Fulminant cerebral demyelination in neuromyelitis optica

Figure 1 MRI at diagnosis of neuromyelitis optica 5 months prior to herniation

(A) Axial fluid-attenuated inversion recovery image shows nonspecific T2 hyperintensities. (B) Cervical sagittal short T1 inversion recovery shows longitudinally extensive myelitis extending into the thoracic cord.

Figure 2 Imaging of fulminant cerebral edema and demyelination

(A) Axial T1-weighted postcontrast image shows multiple ring-enhancing demyelinating lesions bilaterally. (B) Axial T2-weighted image shows extensive vasogenic edema. (C) Diffuse vasogenic edema on head CT (C) with loss of ambient cisterns and uncal herniation (D).

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.

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