Neuromielite Óptica *Spectrum Disorders* (NMOSD) - Síndrome de Devic

Doença desmielinizante inflamatória idiopática recorrente, caracteristicamente envolvendo nervos ópticos e medula espinhal, possível em crianças e adultos.

- ✓ Não é considerada uma variação da esclerose múltipla, mas sim **outra entidade clínica.**
- ✓ Lesões cerebrais geralmente restritas a uma distribuição geográfica particular: órgãos circunventriculares.
- ✓ Subst. branca cerebral relativamente poupada.
- ✓ Anticorpos anti-aquaporina 4: NMO-IgG (S 75%, E 90%).
- ✓ Dentre os pacientes que não tem NMO-IgG+, boa parte tem anti-MOG IgG, com curso clínico geralmente menos grave.
- ✓ Pode estar associada a outras manifestações autoimunes, como lúpus e síndrome de Sjögren.

Critérios diagnósticos

Table 1: 2015 IPND Criteria for NMOSD

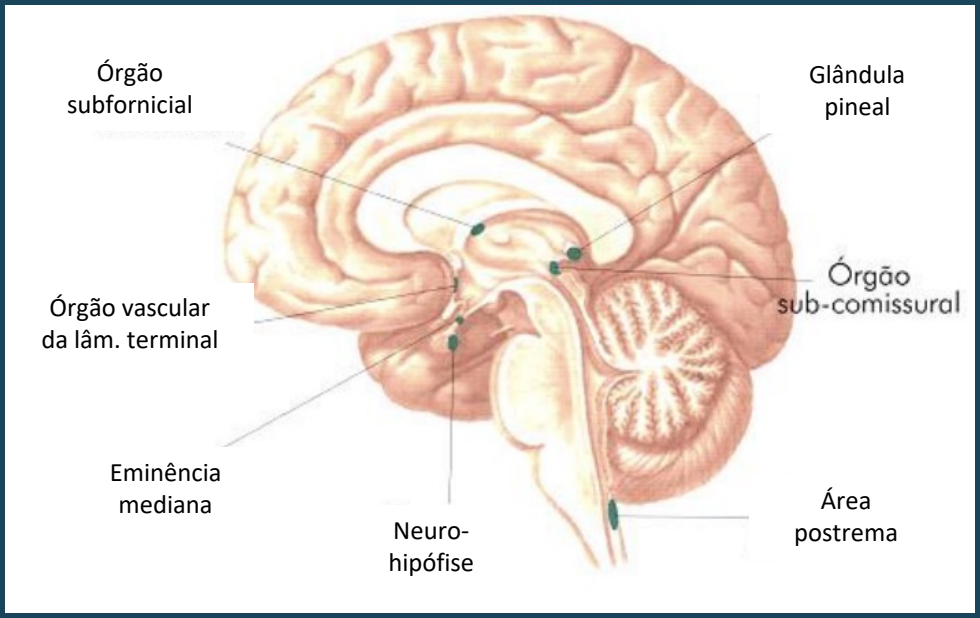
Diagnostic criteria for patients with AQP4-IgG–positive status
At least one core clinical characteristic
AQP4-IgG–positive status
Exclusion of alternative diagnoses
Diagnostic criteria for patients with AQP4-IgG–negative or unknown status
At least two core clinical characteristics occurring as a result of one or more clinical attacks and all of the following requirements:
At least one core clinical characteristic must be optic neuritis, acute myelitis with LETM, or area postrema syndrome
Dissemination in space (two or more different core clinical characteristics)
MR imaging requirements, as applicable
AQP4-IgG–negative status or testing unavailable
Exclusion of alternative diagnoses
Core clinical characteristics
Optic neuritis; acute myelitis; area postrema syndrome (hiccups; nausea and vomiting); acute brainstem syndrome; symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MR imaging lesions; symptomatic cerebral syndrome with NMOSD-typical brain lesions
MR imaging requirements for patients with AQP4-IgG–negative or unknown status
Acute optic neuritis (requires one of the following):
Brain MR imaging showing normal findings or only nonspecific white matter lesions
Optic nerve MR imaging showing a T2-hyperintense lesion or a T1 gadolinium-enhancing lesion extending over more than one-half of the optic nerve length or involving the optic chiasm
Acute myelitis (requires one of the following):
Lesion extending over three or more contiguous segments (LETM)
Spinal cord atrophy involving three or more contiguous segments
Area postrema syndrome
Dorsal medulla/area postrema lesion
Acute brainstem syndrome
Periependymal brainstem lesions

