

European Course on
Pediatric Neuroradiology in
Latin America (ECPNR-LATAM)

3rd Cycle - Module 1

CNS Inflammatory Diseases - Part I

Idealização:



Organização:



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Pediatric Neuroradiology in
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Idealização:



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There is no conflict of interest in this presentation

CNS Inflammatory Diseases - Part I

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Part 1 – Overview

Part 2 – Differential Diagnosis (Andrea Righini)

CNS Inflammatory Diseases - Part I

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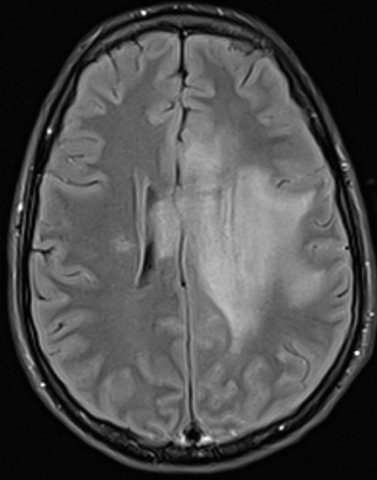
- Immune-mediated inflammatory CNS diseases
- Diagnostic Criteria

Multiple Diseases – Similar Appearances

- Multiple Sclerosis (MS)
- Acute Disseminated Encephalomyelitis (ADEM)
- Neuromyelitis Optica (NMO)
- Myelin Oligodendrocyte Glycoprotein Antibody-Associated Disease (MOGAD)

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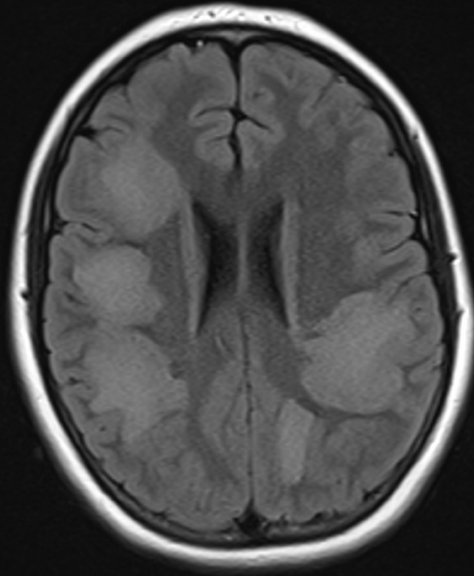
ADEM

3 children presented with acute neurological deficits
3 different diseases

Vasculitis

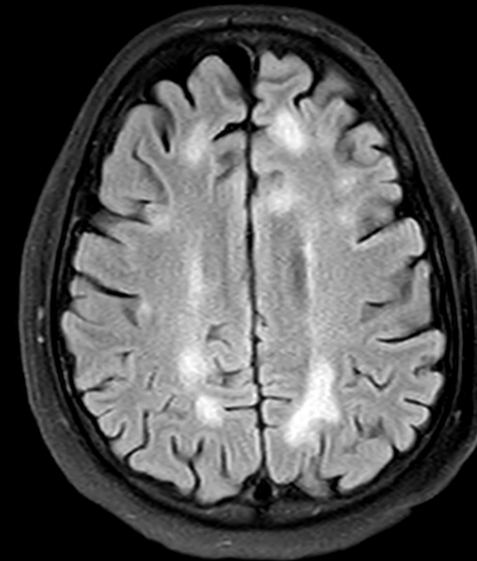
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MS



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Multiple Sclerosis - Background

- MS is a chronic inflammatory auto-immune disease
- Most common in young adults
- Can affect children- Pediatric Onset MS (**POMS**)
- 3-10% of MS patients present under the age of 16

Paediatric Onset Multiple Sclerosis

- Disease course in children is different
- 98% present with relapsing- remitting MS (RR)
- More aggressive onset
- Higher relapsing rate

Clinical Types

Relapsing-Remitting MS (RRMS)

Relapses of new or worsening neurological symptoms followed by periods of partial or complete remissions

Secondary Progressive MS (SPMS)

Relapses and remissions to a more steadily progressive phase.

Primary Progressive MS (PPMS)

Steady worsening of neurological function from the onset without early relapses or remissions.

Clinical Types

Two other related conditions:

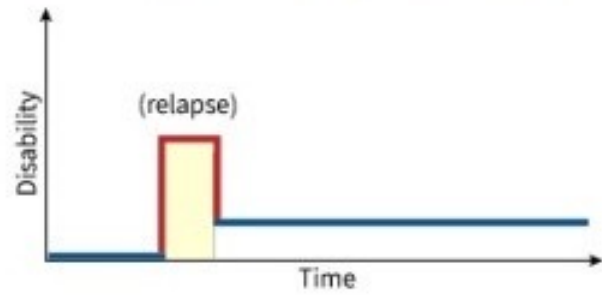
Clinically Isolated Syndrome (CIS)

Single episode of neurological symptoms suggestive of MS but does not meet diagnostic criteria

Radiologically Isolated Syndrome (RIS)

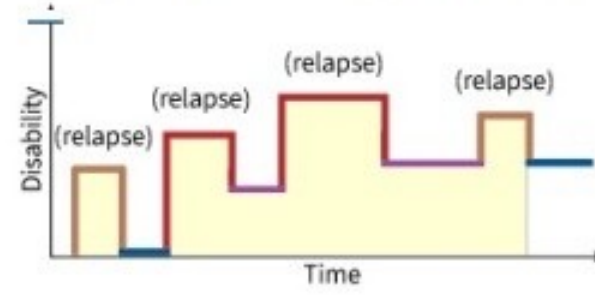
MS-like lesions brain or spinal cord without corresponding neurological symptoms

Clinically Isolated Syndrome (CIS)



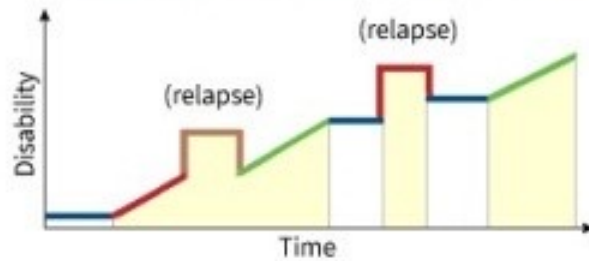
- Active with progression (active MRI)
- Not active without progression
- Active MRI (relapse or progression)

Relapsing Remitting MS (RRMS)



- Active worsening (active MRI)
- Active not worsening (active MRI)
- Not active worsening (active MRI)
- Not active not worsening
- Active MRI (relapse or progression)

Secondary Progressive MS (SPMS)



- Active with progression (active MRI)
- Active without progression (active MRI)
- Not active with progression (active MRI)
- Not active without progression
- Active MRI (relapse or progression)

Primary Progressive MS (PPMS)



- Active with progression (active MRI)
- Active without progression (active MRI)
- Not active with progression (active MRI)
- Not active without progression
- Active MRI (relapse or progression)

Clinical Presentation

- Intermittent neurological deficits
- CIS- 1st episode of MS:
 - 20% chance to develop MS
 - 50% with ON will develop MS
- Frequently mild clinical events are ignored, leading for the delay of the diagnosis

Diagnostic Criteria

Pediatric International Study Group 2013

- 2 or more clinical events
- >30 days and involving more than 1 CNS area
- 1 episode typical of MS + MRI findings consistent with the McDonald criteria with dissemination of space or time
- 1 ADEM attack followed by a clinical event+ MRI lesions
- First event not meet ADEM criteria and MRI lesions according to McDonald criteria

2017 McDonald Criteria

Dissemination in space (DIS): at least 2/4

1 or more periventricular lesion

1 or more juxtacortical/cortical lesion

1 or more infratentorial lesion

1 or more spinal lesion (symptomatic or not)

Dissemination in time (DIT):

- **New lesion**
- Simultaneous presence of **enhancing and non enhancing lesions**
- **CSF oligoclonal bands (OCBs):** indicate inflammation in the CNS. Satisfy the DIT requirement without a second clinical attack or a follow-up MRI

2024 McDonald Criteria



Diagnosis of multiple sclerosis: 2024 revisions of the McDonald criteria

Xavier Montalban, Christine Lebrun-Frény, Jiwon Oh, Georgina Arrambide, Marcello Moccia, Maria Pia Amato, Lilyana Amezcua, Brenda Banwell, Amit Bar-Or, Frederik Barkhof, Helmut Butzkueven, Olga Ciccarelli, Jeremy Chataway, Jeffrey A Cohen, Giancarlo Comi, Jorge Correale, Florian Deisenhammer, Massimo Filippi, Julie Fiol, Mark S Freedman, Kazuo Fujihara, Cristina Granziera, Ari J Green, Hans-Peter Hartung, Kerstin Hellwig, Ludwig Kappos, Dorlan Kimbrough, Joep Killestein, Fred Lublin, Romain Marignier, Ruth Ann Marrie, Aaron Miller, Susana Otero-Romero, Daniel Ontaneda, Sudarshini Ramanathan, Daniel Reich, Maria A Rocca, Àlex Rovira, Shiv Saidha, Amber Salter, Jaume Sastre-Garriga, Deanna Saylor, Andrew J Solomon, Maria Pia Sormani, Bruno Stankoff, Mar Tintore, Helen Tremlett, Anneke Van der Walt, Shanthi Viswanathan, Heinz Wiendl, Brigitte Wildemann, Bassem Yamout, Paola Zoratti, Peter A Calabresi, Timothy Coetzee, Alan J Thompson

Lancet Neurol 2025; 24: 850–65

See [Comment](#) pages 807 and 808

See [Personal View](#) pages 866 and 880

Multiple Sclerosis Centre of Catalonia and Department of Neurology

(Prof X Montalban MD,

G Arrambide MD,

S Otero-Romero MD,

J Sastre-Garriga MD,

Prof M Tintore MD) and Section

Advances in the understanding of multiple sclerosis and the development of biomarkers of pathophysiology prompted a substantial revision of the 2017 McDonald diagnostic criteria. The new 2024 McDonald criteria provide a unified approach for diagnosing multiple sclerosis in individuals with relapsing or progressive courses throughout the lifespan (ie, from paediatric to late-life presentations). The optic nerve can now serve as a fifth anatomical location within the CNS for diagnosis. The central vein sign, paramagnetic rim lesions, and kappa free-light chain concentrations in CSF can be used, when available, to provide supportive evidence and confer specificity for a diagnosis of multiple sclerosis in specific situations. In certain cases, radiologically isolated syndrome or neurological symptoms that do not constitute a clear attack or progression of disability can fulfil the criteria for a multiple sclerosis diagnosis. We also provide guidance for the diagnosis of multiple sclerosis in older individuals (≥ 50 years) and those with comorbidities. The 2024 revised criteria should expedite the diagnosis of multiple sclerosis, while maintaining specificity.

Rationale for updating the McDonald criteria

1. Not all presentations of MS are “typical”
2. New data on value of novel imaging and other para-clinical tests in MS diagnosis e.g. **optic nerve MRI, optical coherence tomography (OCT), central vein sign**
3. Increasing knowledge about **MS misdiagnosis**, and new disorders that can mimic MS e.g. MOGAD
4. Desire to move towards a **biological** rather than clinical definition of MS, and recognition of **MS phenotypes as a continuum** rather than distinct entities

McDonald 2024 criteria: Key revisions

1. Diagnosis of MS in patients with atypical presentations
2. Changes to criteria for dissemination in space
3. New ways to diagnose MS in patients with dissemination in space, without requiring dissemination in time
4. Kappa-free light chains – the new oligoclonal bands?
5. Diagnosis of PPMS – moving towards a single set of MS diagnostic criteria

2024 McDonald Criteria

- **Optic nerve as a 5th anatomical site**

Alongside periventricular, juxtacortical/cortical, infratentorial, and spinal cord
Optic nerve involvement can be shown with **MRI, optical coherence tomography (OCT), or visual evoked potentials (VEP)**

- **Dissemination in time (DIT) no longer mandatory**

Previously required to confirm MS, but now optional

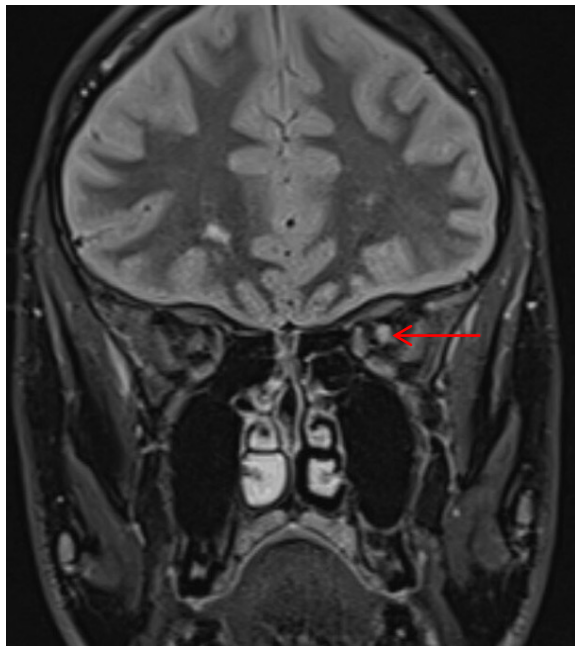
Can be replaced by CSF biomarkers (oligoclonal bands or kappa free light chains)

- **New imaging markers**

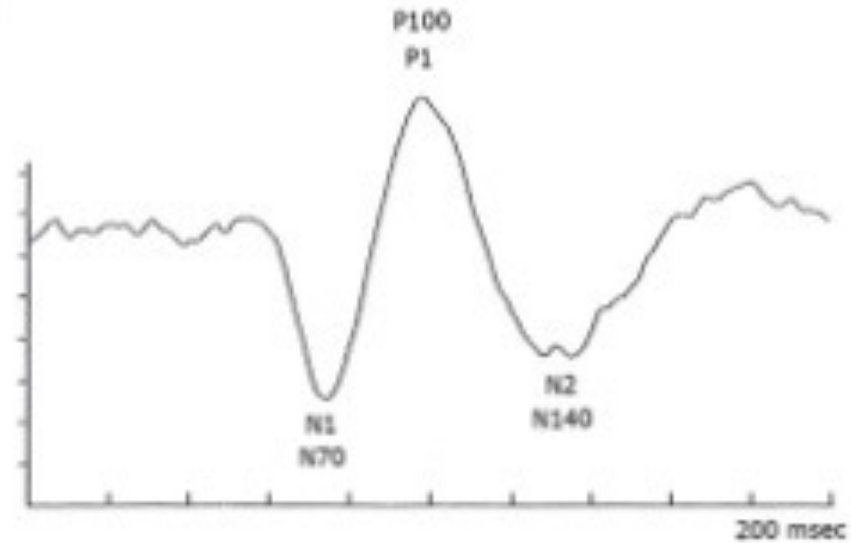
Central vein sign (CVS) and **paramagnetic rim lesions (PRLs)** on MRI can be used in specific cases to increase diagnostic specificity.

Evaluation of the optic nerve

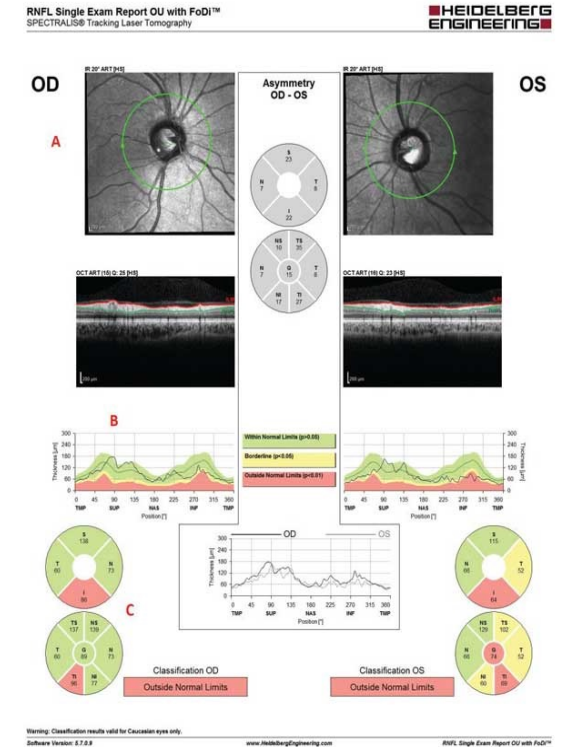
Optic nerve MRI



Visual evoked potentials



Optical coherence tomography



Central Vein Sign (CVS)

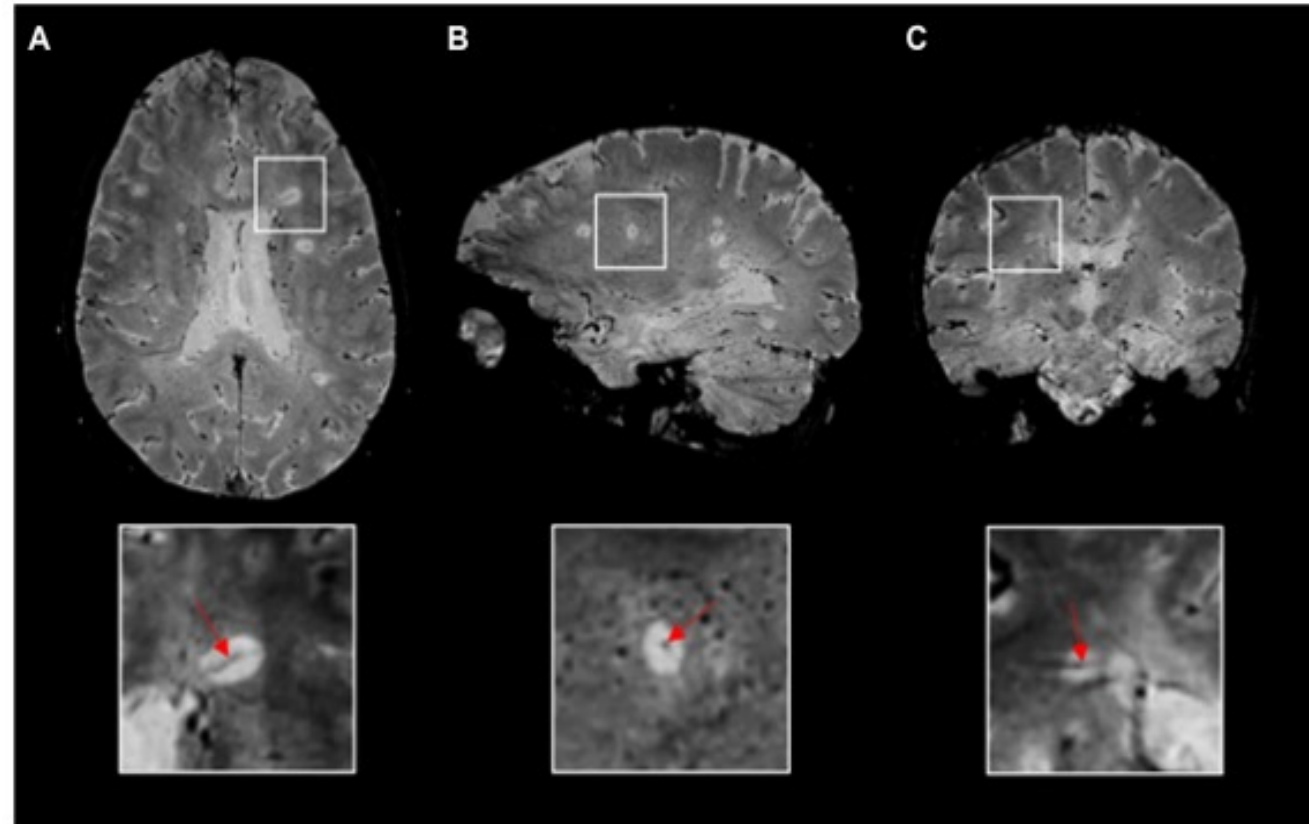
MS lesions are typically form around veins

