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Clinical Observations

Post-Anoxic Reticular Reflex Myoclonus in a Child and Proposed Classification of Post-Anoxic Myoclonus

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ABSTRACT

OBJECTIVE: We describe a child with post-anoxic myoclonus of the reticular reflex type and discuss the classification of post-anoxic myoclonus. **PATIENT DESCRIPTION:** A nine-year-old boy with severe hypoxic-ischemic encephalopathy due to submersion developed early epileptic spasms followed by stimulus sensitive multifocal generalized myoclonus and later dystonia. Video electromyography (EMG) polygraphy performed before treatment demonstrated that the discharges associated with the myoclonus lasted less than 50 milliseconds. Cortical myoclonus was excluded by jerk-locked averaging using arm muscles, which showed no cortical correlates. The recruitment order on EMG polygraphy was consistent with a brainstem generator for the myoclonus, characteristic of reticular reflex myoclonus. Both myoclonus and dystonia responded to clonazepam. He remains in a persistent vegetative state. **CONCLUSIONS:** Reticular reflex myoclonus can be demonstrated by detailed neurophysiological assessment in children as in adults, and it has a similar poor prognosis in children. Post-anoxic myoclonus can have several mechanisms and should not be considered synonymous with Lance-Adams myoclonus.

Keywords: post-anoxic myoclonus, reticular reflex myoclonus, Lance-Adams syndrome, myoclonus

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Introduction

Survivors of anoxic brain injury can develop myoclonus. The incidence of post-anoxic myoclonus (PAM) is unknown but it is considered rare.[1] Lance and Adams[2] first described four patients with intention myoclonus associated with post-anoxic encephalopathy in 1963. In 2000 Hallett[1] classified PAM into acute and chronic forms with the Lance-Adams -type as a chronic form. Most reported patients have been adults and different myoclonus types can coexist in a given individual at the same time.[3] A very rare form of PAM is reticular reflex myoclonus (RRM). Only four individuals with post-anoxic RRM have been documented in detail, and all were adults.[3-6] A neonate reported by Kakisaka et al.[7] had RRM secondary to presumed prenatal hypoxic insult, but the authors did not provide the electrophysiological evidence required to demonstrate the generator and the characteristic electromyographic finding of the myoclonus (i.e., very short durations) of this extremely rare form of myoclonus. The presentation, response to treatment, and prognosis differ between types of PAM.[8,9]

We describe a child with post-anoxic RRM whose myoclonus responded initially to clonazepam despite a poor overall outcome. We demonstrate RRM with electrophysiological evidence.

Patient Description

Clinical details

This previously healthy nine-year-old boy developed severe hypoxic-ischemic encephalopathy and transient multiorgan injury following ten minutes of cold water immersion and subsequent resuscitation for 66 minutes before return of spontaneous circulation. He then received

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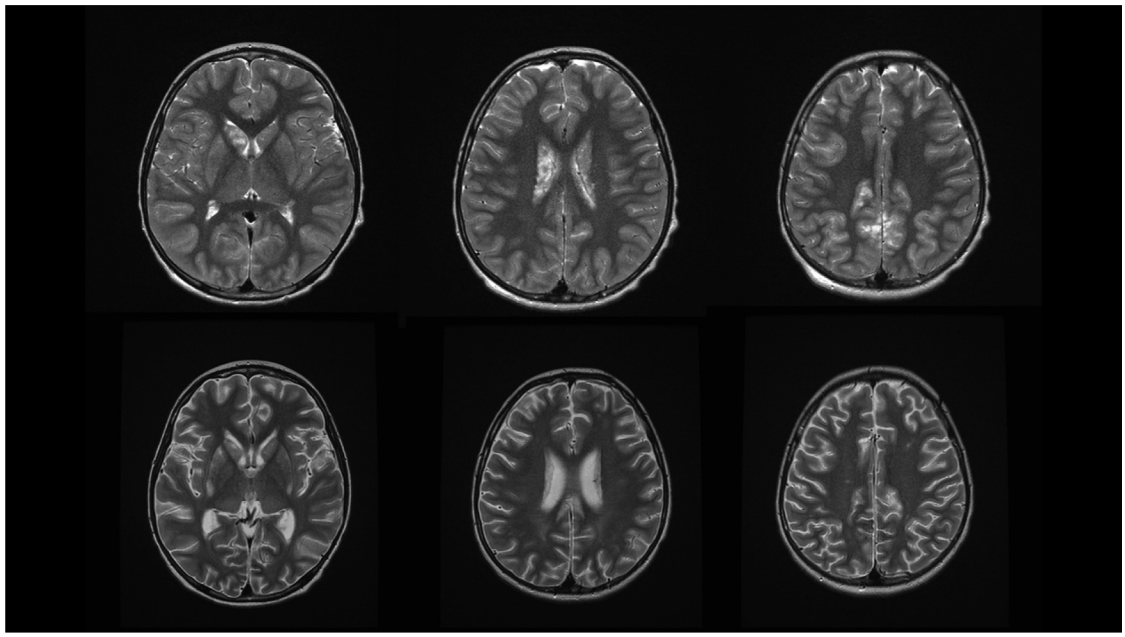