Source.—Reference 5.
Note.—IPND = International Panel for NMO Diagnosis, LETM = longitudinally extensive transverse myelitis.

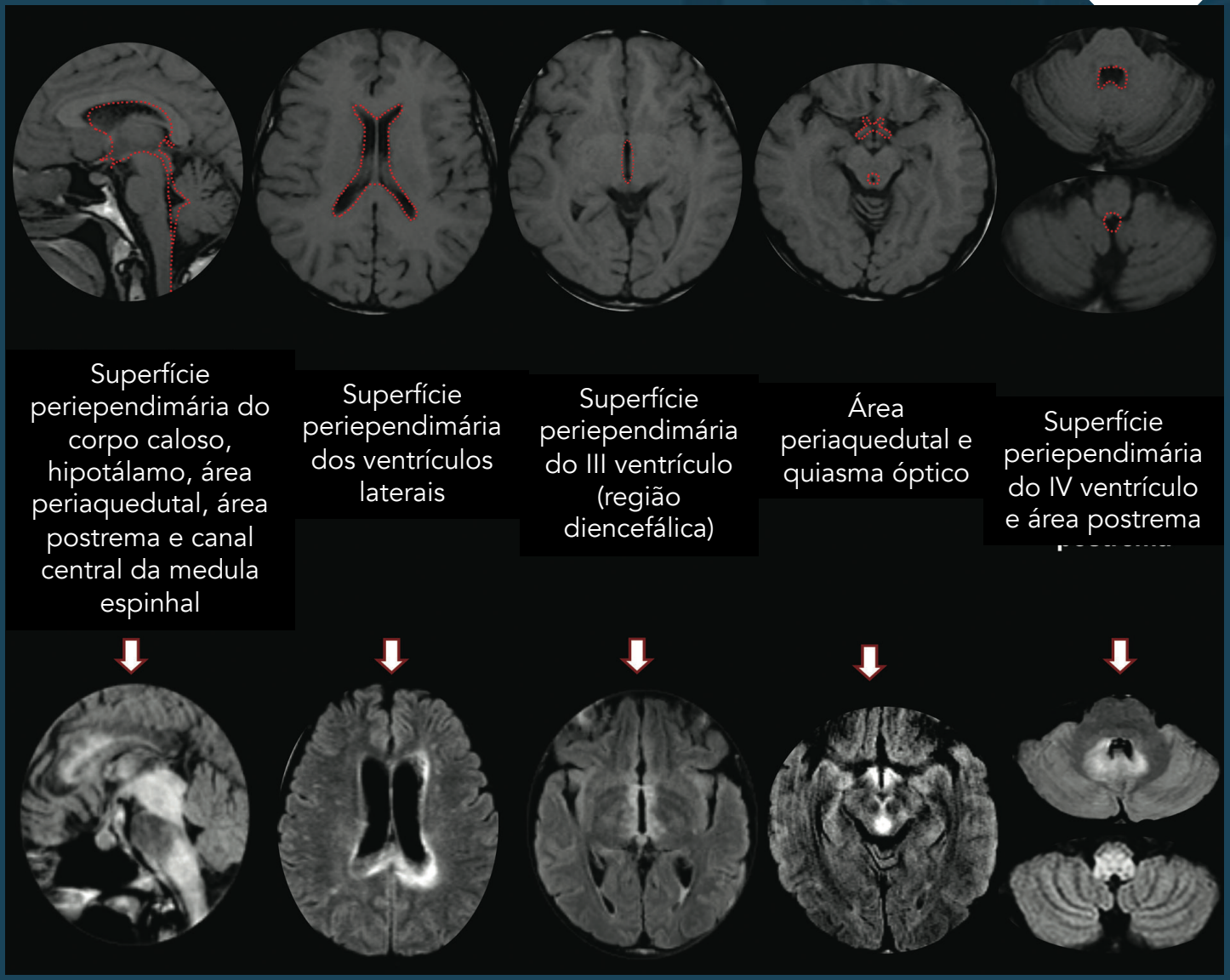
Órgãos circunventriculares

Áreas cerebrais sem barreira hematoencefálica, que permitem que o cérebro monitore mudanças homeostáticas na circulação sistêmica:

- ✓ neuro-hipófise
- ✓ glândula pineal