MS white matter lesions typically have a higher proportion of CVS+ lesions than in common mimics

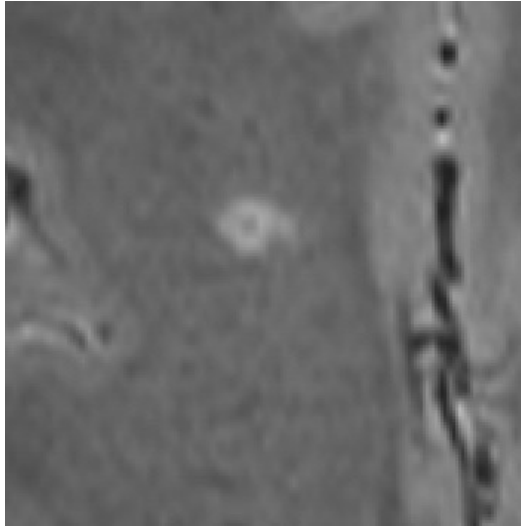
- Small vessel disease
- Migraine
- NMOSD/MOGAD etc

Requires dedicated MRI sequences e.g. SWI

Cut-off of 40% lesions with a central vein has high specificity for MS



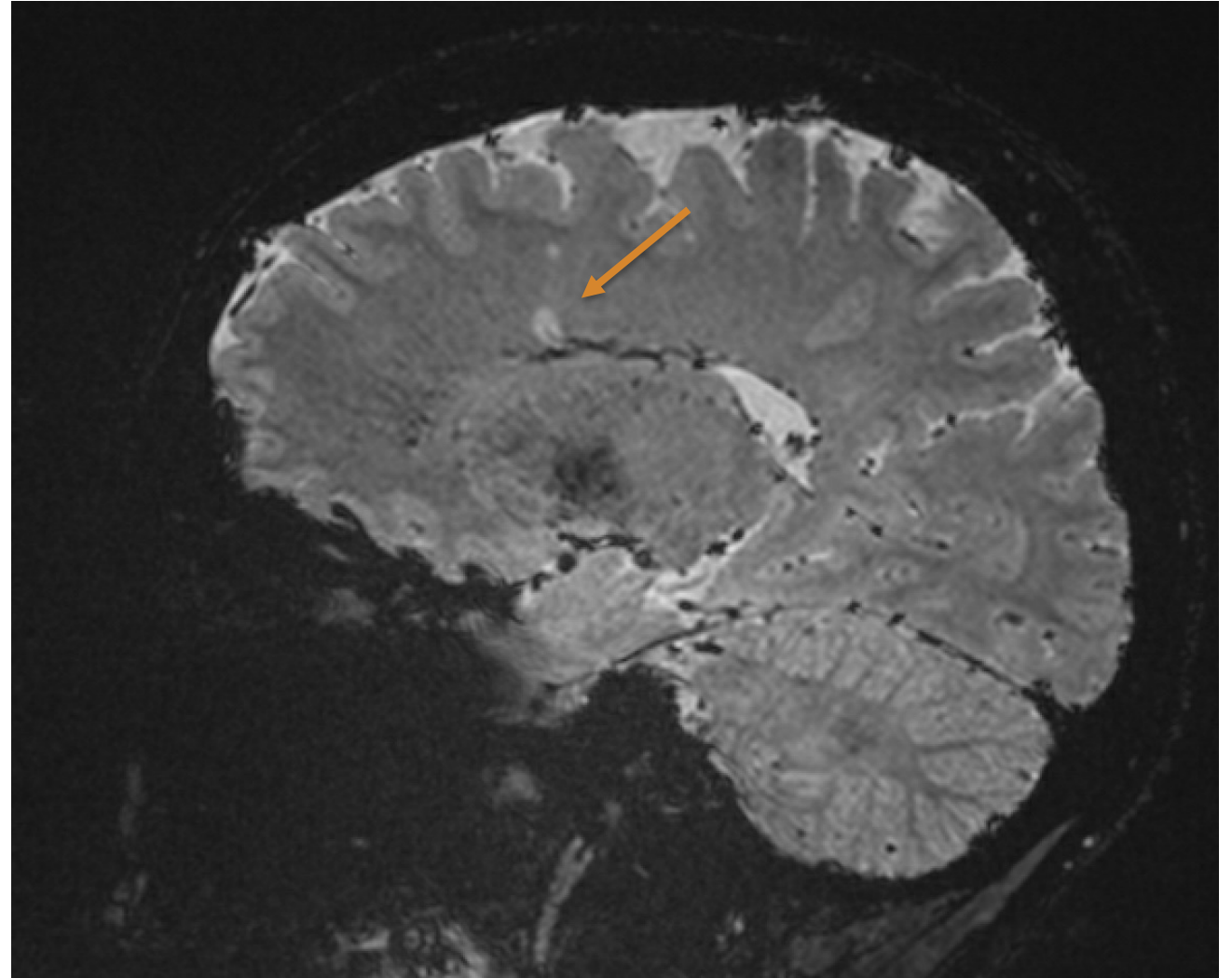
Central Vein Sign



3D T2* EPI



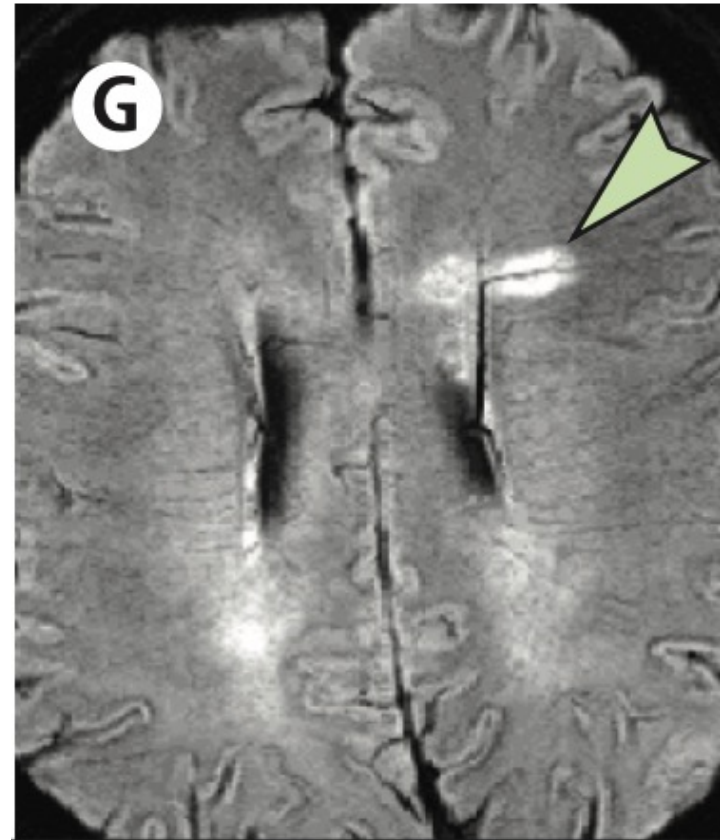
3D FLAIR



PRL



CVS

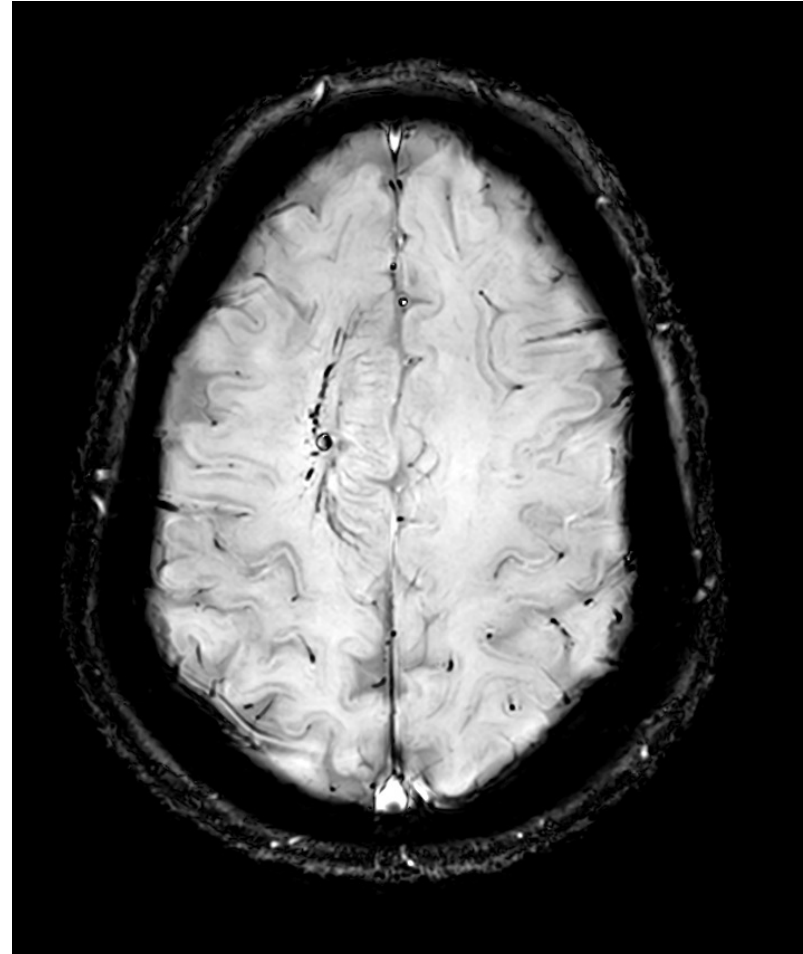
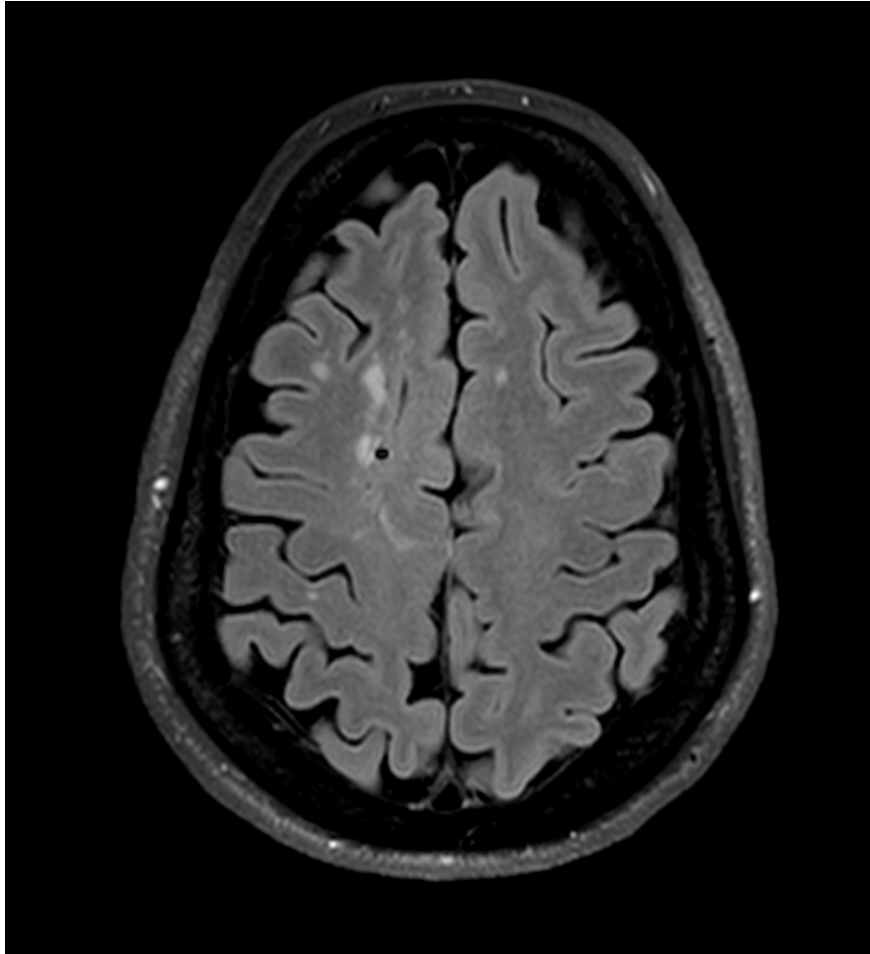


CVS

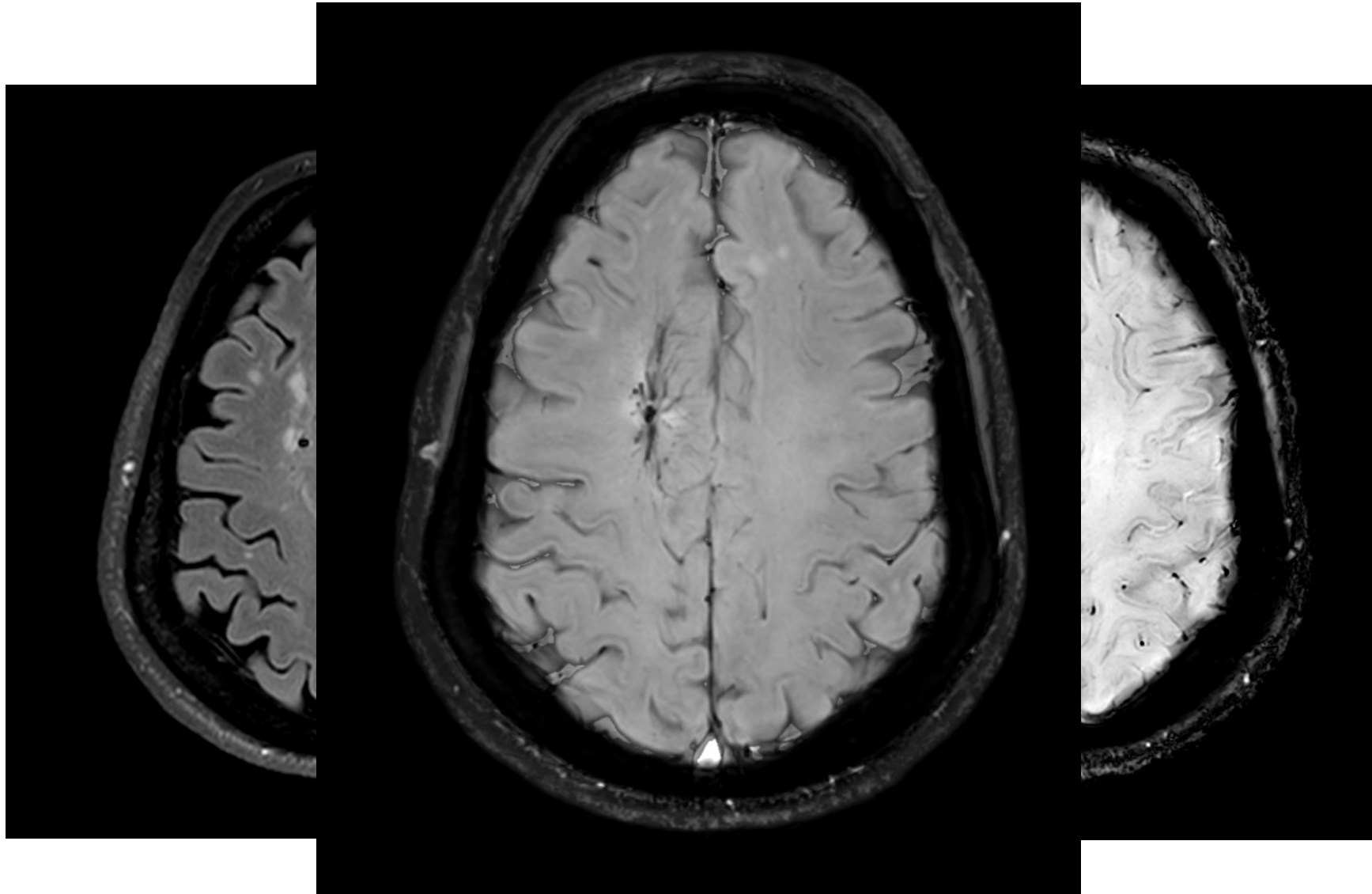
- Flair*: A Combined MR Contrast Technique for Visualizing White Matter Lesions and Parenchymal Veins
- FLAIR* imaging at 3.0 T provided high-quality cerebrospinal fluid-suppressed brain images demonstrating conspicuous [MS] lesions and veins
- Central vein sign



