Neuromyelitis optica spectrum disorder

- Closely related severe demyelinating diseases caused by an autoantibody to the aquaporin-4 water channel.
- Classic presentation of NMO is with the triad of optic neuritis, longitudinally extensive myelitis, and positive anti-AQP4 antibody
  - although a far wider range of manifestations are now recognized as part of NMOSD
- Typically found in patients older than those with multiple sclerosis (MS), average age of presentation of 41.
- Even stronger female predilection (F:M 6.5:1).
- It is found more frequently in patients of Asian, Indian, and African descent

Brain

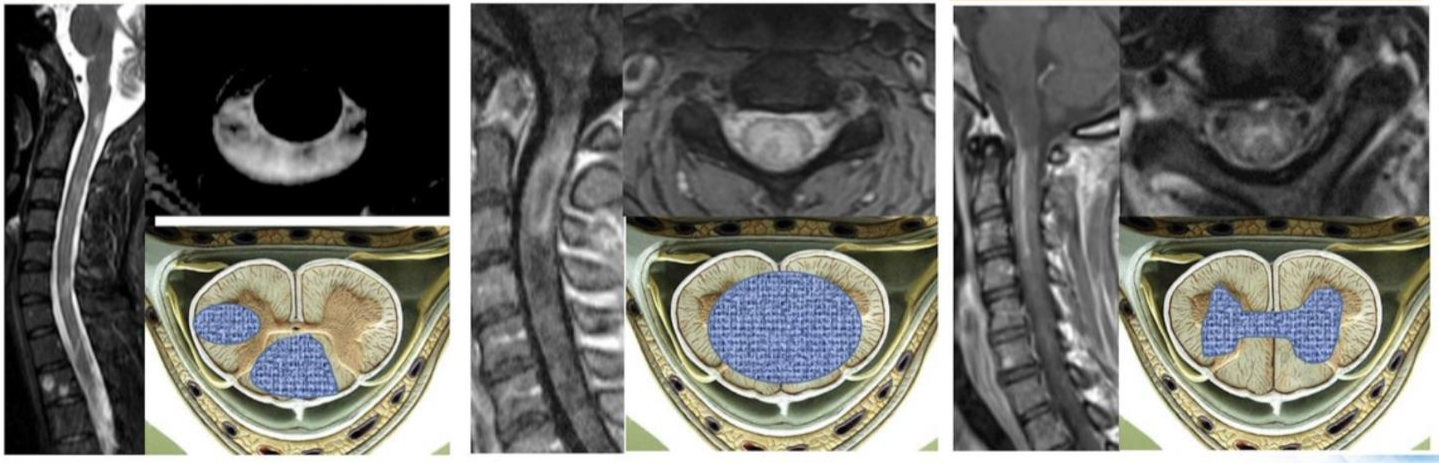
- Lesions which mirror the distribution of aquaporin 4 in the brain,
  - Particularly found in the periependymal regions abutting the ventricles
- Periventricular (hemispheric) confluent smooth sessile white matter involvement (unlike MS, there are usually no Dawson's fingers)
- Periaqueductal grey matter
- Hypothalamus/medial thalamus
- Dorsal pons/medulla
- Corpus callosum
  - multiple callosal lesions with heterogeneous signal leading to a marbled pattern
  - the splenium may be diffusely involved and expanded