- ✓ órgão subcomissural
- ✓ eminência mediana
- ✓ órgão subfornicial
- ✓ órgão vascular
- ✓ área postrema



Lesões cerebrais típicas



Superfície periependimária do corpo caloso, hipotálamo, área periaquedutal, área postrema e canal central da medula espinhal

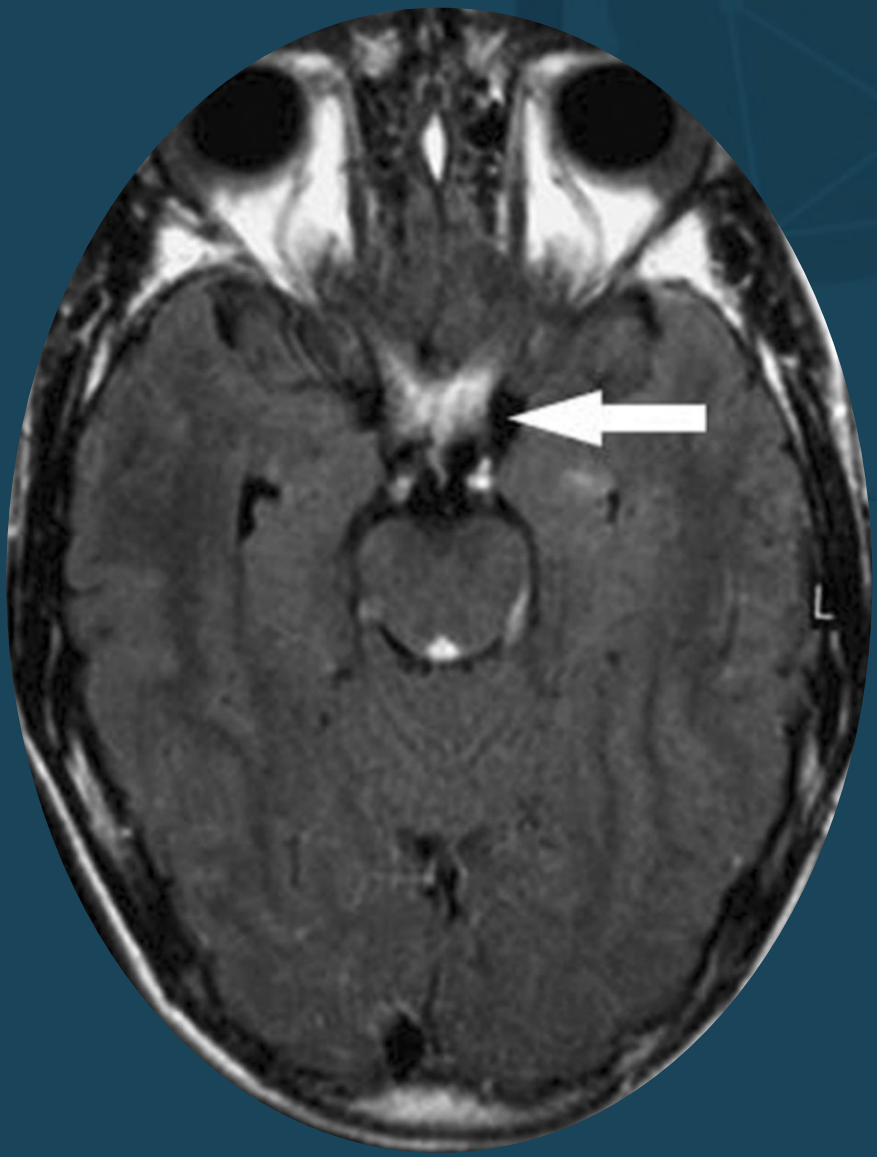
Superfície periependimária dos ventrículos laterais

