Fulminant cerebral demyelination in neuromyelitis optica

A 43-year-old woman with a 2-year history of neuromyelitis optica (NMO; typical imaging [figure 1] and NMO–immunoglobulin G [IgG] antibody), previously treated with plasmapheresis and steroids, presented somnolent. Admission neuroimaging showed fulminant cerebral demyelination (figure 2, A and B), and she rapidly progressed to herniation (figure 2, C and D) and brain death despite 2 courses of plasma exchange, mannitol, hypertonic saline, and methylprednisolone.

NMO-IgG–positive demyelinating disease may include fulminant edematous cerebral demyelination.1 The mechanism may involve deficient clearance of vasogenic edema via the aquaporin-4 water channel.2 Early evaluation of NMO-IgG in atypical demyelinating cases may permit early aggressive treatment with steroids, plasmapheresis, or chemo-immunosuppression.

Christopher R. Newey, DO, MS, Robert A. Bermel, MD, Cleveland, OH

Author contributions: Dr. Newey contributed equally to the writing of the case and formatting the images. Dr. Bermel contributed equally to the writing of the case and formatting the images.

Disclosure: Dr. Newey reports no disclosures. Dr. Bermel serves as a consultant and on the speakers’ bureaus for Biogen Idec and Teva Pharmaceutical Industries Ltd. and receives research support from the National Multiple Sclerosis Society.

Address correspondence and reprint requests to Dr. Christopher R. Newey, Department of Neurology, Cleveland Clinic Foundation, 9500 Euclid Avenue, Cleveland, OH 44195; neweyc@ccf.org


Fulminant cerebral demyelination in neuromyelitis optica
Christopher R. Newey and Robert A. Bermel

Neurology 2011;77;193
DOI 10.1212/WNL.0b013e3182242d6e

This information is current as of July 11, 2011

Updated Information & Services
Including high resolution figures, can be found at:
http://n.neurology.org/content/77/2/193.full

References
This article cites 2 articles, 1 of which you can access for free at:
http://n.neurology.org/content/77/2/193.full#ref-list-1

Subspecialty Collections
This article, along with others on similar topics, appears in the following collection(s):
All Demyelinating disease (CNS)
http://n.neurology.org/cgi/collection/all_demyelinating_disease_cns
CT
http://n.neurology.org/cgi/collection/ct
Devic's syndrome
http://n.neurology.org/cgi/collection/devics_syndrome
MRI
http://n.neurology.org/cgi/collection/mri

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

Reprints
Information about ordering reprints can be found online:
http://n.neurology.org/subscribers/advertise