- Deep (or less frequently subcortical) punctate white matter lesions (which may appear like those seen in MS)
- Larger >3 cm diameter hemispheric white matter lesions

Pathophysiology	- Chronic CNS inflammation with antibodies against AQP4 channels
Epidemiology	- Prevalence: 0.5 – 4.4/100 000 - Ratio female/male: up to 9:1
Clinical features	- Optic neuritis, acute myelitis, area postrema syndrome, acute brain stem, or diencephalic syndrome
Diagnosis	- Combination of clinical and/or MRI in patients with and without AQP4 antibodies - Differential diagnoses: MOG-ab disease, other causes of LETM or optic neuritis (e.g., MS, ADEM, sarcoidosis)
Treatment	- Acute relapses: high-dose intravenous steroids and plasmapheresis - B-cell depleting (Rituximab) or other immunosuppressive medication
Prognosis	- Severely disabling CNS attacks may occur with incomplete recovery - The rise of modern treatment strategies may improve outcomes
When to test for AQP4 antibodies?	- In patients with LETM or extensive optic neuritis - In patients with atypical brain or brainstem lesions

Biomarkers and Imaging Findings in Myelitis

Majda M Thurnher, MD, EDiNR

Multiple Sclerosis (MS)	Neuromyelitis Optica Spectrum Disorder (NMOSD)	Myelin Oligodendrocyte Glycoprotein-associated disease (MOGAD)