FIGURE 1.
Axial T2-weighted brain MRI images. Top row: Imaging at presentation showing focal high signal change at presentation involving corpus striatum bilaterally and parafalcine parietal lobes bilaterally. Bottom row: Day 20 imaging showing volume loss with gliosis in the affected areas. MRI, magnetic resonance imaging.

full intensive care, with therapeutic hypothermia for 24 hours. His initial Glasgow Coma Scale was 3. On day three he developed spasms, which were treated with phenytoin and midazolam infusion. His electroencephalogram (EEG) showed bursts of spikes, polyspikes, and sharp waves interspersed with periods of relative suppression. Magnetic resonance imaging (MRI) showed focal signal changes involving the corpus striatum bilaterally, hippocampi, and cortex of the parietal and occipital lobes bilaterally (Fig 1).

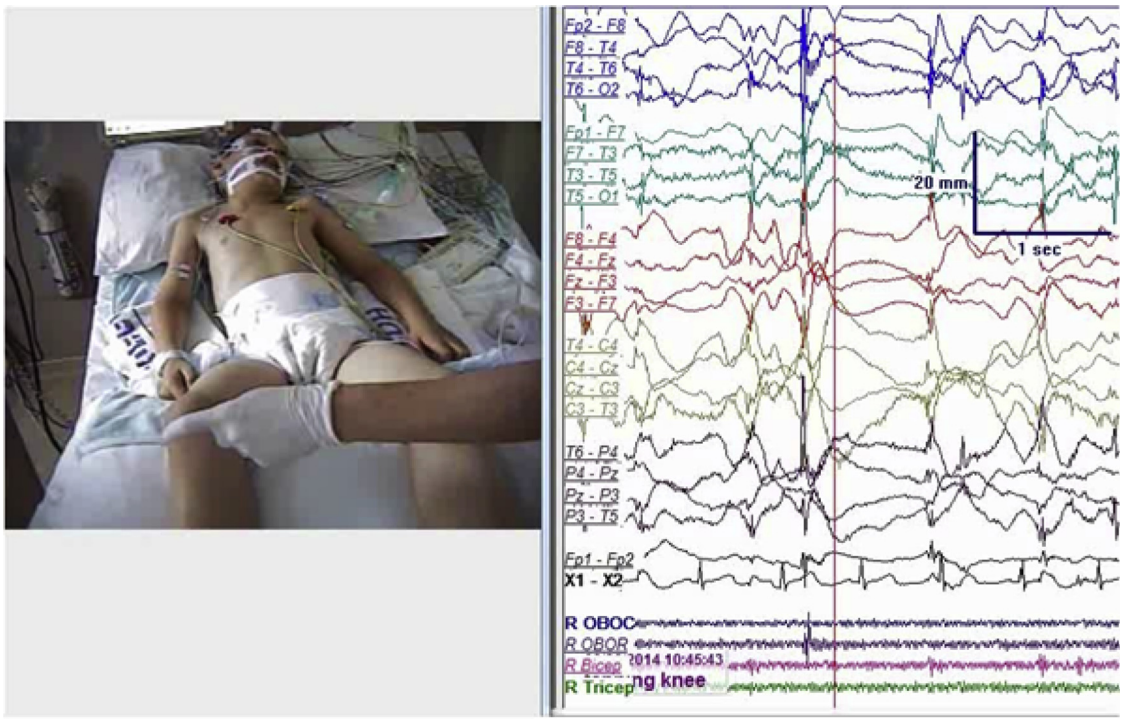

FIGURE 2.
The video EEG with limited EMG leads from day 14 demonstrates muscle stretch in all limbs, sternal tapping, and forehead tapping often induced a generalized myoclonic response. Another video EEG and EMG polygraphy with subsequent change in sweep speed (250 milliseconds/division) from day 22. The EMG polygraphy shows a recruitment order that reveals a consistent lower brainstem lead (sternocleidomastoid and trapezius muscles are recruited first) of the myoclonus with subsequent fast rostral and caudal spread. The EMG discharges are of very short duration, less than 50 milliseconds. The constellation of the electrophysiological findings is consistent with RRM. X1-X2 refers to single channel electrocardiogram. EEG, electroencephalogram; EMG, electromyogram; RRM, reticular reflex myoclonus. The video related to this figure can be found at http://dx.doi.org/10.1016/j.pediatrneurol.2016.12.014. (The color version of this figure is available in the online edition.)