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Tonic status and electrodecremental paroxysms in an adult without epilepsy

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ABSTRACT – Electrodecremental status epilepticus is classically described in infants and children with severe refractory epilepsy, mental retardation, and structural brain abnormalities. We describe a 24-year-old woman who presented to the emergency department with prolonged tonic status epilepticus which evolved into stimulus-induced diffuse voltage attenuation (SIDVA) pattern in the setting of aseptic meningoencephalitis. There was no history of seizures or further events after recovery. This is the first report of a SIDVA pattern in an adult without a history of epilepsy.

Key words: tonic status, electrodecremental, meningoencephalitis, adult, stimulus-induced

Case report

A 24-year-old woman from Central America experienced fever, headaches and vomiting over two days. On the day of admission, her husband found her unresponsive, foaming at the mouth, and making sounds that resembled choking. Her arms and legs were tense and her fists clenched, initially with elbows flexed and then extended. In the emergency department, she was given lorazepam, but had persistent opisthotonic posturing, tonic extension of all extremities, diffuse trembling, and upward gaze deviation. She was intubated and loaded with fosphenytoin. Within five minutes, her limbs stopped shaking, gaze returned to midline, and her tone returned to normal. Total time in sustained tonic status was estimated to be three to four hours. After tonic posturing ceased, examination revealed a temperature of 100.0°F, pulse 142 bpm. Brainstem reflexes were intact, but she was unresponsive to commands with intermittent semi-purposeful movements, such as trying to grab her endotracheal tube. White blood cell (WBC) count was 19,240/cu mm and cerebrospinal fluid (CSF) protein was 83 mg/dL. A lymphocytic predominance was detected with 12 WBCs/cu mm and no red blood cells. Serum glucose was 175 mg/dL and CSF glucose was 105 mg/dL. Brain CT and enhanced MRI were normal. CSF culture and viral PCR tests were negative.

During continued clinical observation in the emergency department, after the three to four hours of sustained tonic status had ceased, she

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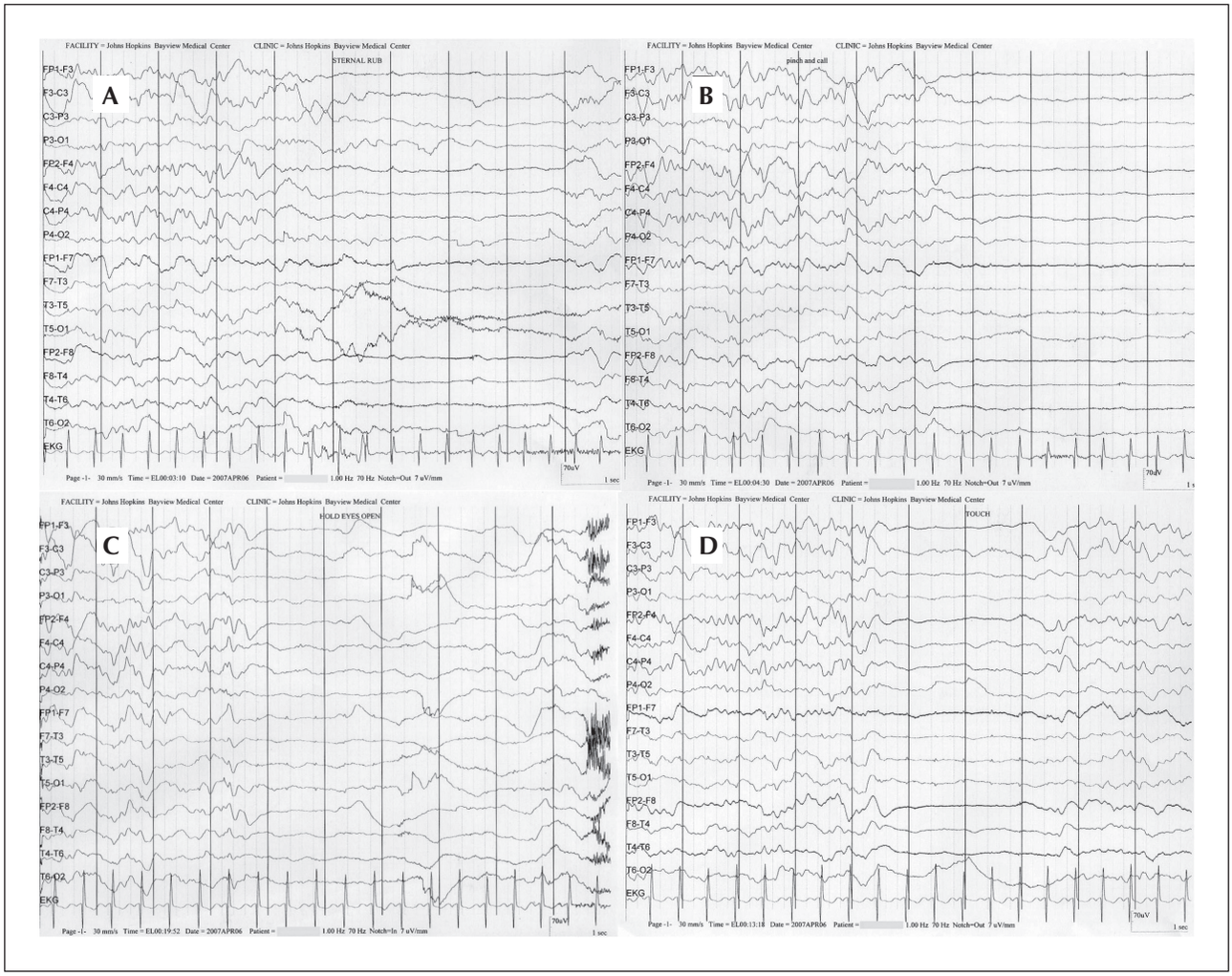

