Epileptic Disord 2011; 13 (2): 188-92

Complex partial status epilepticus: an unusual presentation

Samir Kumar Praharaj [1], Sujit Sarkhel [2], Imon Paul [3], Anshuman Tripathi [3], Mohammad Zia-ul-Haq Katshu [4], Vinod Kumar Sinha [3]

[1] Department of Psychiatry, Kasturba Medical College, Manipal, Karnataka
[2] Institute of Psychiatry, Institute of Postgraduate Medical Education and Research (IPGMER), Kolkata
[3] Central Institute of Psychiatry, Kanke, Ranchi, Jharkhand, India
[4] School of Psychology, Brigantia Building, Bangor University, Bangor, Wales, United Kingdom

Received October 18, 2010; Accepted March 22, 2011

ABSTRACT – Nonconvulsive status epilepticus (NCSE) is not uncommon, especially in the elderly population. It can occasionally present as confusion, personality change, coma, subtle motor activity and psychosis, thus causing diagnostic difficulties. We report an unusual case of NCSE which presented as psychosis-like episodes at regular intervals. The confirmation of diagnosis was aided by serial electroencephalography.

Key words: nonconvulsive status epilepticus, complex partial status epilepticus, psychosis, hallucinations, confusion, elderly

Status epilepticus (SE) is defined as either recurrent seizures without intermediate complete recovery of neurological function or continuous clinical or electrical seizure activity lasting for at least 30 minutes, with or without impairment of consciousness (Treiman, 1993). The recognized forms of SE include recurrent generalised tonic-clonic seizures without full consciousness (convulsive SE), prolonged seizure activity in the absence of major motor manifestations where the patient presents an "epileptic twilight state" (non-convulsive SE or NCSE), and continuous or repetitive focal seizure activity without alteration of consciousness (epilepsia partialis continua) (Treiman, 1993).

NCSE comprises almost a quarter of all cases of SE (Treiman *et al*., 1998). In various studies, the incidence of NCSE ranges from 2-20 cases per 100,000 (Meierkord and Holtkamp, 2007). The rate of NCSE has been shown to increase with age as focal seizures tend to generalise less often in old age (DeToledo, 1999; Waterhouse and DeLorenzo, 2001). The clinical manifestations of NCSE may pose diagnostic difficulties in elderly patients.

Here, we report an unusual case of NCSE with psychosis-like episodes at regular intervals. The confirmation of diagnosis was aided by serial electroencephalography (EEG).

Case study

A 55-year-old male presented with an episodic illness of 35 years, each

Correspondence:
S.K. Praharaj
Department of Psychiatry,
Kasturba Medical College,
Manipal,
Karnataka,
India 576104
<samirpsyche@yahoo.co.in>

doi:10.1684/epd.2011.0429

episode lasting for three to four days and occurring every month. The episodes were characterised by muttering and smiling to himself, an inability to work, decreased sleep and abusive and assaultive behaviour towards his wife. Onset and resolution of each episode was abrupt. During these periods, he would not recognize family members, speak less than usual and remain housebound. He would have continuous hallucinations of derogatory voices, although his feeding and toilet habits remained intact. He had partial memory of his episodic abnormal behaviour and occasionally expressed remorse for these activities. These episodes, occurring every month, were so predictable and stereotyped that his wife would leave home preemptively to avoid violence. One month prior to the onset of the illness, he sustained trauma over the occipital region after being hit by a blunt object and was unconscious for four to five hours. For the initial three weeks, his behaviour in the ward remained unremarkable. Subsequently, he had an episode characterised by hallucinations, confusion and lethargy which aborted following administration of lorazepam, at 4 mg intramuscularly. There was complete amnesia for the episode. His younger brother had bipolar disorder.

Routine investigations including complete haemogram, renal and liver function tests, serum electrolyte analysis, as well as ECG and thyroid profile, were normal. Brain CT and MRI revealed no abnormality. PGI Battery for Brain Dysfunction (PGI-BBD; Pershad, 1990) and Luria-Nebraska Neuropsychological Battery (LNNB) revealed no neuropsychological deficit. The EEG was recorded thrice; at baseline, during the episode and following its resolution, using 21 Ag-AgCl electrodes placed according to the international 10-20 system with Neurofax EEG-9000 (Nihon Kohden, Japan) amplifiers (sampling rate: 512 Hz, TC: 0.1 seconds, HFF: 70 Hz) and linked ear electrodes as reference. Each recording was performed for 30 minutes, with the patient remaining awake and with eyes closed. The EEG showed a change in background from well regulated, predominantly alpha activity at baseline to desynchronized activity during the episode, reverting back to baseline state after resolution of the episode *(figure 1A-C)*. The postictal EEG also showed bilateral fronto-centro-parietal spike-slow wave activity *(figure 1C)*. The spike-slow wave events were selected manually for source reconstruction using Multiple Signal Classification (MUSIC) algorithm which localised these abnormalities to the right medial frontal region *(figure 2)*.