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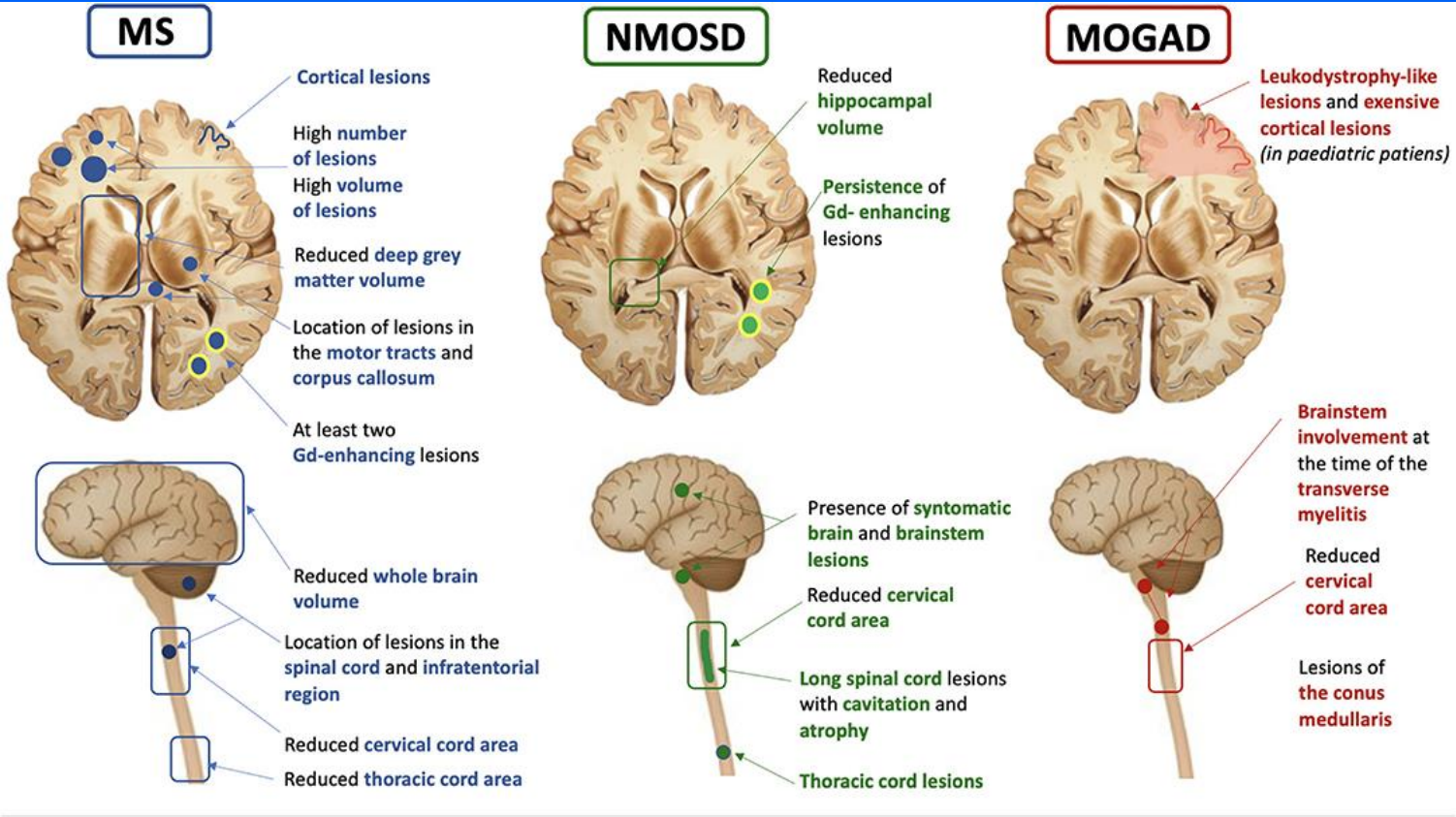
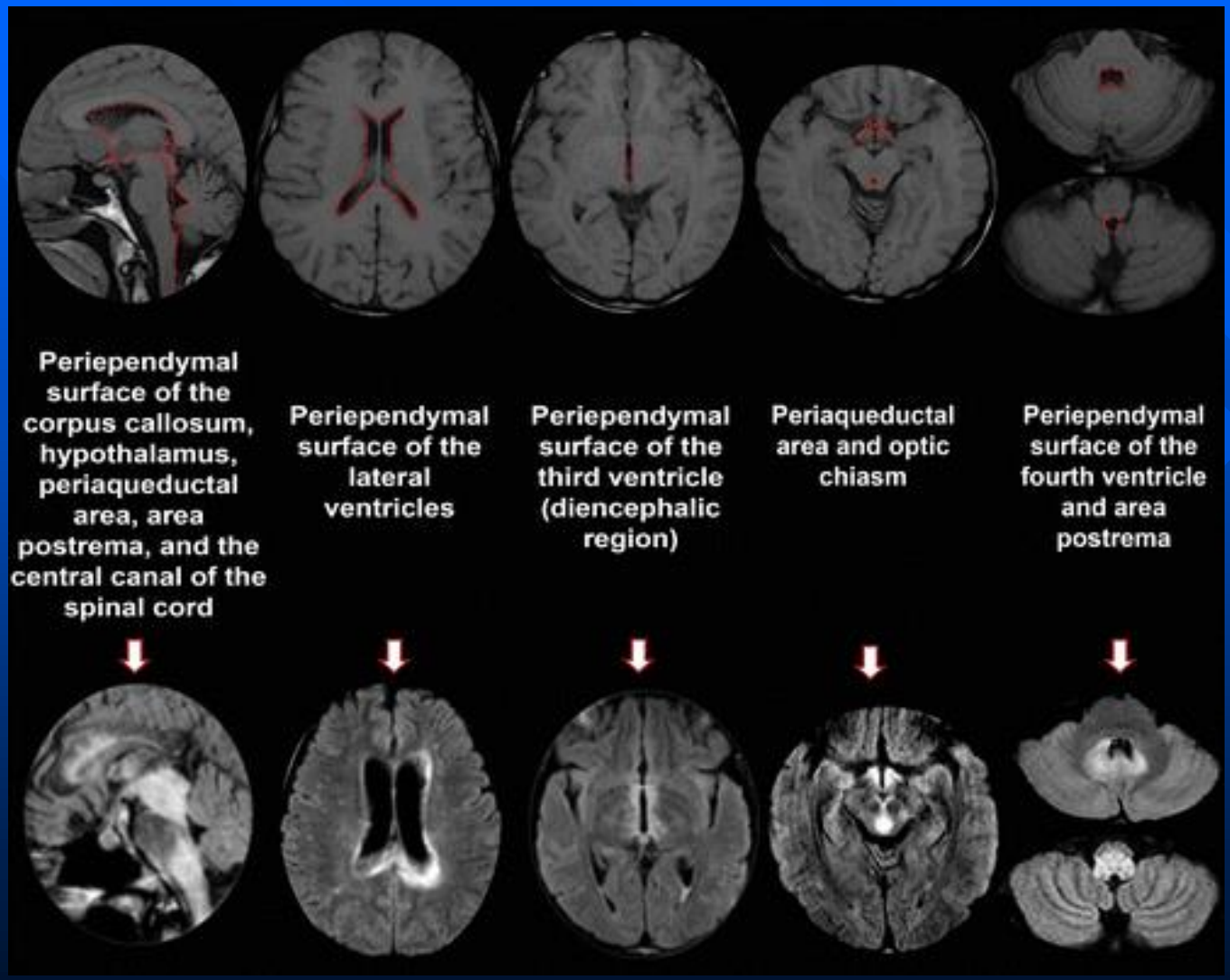
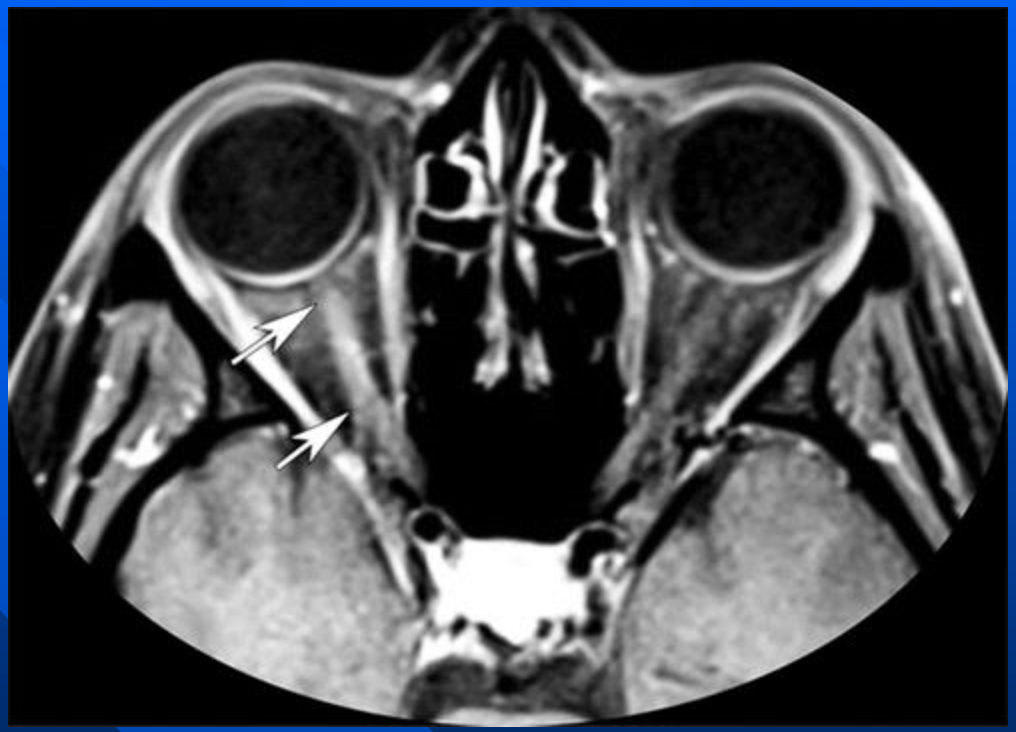
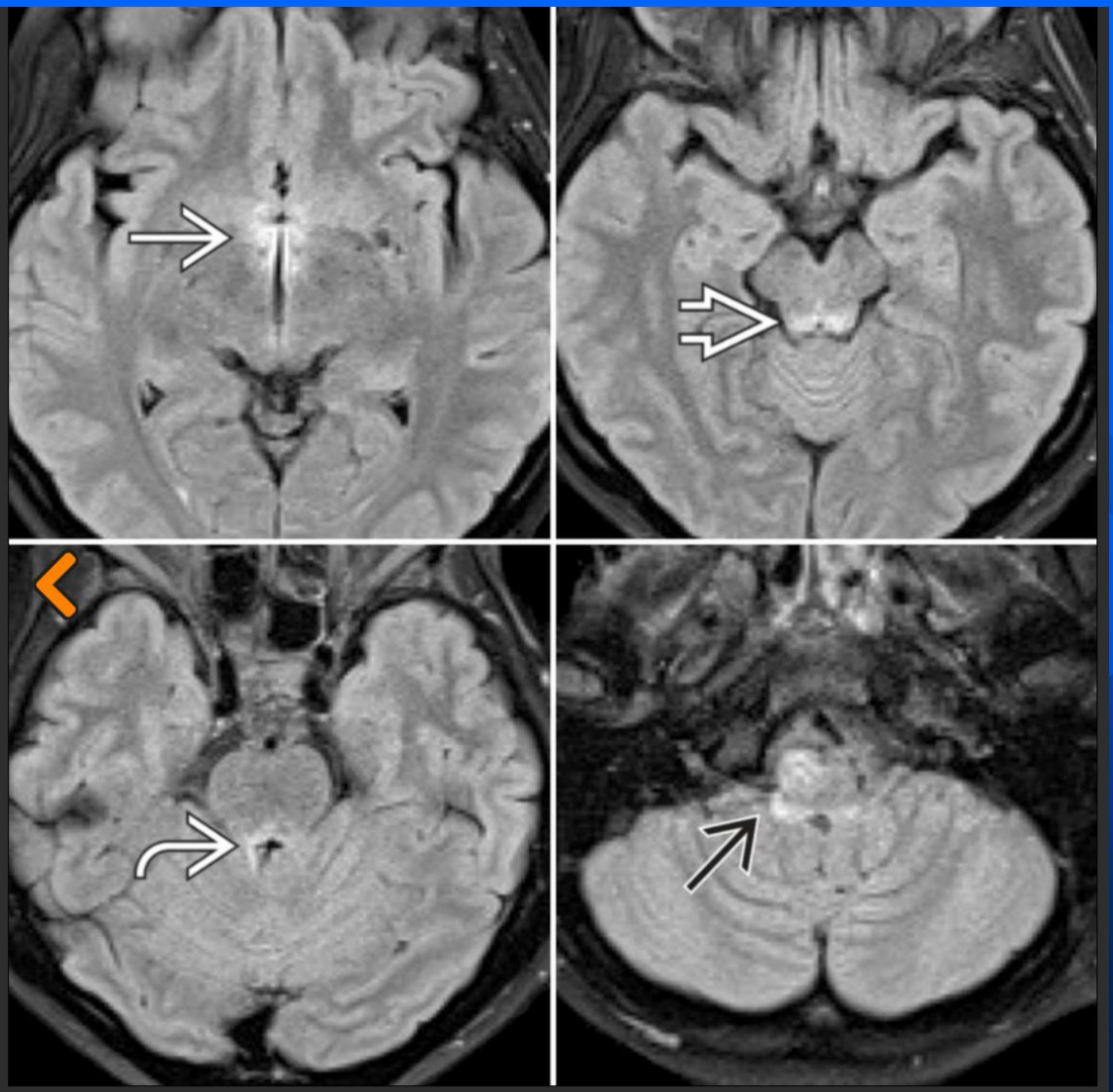
CHICAGO
TRANSFORMING THE FUTURE OF NEURORADIOLOGY


MS
Cortical lesions
High number of lesions
High volume of lesions
Reduced deep grey matter volume
Location of lesions in the motor tracts and corpus callosum
At least two Gd-enhancing lesions
Reduced whole brain volume
Location of lesions in the spinal cord and infratentorial region
Reduced cervical cord area