Figure 1. Stimulus-induced diffuse voltage attenuation (SIDVA). The background prior to the voltage attenuation episodes resembled either sleep with delta and superimposed faster alpha/beta activity or diffuse theta/delta activity suggesting diffuse encephalopathy. These are representative frames showing electrographic attenuation in all derivations induced by (**A**) sternal rub, (**B**) pinching the arm and calling the patient's name, (**C**) the examiner holding the patient's eyes open, or (**D**) lightly touching the patient's forearm. In all cases, the beginning of the electrodecrement corresponded to the stimulus.

did not regain consciousness, instead fluctuating between two modes of activity. In one mode, her heart rate was 140-150 bpm, with agitation, limb thrashing, and brisk movement to painful stimuli. In the second mode, which eventually predominated, heart rate was 110-120 bpm and she was still. EEG was initiated in this second mode and showed symmetric high-voltage delta with superimposed alpha frequencies in a pattern suggestive of sleep K complexes. On stimulation by touch, sound, or passive eye opening, she had stereotyped, abrupt, diffuse electrographic attenuation in all derivations to < 5-15 microvolts for 2 to 10 seconds, ending in a spindle-like burst and high-voltage fronto-central delta activity *(figure 1)*. The end of the electrodecremental events correlated with brief episodes of tonic posturing, extension of the arms and legs, retroflexion of the neck, and diffuse trembling for one to two seconds before returning to quiet flaccidity. The episodes of stimulus-induced diffuse voltage attenuation (SIDVA) were elicited by sternal rub, pinching an extremity, lightly touching an arm, holding the eyes open, or by auditory stimuli such as clapping or calling the patient's name. They occurred more than ten times, most strikingly with mild auditory stimulation (softly saying the patient's name in her ear).