T2 FLAIR*



T2 FLAIR*



2024 McDonald Criteria

- **Biomarkers**

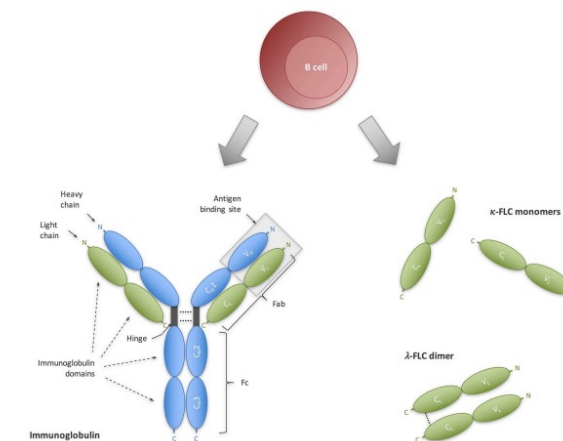
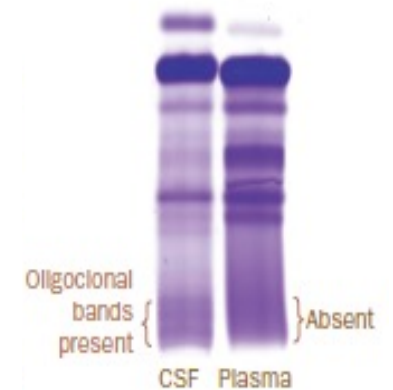
Kappa free light chain (kFLC) index is now considered interchangeable with oligoclonal bands in CSF

- **Radiologically isolated syndrome (RIS)**

People with incidental MRI findings typical of MS can meet criteria for MS diagnosis if other supportive features are present (e.g., positive CSF, CVS, or PRLs)

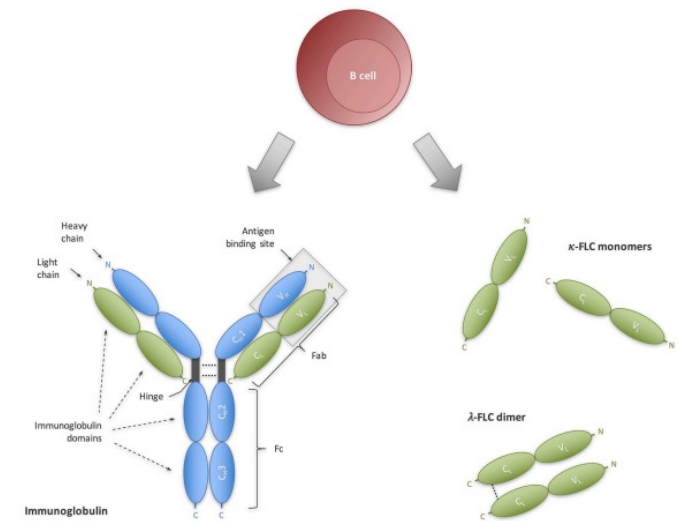
Oligoclonal bands vs κ -FLC index

- Detection of oligoclonal bands is **time-consuming and rater-dependent**
- **Plasma cells also secrete kappa and lambda free light chains** in excess of formed immunoglobulins, which can be easily quantified in a rater-independent fashion in the serum and CSF
- Previous studies have suggested that **kappa free light chains (κ -FLC) index may differentiate MS from healthy controls**, and at higher index levels from other neuroinflammatory disorders



When is κ -FLC index likely to be useful?

Because it does not require any specialist laboratory expertise it might provide information on intrathecal antibody production more quickly (within 24hrs), particularly in regional hospitals



2024 McDonald Criteria

- **Unified framework across all ages and disease types**

Same diagnostic criteria now apply to **paediatric and adult-onset MS**, and to **relapsing and progressive forms**

Specific guidance given for children, older patients (≥ 50 years), and those with comorbidities

- **Safeguards against misdiagnosis**

Strong emphasis on ruling out mimics such as migraine, small-vessel disease, NMOSD, and MOGAD.

Additional confirmatory evidence is recommended for older individuals and those with vascular risk factors.

2024 McDonald Criteria Children (Paediatric MS)

Unified framework: Same criteria as adults, but with added safeguards

ADEM presentations: If a child presents with acute disseminated encephalomyelitis, diagnosis of MS requires either a second attack or new T2 lesions appearing ≥ 90 days later

Biomarkers: *MOG-IgG testing* is **mandatory for all children <12 years** with first demyelinating events (to distinguish from MOGAD) ≥ 12 years, MOG-IgG testing is recommended only when features are atypical for MS or suggestive of MOGAD

2024 McDonald Criteria Children (Paediatric MS)

MRI features:

Central vein sign (CVS) supports diagnosis, but in children <18 years, it should be applied cautiously—requires $\geq 50\%$ of lesions showing CVS

Note: Paediatric MS tends to have higher inflammatory activity; early treatment is crucial

Imaging

- **Location:**

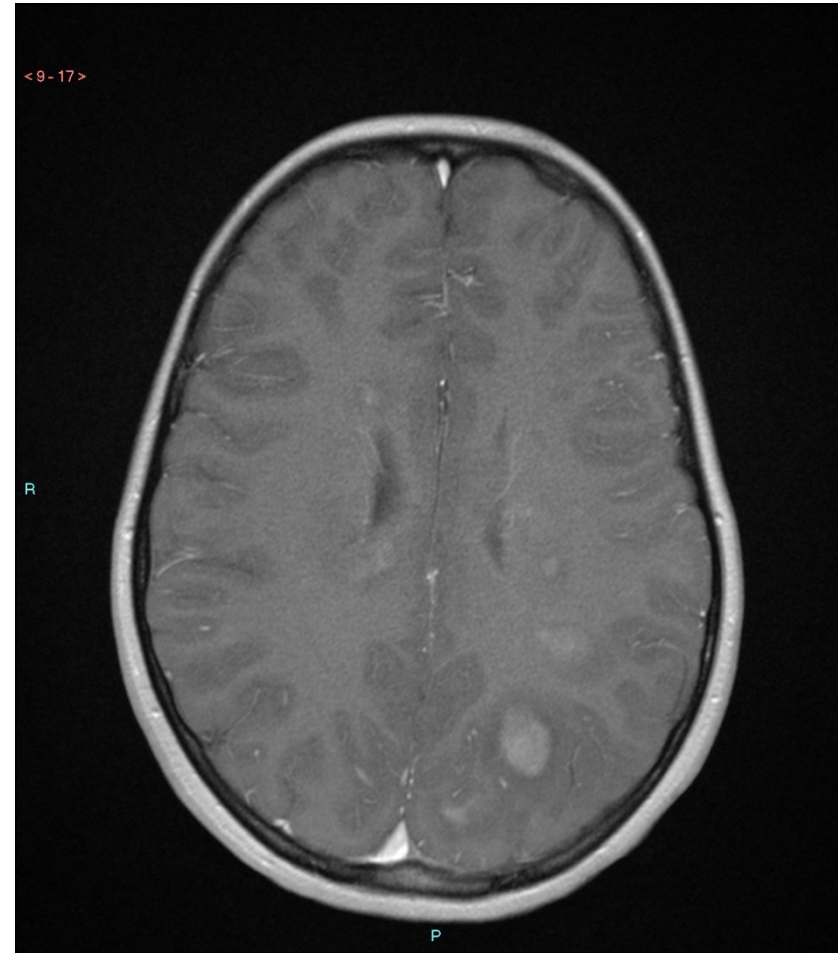
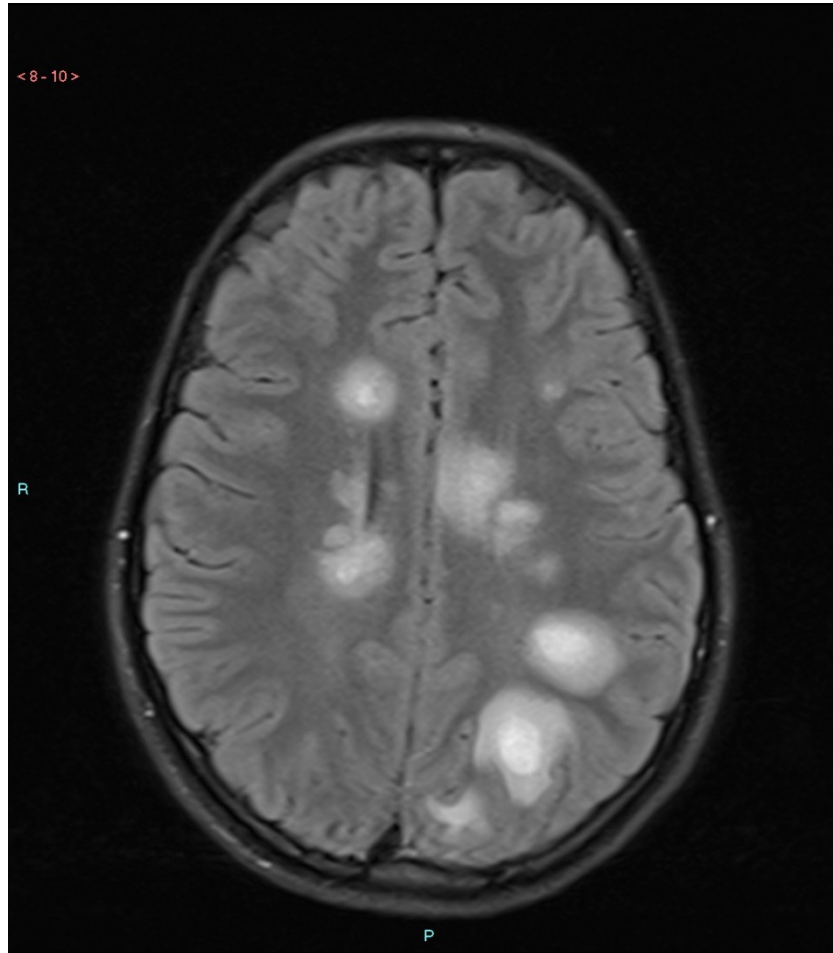
1. Supratentorial > infratentorial
2. Periventricular - Dawson fingers
3. Deep cerebral WM
4. Juxtacortical

Imaging

- **Size and Number**

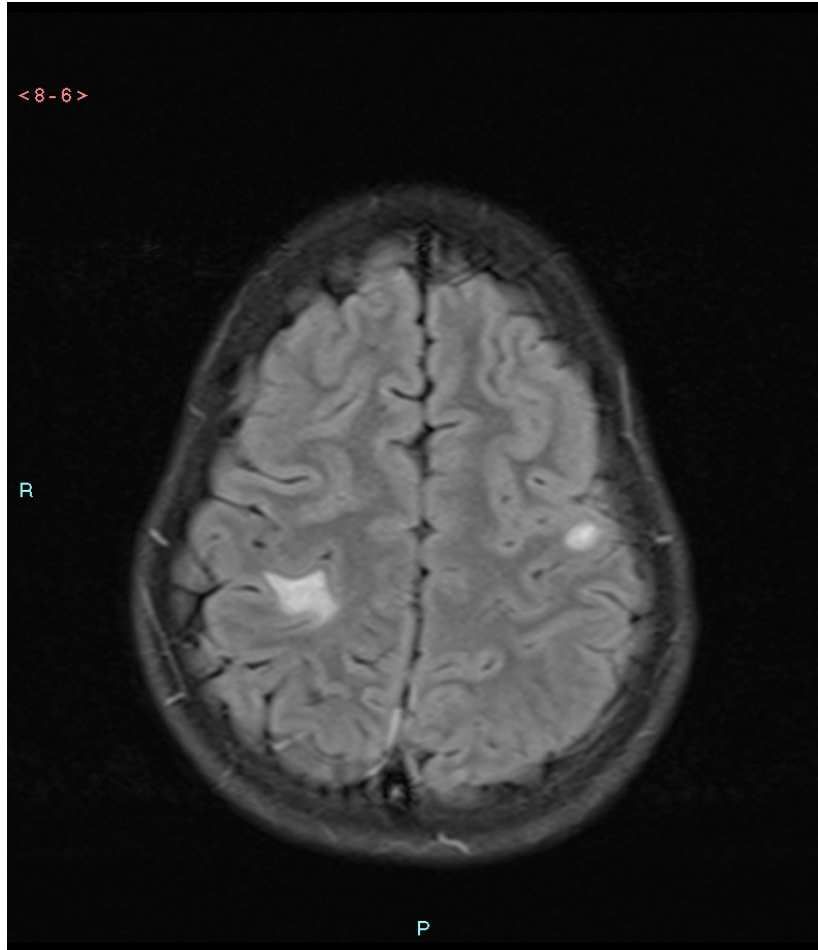
1. Multiple > solitary
 2. Small lesions usually 5-10 mm
 3. Giant- tumefactive lesions with surrounding edema (can be solitary)
- Active plaques can enhance
 - GAD accumulations is slow, steroid treatment suppress enhancement

