Extreme delta brushes in a 14-year old girl with anti-NMDAR encephalitis

Sir,

A previously healthy, 14-year old girl presented with an explosive onset of seizures (generalized tonic-clonic and left focal) and new-onset behavioral change. She was treated with multiple antiepileptic drugs. Given the high seizure burden and altered behavior, a continuous electroencephalogram (EEG) monitoring was commenced [Figures 1-3].

Laboratory investigations for infections and metabolic derangements were unremarkable. Magnetic resonance imaging (MRI) of the brain revealed no abnormalities. Serum and cerebrospinal fluid (CSF) anti- N-methyl-D-aspartate (NMDA) antibodies were both positive. MRI of the pelvis and abdomen were normal. A diagnosis of anti-NMDA-receptor (NMDAR)-mediated encephalitis was made.

While in the hospital, she received treatment with intravenous immunoglobulins (IVIG), steroids, and rituximab. She showed a good clinical response to therapy and was discharged home after 2 months in hospital. An electroencephalogram (EEG) done before discharge revealed persistent delta slowing over the right frontal head region but no extreme delta brushes (EDB).

She continued to have relapses intermittently. These relapses were similarly characterized by altered behavior and clinical

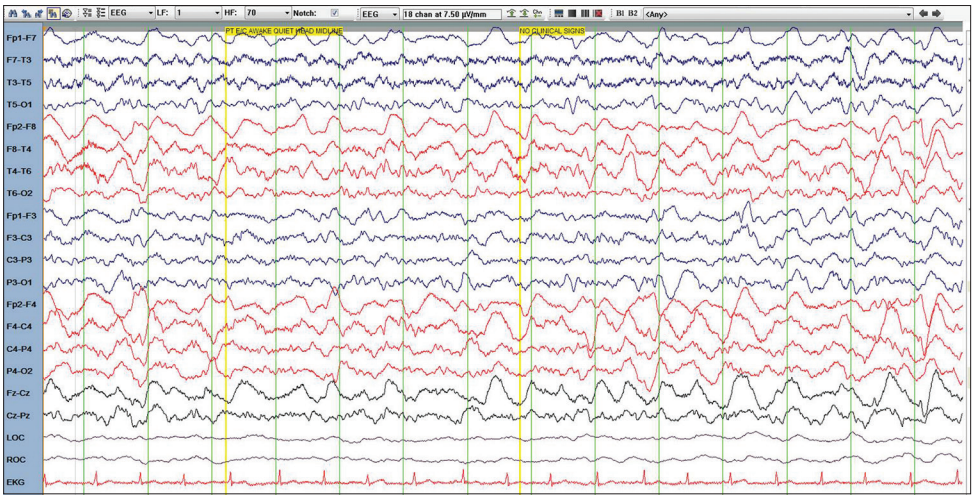

Figure 1: EEG on day 1 of hospitalization: The record shows nearly continuous 1-2 Hz polymorphic delta activity over the right fronto-temporal head region (longitudinal bipolar montage with a timebase 30 mm/sec, sensitivity 7 μV/mm, high frequency filter at 70 Hz, low frequency filter at 1 Hz and sampling rate of 256 Hz)

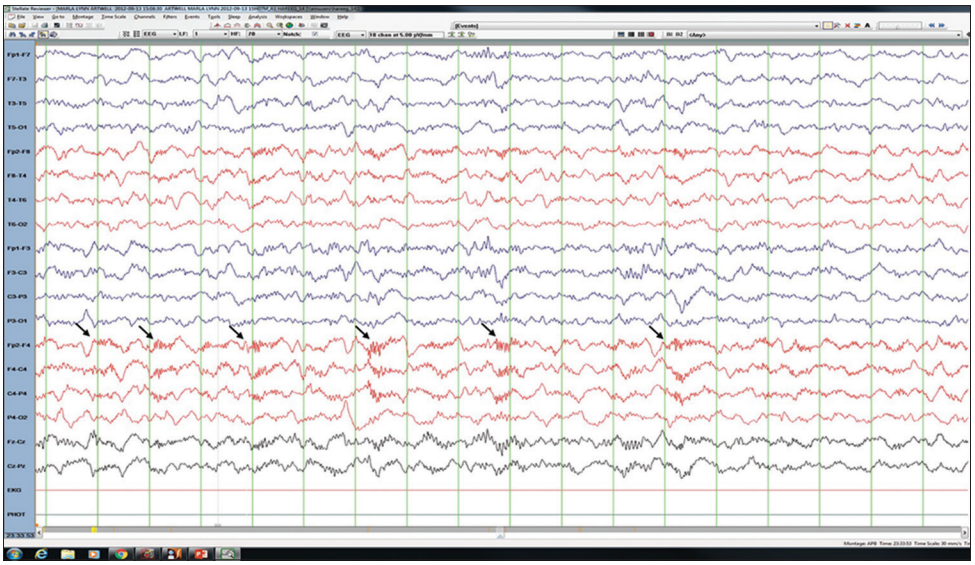

Figure 2: EEG on day 2 of hospitalization: The record shows 1-2 Hz delta activity at Fp2/F4/C4 with superimposed 25-30 Hz beta activity (black arrows) suggestive of extreme delta brushes (longitudinal bipolar montage with a timebase 30 mm/sec, sensitivity 7 μV/mm, high frequency filter at 70 Hz, low frequency filter at 1 Hz and sampling rate of 256 Hz)

