neurologists not trained in the use of SC and the expert panel. The "Possible NCSE" category is a grey zone and further studies are needed.

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