Imaging

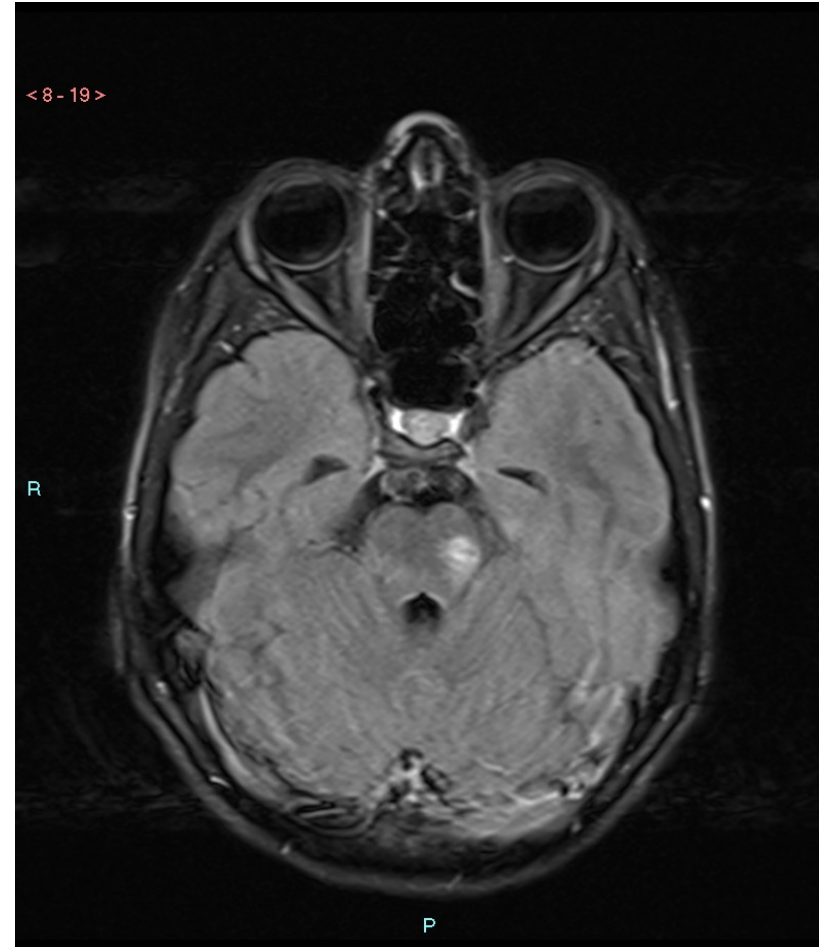


13 YO girl/ 1st episode of weakness, numbness

Imaging

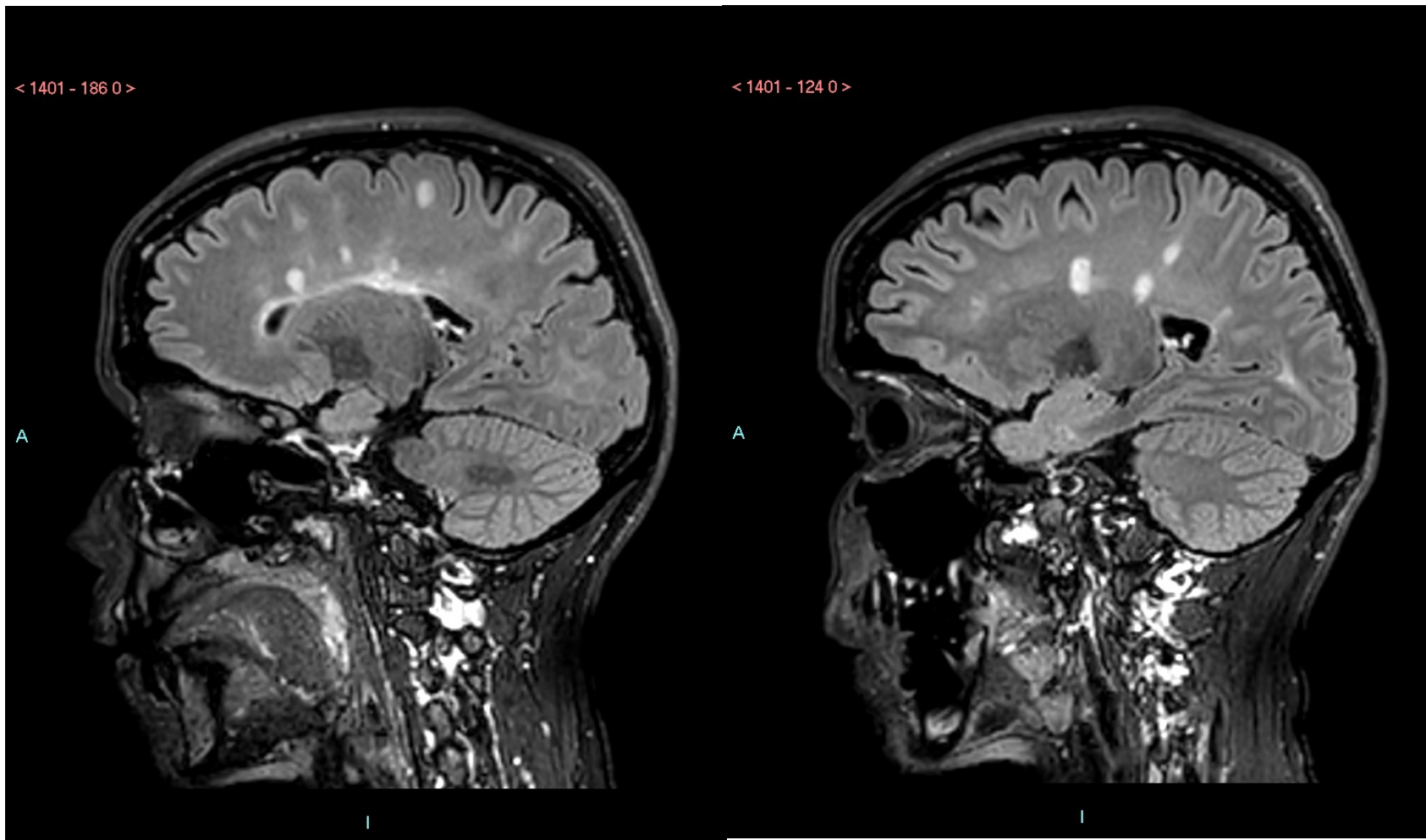


Juxtacortical

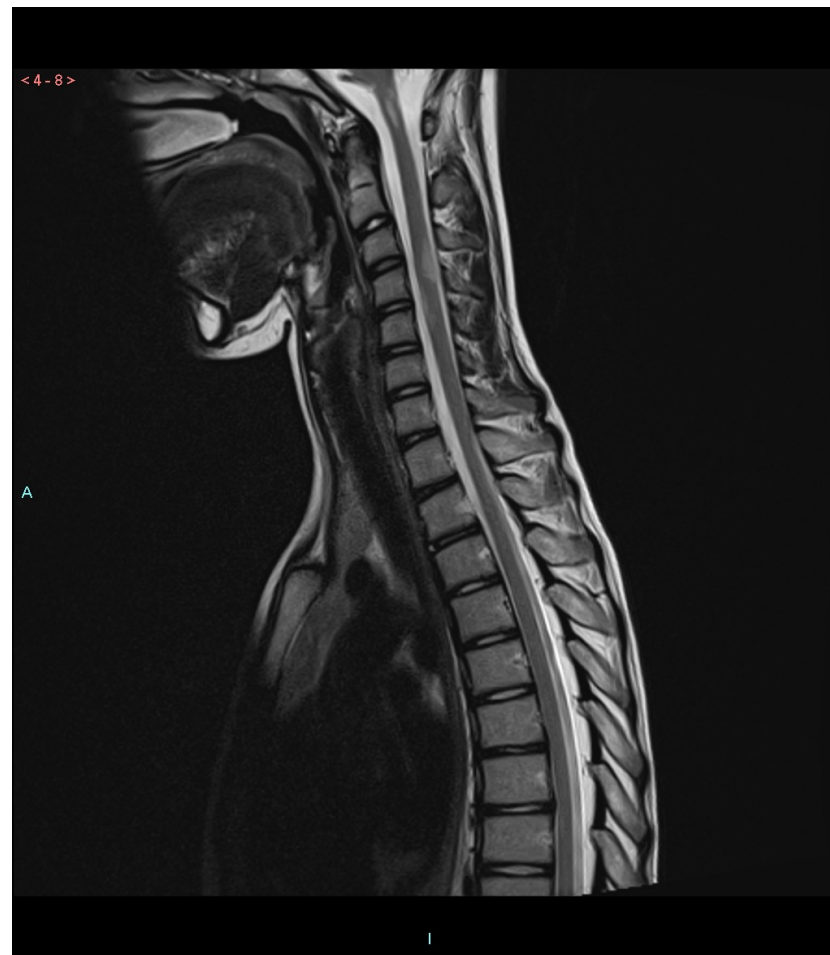
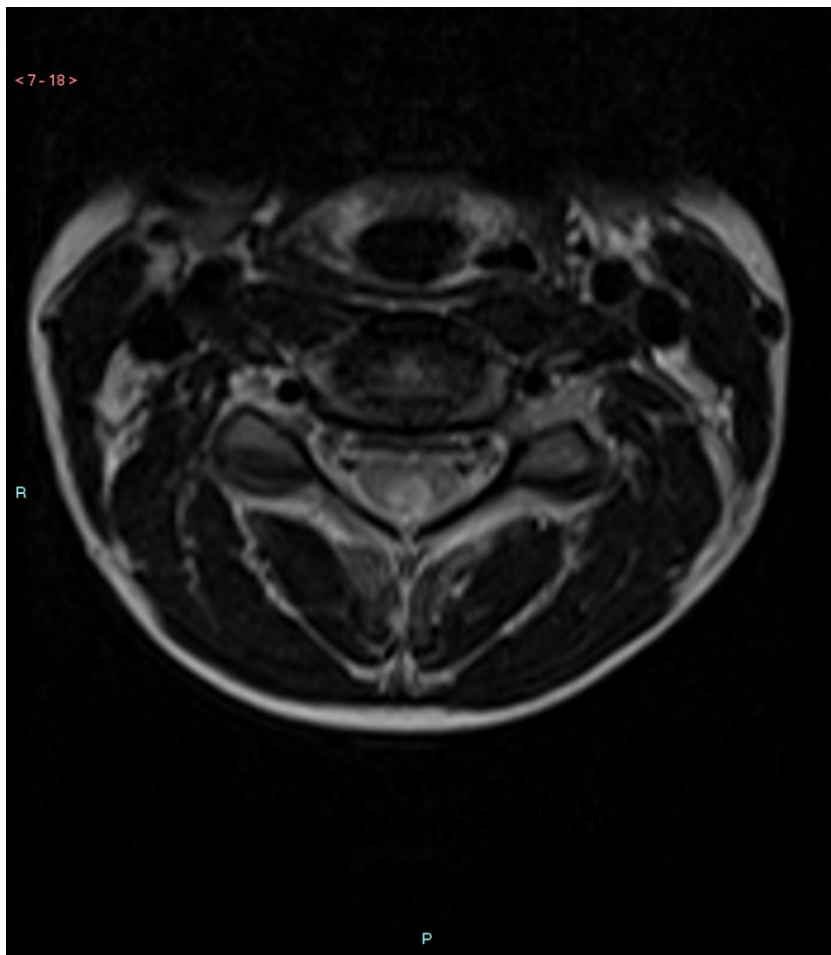


Posterior fossa- pons

Dawson Fingers

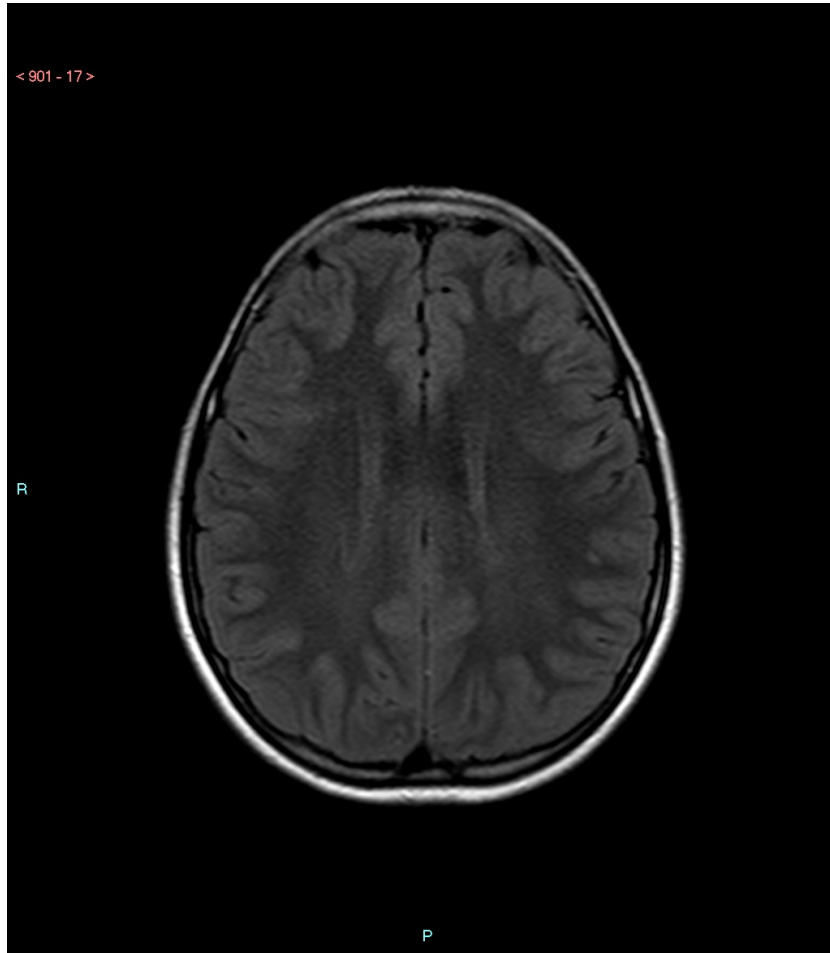


Imaging

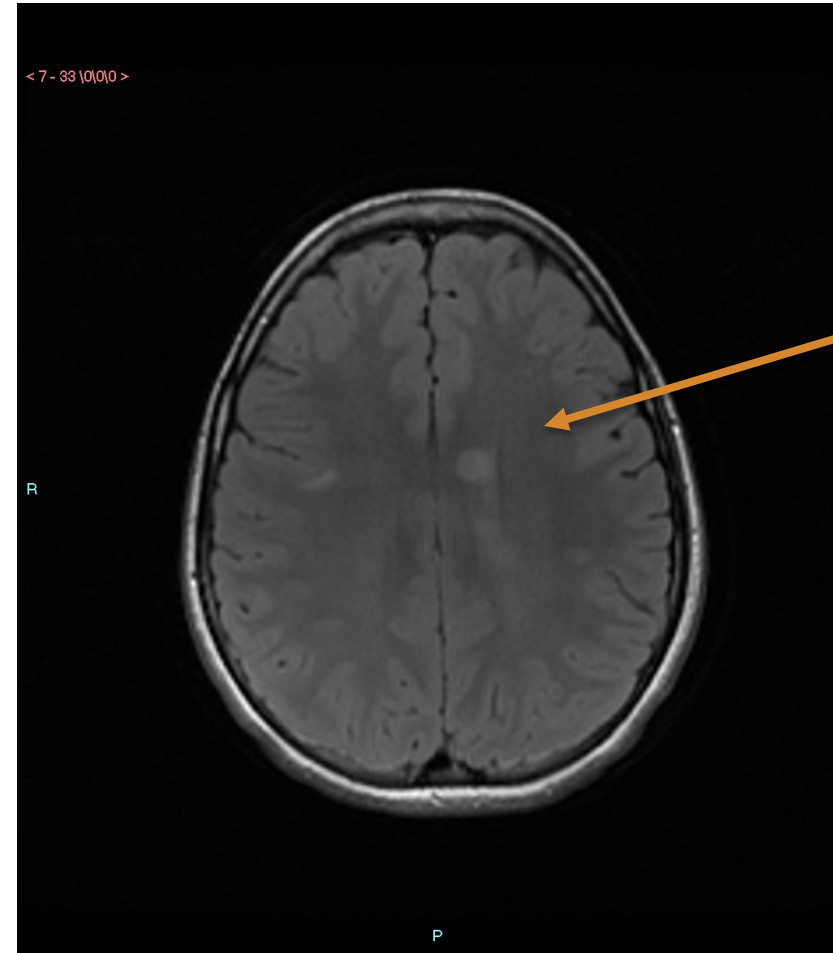


6 month later routine spine study

Follow up



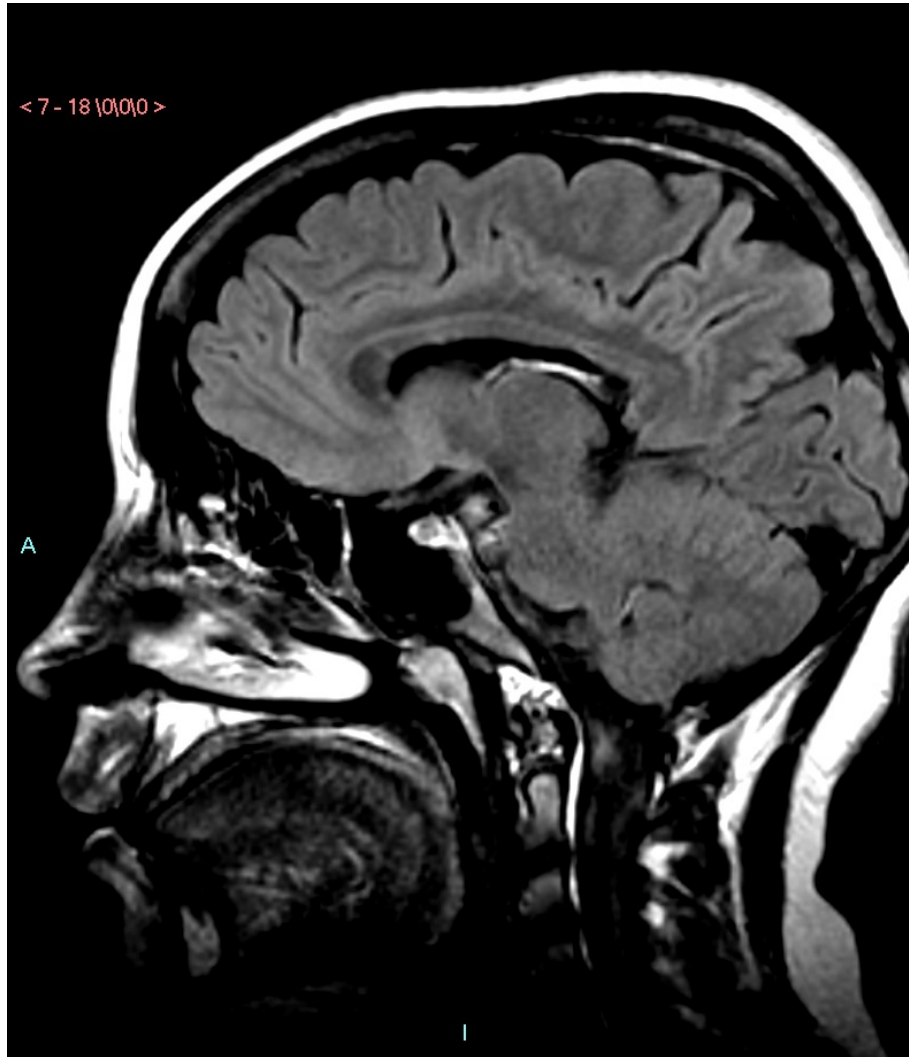
1st episode



New
Lesion

1 year later

POMS- tend to involve the CC



- CC involvement is demonstrated in the sagittal view
- Difference in Disease Burden and Activity in Pediatric Patients on Brain Magnetic Resonance Imaging at Time of Multiple Sclerosis Onset vs Adults

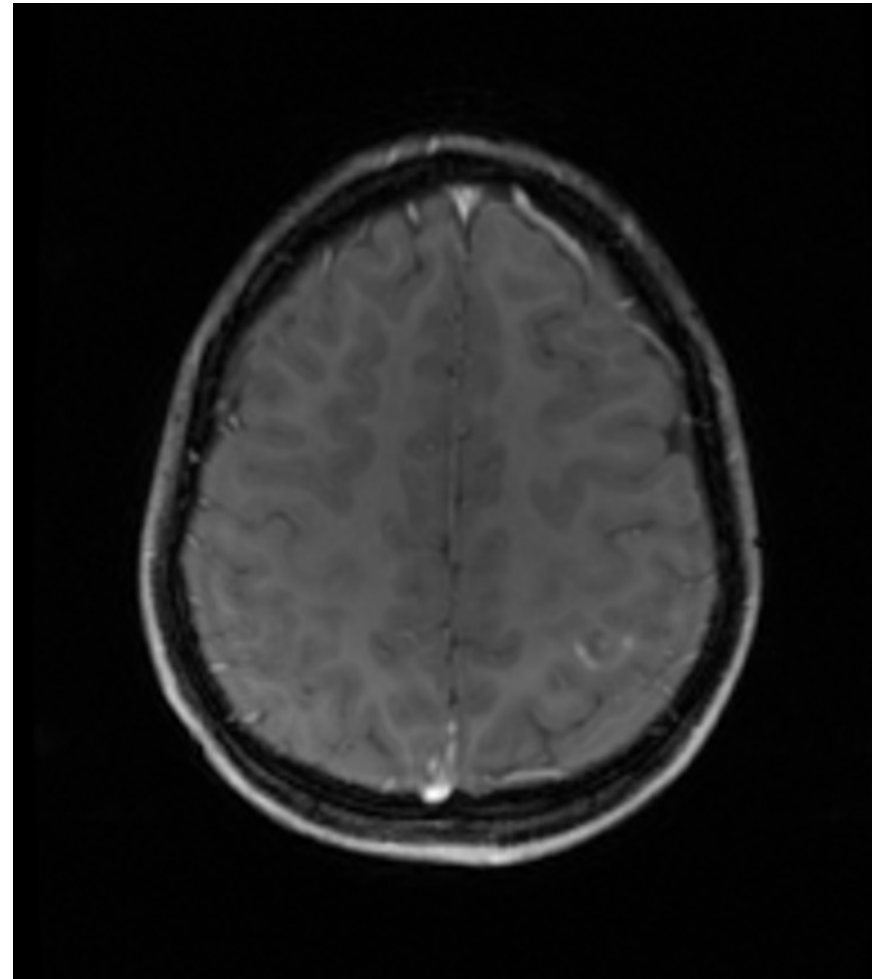
Waubant E, et al. Difference in disease burden and activity in pediatric patients on brain magnetic resonance imaging at time of multiple sclerosis onset vs adults. Arch Neurol. 2009 Aug;66(8):967-71. doi: 10.1001/archneurol.2009.135. PMID:



