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. The mechanism may involve deficient clearance of vasogenic edema via the aquaporin-4 water channel. Early evaluation of NMO-IgG in atypical demyelinating cases may permit early aggressive treatment with steroids, plasmapheresis, or chemo-immunosuppression.

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