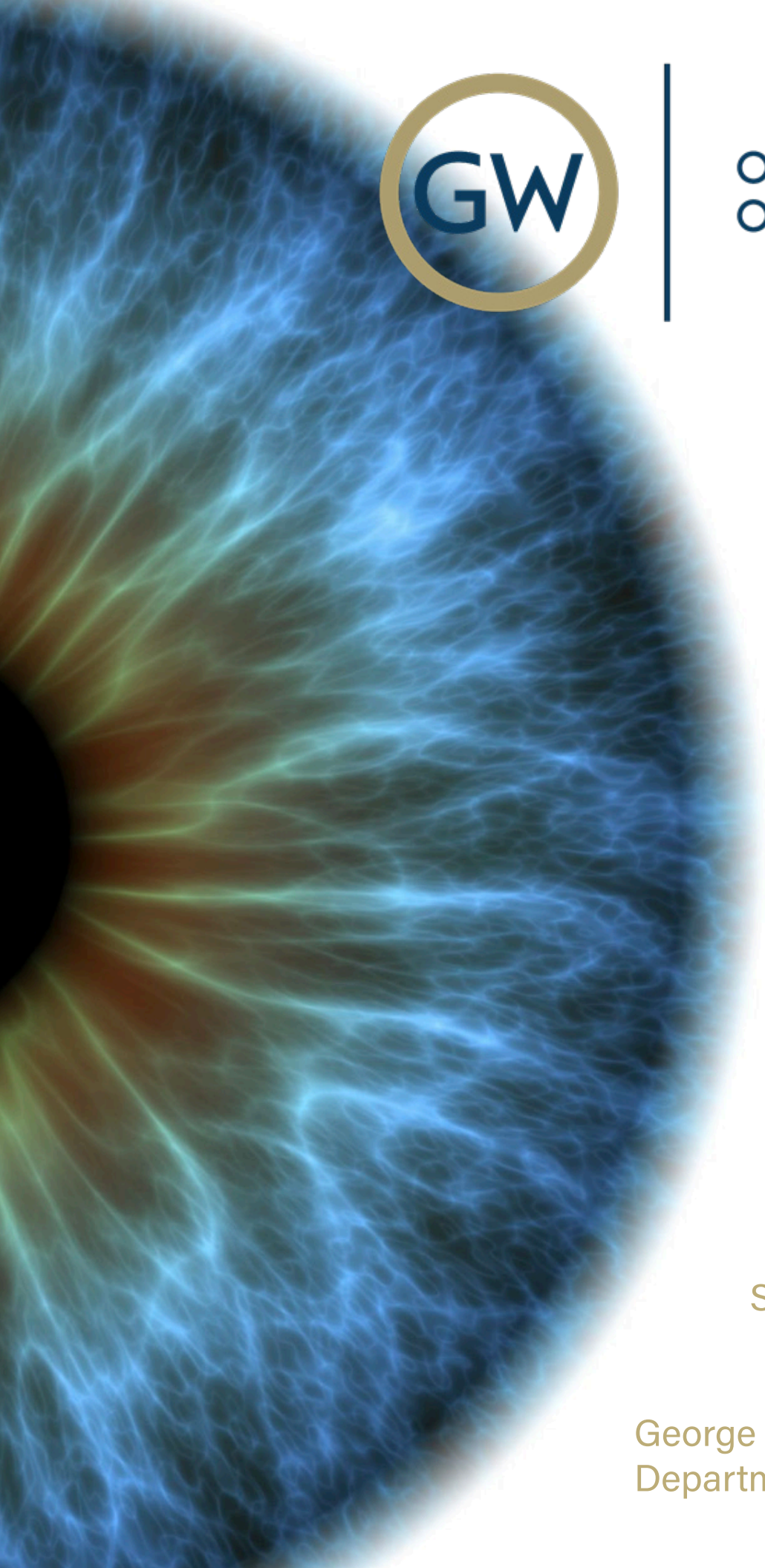
9-27-2024

GW Ophthalmology On-Call: September 2024

Adam Dawer

Jenny Lemus



OPHTHALMOLOGY ON CALL

September 2024
Edition 1

George Washington University
Department of Ophthalmology

Table of Contents

Introduction

GW Ophthalmology On Call is a monthly case report publication authored by medical students and residents in the GW Department of Ophthalmology. This publication aims to present educational cases encountered in the department, offering valuable learning opportunities for clinicians and students alike. The content is intended solely for educational purposes and is not meant to guide patient treatment. All cases adhere to HIPAA guidelines, with patient details modified to ensure privacy.

