Scotosensitive myoclonic seizures in MERRF

Figure EEG revealing eye-closure-induced bioccipital spike-and-wave discharges

Eye-closure-induced bioccipital spike-and-slow wave activity at approximately 10 Hz (A) was frequently associated with bilateral arm myoclonus. Attempting to close eyes against enough resistance to keep them open did not elicit such a pattern (B, arrow), whereas preventing the light from entering the eyes, with the eyes still open, triggered the ictal discharge (C, arrow).

Supplemental data at
www.neurology.org

A 42-year-old woman presented with a history of intractable eye-closure-sensitive myoclonic and grand mal seizures since age 22, resulting in falls and fractures. She also had five episodes of status epilepticus. She denied visual phenomena, eyelid myoclonus, and absence seizures. She had normal cognitive function, but proximal muscle weakness of all extremities. Her EEG revealed eye-closure-induced bioccipital spike-and-wave discharges. These were scotosensitive,¹ i.e., induced by lack of visual input, but not by eyelid closure (figure and video [on the *Neurology*[®] Web site at www.neurology.org]). Intractability, late age at onset, and complications are unusual features of eye-closure-induced seizures, and the concomitant muscle weakness suggested mitochondrial disease.² Mitochondrial DNA analysis identified the myoclonic epilepsy associated with ragged red fibers—associated A8296G mutation in the tRNA(Lys) gene.

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