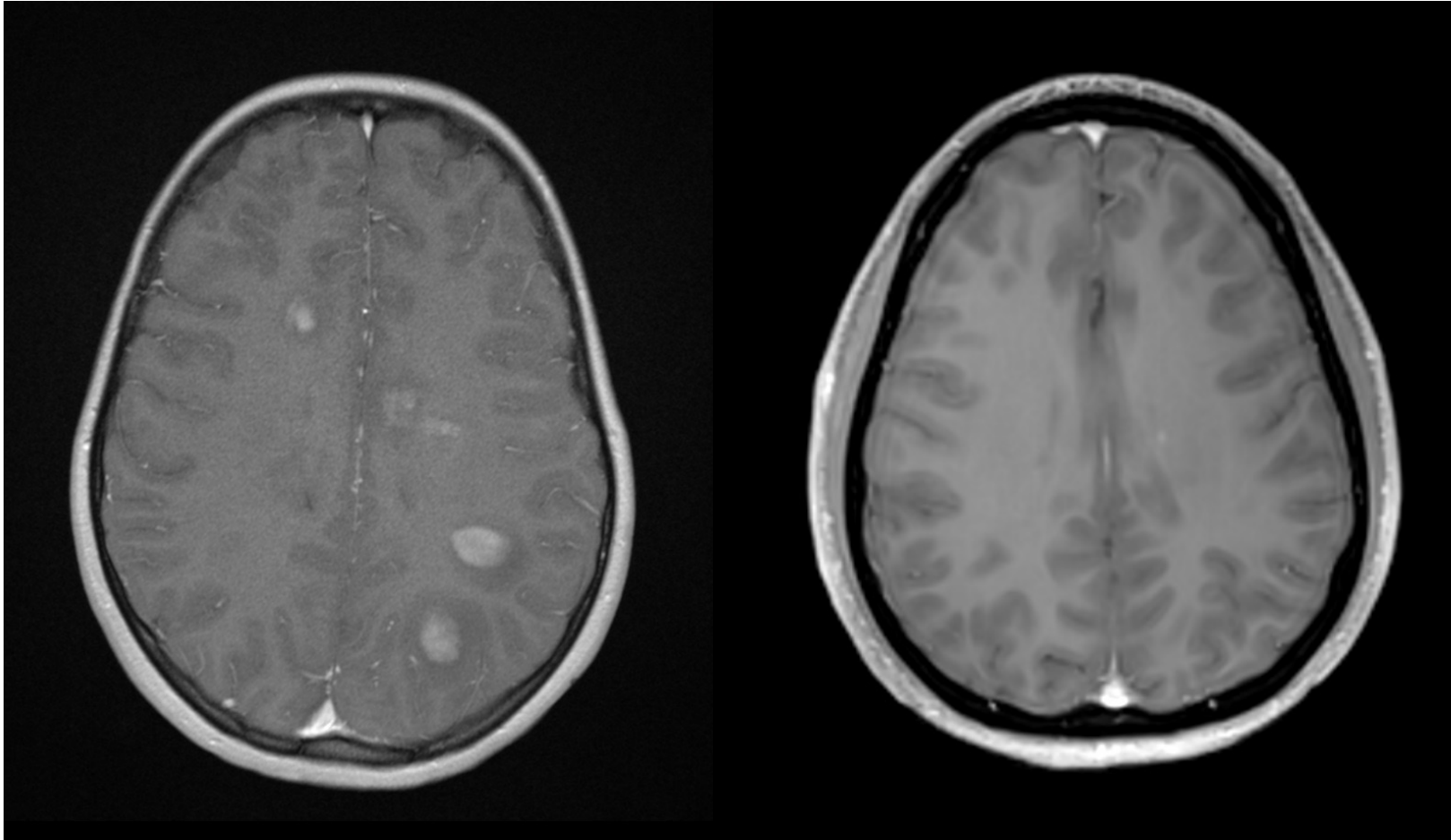
Sag view- look at the cervical cord

Gd Enhancement

- Types of enhancement:
Punctate
Nodular
Ring (incomplete)
- Same child 2 years later, relapse



Gd Enhancement



Nodular

Punctate

DWI/DTI

- DWI has no significant role in the diagnosis
- ADC can be normal or high
- Occasionally can show restriction
- DTI in normal appearing WM can show reduced diffusivity

MS a focal or diffuse disease?

J Neurol (2005) 252 [Suppl 5]: V/16–V/24
DOI 10.1007/s00415-005-5004-5

Massimo Filippi
Maria Assunta Rocca

MRI evidence for multiple sclerosis as a diffuse disease of the central nervous system

MRI evidence for diffuse pathology in the normal-appearing white matter (NAWM)

Post-mortem studies have shown subtle and widespread abnormalities in the NAWM from MS patients, which include diffuse astrocytic hyperplasia, patchy edema, and perivascular cellular infiltration, as well as axonal damage [2, 4, 27]. The application of modern MR-based techniques to the assessment of MS patients has allowed the

MRI evidence for pathology in the GM

Several *post-mortem* studies [15, 68, 73, 88] have shown the presence of MS-related damage, including axonal and neuronal loss, in the cortical and deep GM of MS patients. Such abnormalities usually go undetected when using cMRI because of their relatively small size, their relaxation characteristics which result in poor contrast with the surrounding normal GM and because of partial volume effects with the surrounding cerebro spinal fluid (CSF). High-field strength MR scanners are improving our ability to image focal GM lesions, and the application of modern MR-based techniques for the assessment of GM pathology has undoubtedly allowed to be overcome some of the limitations of cMRI.

Using ROI [9] and histogram analysis [6, 9, 24, 50, 51, 104], MT and DW MRI abnormalities have been shown in the GM of MS patients, including those with PPMS [24, 104], whereas no MD abnormalities have been detected in the GM of patients with early RRMS [54]. GM changes are more pronounced in patients with SPMS than in those with RRMS [6, 51], while no difference in the extent and severity of GM involvement were found between patients with SPMS and those with PPMS [104]. In patients with SPMS and PPMS [83], as well as in those with RRMS [82], these changes worsen over time. This suggests a progressive accumulation of GM damage al-

Cortical Atrophy

Neuroradiology (2018) 60:179–187
<https://doi.org/10.1007/s00234-017-1964-2>

DIAGNOSTIC NEURORADIOLOGY



Focal cortical thinning in patients with stable relapsing-remitting multiple sclerosis: cross-sectional-based novel estimation of gray matter kinetics

Lior Orbach^{1,2} · Shay Menascu^{1,2} · Chen Hoffmann^{1,2} · Shmuel Miron¹ · Anat Achiron^{1,2}

Received: 30 October 2017 / Accepted: 13 December 2017 / Published online: 28 December 2017
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Abstract

Purpose The aim of our study is to identify radiological patterns of cortical gray matter atrophy (CGMA) that correlate with disease duration in patients with relapsing-remitting multiple sclerosis (RRMS).

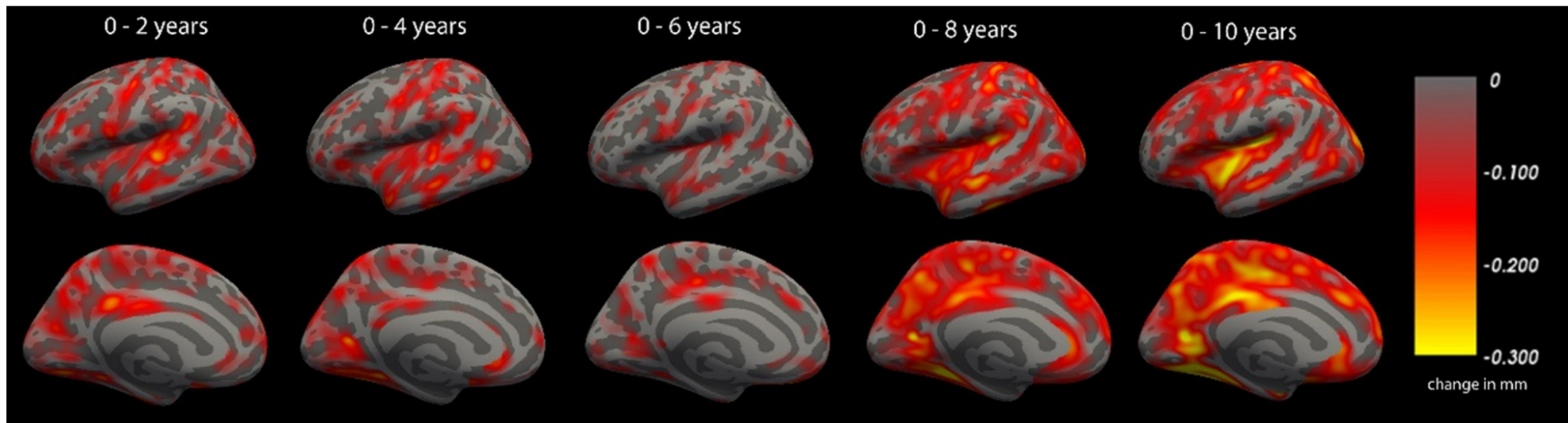
Methods RRMS patients were randomly selected from the Sheba Multiple Sclerosis (MS) center computerized data registry based on stratification of disease duration up to 10 years. Patients were scanned by 3.0 T (Signa, GE) MRI, using a T1 weighted 3D high resolution, FSPGR, MS protocol. Neurological disability was assessed by the Expanded Disability Status Scale (EDSS). FreeSurfer was used to obtain brain volumetric segmentation and to perform cortical thickness surface-based analysis. Clusters of change in cortical thickness with correlation to disease duration were produced.

Results Two hundred seventy-one RRMS patients, mean \pm SD age 33.0 ± 7.0 years, EDSS 1.6 ± 1.2 , disease duration 5.0 ± 3.4 years. Cortical thickness analysis demonstrated focal areas of cerebral thinning that correlated with disease duration. Seven clusters accounting for 11.7% of the left hemisphere surface and eight clusters accounting for 10.6% of the right hemisphere surface were identified, with cluster-wise probability of $p < 0.002$ and $p < 0.02$, respectively. The clusters included bilateral involvement of areas within the cingulate, precentral, postcentral, paracentral, superior-parietal, superior-frontal gyri and insular cortex. Mean and cluster-wise cortical thickness negatively correlated with EDSS score, $p < 0.001$, with stronger Spearman rho for cluster-wise measurements. **Conclusions** We identified CGMA patterns in sensitive brain regions which give insight and better understanding of the progression of cortical gray matter loss in relation to dissemination in space and time. These patterns may serve as markers to modulate therapeutic interventions to improve the management of MS patients.

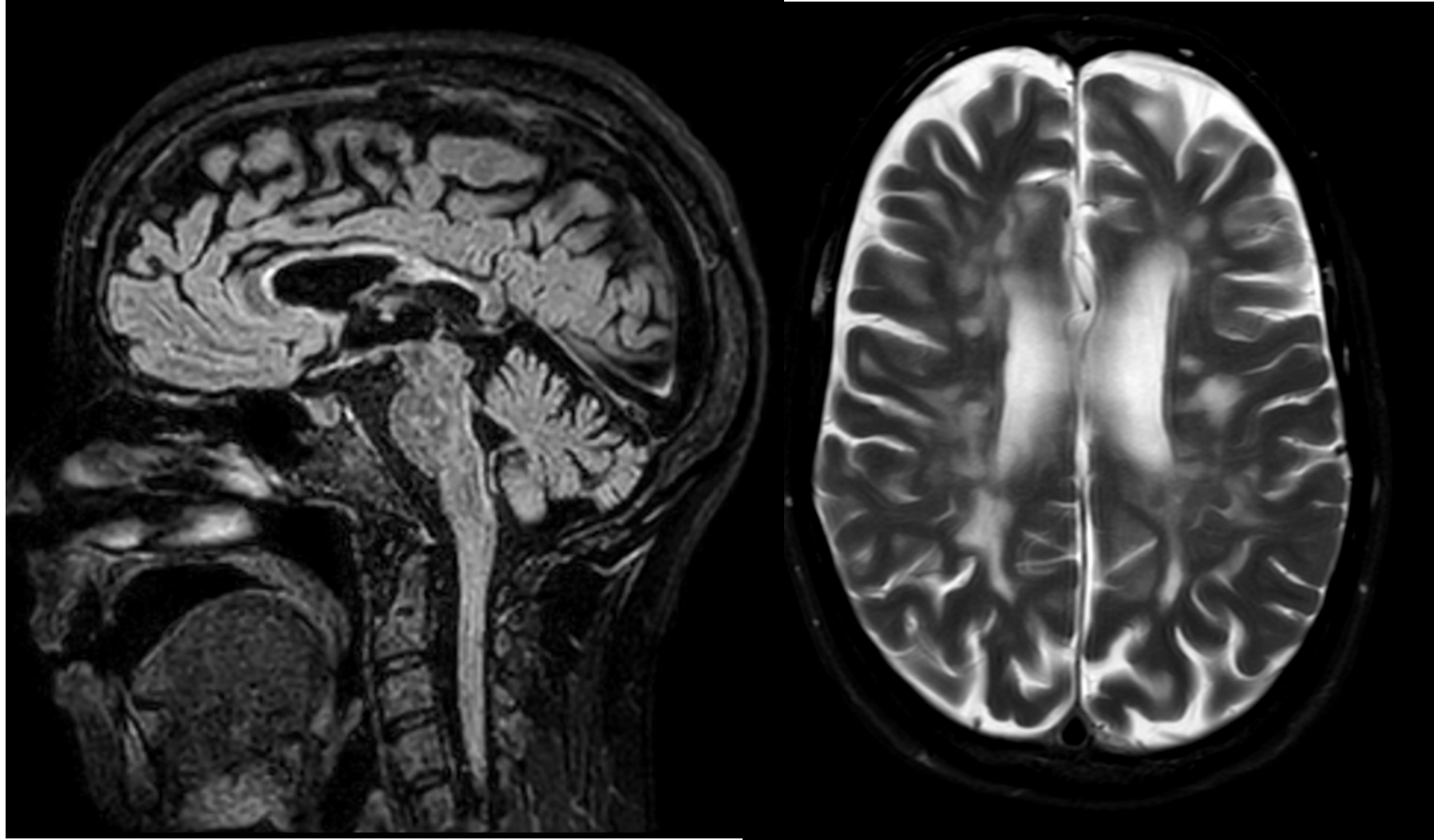
Keywords Multiple sclerosis · Gray matter atrophy · Cortical thickness · MRI · FreeSurfer

- As the disease progresses the cortex undergoes atrophy
- We measured the cortical thickness in acute diagnosed MS patients and chronic MS patients
- The value was 2.51 mm in the newly diagnosed and 2.42 mm in 10 years disease duration ($p < 0.001$)

Cortical Atrophy

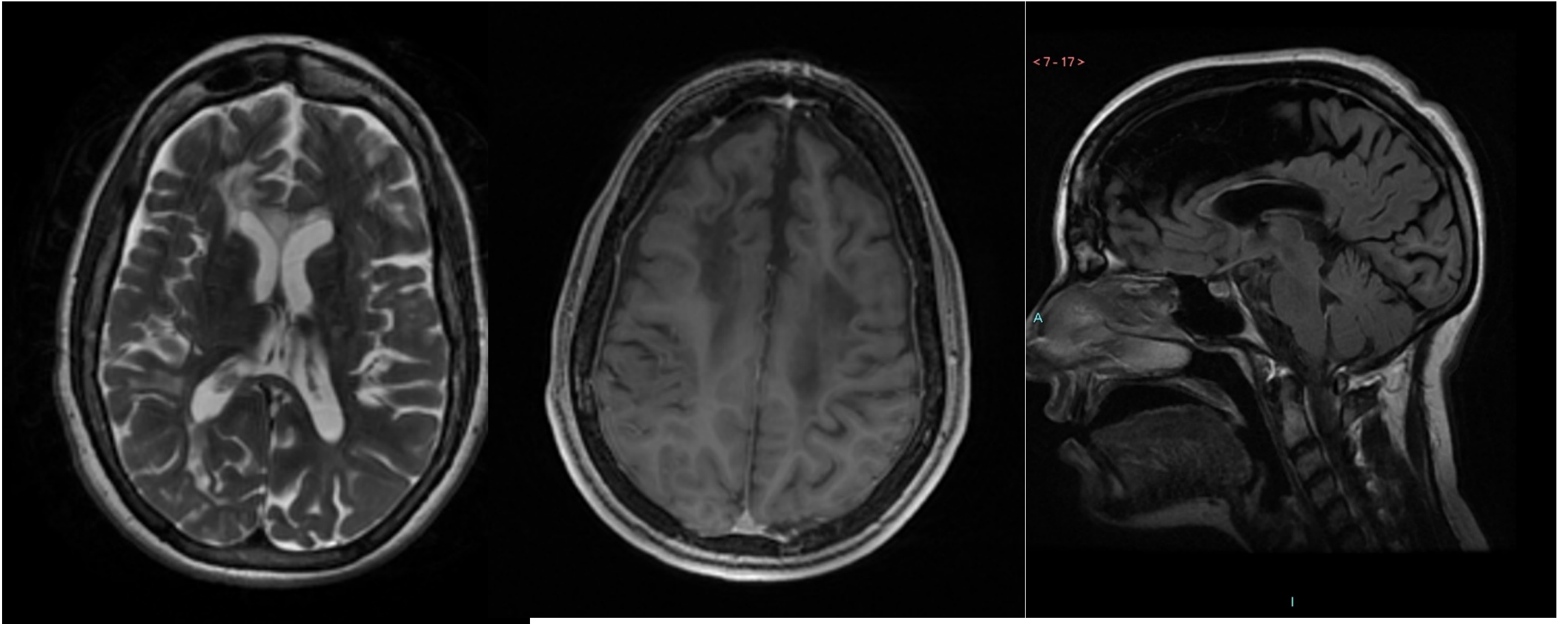


Chronic MS



34 YO female, diagnosed at the age of 14
Severe atrophy of the white matter

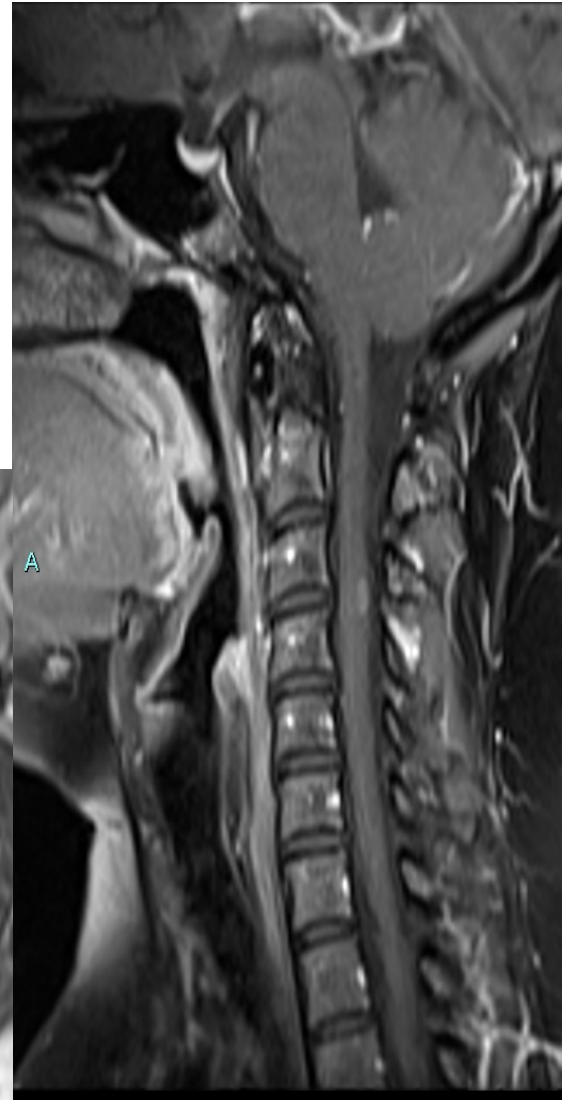
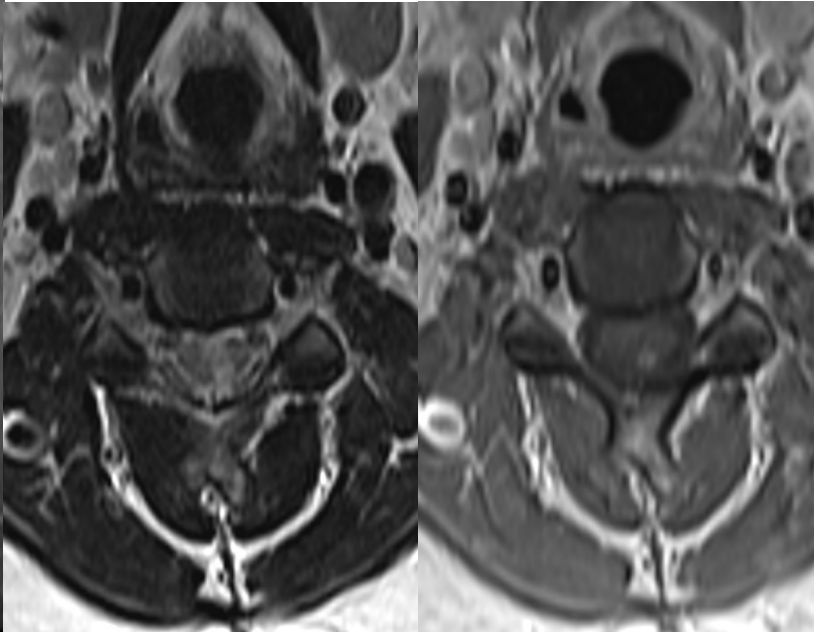
Chronic MS