Published online at
https://ophthalmology.smhs.gwu.edu/news/topic/ophthalmology-call

Publication Date
27 September 2024

Disclosures
Disclosure forms are available with the article online

Corresponding Authors
Adam Dawer, BS. Jenny Lemus, BS

Medical Student Editor
Shaleen Arora, BS

Resident Editors
William Foos, MD
Julie Thomasian, MD

Attending Reviewer
David Belyea, MD MBA

Editors in Chief
Jason Dossantos, BS
Yusuf Bade, BA
Hannah Becker, MFA

How to Cite
Dawer A, Lemus J, et.al. MOG Antibody Disease. GW On-Call Clinical Cases. 2024

MOG Antibody Disease

Adam Dawer, BS[1]; Jenny Lemus, BS[1]
[1]George Washington Hospital/GW School of Medicine and Health Sciences

History of Present Illness

A 9-year-old female presented to the emergency department with a one month history of headaches with associated sleep disturbance, right sided vision loss, and an episode of right upper extremity paralysis. She also reported pain with extraocular movement OS, but denied flashes, floaters, curtaining, or tinnitus.

She has no previous ocular history, no past medical history, takes no medications, has no allergies, no significant family medical history and does not use illicit drugs, tobacco or alcohol.

During review of systems, she endorsed nausea and headaches but all other systems are negative.

Ocular Exam

Physical Exam		
	OD	OS
Visual Acuity	20/125	20/25
Intraocular Pressure (mmHg)	16	13
Pupils	+APD	Round and reactive
Extraocular Motility	EOMI	EOMI

Slit Lamp Exam		
	OD	OS
External	Normal	Normal
Lids and Lashes	Normal	Normal
Conjunctiva/ Sclera	White and quiet	White and quiet
Cornea	Clear	Clear
Anterior Chamber	Deep and quiet	Deep and quiet
Iris	Normal	Normal
Lens	Clear	Clear

Dilated Fundus Exam		
	OD	OS
Cuo to Disc	Normal	Normal
Optic Nerve	Edema grade 3	Edema grade 2
Vessels	White and quiet	White and quiet
Macula	Clear	Clear
Periphery	Deep and quiet	Deep and quiet
Vitreous	Normal	Normal

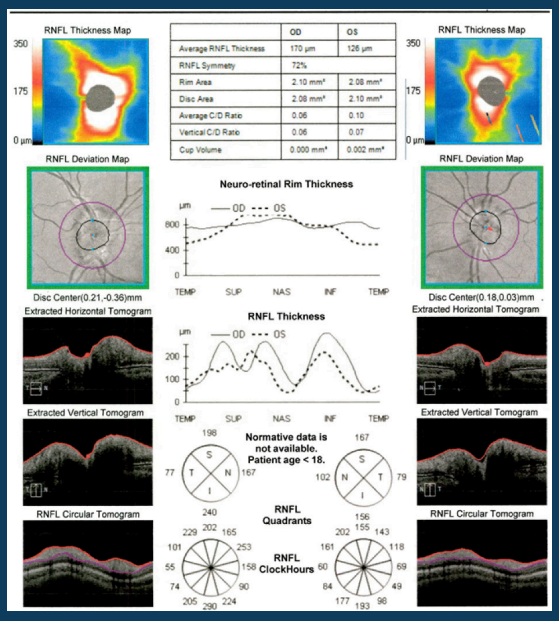


Figure 1: Initial OCT showing retinal nerve fiber layer thickening.

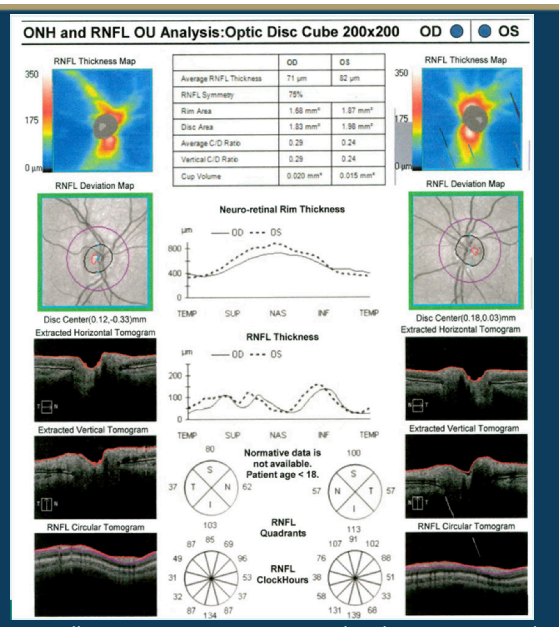


Figure 2: Follow-up OCT RNFL at 3 months, demonstrates a decrease in RNFL thickness compared to initial presentation, with the right eye showing further loss and abnormal thinning.