Considering the possibility of complex partial status epilepticus, carbamazepine treatment was started. During the course of optimisation of carbamazepine treatment, he had two more episodes, identical to the previous episodes, which responded to injection of lorazepam. After episodes were controlled with 800 mg per day of carbamazepine for two consecutive months, there was another recurrence following reduction in dosage to 400 mg per day by the patient himself. He was finally maintained on 800 mg per day of carbamazepine without recurrence of such episodes over the next six months.

Discussion

In this case, our patient presented with prominent hallucinatory psychotic episodes lasting for a few days with confusion and amnesia for the episodes. Although psychotic manifestations were in the fore, the recurrent and stereotyped nature of the episodes raised a suspicion of NCSE. However, the two initial EEGs did not reveal any obvious epileptiform abnormality, although desynchronization of background was evident during ictal EEG which normalised subsequently postictally. The last EEG also showed bilateral fronto-centro-parietal spike-slow wave activity. NCSE may present clinically with myriad manifestations. Confusion, personality change, psychosis, a prolonged postictal period (greater than 30 minutes), coma, subtle motor activity or nystagmus are some of the presentations of NCSE that may mislead clinicians to correctly diagnose NCSE (Kaplan, 1999; Walker, 2001).

NCSE is often recurrent, self-limiting and responsive to benzodiazepines (Treiman and Walker, 2006). Among first-line agents, lorazepam shows the maximum response rate (in two thirds of cases), followed by phenobarbital, a combination of diazepam and phenytoin and phenytoin alone (Bleck, 1999; Treiman, 2007). Our patient also showed prompt response to lorazepam. Moreover, carbamazepine prevented recurrence of attacks, which re-emerged after fortuitous dose reduction.

It has been suggested that diverse components, such as impaired consciousness, ictal EEG changes and response to treatment, may be considered in the definition of NCSE (Mayeux and Lueders, 1978; Treiman and Delgado-Escueta, 1983; Tomson *et al*., 1992; Cockerell *et al*., 1994; Niedermeyer and Ribeiro, 2000), in order to broaden its ambit and include atypical presentations. Interestingly, clinical features and response to treatment were suggestive of the diagnosis of NCSE in our case. Although the EEG was not diagnostic in our case, it provided presumptive evidence. Such stereotypical presentations, especially in older individuals, should raise the suspicion of NCSE. In such cases, three sequential EEG recordings (preictal, ictal and postictal), rather than a single recording, may be more revealing. EEG criteria for NCSE have been proposed (Kaplan, 2007), although they are not diagnostic.

