Anti-Myelin Oligodendrocyte Glycoprotein Antibody Associated Disease in Chile: a longitudinal study

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Introduction:

MOGAD is an emerging disorder recognised as a clinical entity distinct from MS and AQP4+NMOSD and its phenotypic spectrum continues to expand. Recently, the 2023 MOGAD diagnostic criteria were published.

Objective: To describe the clinical and paraclinical characteristics of monophasic and relapsing, paediatric and adult patients with MOGAD in Chile

Methods:

Observational, longitudinal retrospective and prospective multicentre study including patients
with positive anti-MOG-Ig using live (Mayo Clinic, Oxford University) or fixed (Euroimmun)
Cell-Based Assays with antibody titres since February 2023

Results:

We found 68 patients, 68% women, with a median age at onset 30 years (range 1–68), (23% onset < 18 years old), with a median disease duration of 29 months (range 4–1501). Patients from the Public healthcare system had a significantly longer diagnostic delay compared to Private health (median 13 months vs 1 month, p=0.04). In the whole cohort, the most frequent symptoms at onset were isolated optic neuritis (ON) (47%) and myelitis (18%). Encephalitis with seizures or encephalomyelitis was the most common presentation in pediatric-onset patients (60%), compared to 6% of adult-onset patients (p<0.001). A relapsing course was observed in 31%, these patients were younger at disease onset (24 vs. 33 years, p = 0.05) and with a trend in longer disease duration compared to monophasic patients (99 vs. 22 months, p=0.05). Three patients (2 pediatric, 1 adult) developed encephalitis with
seizures/status epilepticus, with concomitant positive CSF anti-NMDAR-IgG. The median EDSS at the last follow-up was 1.5 (range 1–6) for the relapsing cohort and 0 (range 0-3) for the monophasic cohort.
The diagnostic performance of the 2023 criteria was sensitivity 100%, specificity 99.7%, positive predictive value 97% and negative predictive value 100%

Table 1: Baseline characteristics of included patients N=68	
Median age at onset years(range)	34 (1-68)
Pediatric onset n(%)	14(21)
Female sex n(%)	47(68)
Healthcare private(%):public(%)	62:38
Median diagnostic delay months (range)	1 (0-379)
Initial symptoms	
Optic neuritis	64% (37) (46% un- 54% bilat)
Transverse myelitis	20% (14) (64% short-36% LETM)
Encephalitis	6%
Brainstem	7%
ADEM	15%
Paraclinical Findings	
Positive oligoclonal bands	9/51 (18%)
Concomitant CSF anti NMDAr-IgG	3 patients with encephalitis/FLAMES

Table 2: Follow-up characteristics of included patients	
Acute treatment n(%)	61(90)
Oral steroids	75%
Intravenous steroids	89%
IVIg	11%
PLEX	5%
Relapsing course n(%)	**19(28)**
Median number of relapses (range)	2 (2-6)
Median follow-up months (range)	104 (14-380)
Chronic treatment n(%)	**75% (51)**
Oral steroids	14%
IVIg	4%
Rituximab	20%
Azathioprine	22%
Mycophenolate	47%
EDSS at the last visit (range)	**1.5(0-6.0)**

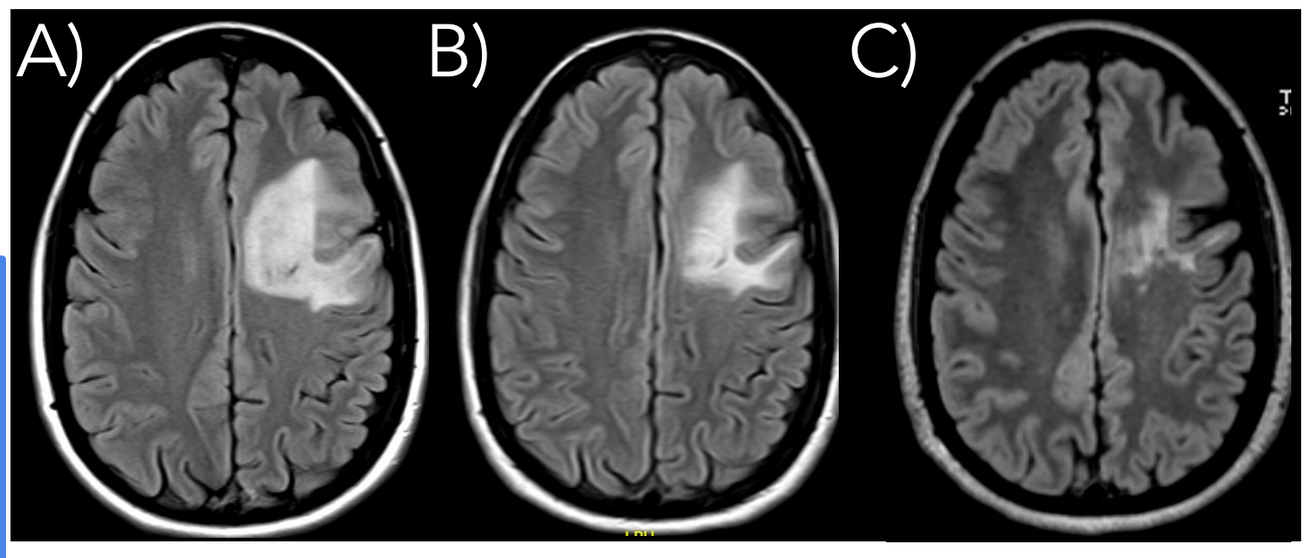

Figure 1. *Representative axial FLAIR images of a female patient with relapsing tumefactive demyelination with epilepsy.*
First relapse at 15 years old, biopsy consistent with tumefactive demyelination. Diagnosis of MOGAD (live-cell based assay) during her 4th relapse at 39 years old. A) May, 2019. B) June, 2019. C) January, 2020.

CONCLUSIONS:

In Chile, patients with MOGAD exhibit a broad spectrum of clinical presentations at disease onset and during relapses, consistent with previous reports in pediatric and adult populations, with a higher proportion of female patients. Interestingly, patients from the Public Health System, had significantly longer diagnostic delays compared to Private Health patients, raising concerns about the education of the healthcare professionals concerning this emerging entity. Close monitoring is needed, particularly in younger patients with short follow- up periods, as a relapsing course was observed in about 30% the cohort.

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