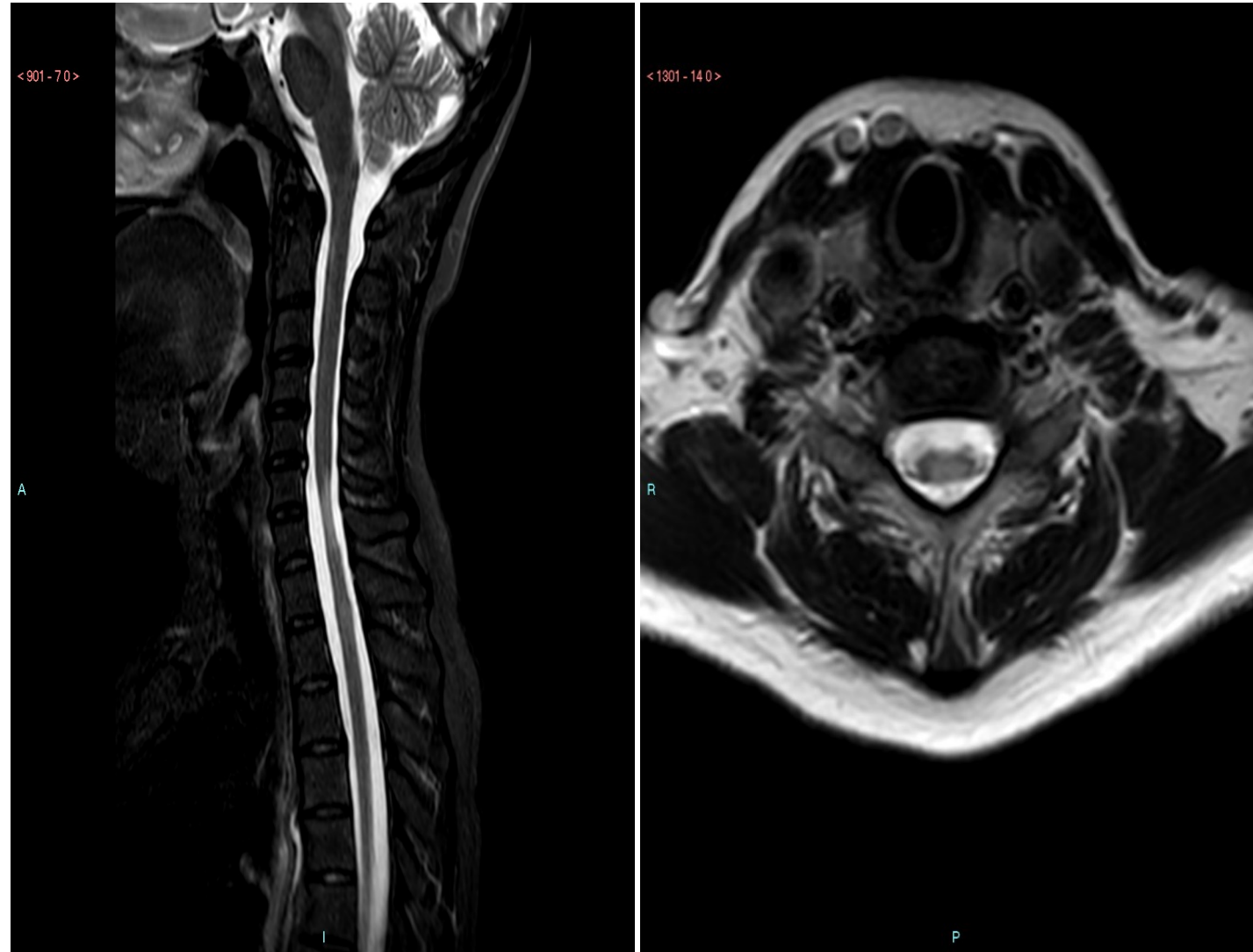
MS spinal lesions- acute phase



Multiple plaques levels C2, C4, C5
The lesion C3-C4 is enhancing with
Surrounding edema
The old plaques are not enhancing



MS- chronic phase



Multiple Diseases – Similar Appearances

- Multiple Sclerosis (MS)
- **Acute Disseminated Encephalomyelitis (ADEM)**
- Neuromyelitis Optica (NMO)
- Myelin Oligodendrocyte Glycoprotein Antibody-Associated Disease (MOGAD)

Acute Disseminated Encephalomyelitis (ADEM)

- 2nd to MS acquired idiopathic demyelinating disease
- Peak age group 5-8y
- Clinical symptoms: fever prior to the neurological presentation few days to weeks before (infection, immunization)
- Usually monophasic, recurrent type is involving the same areas
- Multiphasic involves different areas
- Most recover completely, 20% have residual neurological deficits
- Recovery in 50% within 2 months

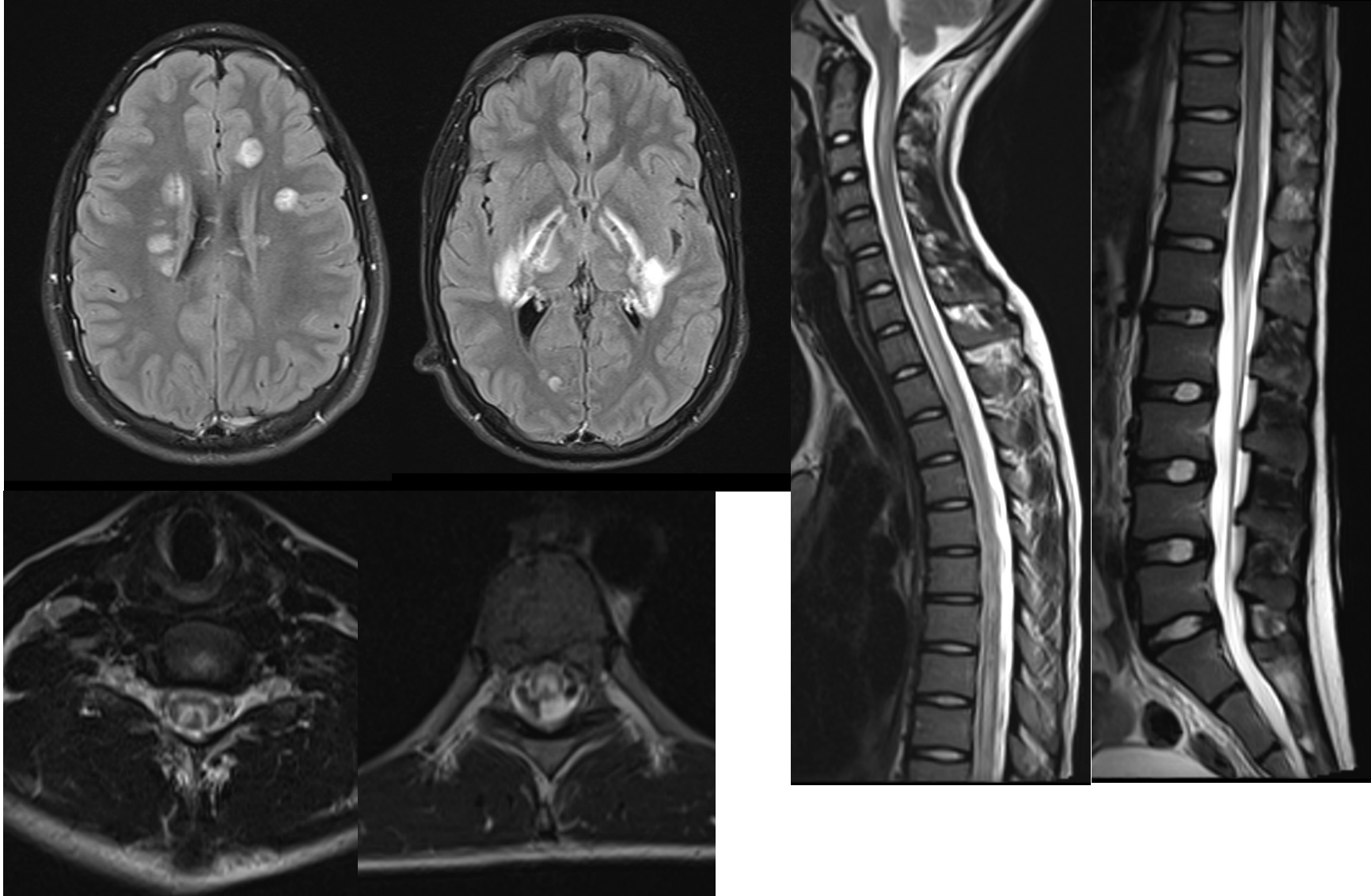
Acute Disseminated Encephalomyelitis (ADEM)

Diagnostic Criteria

Pediatric International Study Group:

- Encephalopathy
- 1st polyfocal clinical+ inflammatory demyelinating event
- No new lesions emerge in consecutive MRI (3months)
- Abnormal MRI during acute phase:
 - Diffuse large T2 lesions involving mostly the WM
 - Deep grey matter lesions
 - T1 hypointense lesions are rare

