Background: Posterior reversible encephalopathy syndrome (PRES) associates various neurological manifestations (headaches, seizures, altered mental status, cortical blindness, focal neurological deficits, vomiting) and transitory changes on neuroimaging consistent on cerebral edema. It has been associated with hypertension and immunosuppressive treatments, among other factors. In aditrion, epileptic seizures appeares in the majority of cases and 20% of them show status epilepticus.

Methods: We report a case of PRES occuring in the setting of a metastasic brain tumor treated with chemotherapy and radiosurgery admitted to the intensive care unit (ICU) that develops status epilepticus.

Results: 53-year-old man with cerebellar metastases secondary to lung carcinoma treated with chemotherapy and radiosurgery the previous month. After accidental fall down the stairs he presented complex seizures. The CT showed no changes in his previous brain lesions and we started treatment with levetiracetam. Due to the persistence of the crisis, admission to the ICU was decided and valproate was added to the treatment. The electroencephalogram (EEG) showed intercritical abnormalities and moderate encephalopathy. The patient presented drowsiness, bradypsychia and right hemiparesis. After 4 days he was agitated and the EEG showed status epilepticus. Treatment with phenytoin was started. Daily EEG were performed, showing an improvement in the registry. The lumbar puncture did not show tumor cellularity and the MRI reported findings compatible with PRES.

Conclusions: PRES is a rare dissease secondary to vasogenic edema. In the majority of cases changes are localized in posterior irrigation areas of the brain. CT imaging is typically normal. The classic imaging finding of PRES in RMI are occipital subcortical vasogenic edema without signs of stroke.

The possible cause of the syndrome developed by our patient is vascular disease with endothelial damage and rupture of the blood-brain membrane secondary to chemotherapy and/or radiosurgery. The main pilars of the treatment includes blood pressure regulation, control of seizures and anti-edema therapy. The treatment of epileptic crisis and status epilepticus should be performed in an ICU with monitorization of cerebral electrical activity.

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Everyday nonconvulsive status epilepticus episodes in a 12-year-old girl with pharmacoresistant epilepsy of unknown etiology admitted for presurgical evaluation

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We would like to discuss a case of a girl who has been admitted to our hospital for presurgical evaluation and remains a diagnostic and therapeutic challenge.

She's been suffering from epilepsy since being 18 months – at first presenting with unprovoked, sporadic GTCS. No correlation with fever or infection. Until the age of 10 no AED was introduced and no seizures were reported. Her intellectual development was rated as above-average. After the seizure-free period the seizures recurred, described by the mother as: 1 - difficulties speaking and eyelid fluttering, lasting 20-30 minutes every evening; 2- tremor of the left hand with preserved consciousness, occurring couple of times a week. In the medical files we found description of episodes of strange feelings like elongation of her nose or the right hand. The EEG recordings (interictal) always showed generalized changes, also with a pattern of typical 3Hz discharges. The MR exams at first showed no changes. Metabolic tests were not significant. No genetic testing performed. She has been treated with VPA, LEV, TPM, LAC, LTG, CLB and solumedrol pulses with no significant effect. The last MR exam (2018) suggested pathological focus in the right parahippocampal gyrus. In FDG-PET exam decreased FDG uptake in the medial part of the right temporal lobe was described.

In our department we performed a 15-hour video-EEG (on VPA + LTG + CLB treatment) with registration of three clinical episodes lasting about 30 minutes each. During those events the girl was responsive, but had difficulties speaking with lower reaction time and slower speed of speech. When in standing position she had tendency to fall down, like having "week knees". No lateralizing signs could be noticed. At the time of clinical symptoms we registered runs of generalized waves around 14Hz and amplitude 30-80uV with superimposed synchronized sharp waves and sharp-slow wave discharges, evolving into continuous generalized discharges of sharp and slow wave complexes 2-2,5Hz and amplitude up to 320uV. No significant effect after diazepam application.

We introduced MAD with uncompliance. Now the girl is put on ESM. We expect the results of genetic testing (1000 genes panel) in April.

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Penelope syndrome: Exploring the Pandora's Box of Genetic Associations

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Background: Penelope syndrome is identified by the defining characteristic EEG features of continuous spikes and waves, or status epilepticus, during sleep (SES). SES is associated with acquired epileptic aphasia, cognitive and behavioral disturbance, as well as motor impairment. In addition to organic brain lesions, genetic underpinnings are being identified.

Methods: We present two siblings, their father who were identified with features of epileptic aphasia, language disturbance and focal epilepsy, and a child with chromosomal aberration. In 2 of the 3 children a distinctive pattern consistent with SES was identified during EEG monitoring.

Results: The first patient presented with focal onset seizures at age 3 years accompanied by language and cognitive regression, EEG features consistent with SES. She was treatment resistant with conventional anti-epileptic medications, but responded well to steroids regaining language developmental milestones. Her 5-year-old sister presented with focal onset seizures. Her language development shows impairments in receptive language functions. A previously unreported potentially pathogenic GRIN2A variant was identified in the 2 siblings and their biological father. The third child initially presented with focal seizures with EEG correlates, developmental impairments in language, cognition and behavior. During follow up, she too exhibited EEG features of SES and treatment resistance. She was identified to have a 16p11.2 microduplication on a chromosomal microarray.

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 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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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