Reduced thoracic cord area
NMOSD
Reduced hippocampal volume
Persistence of Gd- enhancing lesions
Presence of syntomatic brain and brainstem lesions
Reduced cervical cord area
Long spinal cord lesions with cavitation and atrophy
Thoracic cord lesions
MOGAD
Leukodystrophy-like lesions and exensive cortical lesions (in paediatric patiens)
Brainstem involvement at the time of the transverse myelitis
Reduced cervical cord area
Lesions of the conus medullaris


Periependymal surface of the corpus callosum, hypothalamus, periaqueductal area, area postrema, and the central canal of the spinal cord
Periependymal surface of the lateral ventricles
Periependymal surface of the third ventricle (diencephalic region)
Periaqueductal area and optic chiasm
Periependymal surface of the fourth ventricle and area postrema





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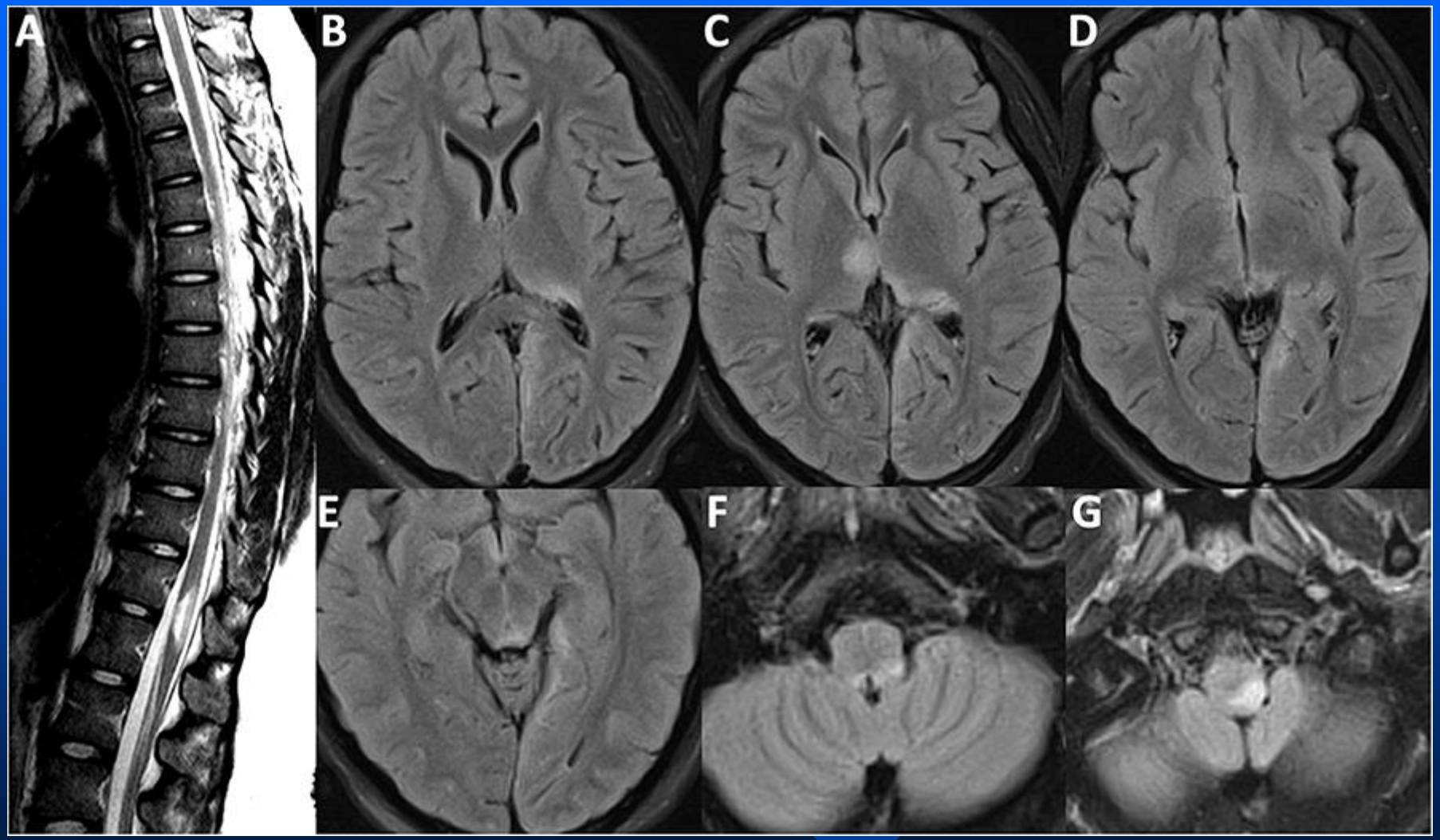
Axial FLAIR MR images show the typical distribution pattern of brain lesions in NMOSD, including the periependymal 3rd ventricle →, adjacent to the aqueduct ⇨, the periependymal 4th ventricle, ↗ and the medulla ⇨.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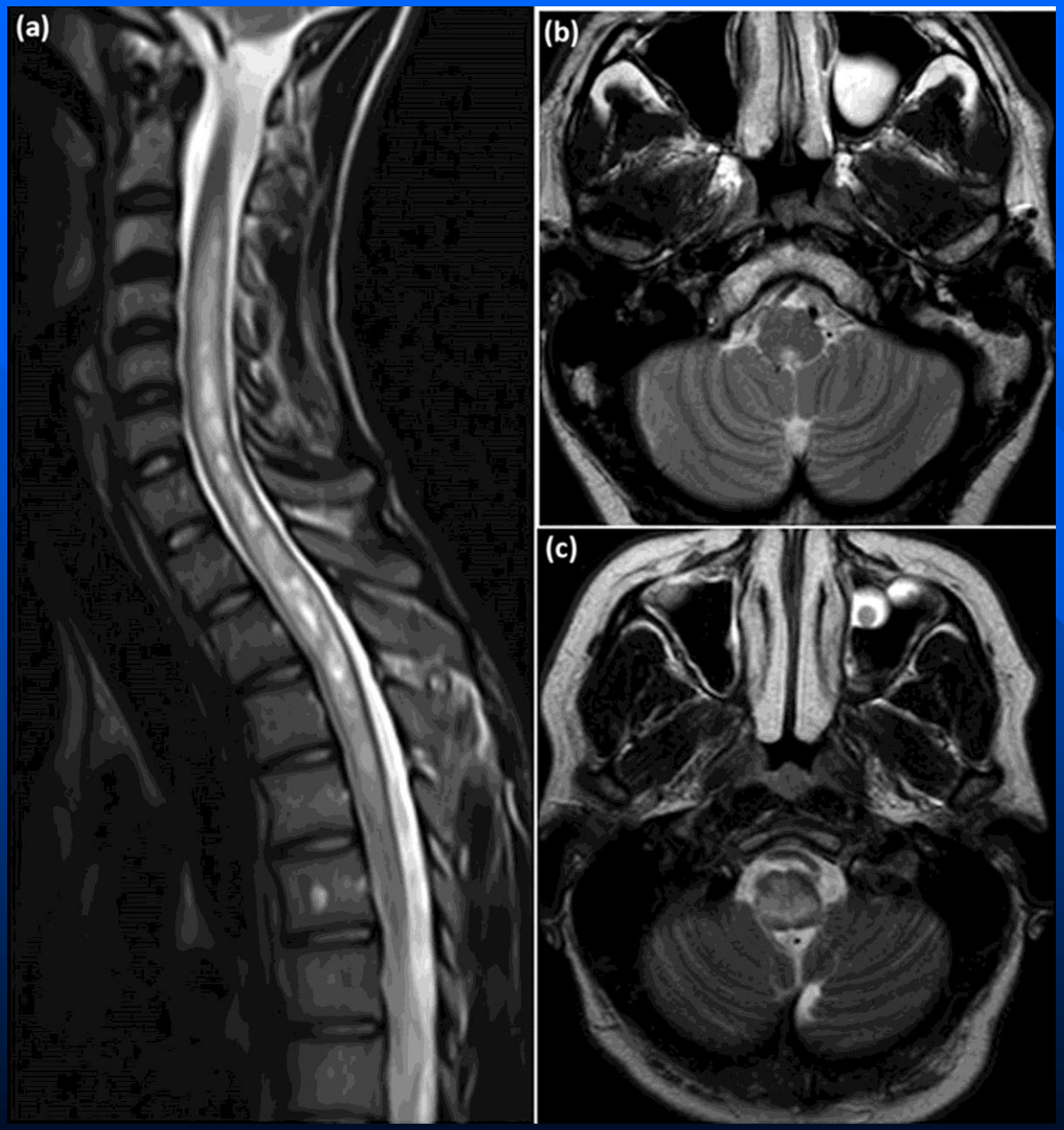
(a)
(b)
(c)