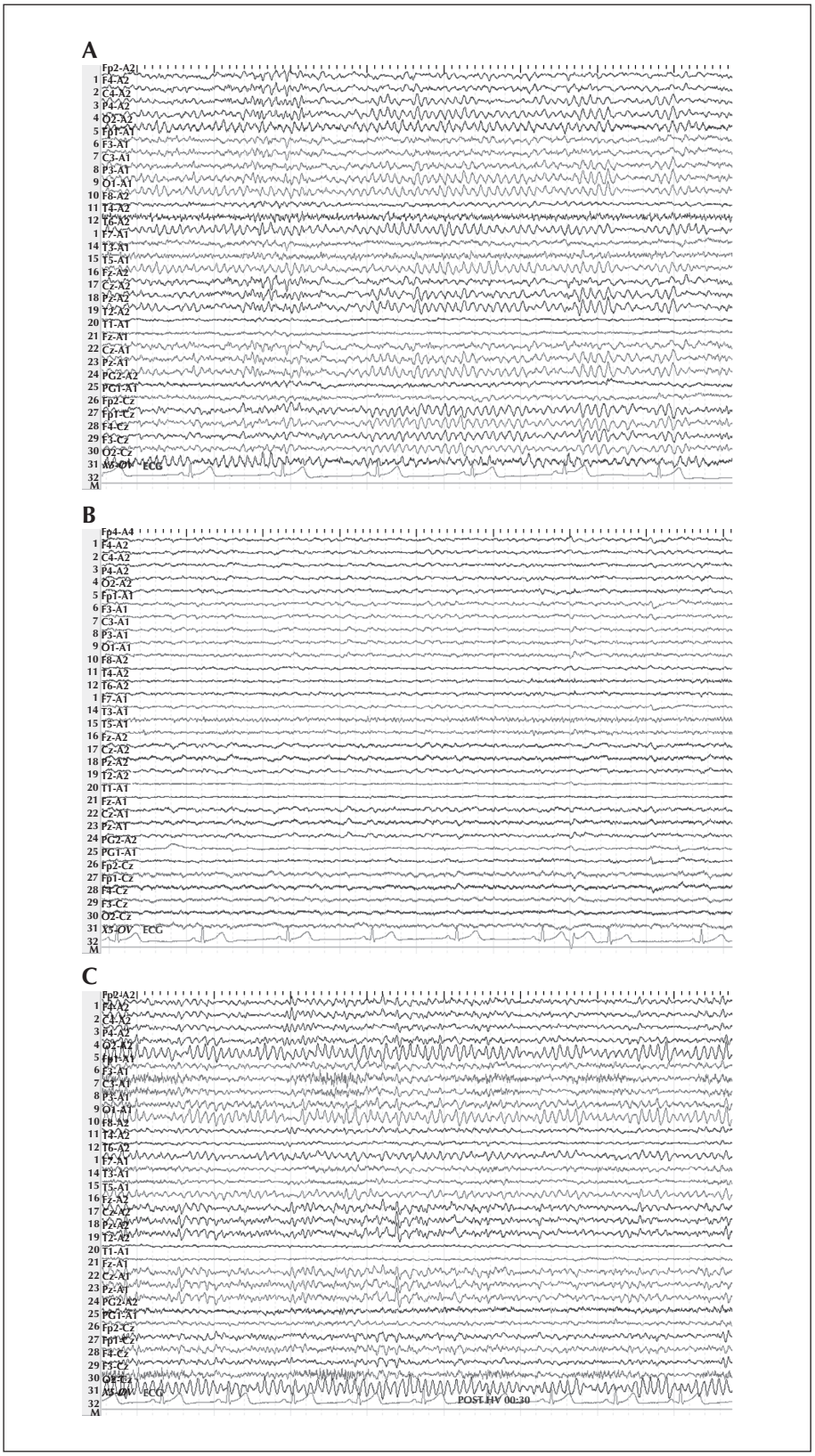

Figure 1. EEG during wakefulness showing (**A**) normal background at baseline, (**B**) desynchronized background during the episode, and (**C**) normal background postictally with an episode of bilateral fronto-centro-parietal spike-slow waves.

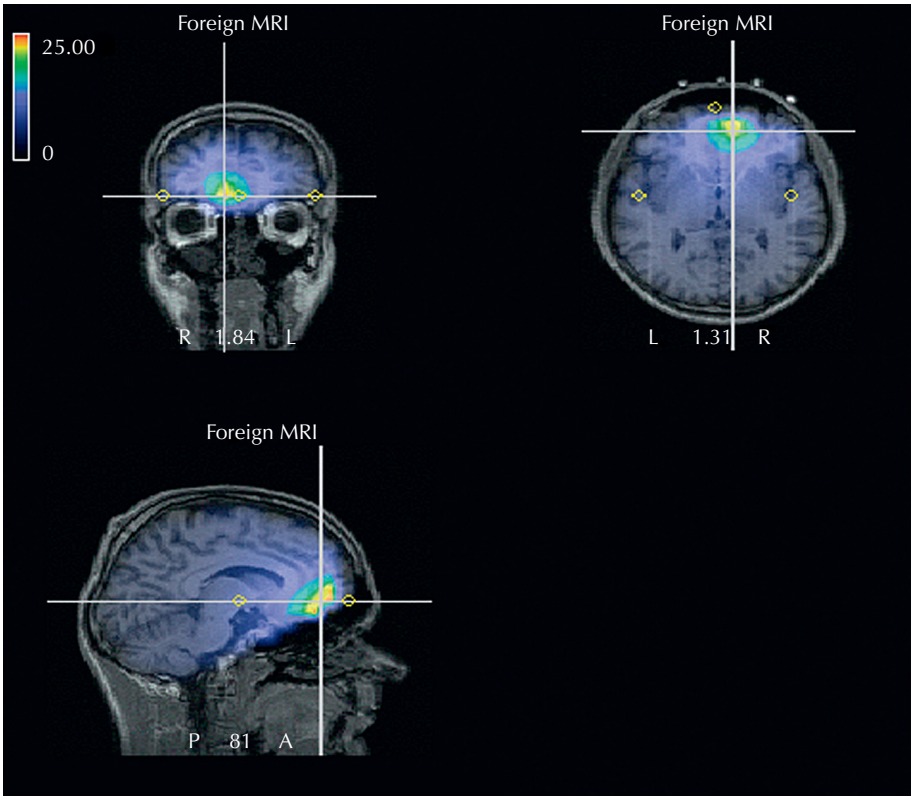

Figure 2. MUSIC analysis of spike-slow wave events localised these abnormalities to the right medial frontal region.

