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.

doi:10.1016/j.yebeh.2019.08.040

Epilepsy & Behavior 101 (2019) 106766

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.

doi:10.1016/j.yebeh.2019.08.041

Epilepsy & Behavior 101 (2019) 106767

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 theese events he had difficult to: a) find objetcs 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 frequence with progressive prolonging of normal pattern intervals (from 2" until 20" and more).

Brain TC, routinary haematochemical examination, EKG were normal. No fever or use of psycotropic 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 compromission of daily performances?

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doi:10.1016/j.yebeh.2019.08.042

Epilepsy & Behavior 101 (2019) 106768

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