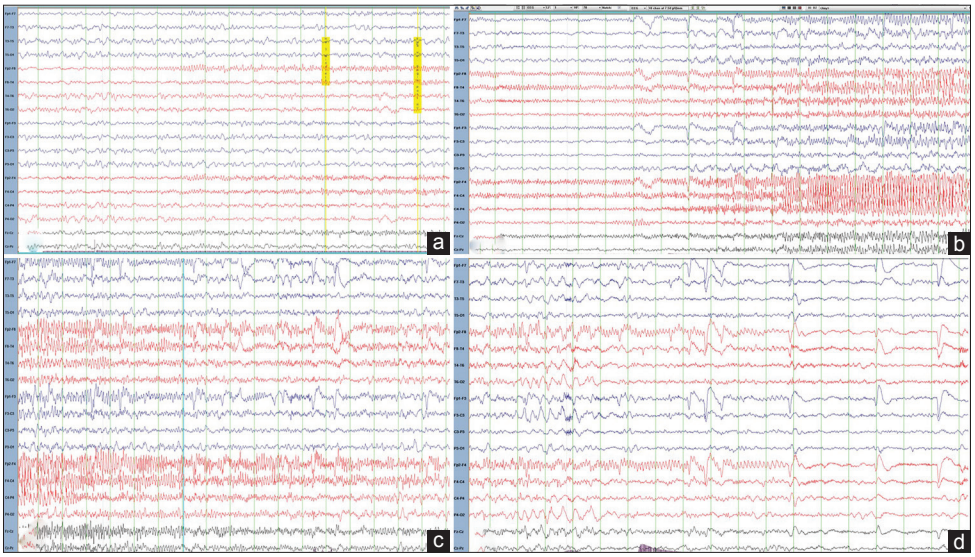

Figure 3: EEG on day 2 of hospitalization: The record shows sudden onset of 25 Hz beta activity at Fp2/F4 which evolves in frequency, location (right hemispheric spread and then left hemisphere involvement), amplitude and morphology (a-d). The seizure lasted for 45 seconds and there was no clinical correlate. Continuous EEG captured multiple similar electrographic seizures (longitudinal bipolar montage with a timebase 30 mm/sec, sensitivity 7 μV/mm, high frequency filter at 70 Hz, low frequency filter at 1 Hz and sampling rate of 256 Hz)

seizures with persistently positive CSF anti-NMDA antibodies. Serial oncological imaging was negative. The relapses were managed with steroids and additional cycles of rituximab.

Seizures are a common presentation in anti-NMDA encephalitis and have been estimated to affect up to 77% of the children.[1,2] The associated EEG patterns described are usually nonspecific. One unique EEG pattern described is EDB. Ikeda *et al*.,[3] described this EEG pattern in a 19-year old woman with anti-NMDA receptor encephalitis with a high seizure burden. Schmitt *et al*.,[4] later described a similar pattern in 30% of the patients with anti-NMDA encephalitis. The pattern was characterized by rhythmic 1–3 Hz delta activity with superimposed 20–30 Hz beta activity.

The presence of EDB on EEG has been associated with a more protracted hospital course, abnormal neuroimaging, and a trend towards a worse outcome at discharge in individuals with anti-NMDA encephalitis.[4] Notably, the resolution of EDB on EEG has been associated with clinical improvement.

The mechanism of EDB is unknown. It has been postulated that in patients with anti-NMDA-receptor encephalitis, the levels of synaptic NMDA-receptor and NMDA-receptor mediated currents are reduced leading to altered rhythmic neuronal activity, which may produce the distinct EEG pattern.[4]

Although EDB has been described in the EEG of individuals with anti-NMDA encephalitis, its specificity is not known.

Recently, EDB has been described among patients with other diagnoses, such as stroke, brain tumor, metabolic disturbances, and hypoxic encephalopathy.[5] Regardless, the presence of EDB on EEG, especially in the context of other clinical symptoms such as behavioral change, seizures, autonomic disturbances and movement disorders should alert the clinician about a possible diagnosis of anti-NMDAR encephalitis. Recognition of this unique EEG pattern may aid in the diagnosis and prevent delays in treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

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