The most frequent form of extratemporal NCSE is associated with frontal localisation, as in our case. Rohr-le Floch *et al*. (1988), described the association of NCSE of frontal origin with prolonged periods of cognitive disturbance, occasionally with visible focal ictal signs and a consistent epileptiform EEG pattern localised over one or both frontal regions at the onset of seizures. Compared to NCSE of temporal origin, frontal NCSE cases are associated with less impairment of consciousness, but prominent confabulation, smiling, inappropriate laughter or an "ironic" appearance (Rohr-le Floch *et al*., 1988). Patients may appear indifferent or brooding, occasionally fearful, negativistic, angry, irritable, aggressive, anxious and agitated with simple and complex automatisms (Rohr-le Floch *et al*., 1988). Thomas *et al*. (1999) reported two forms of frontal NCSE. In type 1, mood disturbances with affective disinhibition or affective indifference were associated with subtle impairment of cognitive functions without overt confusion, and EEG showed a unilateral frontal ictal pattern and normal background activity. In contrast, in type 2, a cyclic severe confusional state with major behavioural disturbances was associated with bilateral, asymmetric frontal EEG discharges occurring on an abnormal background. Behavioural features associated with NCSE in the elderly include mutism and perseveration, agitation, emotional lability, aggressiveness, and hallucinosis with face, eye, and limb myoclonias (Kaplan, 2005). Our case presented with features suggestive of the rarer type 2 frontal NCSE of Thomas *et al*. (1999), *i.e*. a cyclic, acute, confusional state with hallucinations. It is imperative to rule out NCSE in such unusual presentations in the elderly. □

Disclosure.
None of the authors has any conflict of interest or financial support to disclose.

References

Bleck TP. Management approaches to prolonged seizures and status epilepticus. *Epilepsia* 1999; 40: S59-63.

Cockerell OC, Walker MC, Sander JW, Shorvon SD. Complex partial status epilepticus: a recurrent problem. *J Neurol Neurosurg Psychiatry* 1994; 57: 835-7.

DeToledo JC. Changing presentation of seizures with aging: clinical and etiological factors. *Gerontology* 1999; 45: 329-35.

Kaplan PW. Assessing the outcome in patients with nonconvulsive status epilepticus. NCSE is underdiagnosed, potentially overtreated and confounded by comorbidity. *J Clin Neurophysiol* 1999; 16: 341-52.

Kaplan PW. The clinical features, diagnosis, and prognosis of nonconvulsive status epilepticus. *Neurologist* 2005; 11: 348-61.

Kaplan PW. EEG criteria for nonconvulsive status epilepticus. *Epilepsia* 2007; 48: 39-41, Erratum in: *Epilepsia* 2007; 48: 2383.

Mayeux R, Lueders H. Complex partial status epilepticus: case report and proposal for diagnostic criteria. *Neurology* 1978; 28: 957-61.

Meierkord H, Holtkamp M. Non-convulsive status epilepticus in adults: clinical forms and treatment. *Lancet Neurol* 2007; 6: 329-39.

Niedermeyer E, Ribeiro M. Considerations of nonconvulsive status epilepticus. *Clin Electroencephalogr* 2000; 31: 192-5.

Pershad D. *Handbook of PGI battery of brain dysfunction (PGI-BBD)*. Agra: National Psychological Corporation, 1990.

Rohr-le Floch J, Gauthier G, Beaumanoir A. Etats confusionelles d'origine eĭpileptique: intereˆt de leg fait en urgence. *Rev Neurol (Paris)* 1988; 144: 425-36.

Thomas P, Zifkin B, Migneco O, Lebrun C, Darcourt J, Andermann F. Nonconvulsive status epilepticus of frontal origin. *Neurology* 1999; 52: 1174-83.

Tomson T, Lindbom U, Nilsson BY. Nonconvulsive status epilepticus in adults: thirty-two consecutive patients from a general hospital population. *Epilepsia* 1992; 33: 829-35.

Treiman DM. Generalized convulsive status epilepticus in the adult. *Epilepsia* 1993; 34: S2-S11.

Treiman DM. Treatment of convulsive status epilepticus. *Int Rev Neurobiol* 2007; 81: 273-85.

Treiman DM, Delgado-Escueta AV. Complex partial status epilepticus. *Adv Neurol* 1983; 34: 69-81.

Treiman DM, Walker MC. Treatment of seizure emergencies: convulsive and non-convulsive status epilepticus. *Epilepsy Res* 2006; 68: S77-82.

Treiman DM, Meyers PD, Walton NY, *et al*. A comparison of four treatments for generalized convulsive status epilepticus. Veterans Affairs Status Epilepticus Cooperative Study Group. *N Engl J Med* 1998; 339: 792-8.

Walker MC. Diagnosis and treatment of nonconvulsive status epilepticus. *CNS Drugs* 2001; 15: 931-9.

Waterhouse EJ, DeLorenzo RJ. Status epilepticus in older patients: epidemiology and treatment options. *Drugs Aging* 2001; 18: 133-42.