Superfície periependimária do III ventrículo (região diencefálica)

Área periaquedutal e quiasma óptico

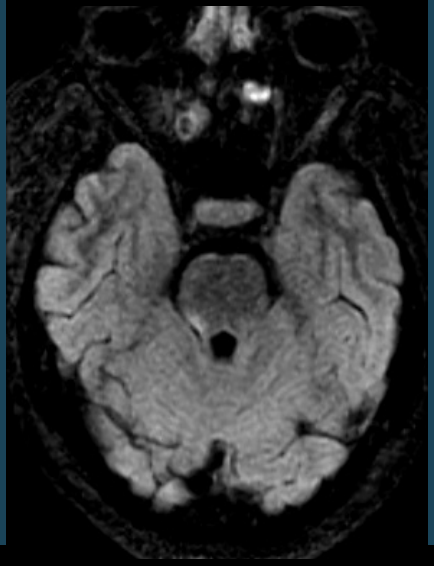
Superfície periependimária do IV ventrículo e área postrema

Neurite óptica: tipicamente bilateral

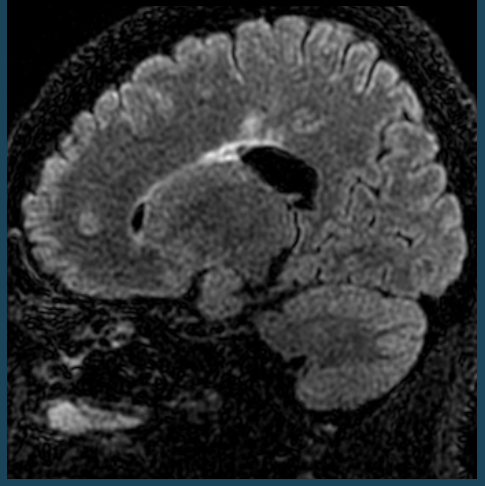


Lesões encefálicas e mielite longitudinalmente extensa (extensão de > 3 corpos vertebrais)

