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Refractory non-convulsive status epilepticus with favorable outcome in a patient with Marchiafava–Bignami disease

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Background: Marchiafava–Bignami disease (MBD) is a rare condition mainly associated with chronic alcoholism, which is characterized by demyelination of the corpus callosum. MBD results in a variety of neurological symptoms including altered mentality, gait difficulty, cognitive dysfunction, and seizure. Herein, we report a patient showing a favorable outcome after refractory non-convulsive status epilepticus (NCSE) as an initial manifestation of MBD.

Methods: A case report.

Results: A 58-year-old man presented with an acute confusional state with intermittent upward eyeball deviation, which had been developed a few hours. He had a history of chronic heavy alcohol consumption. The amount of alcohol intake was about 2 bottles of Korean Soju per day. Upon neurological examination, he was disoriented and his level of consciousness fluctuated. Considering the possibility of alcohol-related symptoms, he was promptly administered intravenous (IV) thiamine 50 mg with normal saline. Routine blood tests, including tests of thiamine levels, and cerebrospinal fluid studies revealed no abnormalities. Brain magnetic resonance imaging (MRI) on admission showed hyperintense lesions involving the splenium and genu of the corpus callosum and the cerebral cortex. Electroencephalography revealed periodic rhythmic delta activities suggestive of NCSE. IV lorazepam (0.1 mg/kg) followed by IV fosphenytoin (30 mg/kg loading doses) were administered, but his clinical and electrographic seizure persisted. Seizure control was achieved on day 4, after adding levetiracetam (2000 mg/day) and lacosamide (400 mg/day). On day 7, he was oriented and was able to name objects, follow commands, and to walk with some assistance. He received IV thiamine 200 mg/day for 28 days, followed by oral thiamine 30 mg per day. Follow-up MRI at 1 month after the onset of symptoms showed persistent hyperintense lesions involving the splenium and genu of the corpus callosum, with some atrophic changes. At the 2-month follow-up, he was able to carry out many of his usual activates without assistance. He did not experience any symptom that indicated a seizure while on maintenance levetiracetam (300 mg/day) and lacosamide (400 mg/day).

Conclusion: MBD can be involved in the etiology of NCSE. Also early treatment with thiamine may be necessary for a better prognosis.

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Status epilepticus secondary to extensive pneumocephalus

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Background: Pneumocephalus is a frequent pathology in the postoperative period of a craniotomy. Patients who present neurological deterioration or epileptic seizures in the postoperative period of a craniotomy require a rapid diagnosis. It is important to perform imaging and electroencephalogram tests (EEG) to diagnose surgical complications and epileptic events and initiate an early and aggressive treatment.

Methods: We report a case of status epilepticus occuring in the setting of extensive pneumocephalus after craniotomy for resection of frontal meningioma.

Results: 82-year-old who presented neurological deterioration in the inmediate postoperative period of a frontal meningioma resection. Urgent computerized tomography showed a large frontal air collection (extensive pneumocephalus) with postoperative changes, without bleeding. The patient presented a complex epileptic crisis and the EEG showed status epilepticus. The patient required two anticonvulsant drugs (levetiracetam and phenytoin) and subsequent general anesthesia with midazolam and propofol. Daily EEG showed moderate-severe encephalopathy with "outbreak-suppression". After withdrawal of general anesthesia the patient presented a GCS of 3 and after 14 days the medical staff decided to limit the therapeutic effort taking into account her vital preferences and those of her family.

Conclusions: Pneumocephalus is the consequence of a traumatic violation of the dura and should be considered in all patients who present epileptic crisis after a craniotomy. Gold standard diagnostic test is CT without contrast, which can identify even small amounts of intracranial air and can exclude other diagnoses. Epileptic status is a neurological emergency that requires immediate treatment and an early EEG must be performed.

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Posterior reversible encephalopathy syndrome du to chemotherapy, a case report

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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.