Exploring ADEM in MOGAD: A Case Study in a Young Adult

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HPI

- **21 y/o male** with PMHx of **epilepsy** presents to ED with **hypersomnolence, L-sided facial weakness, and dysarthria**
- Progressively worsening **malaise, fever, and fatigue the week prior**
- After admission, worsening hypoxia leading to intubation and sedation

Laboratory Findings

Cerebrospinal Fluid (CSF) Analysis:

- **Elevated white blood cell (WBC) count: 14 cells/µL (94% lymphocytes).**
- Glucose: 60 mg/dL (N = 50-80 mg/dL)
- Protein: 40 mg/dL (N = 15-60 mg/dL)
- Xanthochromia: None.

Serum Analysis:

- **Neutrophilic leukocytosis.**
- **Elevated ESR and CRP**

Infectious Disease Workup:

- Infectious workup from CSF (culture, VDRL, Biofire, etc.): Negative.
- **HSV positive penile ulcer yet serum/CSF negative**
- **Diagnosed with infectious mononucleosis at urgent care facility despite a negative mononucleosis test.**
- **Positive Rocky Mountain Spotted Fever (RMSF) IgM from serum**; IgG: Negative.

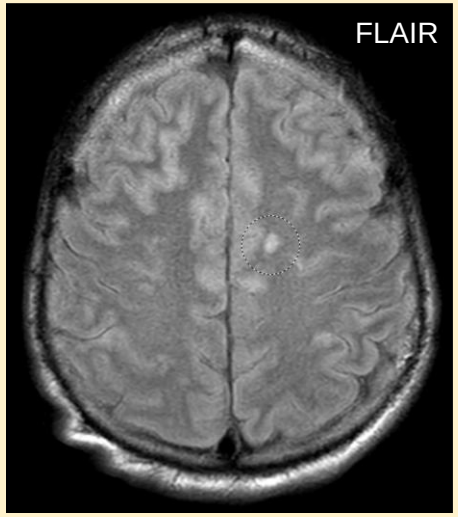
Autoimmune Studies:

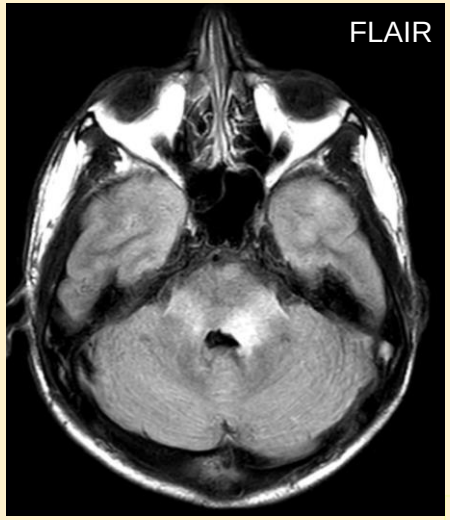
- CSF studies for autoimmune antibodies: **Positive for Myelin Oligodendrocyte Glycoprotein (MOG) antibodies.**

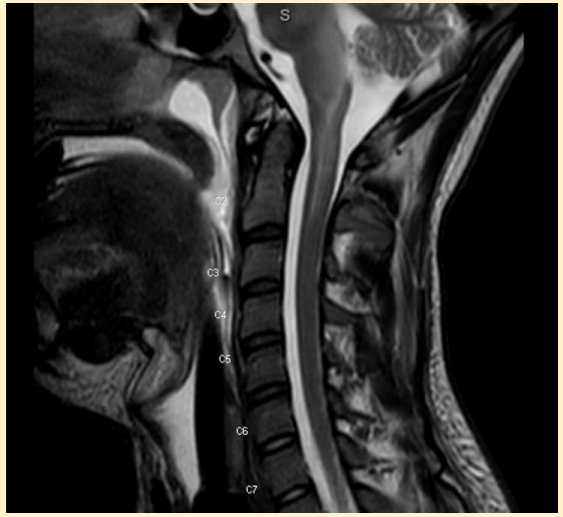
Day 1 Post-Admission

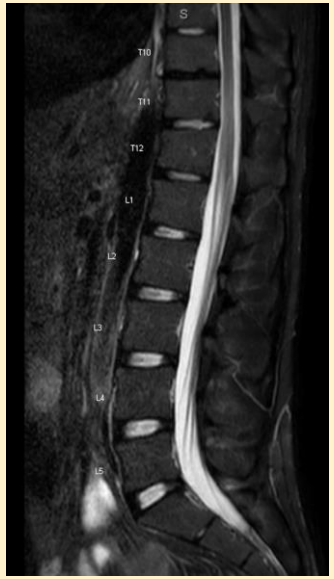
Week 1 Post-Admission (new findings)

Week 3.5 Post-Admission (f/u on initial findings)

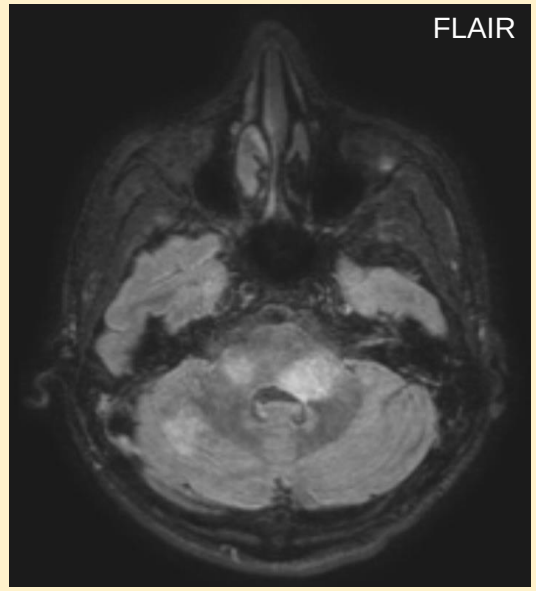


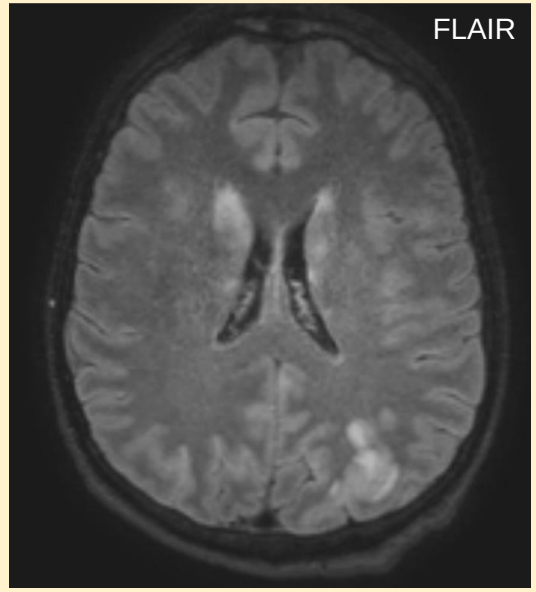


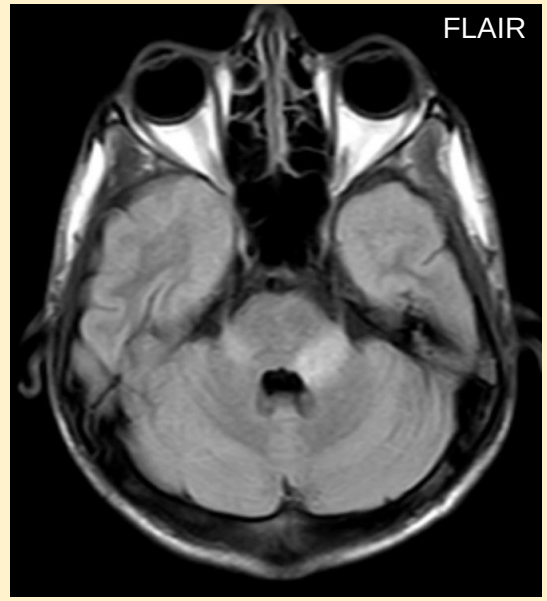


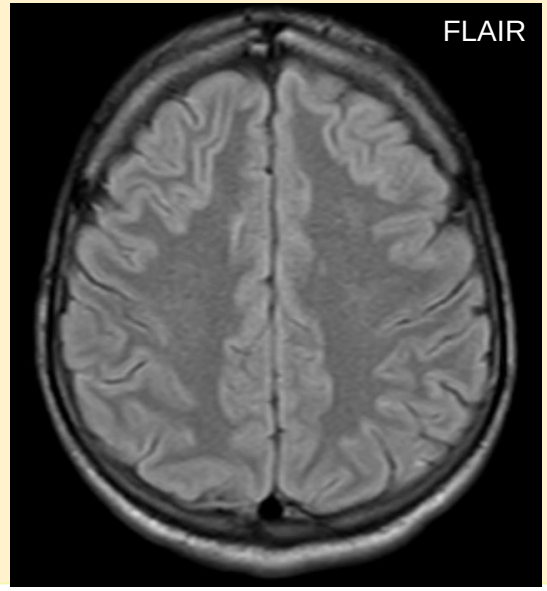


STIR







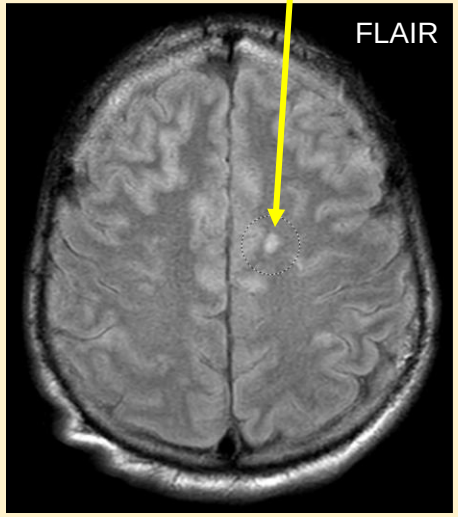


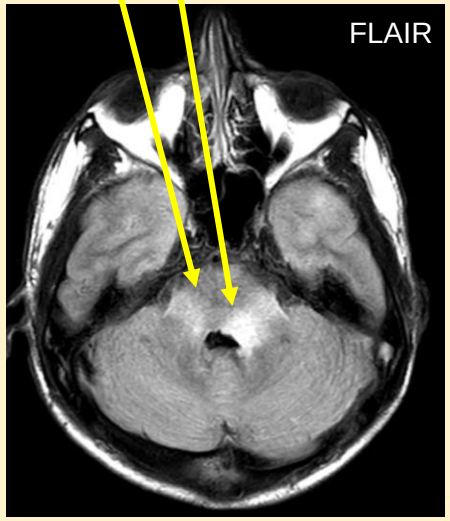
Day 1 Post-Admission

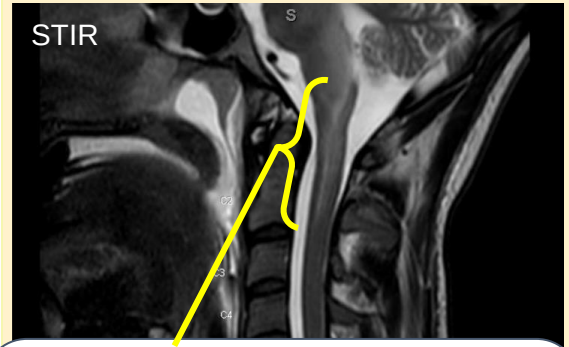
Week 1 Post-Admission (new findings)

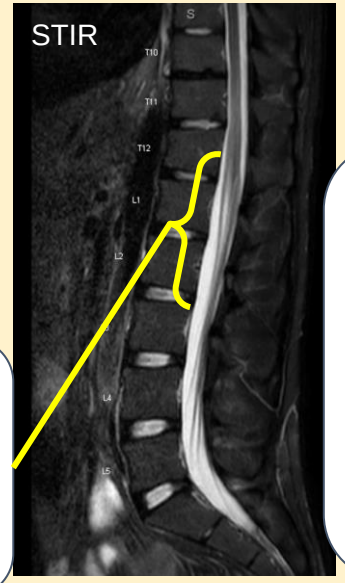
Week 3.5 Post-Admission (f/u on initial findings)

Supra & infratentorial FLAIR hyperintensities of white & grey matter. Cerebellar peduncle involvement. No diffusion restriction or abnormal enhancement

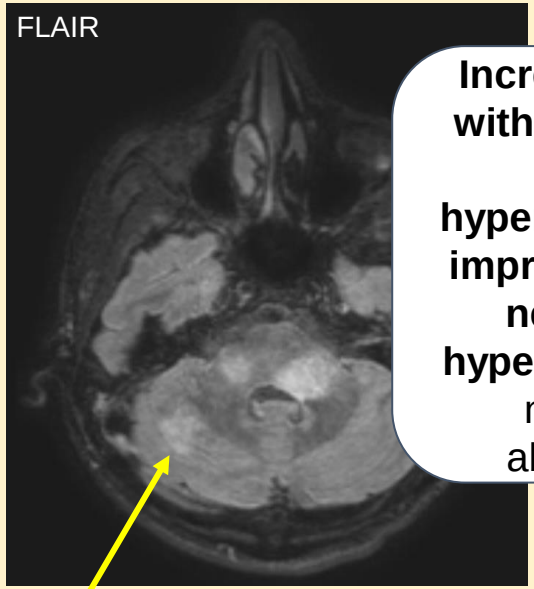




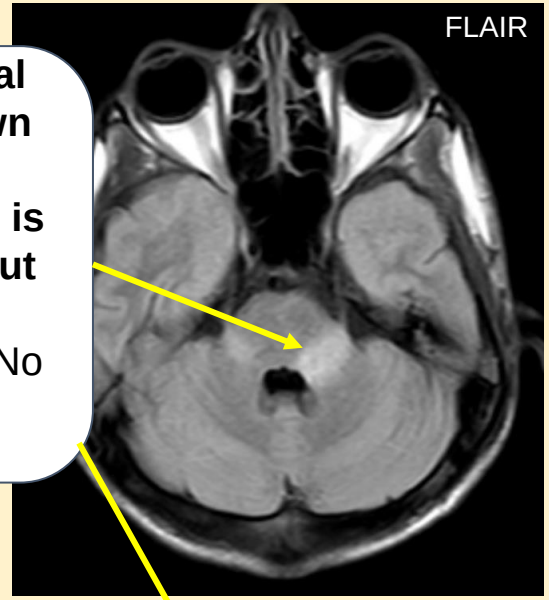




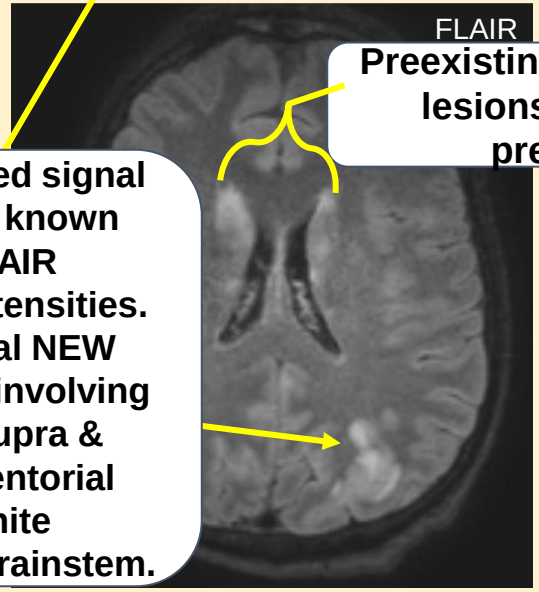
Multiple longitudinally extensive (≥ 3 levels) STIR hyperintensities in spinal cord, conus medullaris involvement. No abnormal post-contrast enhancement.



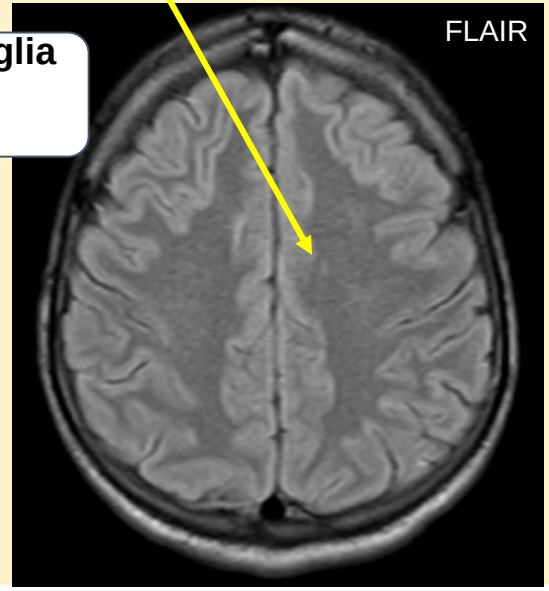
Increased signal within the known FLAIR hyperintensities is improved without new area of hyperintensity. No new acute abnormality.



Increased signal within known FLAIR hyperintensities. Several NEW lesions involving the supra & infratentorial white matter/brainstem.



Preexisting basal ganglia lesions not shown previously



Diagnosis/Intervention/Management

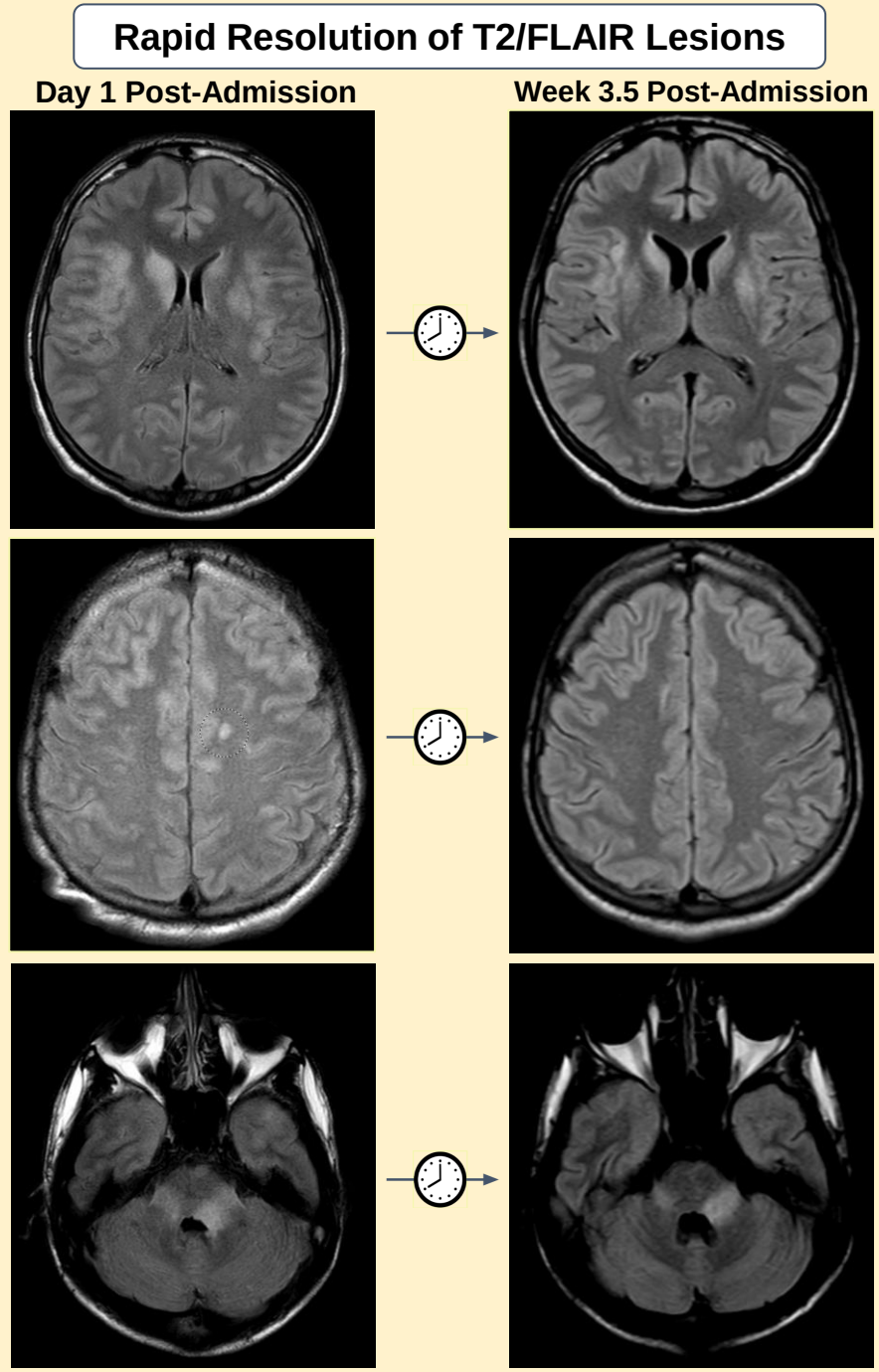
- Initial Differential Diagnosis: ADEM (acute disseminated encephalomyelitis), CNS demyelinating disorders (MOGAD (Myelin Oligodendrocyte Glycoprotein Antibody Disease), NMO, MS), other inflammatory/vascular pathologies
- Working Diagnosis: ADEM-manifestation of MOGAD, possibly precipitated by viral illness, unlikely RMSF-associated.

Diagnostic Criteria[1]	**Present Case**
(A) Core clinical demyelinating event	ADEM, Myelitis, Cerebral monofocal or polyfocal deficits, Brainstem or cerebellar deficits
(B) Positive MOG-IgG test	Clear positive
(C) Supporting clinical or MRI features	Conus lesion, Multiple T2 hyperintense lesions in supratentorial/infratentorial white matter, Deep gray matter involvement
Atypical findings in present case	No optic neuritis (ON; present in 41-63% of patients)[2], ADEM-like presentation (only ~10% of adult cases)[3], all viral testing negative

- Intervention: Steroids for ADEM (upon initial MRI findings), Plasma exchange for MOGAD (~1 week into hospital stay), Week-long course of doxycycline for possible RMSF infection
- Course/Outcome: Slow improvement in mental status over first 4 weeks, 5 weeks post-admission patient was ambulating/mentating normally, discharged week later.

Key MRI Findings suggesting MOGAD[3] over NMOSD/MS

- Multiple longitudinally extensive (≥ 3 levels) STIR hyperintensities in spinal cord
  - also extensive in NMOSD, not MS (short)
  - also multiple in MS, not NMOSD (single)
- Conus medullaris involvement
- T2/FLAIR hyperintensities of white & deep grey matter
- Extensive cerebellar peduncle involvement
- Rapid improvement/resolution (3 to 4 weeks later)



Discussion/Clinical Pearls

- The clinical presentation and imaging results of MOGAD are variable and overlap with other demyelinating conditions, often mimicking ADEM in children (less common in adults)