ADEM

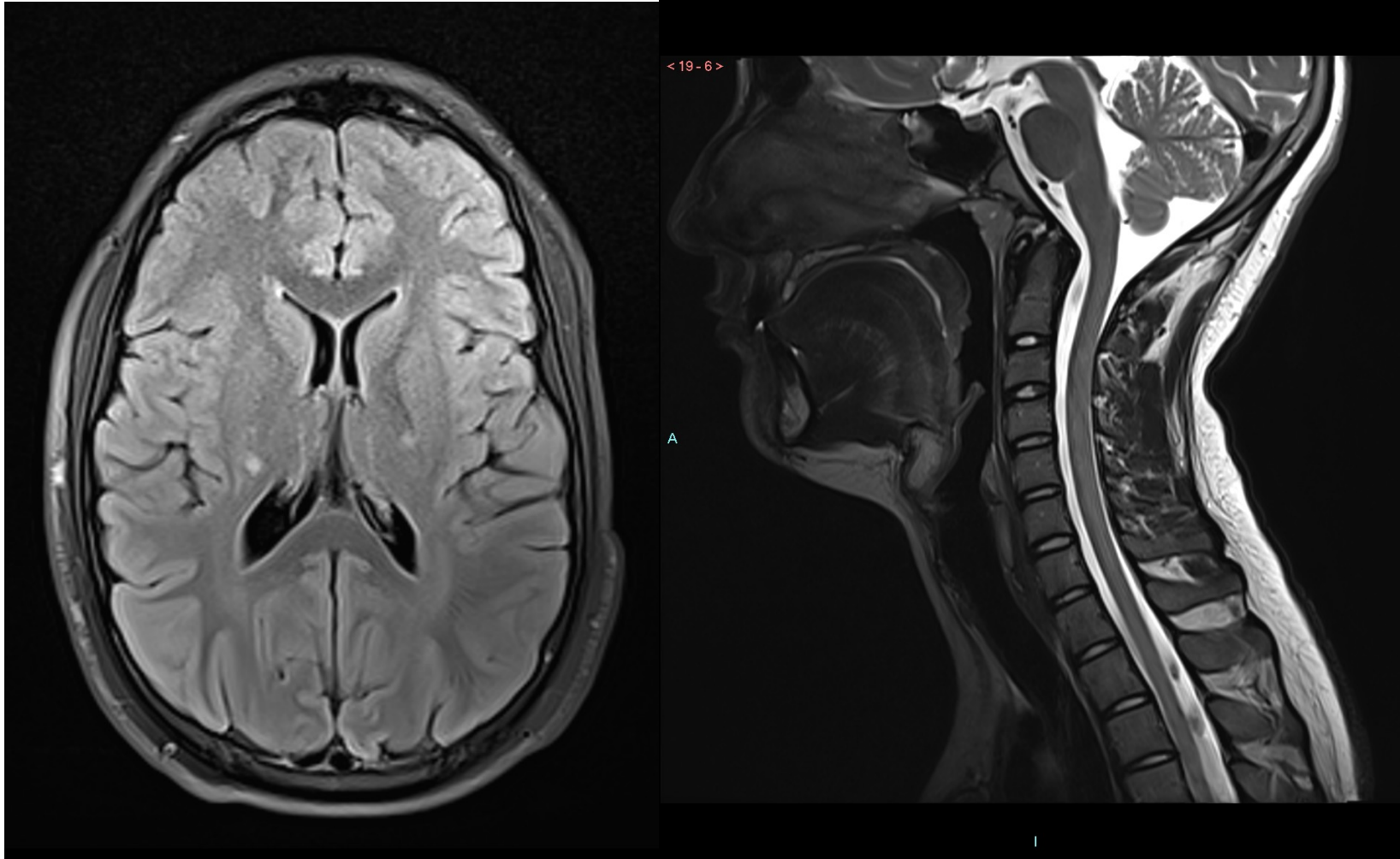


14 YO

febrile viral infection
2W before

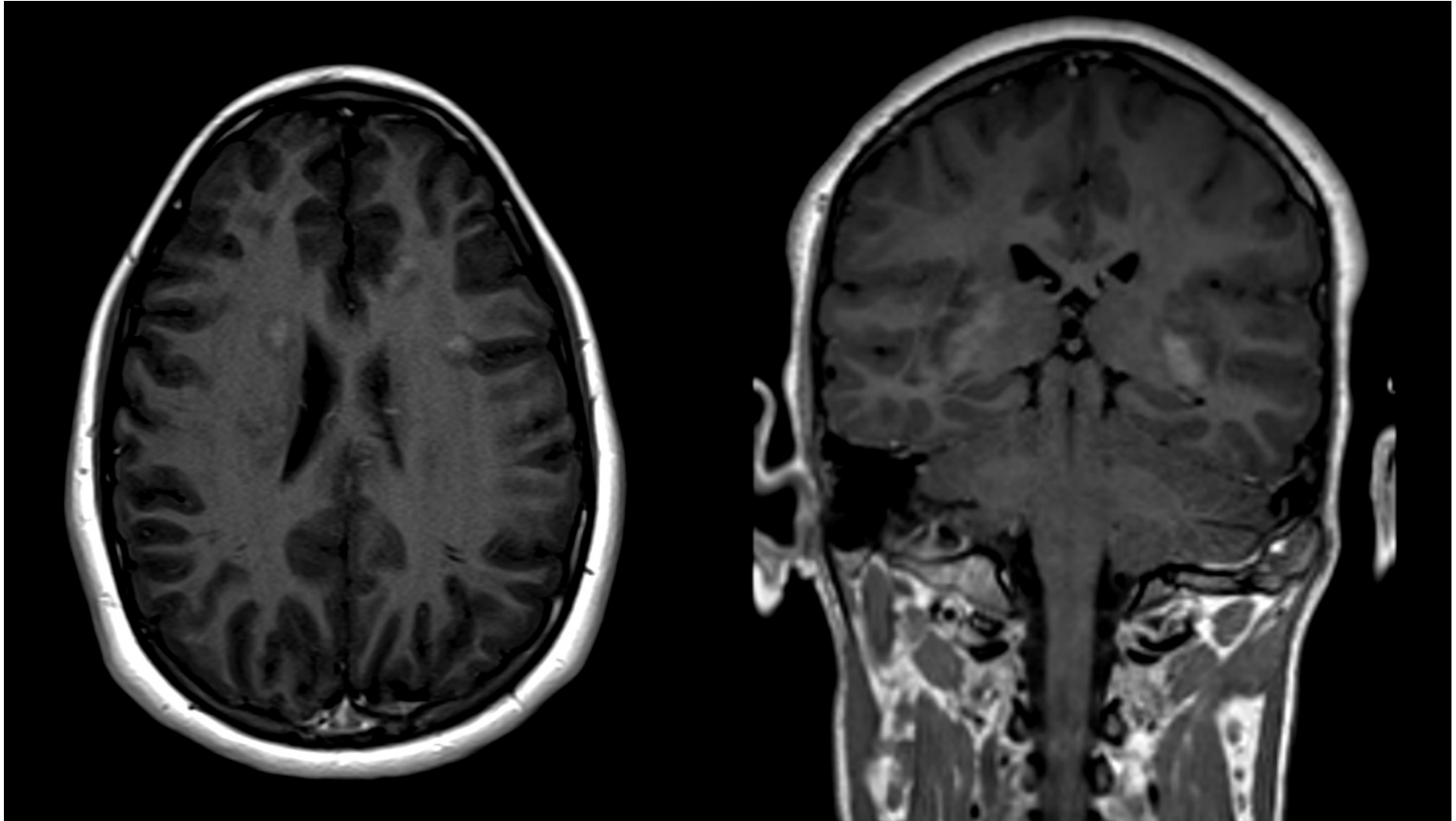
urine retention,
encephalopathy

ADEM F/U

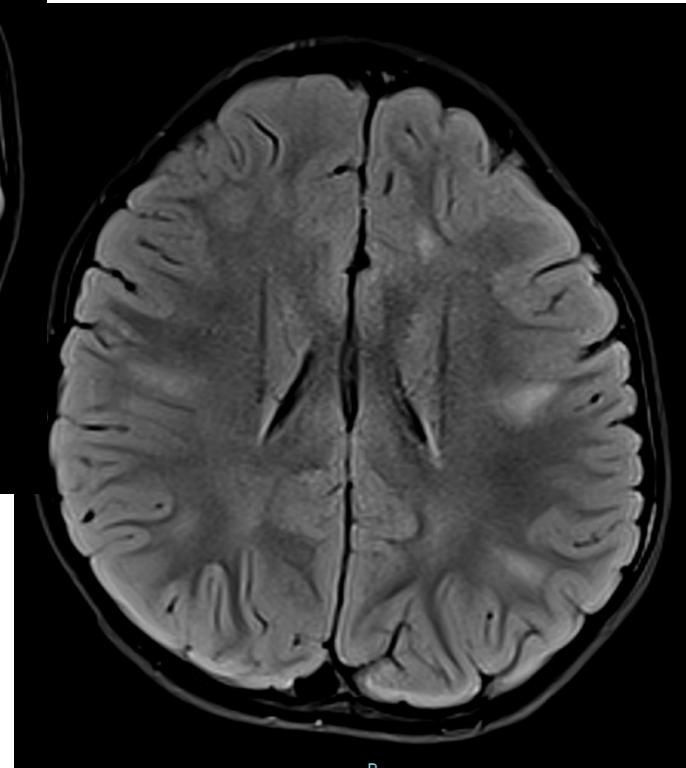
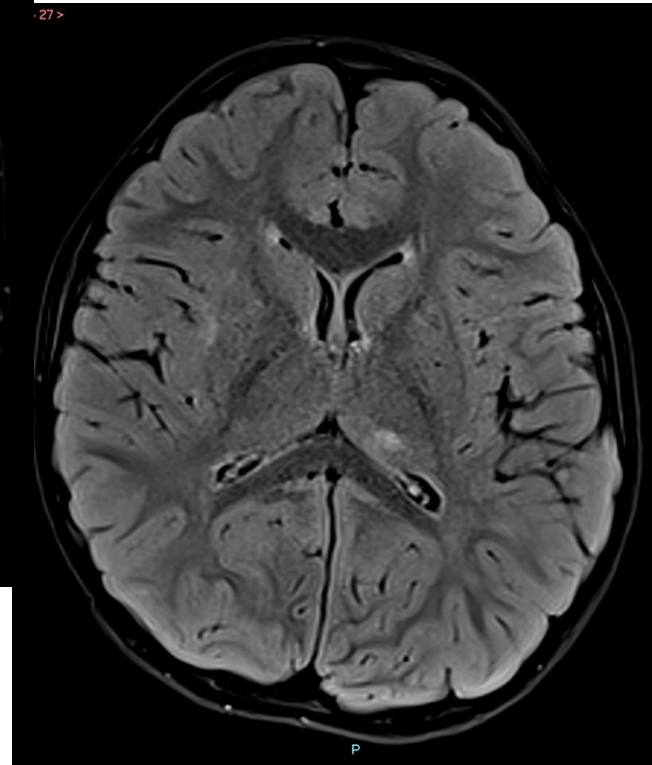


4 month later -Clinically intact

ADEM T1+ GAD



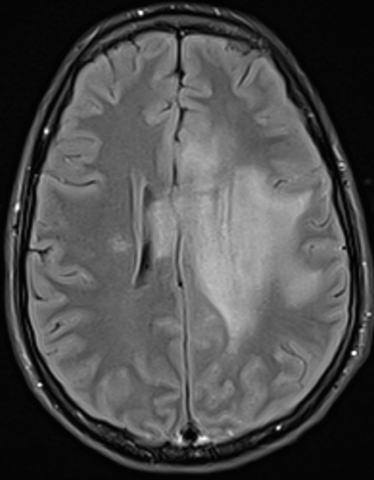
ADEM



2y ataxia, 3w post viral infection

< 56 - 23 >

R



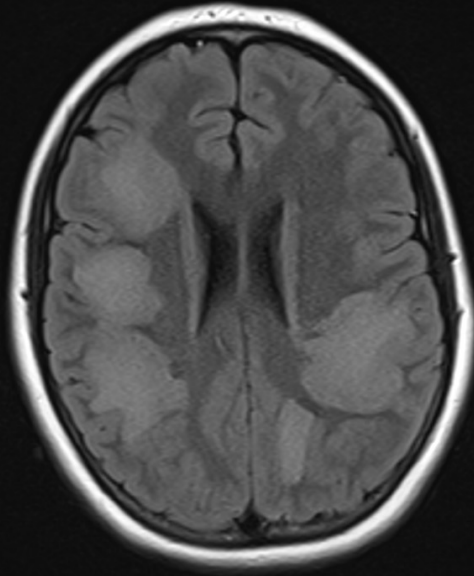
ADEM

All cases were initially diagnosed as ADEM

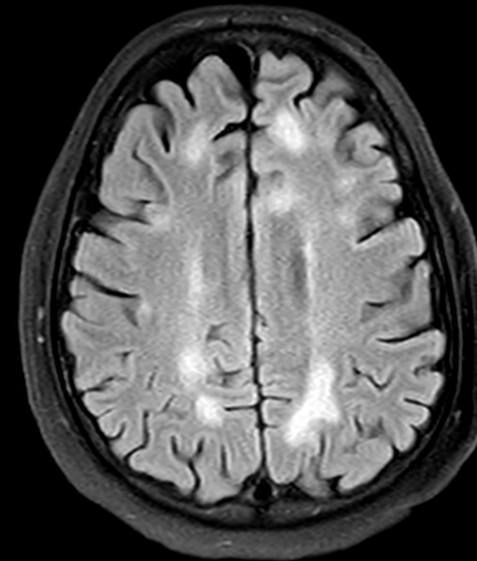
Vasculitis

301 - 24 0 >

R



MS



P

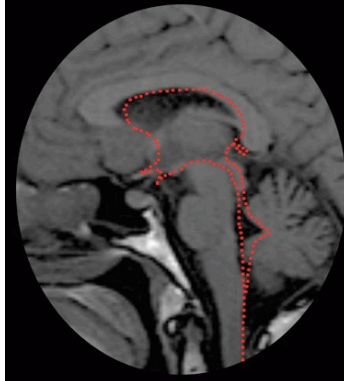
Multiple Diseases – Similar Appearances

- Multiple Sclerosis (MS)
- Acute Disseminated Encephalomyelitis (ADEM)
- **Neuromyelitis Optica (NMO)**
- Myelin Oligodendrocyte Glycoprotein Antibody-Associated Disease (MOGAD)

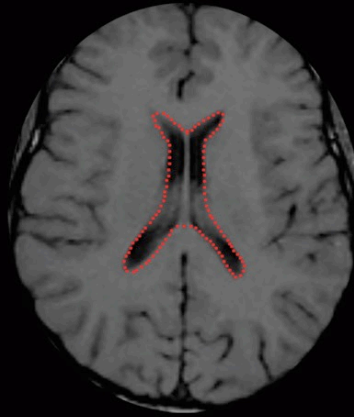
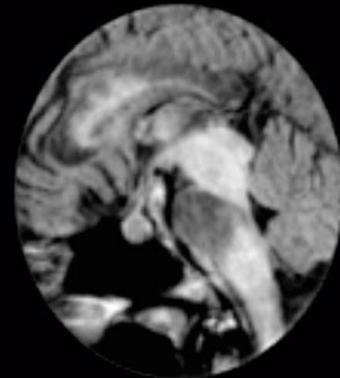
Neuromyelitis Optica (NMO)

- Diagnostic Criteria
Pediatric International Study Group:
 - Optic neuritis
 - Acute myelitis
 - At least 2 out of 3:
 1. Spinal cord lesion 3 segments
 2. Brain MR not meeting the criteria for MS
 3. Aquaporin-4 IgG seropositive status

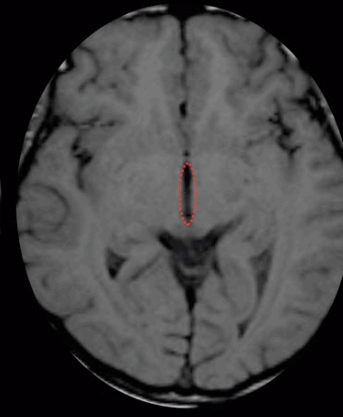
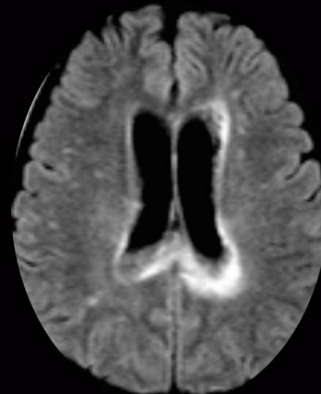
AQP-4



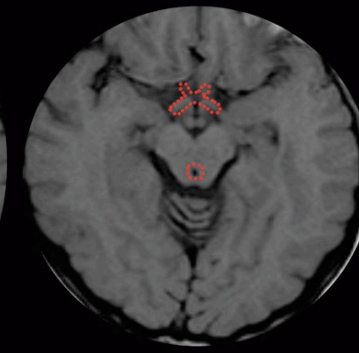
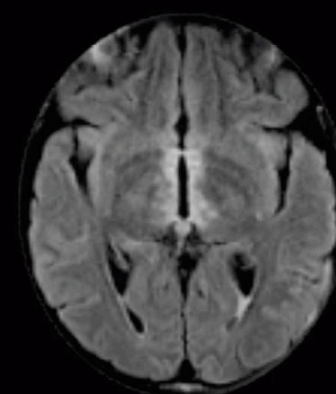
Periependymal surface of the corpus callosum, hypothalamus, periaqueductal area, area postrema, and the central canal of the spinal cord



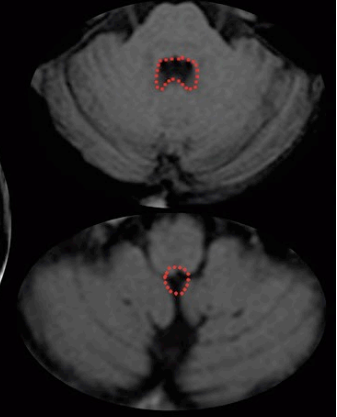
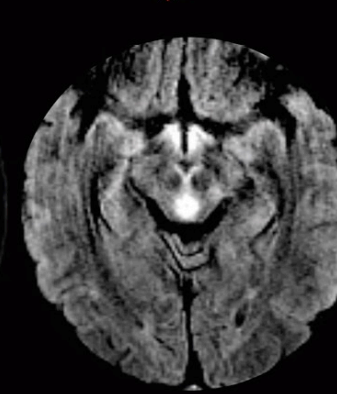
Periependymal surface of the lateral ventricles



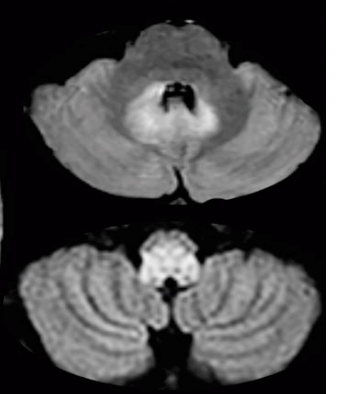
Periependymal surface of the third ventricle (diencephalic region)



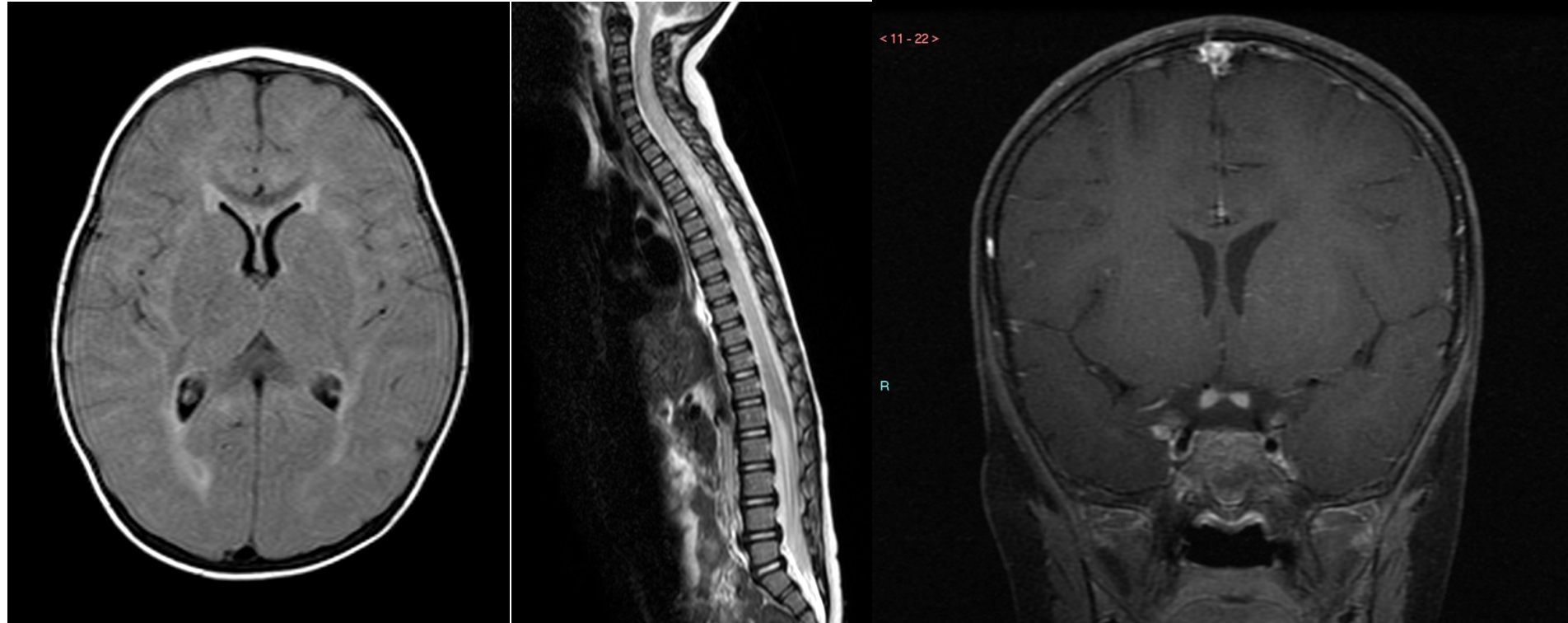
Periaqueductal area and optic chiasm



Periependymal surface of the fourth ventricle and area postrema

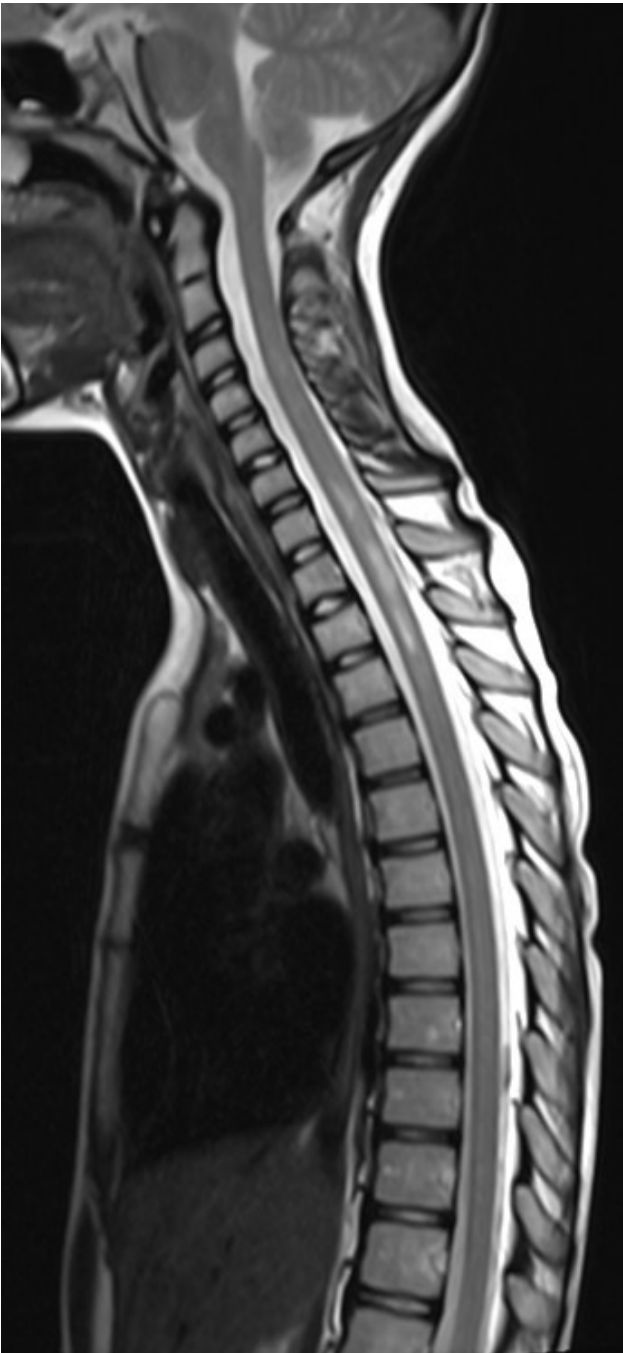


NMO




5y girl, sudden blindness on the R eye.
F/U scan after deterioration of the L eye