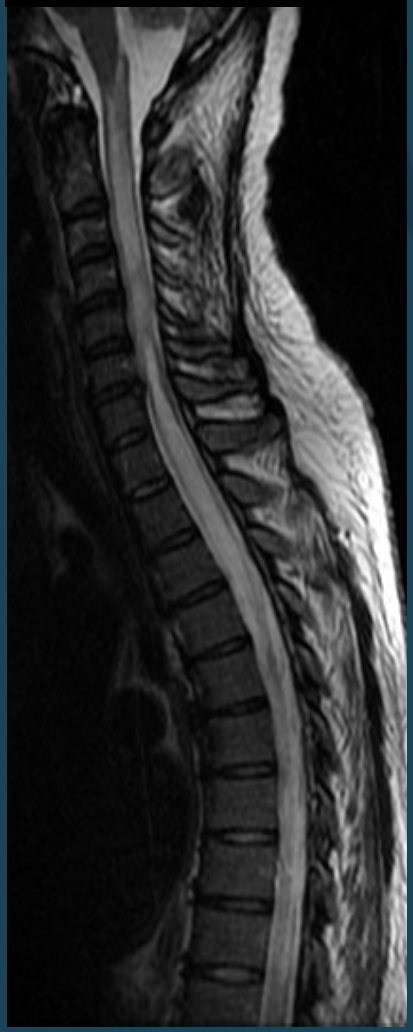
Axial FLAIR



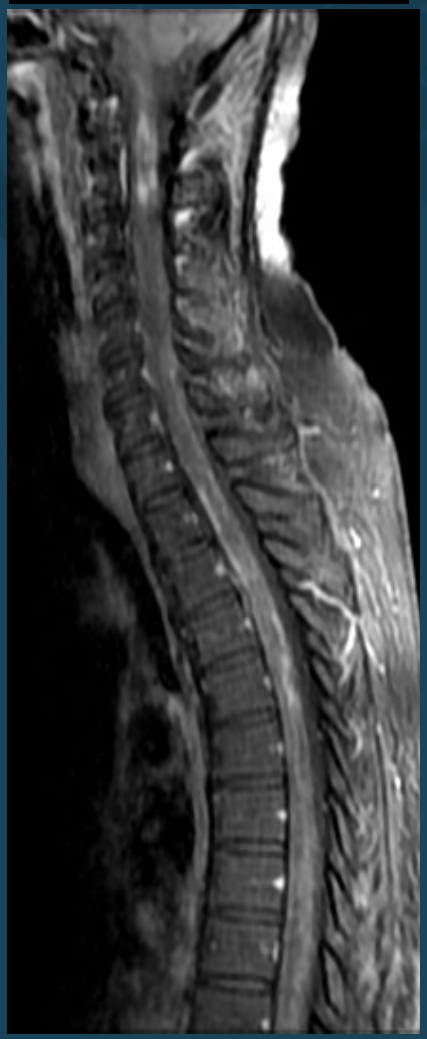
Sagital FLAIR



Sagital T2



Sagital T1-Gd



“Bright spotty lesions”

Research Paper

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“Bright spotty lesions” on spinal magnetic resonance imaging differentiate neuromyelitis optica from multiple sclerosis

