average duration of 27 days (range 14-32). Characterizing features were male gender, young age, previous good health, cerebrospinal fluid pleocytosis (in 7), antecedent febrile illness (in 7), extraordinarily prolonged status epilepticus, failure of extensive investigations to reveal an underlying cause, catastrophic outcome as well as T2 hyperintense signal in temporal lobes(in 5), BL symmetrical putaminal T2/flair high signals(in 1)and leptomeningeal enhancement (in 1) on brain magnetic resonance imaging. Treatment modalities included at least 3antiepileptic drugs (in all patients), anesthetics (in all patients), intravenous immunoglobulin (IVIG, 8 patients), steroids (4 patients), plasmopheresis (1 patient). Five patients improved completely without neurological deficit and three of them survived withmoderate to severe disability. Two patients who were not given early immunotherapy, died from complications associated with prolonged ICU stay. One of the survivors received long term immunotherapy.

Conclusions: The cause of NORSE syndrome may often be difficult to find. NORSE carries a poor prognosis and early recognition and treatment may improve outcomes. Epilepsy and cognitive issues are common among survivors although a small minority of them eventually return to a normal lifestyle. Affected individuals are most often treated for weeks in an intensive care unit because they require prolonged anesthesia to control their seizures. In this case series, we have shown that immunotherapy seems to be a helpful treatment option when conventional therapy fails.

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Association between initial therapy of refractory generalized convulsive status epilepticus in adults and its outcome

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Objective: to analyze outcomes of refractory (R) generalized convulsive status epilepticus (GCSE) in adults.

Materials and Methods: The study included 23 women with RGCSE, aged from 19 to 76 years. Patients presented with RGCSE in acute symptomatic form (n = 5) and as a complication of epilepsy (n = 18), of them 14 with structural focal epilepsy, 3 with focal epilepsy of unknown etiology, 1 with combined idiopathic generalized and structural focal epilepsy. Intravenous (iv) ivLorazepam, ivPhenytoin, ivPhenobarbital are unavailable in Russia, however iv Diazepam (ivDZP), Valproic Acid (ivVPA), Levetiracetam (ivLEV), Lacosamid (ivLCM) and iv anesthetics are available. Initial GCSE treatment for patients at home and in an ambulance was started with ivBZD, with ivVPA, or with combination of ivBZD + ivVPA within 35 min to 150 min after the GCSE onset. The maximum dose (mg/kg) of all types of ivAEDs and iv anesthetics was used.

Results: 3 patients (13%) with acute symptomatic GCSE died due to multiple organ failure not related to GCSE. In the remaining 18 cases, RGCSE was relieved. Non-acute symptomatic RGCSE lasting for 3 to 132 hours was observed mainly in structural focal (frontal, fronto-temporal) epilepsy (60.9%). The main reason for the development of GCSE in adults (43.5%) was an AED withdrawal. In these cases, the basic oral AEDs were continuously administered through a naso-gastric tube. Super-refractory GCSE was confirmed in 11 cases (47.8%). In the prehospital phase of GCSE treatment, ivDZP was used in 6 patients, 5 of them developed super-refractory GCSE; ivVA was used in 8 patients, 2 of them developed super-refractory GCSE; initial therapy with ivDZP + ivVA was used in 7 patients, 3 of them developed super-refractory GCSE. During treatment with iv anesthetics, respiratory depression developed with subsequent invasive ventilation and longer recovery. In one case of RGCSE recurrence, perampanel was successfully administered through a naso-gastral tube at 12 mg/day.

Conclusions: deaths occurred only in cases of acute symptomatic RGCSE. The vast majority of refractory RGCSE in adults (43.5%) was provoked by withdrawal of AEDs. Preliminary results indicate that prehospital (initial) use of ivVPA or a combination of ivDZP + ivVA significantly increase the efficacy of RGCSE therapy.

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Prognosis in patients with brain tumor-associated status epilepticus

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Background: Tumor related epilepsy is common in patients with brain tumors and it usually starts at the onset of the disease. By contrast, status epilepticus is less frequent, occurs at later stages and it is often related with tumor progression. We aimed to investigate prognosis and associated factors in these patients.

Methods: We consecutively included 61 patients that were admitted at our hospital due to brain tumor-associated status epilepticus between 2011-February and 2018-April. Demographic and clinical characteristics (type of tumor, duration and treatment of the status epilepticus) were recorded. We analyzed status epilepticus prognosis at long-term (GOSE – Glasgow Outcome Scale) and related factors.

Results: Mean age was 62.64 years-old (±14.9) and 59% were men. The 34.4% (n = 21) were metastasis, followed by meningiomas (24.6%, n = 15), glioblastoma multiforme (24.6%, n = 15), low grade glioma (4.9%, n = 3) and other (11.7%, n = 7). Mean follow up was 12.6 months and mean survival time 19.6 months (IC 11.4-27.4). On the multivariate analysis the presence of glioblastoma multiforme or metastasis (OR 19 (2.23 – 161.58), p = 0.007) and the status epilepticus duration >18 hours (1.28 – 42), p = 0.025) independently predict worse prognosis at one year follow-up. Mortality at follow up was only associated with the type of tumor (HR 7.26 (2.59-20.37), p < 0.001).

Conclusions: Longer duration of status epilepticus and the presence of glioblastoma multiforme or metastasis predict poor prognosis at long-term.

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