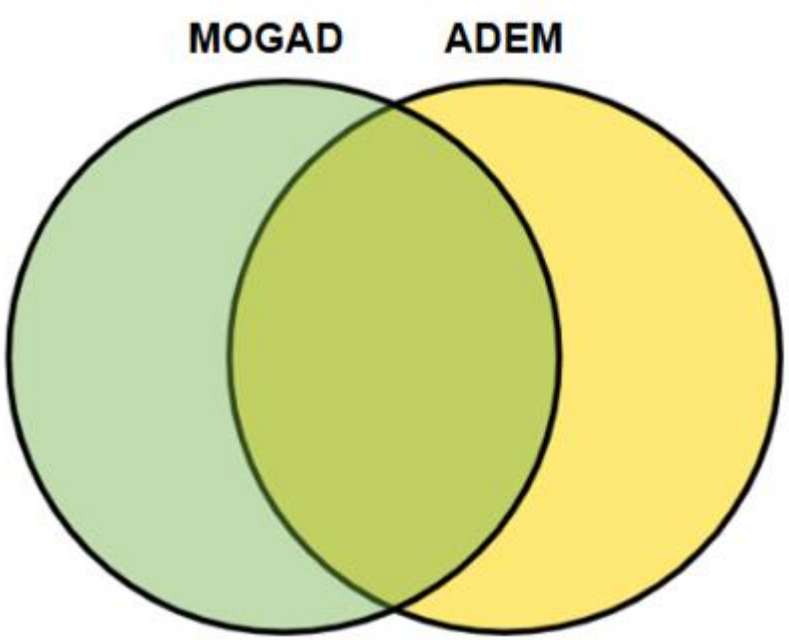
- However, specific clinical and radiological findings help differentiate MOGAD from other disorders. In addition to the MRI findings already discussed…
  - 37.5-70% of MOGAD cases begin with a viral prodrome, compared to NMOSD (15-35%) and MS (27-48%).[4]
  - In MOGAD, ON tends to be bilateral and involves a longer segment of nerve compared to MS. The anterior optic pathway is more frequently affected in MOGAD in contrast to the posterior in NMOSD.[4]

- MOGAD represents a spectrum of autoimmune disorders characterized by presence of MOG antibodies. ADEM is a clinical syndrome with more defined symptoms and progression.
  - The considerable overlap in each condition's presentation, particularly in children, highlights the complexity in distinguishing these conditions based solely on clinical symptoms.

Population-specific Relationship Between ADEM and MOGAD

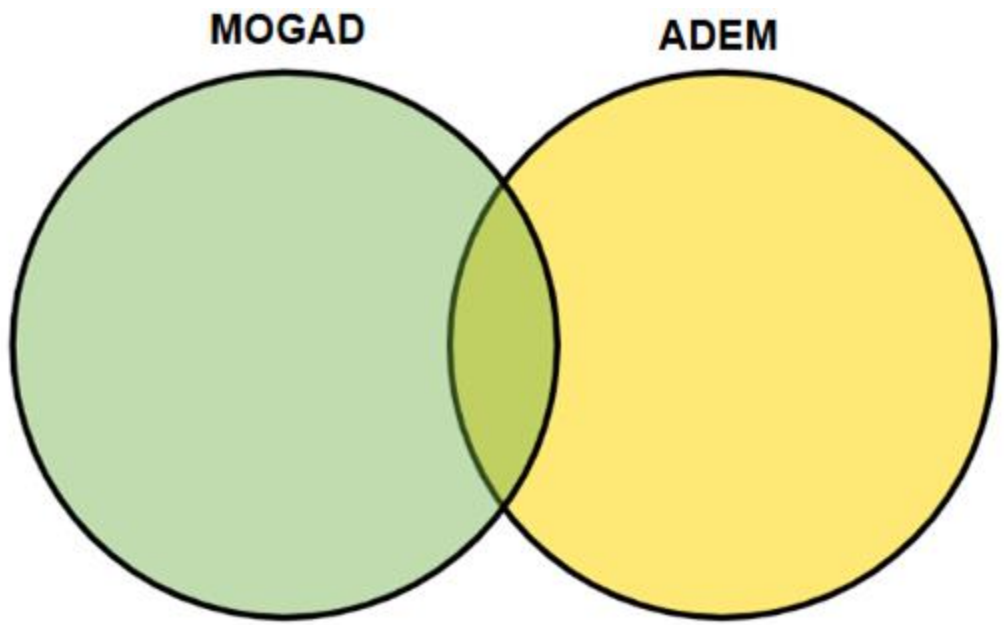
Children

- 40-50% of pediatric MOGAD cases have an ADEM-like phenotype.[5]
- 30-65% of pediatric ADEM patients are found to have MOG-antibodies.[6]



Adults

- Only ~10% of adult MOGAD patients with ADEM-phenotype.[3]
- Specific data not available on frequency of adult ADEM patients with MOG-antibodies



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