NMO



- 10y girl
- Weakness right hand
- Followed by pain in the L eye



Neuromyelitis Optica Spectrum Disorders: Spectrum of MR Imaging Findings and Their Differential Diagnosis¹

Bruna Garbugio Dutra, MD
Antônio José da Rocha, MD, PhD
Renato Hoffmann Nunes, MD
Antônio Carlos Martins Maia Júnior,
MD, PhD

Abbreviations: AQP4 = aquaporin-4, CLIP-PERS = chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids, CNS = central nervous system, CSF = cerebrospinal fluid, FLAIR = fluid-attenuated inversion-recovery, IgG = immunoglobulin G,

Neuromyelitis optica (NMO) is an autoimmune demyelinating disorder for which the aquaporin-4 (AQP4) water channels are the major target antigens. Advances in the understanding of NMO have clarified several points of its pathogenesis, clinical manifestations, and imaging patterns. A major advance was the discovery of the AQP4 antibody, which is highly specific for this disorder. Descriptions of new clinical and radiologic features in seropositive patients have expanded the spectrum of NMO, and the term *NMO spectrum disorder* (NMOSD) has been adopted. NMOSD is now included in a widening list of differential diagnoses. Acknowledgment

Multiple Diseases – Similar Appearances

- Multiple Sclerosis (MS)
- Acute Disseminated Encephalomyelitis (ADEM)
- Neuromyelitis Optica (NMO)
- **Myelin Oligodendrocyte Glycoprotein Antibody-Associated Disease (MOGAD)**

Myelin Oligodendrocyte Glycoprotein Antibody-Associated Disease (MOGAD)

- Antibodies mistakenly attack myelin oligodendrocyte glycoprotein (MOG), a protein in the myelin sheath
- Often triggered by an infection or vaccination
 - cases reported following SARS-CoV-2 infections and vaccinations

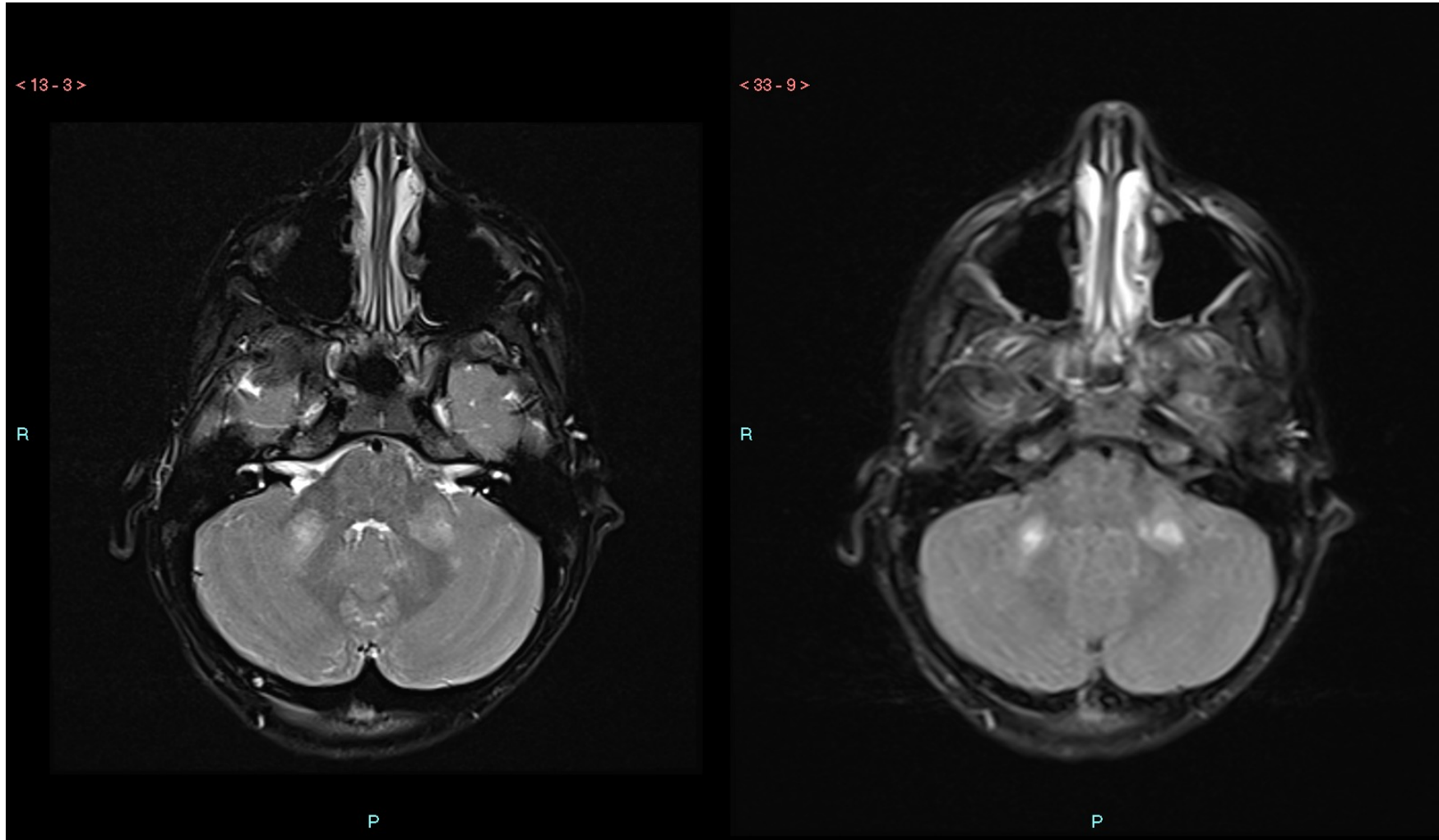
Myelin Oligodendrocyte Glycoprotein Antibody-Associated Disease (MOGAD)

- Optic neuritis (41-63%)
- Transverse myelitis (30%) including conus medullaris syndrome
 - associated with longitudinally extensive spinal cord lesions
- ADEM-like encephalomyelitis (2-6%)
- Aseptic meningitis (4%)
- Cortical encephalitis with seizures
- Infratentorial syndromes (i.e. brainstem or cerebellar syndromes)

MOGAD

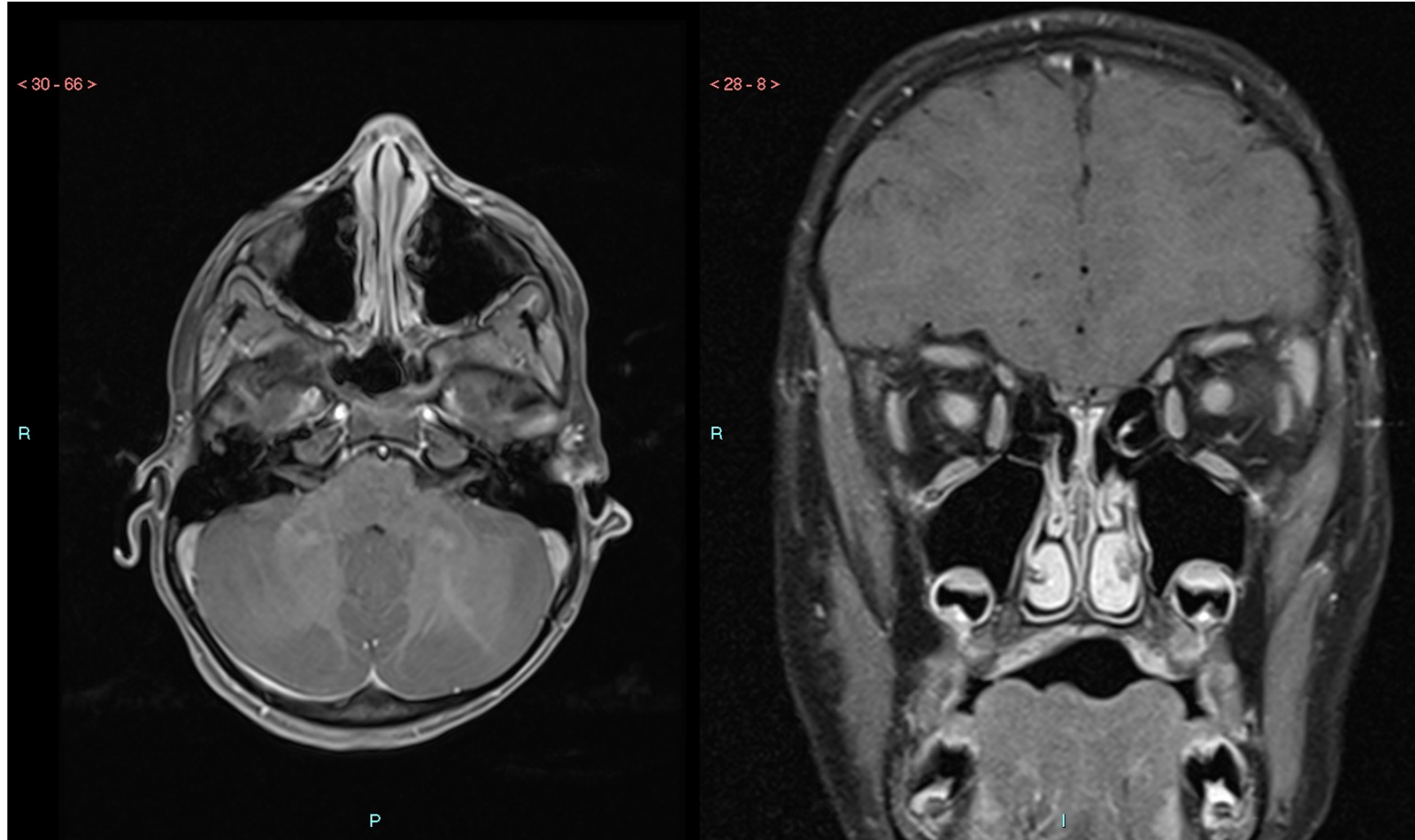
- 3y with a gradual loss of vision during last 3 weeks
- Transferred from another hospital with the diagnosis of bilateral optic glioma and NF1 (?)

Anti MOG Ab



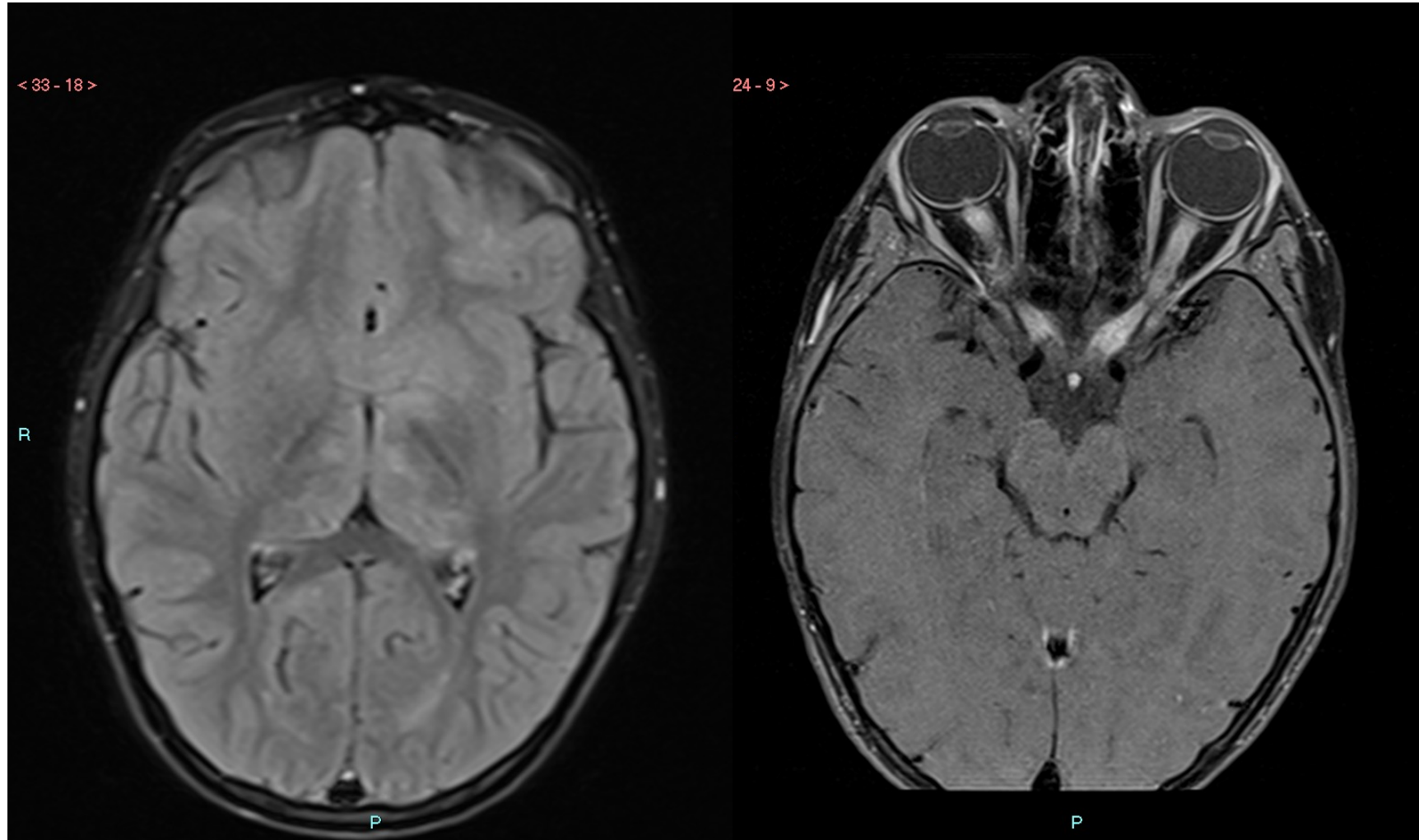
Enhancement bilaterally cerebellar peduncles

Anti MOG Ab



Both optic nerves enhancing

Anti MOG Ab



The enhancement is all along the nerves
Abnormal signal on both thalami



Radiological features in pediatric myelin oligodendrocyte glycoprotein antibody-associated disease—diagnostic criteria and lesion dynamics

Li-tal Pratt^{1,2} · Hadas Meirson^{2,3} · Mika Shapira Rootman⁴ · Liat Ben-Sira^{1,2} · Shelly I Shiran^{1,2}

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Radiological features and lesion dynamics in pediatric myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD), pediatric onset multiple sclerosis (POMS) and aquaporin 4-IgG-seropositive neuromyelitis optica spectrum disorder (AQP4+NMOSD)

	MOGAD	POMS	AQP4+NMOSD
MRI typical and/or common patterns at initial presentation			
Brain	<ul style="list-style-type: none"> ADEM features <ul style="list-style-type: none"> Multiple ill-defined T2 hyperintense lesions in supratentorial and often infratentorial white matter* Ill-defined T2 hyperintensity involving pons, middle cerebellar peduncle or medulla* Deep gray matter involvement* Leukodystrophy-like pattern in younger children -/+ nodular enhancement Cortical lesion with or without leptomeningeal enhancement* 	<ul style="list-style-type: none"> Multiple, well-defined, ovoid T2 hyperintense lesions in periventricular white matter (Dawson fingers) and juxta/intracortical regions Involvement of the corpus callosum - frequent -/+ Ring or open ring enhancement T1 hypointense lesions ('black holes') 	<ul style="list-style-type: none"> T2 hyperintense lesions in regions with high AQP4 expression: diencephalic regions surrounding the third ventricle and aqueduct, dorsal brainstem abutting the fourth ventricle – specifically the area postrema, and peripendymal circumventricular areas -/+ Patchy, cloud-like or pencil-thin linear peripendymal enhancement
Schematic representations of supratentorial brain cross-sectional involvement ^b			
Schematic representations of infratentorial brain cross-sectional involvement ^b			
Optic nerves	<ul style="list-style-type: none"> Bilateral simultaneous optic neuritis* Longitudinally extensive* (involving more than 50% of the optic nerve length) Anterior segments with sparing of optic chiasm and optic tracts Perineural optic sheath enhancement* Optic disc swelling* 	<ul style="list-style-type: none"> Unilateral optic neuritis Short segment Anterior segments 	<ul style="list-style-type: none"> Bilateral simultaneous optic neuritis Longitudinally extensive Posterior segments, including optic chiasm and optic tracts
Schematic representations of optic nerves cross-sectional involvement ^b			
Spinal cord	<ul style="list-style-type: none"> Longitudinally extensive transverse myelitis* <ul style="list-style-type: none"> Short-segment lesions may co-exist On axial images: central cord lesion or H-sign* >>> 'snake-eyes' sign Conus lesion* >= 2 lesions Enhancement: nodular (25%), diffuse nerve root enhancement (34%), leptomeningeal enhancement (69%) 	<ul style="list-style-type: none"> Short-segment lesions On axial images: peripheral involvement of lateral and dorsal columns Multiple lesions 	<ul style="list-style-type: none"> Longitudinally extensive transverse myelitis On axial images: central cord involvement with bright spotty lesion Cervical and upper thoracic cord Single lesion Enhancement: frequent, patchy
Schematic representations of spinal cord cross-sectional involvement ^b			
Lesion dynamics			
Intra-attack changes	<ul style="list-style-type: none"> Radiological lag – common Resolution of T2-lesions – uncommon Initial MRI may be normal in 10% of brain and spinal cord attacks 	<ul style="list-style-type: none"> Radiological lag – uncommon Resolution of T2-lesions – no 	<ul style="list-style-type: none"> Radiological lag – uncommon Resolution of T2-lesions – no
Disease course	Monophasic or relapsing (in 20–30%)	Relapsing	Relapsing
Follow-up T2-lesion resolution	Common	Rare	Rare
Follow-up new silent T2-lesions	Uncommon	Common	Rare

Conclusion

- MS of the pediatric age group is frequent
- The appearance and the clinical course are different than the adult type (more intense onset)
- Differential diagnosis – not always easy
- Correct diagnosis is needed for treatment decisions

Acknowledgements

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Queen Square MS Centre

European Course on
Pediatric Neuroradiology in
Latin America (ECPNR-LATAM)
3rd Cycle - Module 1

Questions?

Contact

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Idealização:



Organização:

