XLVIII CONGRESSO NAZIONALE
14-17 OTTOBRE 2017 – NAPOLI

Pitfalls in non convulsive status epilepticus diagnosis: Creutzfeldt-Jakob disease

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Background Non convulsive status epilepticus (NCSE) is a diagnostic challenge. Clinical presentation is not always diagnostic when the disturbance of consciousness is the predominant feature, without focal or subtle epileptic phenomena. In these cases EEG is crucial for the diagnosis. A consensus panel published in 2013 proposed working criteria for EEG diagnosis of NCSE (Salzburg criteria[1]).

Case presentation We present two older patients admitted for subacute onset of mental status alteration. At examination Patient 1 showed bilateral blindness, postural and action tremor in the head and in both arms with myoclonic jerks of the left arm; patient 2 arrived in a stupor state with erratic jerks. EEG monitoring revealed a 1.5-2 Hz lateralized periodic discharges (LPDs) pattern in both patients (Fig. 1A-2A). Intravenous antiepileptic drugs (AEDs) administration induced resolution of epileptiform abnormalities (EA) (Fig. 1B-2B) with a partial improvement of alertness state, not well evaluable because patients fell into iatrogenic sedation. In patient 1 myoclonic jerks disappeared. After 60 minutes, EA reappeared and the patients returned to the basal condition. According to Salzburg criteria algorithm for EEG diagnosis of NCSE in clinical practice[2] (Fig.3), since EA frequency was ≤ 2.5 Hz, at least one secondary criteria should have been fulfilled among typical spatiotemporal evolution, subtle ictal phenomenon, EEG and clinical improvement after IV AEDs, to have a definite or possible NCSE diagnosis. Given the EEG response to IV AEDs, both patients met Salzburg criteria for possible NCSE. We started an antiepileptic therapy, but no clinical or EEG improvement was appreciated. Subsequent EEGs revealed in both patients, a continuous pattern of bilateral asymmetric 1.5 Hz periodic sharp wave complexes (PSWC) which fulfilled EEG criteria for CJD. The diagnosis of CJD was then confirmed by neuroimaging, liquor and genetic analysis.

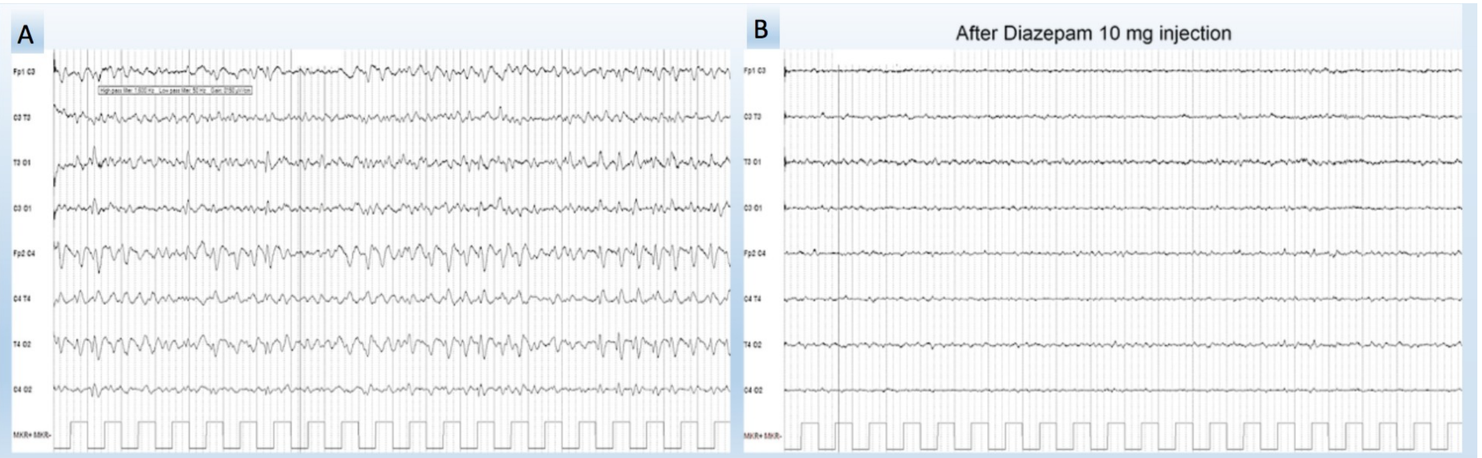

Figure 1 *EEG of patient 1:* ***A)*** *showed an epileptiform activity with periodic 1.5 Hz sharp-wave complexes better represented over the central, temporal and parietal regions, with sporadic contralateral spread.* ***B)*** *10 mg Diazepam IV injection induced a prompt resolution of epileptiform activity after six minutes.*

Figure 2 *EEG of patient 2:* ***A)*** *revealed a 1.5-2 Hz bilateral asymmetric periodic discharges with right prevalence,* ***B)*** *after administration of IV diazepam 10 mg there was a significant reduction of epileptiform discharges with a reappearance of alpha symmetric background activity.*

