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, 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. Mitochondrial DNA analysis identified the myoclonic epilepsy associated with ragged red fibers–associated A8296G mutation in the tRNA(Lys) gene.

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