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.

Mohamad Z. Koubeissi, MD, Chaiyos Khongkhaitum, MD, Annette I. Janus, MD, and Hans Lüders, MD, PhD, Cleveland, OH

Disclosure: The authors report no disclosures.

Address correspondence and reprint requests to Dr. Mohamad Z. Koubeissi, Department of Neurology, University Hospitals Case Medical Center, Case Western Reserve University, 11100 Euclid Avenue, Cleveland, OH 44106-5040; mohamad.koubeissi@uhhospitals.org

ACKNOWLEDGMENT

The authors thank Maha Haddad and Doris Evans for assistance in preparing the video.

REFERENCES

Scotosensitive myoclonic seizures in MERRF
Neurology 2009;72:858
DOI 10.1212/01.wnl.0000343959.15205.52

This information is current as of March 2, 2009

Updated Information & Services
including high resolution figures, can be found at:
http://www.neurology.org/content/72/9/858.full.html

Supplementary Material
Supplementary material can be found at:
http://www.neurology.org/content/suppl/2009/03/02/72.9.858.DC1.htm1

References
This article cites 2 articles, 0 of which you can access for free at:
http://www.neurology.org/content/72/9/858.full.html##ref-list-1

Subspecialty Collections
This article, along with others on similar topics, appears in the following collection(s):
EEG
http://www.neurology.org/cgi/collection/eeg_
Epilepsy semiology
http://www.neurology.org/cgi/collection/epilepsy_semiology
Muscle disease
http://www.neurology.org/cgi/collection/muscle_disease
Myoclonus; see Movement Disorders/myoclonus
http://www.neurology.org/cgi/collection/myoclonus_see_movement_disorders-myoclonus

Permissions & Licensing
Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:
http://www.neurology.org/misc/about.xhtml#permissions

Reprints
Information about ordering reprints can be found online:
http://www.neurology.org/misc/addir.xhtml#reprintsus

Neurology © is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright . All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.