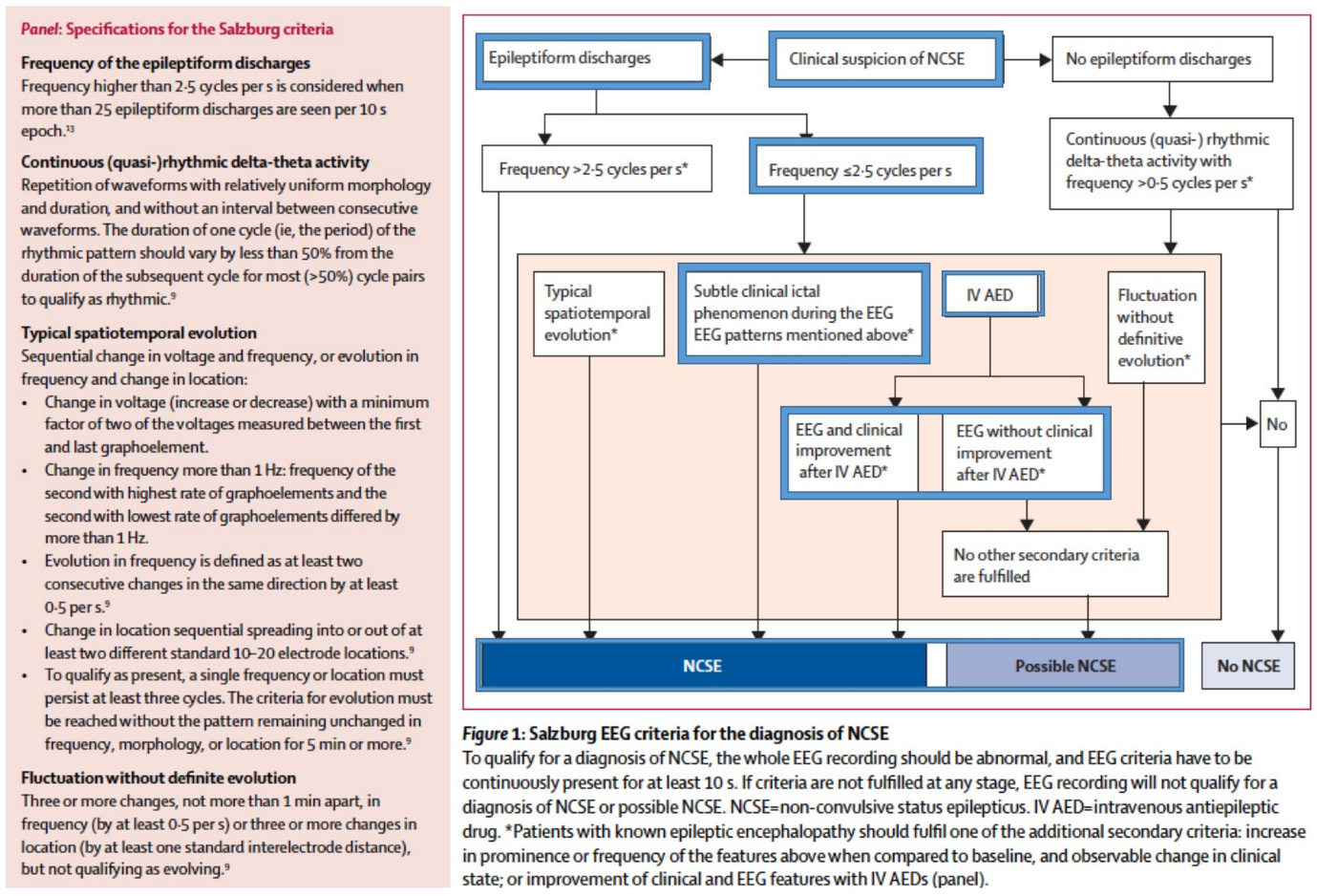

Figure 1: **Salzburg EEG criteria for the diagnosis of NCSE**
To qualify for a diagnosis of NCSE, the whole EEG recording should be abnormal, and EEG criteria have to be continuously present for at least 10 s. If criteria are not fulfilled at any stage, EEG recording will not qualify for a diagnosis of NCSE or possible NCSE. NCSE=non-convulsive status epilepticus. IV AED=intravenous antiepileptic drug. *Patients with known epileptic encephalopathy should fulfil one of the additional secondary criteria: increase in prominence or frequency of the features above when compared to baseline, and observable change in clinical state; or improvement of clinical and EEG features with IV AEDs (panel).

Fig.3 *Algorithm of Salzburg criteria, modified from Leitinger 2016*

Discussion Periodic sharp wave complexes (PSWC) in CJD cannot be typical at first, can be lateralized resembling to LPDs, suggesting a NCSE as occurred in our patients. Many CJD reports showed that intravenous AEDs transiently attenuates or even abolish PSWC, but without clinical improvement[3]. Furthermore, in patients with a decreased vigilance state as well as in elderly patients, a clinical improvement is not easily appreciable given that IV AEDs administration further reduce the consciousness level.

Conclusion We acknowledge the high diagnostic accuracy of Salzburg criteria, however we believe that a slow evolution of clinical symptoms and periodic EEG pattern with frequency ≤ 2.5 Hz should be a red flag for considering the diagnosis of CJD. In these cases, an additional clinical effort should be done in searching anamnestic and objective data focusing on CJD and secondary Salzburg criteria should be critically applied, to avoid diagnostic mistakes and harmful treatments for our patients.

Bibliography

1.Beniczky S, Hirsch LJ, Kaplan PW, Pressler R, Bauer G, Aurlien H, et al. Unified EEG terminology and criteria for nonconvulsive status epilepticus. Epilepsia 2013;54:28–9.
2.Leitinger M, Trinka E, et al. Diagnostic accuracy of the Salzburg EEG criteria for non-convulsive status epilepticus: a retrospective study. Lancet Neurol 2016;15:1054–62.
3.Lapergue B, Demeret S, et al. Sporadic Creutzfeldt-Jakob disease mimicking nonconvulsive status epilepticus. Neurology 2010;74:1995–9.