In the Neuro-Critical Care Unit she was given phenytoin and levetiracetam, benzodiazepines were avoided due to concern of paradoxical worsening of tonic seizures (Sazgar and Bourgeois, 2005; Dimario and Clancy, 1988). Ceftriaxone, vancomycin, ampicillin, and acyclovir were administered until the results of the CSF studies were provided. Her EEG continued to show a diffuse encephalopathic pattern for the first few days, after which her mental status improved. She was

extubated and discharged without sequelae on levetiracetam and had had no recurrence when seen in the clinic a month later. Mild cognitive slowing was noted at the time of discharge and was unchanged at outpatient follow-up, confirmed by her husband to be her baseline.

Discussion

Stimulus-induced diffuse electrodecremental seizures are classically described in infants and children with severe refractory epilepsies and often with mental retardation (MR) and brain abnormalities (Tibussek *et al.,* 2006; Arroyo *et al.,* 1994). On EEG, periods of voltage attenuation vary in duration, usually lasting less than ten seconds, as in our patient. Tonic semiology is most commonly described in diffuse electrodecremental seizures, although atonic, dystonic, and partial dystonic seizures are also described. In ten adult patients with electrodecremental seizures, all had mild MR, a long history of multiple seizure types, and poor seizure control (Fariello *et al.,* 1979). In this series, seizures tended to cluster, with the abrupt onset of many in a single day, followed by cessation within a week.

To differentiate between an epileptic "diffuse electrodecremental pattern" (DEP) and non-specific arousal reactions, Schmitt and colleagues suggested that DEPs begin synchronously with clinical signs of a seizure (Tibussek *et al.,* 2006). In our patient, brief one to two-second bursts of tonic posturing occurred at the end of each period of EEG flattening, thus it is unclear whether these events were epileptic phenomena. However, electro-clinical dissociation may have occurred, given the initial period of prolonged tonic status of three to four hours prior to these episodes without intercurrent recovery of alertness. Alternatively, SIDVA may occur following status but without concurrent clinical seizure activity, as described in a critically ill 15-month-old child with no prior history of seizures or other neurological abnormalities (Losey and Ng, 2009). This patient presented with status epilepticus and was treated. Her initial EEG showed background slowing. She was hospitalized with critical illness affecting multiple organs requiring intubation and ventilation. A few days later, she had a breakthrough seizure prompting a subsequent EEG which showed SIDVA following sternal rub or painful stimulation applied to the extremities. Like our patient, she had a full recovery without further episodes.

The unique aspects of this case are the occurrence of SIDVA in an adult without epilepsy in the setting of aseptic meningoencephalitis. The patient also made a full recovery. Meningitis or encephalitis was suggested as the aetiology for diffuse electrodecremental seizures in 7/39 patients in one case series (Arroyo *et al.,* 1994), while another series included a four-year-old girl with startle-provoked diffuse electrodecremental seizures in whom the aetiology was presumed to be herpes encephalitis; seizures were elicited by sound, and each episode lasted 10-15 seconds (Tibussek *et al.,* 2006). However, all patients in both series had intractable epilepsy since infancy or childhood, and most patients suffered from severe MR and diffuse cerebral abnormalities.

Many anatomical sites of origin have been suggested to be associated with diffuse electrodecremental events. The prevailing theory is that they arise from very high frequency cortical discharges (Arroyo *et al.,* 1994). Hence, either a focal or diffuse cortical process such as meningitis or meningoencephalitis may trigger SIDVA in a susceptible individual, such as those with MR. Our patient's mild cognitive slowing may be indicative of an underlying cortical susceptibility. The only previously published case, a 15-month-old child with SIDVA during critical illness and subsequent full recovery (Losey and Ng, 2009), suggests this may be a rare occurrence. However, such striking electrographic phenomena may frequently go unrecognized when they are not associated with clinical seizure symptoms, and therefore may not be captured on EEG. If SIDVA does indeed arise from a combination of mild diffuse brain damage (*e.g.* MR), acutely triggered by meningitis or encephalitis, it may provide insight into the mechanisms of more severe intractable epilepsies associated with diffuse voltage attenuation patterns. □

Disclosure.
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