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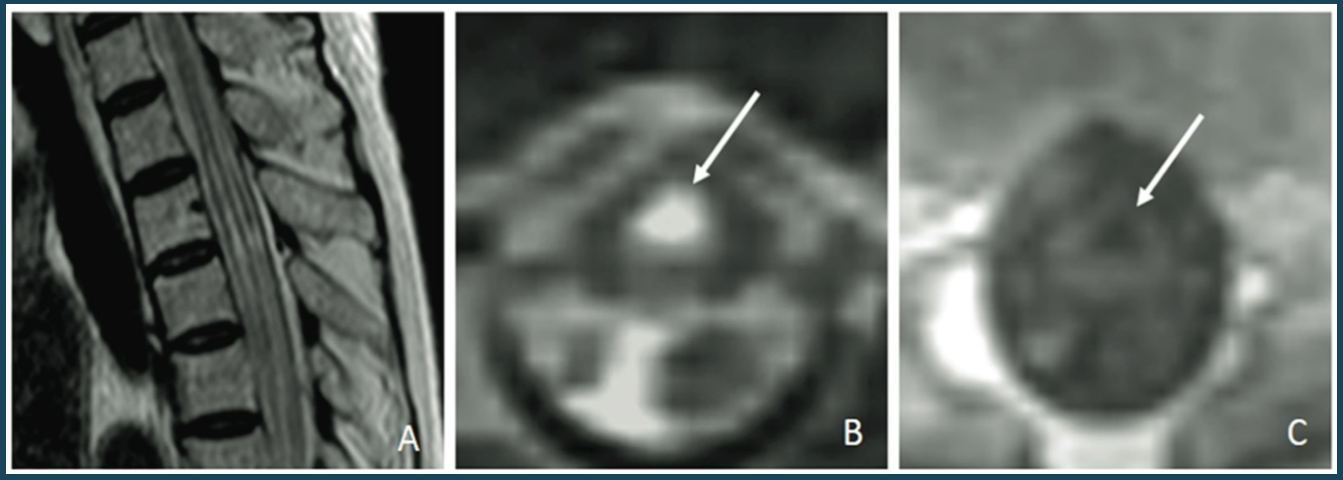
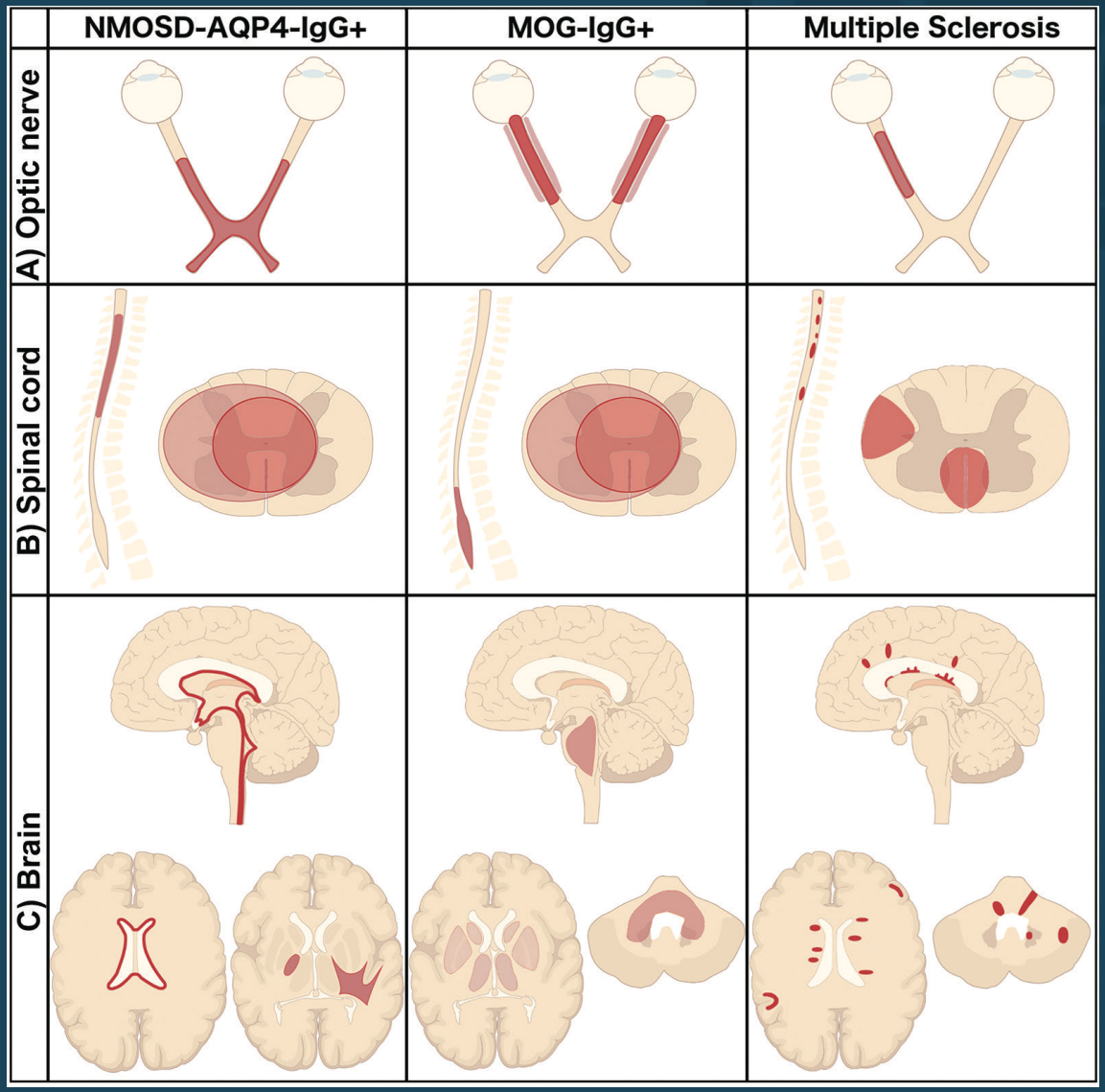


Diagnóstico diferencial



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- *Yonezu T et al. "Bright spotty lesions" on spinal magnetic resonance imaging differentiate neuromyelitis optica from multiple sclerosis. Multiple Sclerosis Journal 2014;20(3):331-7.*