Differential Diagnosis

- Neuromyelitis optica spectrum disorder (NMO)
- Multiple sclerosis (MS)
- Optic neuritis (ON)
- Transverse Myelitis (TM)
- Acute disseminated encephalomyelitis(ADEM)

Clinical Course

A 9-year-old female was admitted to the hospital with a one month history of daily headaches, nausea, and pain with extraocular movement with no reported prodrome. Ophthalmology was consulted and on exam, she was found to have bilateral disc edema (grade 3 OD and grade 2 OS) with loss of color vision, R sided visual field deficit, significant right afferent pupillary defect (APD), and slow mentation. She was given a 5-day course of IV methylprednisolone.

On Day 4, the patient reported a reduction in headaches. A Humphrey visual field (HVF) test revealed significant restriction of her right visual field and an enlarged blind spot in her left eye. A repeat dilated fundus exam showed improvement in bilateral disc edema, now grade 2 OD and grade 1 OS. An MRI revealed bilateral optic and spinal enhancement on several levels. A MOG antibody test was ordered during her hospital stay, and the results confirming the presence of MOG antibodies were received after her discharge. She finished her course of IV methylprednisolone and was discharged with a cholecalciferol prescription and a diagnosis of bilateral ON and transverse myelitis (TM). At time of discharge, her visual acuity had improved to 20/25 and bilateral disc edema had resolved.

On Day 20, she presented with blurred vision and floaters in her right eye as well as pain in her left eye. She was given a 5-day course of IV methylprednisolone and a 2-day course of intravenous immune globulin (IVIG) which

resolved her left eye pain but did not improve right eye vision. At this time her relative APD was still present; however, vision, color vision, and optic nerve elevation/RNFL thickness were significantly improved from initial admission, thus she was discharged with a diagnosis of myelin oligodendrocyte glycoprotein antibody disease (MOGAD) and a 14-day oral Prednisolone prescription.

At her subsequent one month follow up appointment, her visual function had returned to normal with restored central visual acuity. She still had a mild temporal deficit in her right eye; however, HVF reported significant improvement in the rest of her visual field and optic nerve thickness had returned to normal, with OCT RNFL values of 71 microns OD and 82 microns OS. She is scheduled for serial follow up to monitor her progression of symptoms, but there is currently no indication for immunomodulating therapies.

Discussion

Myelin oligodendrocyte glycoprotein antibody disease (MOGAD) is a rare, immune-mediated, demyelinating disorder of the central nervous system which causes inflammation of the optic nerve, spinal cord, and/or the brain.[1,2] Myelin oligodendrocyte glycoproteins (MOG) are found exclusively on the surface of myelin sheaths in the CNS and are targeted by the immune system in patients with MOGAD.[2] MOGAD is generally considered idiopathic, typically presenting without any prodrome.[2] There have been associations with demyelinating encephalitis, post-infectious demyelination from herpes simplex virus, Borrelia, Epstein-Barr infections, and relapsing multiple sclerosis (MS), but it is unclear if MOG antibodies play a pathogenic role or are epiphenomenal.[2]

Patients with related CNS inflammatory demyelinating diseases, such as neuromyelitis optica spectrum disorders (NMOSD) and MS, often present with overlapping clinical phenotypes as MOGAD, such as optic neuritis (ON) and transverse myelitis (TM), making diagnosis and management challenging.[3,5] MOGAD is unique however since it typically follows a monophasic course of acute disseminated encephalomyelitis (ADEM) with distinct lesions, as well as perivascular infiltrate with MOG-laden macrophages and CD4 T-cells[6]; it also does not link to other autoimmune diseases, unlike NMOSD.[4] Moreover, MOGAD lacks a clear sex predilection, has a higher incidence in children compared with adults, and is more frequent in Caucasians.[4,5]

The initial clinical signs of MOGAD often include eye pain with extraocular movement and optic disc edema, as observed on dilated fundus examination.[7] Furthermore, 95% of patients experience complete visual field loss or a central scotoma[8]. Blurred vision, loss of color vision, seizures, and partial paralysis are common additional symptoms.[4,9]

MRI is the preferred imaging modality for diagnosing MOGAD, as it helps identify demyelinating lesions in the brain and spinal cord. Characteristic findings include hyperintense lesions often located in the pons near the fourth ventricle that have a distinctive fluffy appearance, as well as spinal cord lesions affecting central and lateral sides.[5] Diagnosis of MOGAD is made through a combination of clinical evaluation, imaging studies and laboratory tests. A key diagnostic element involves testing for MOG-antibodies through specific assays.[1] Additionally, testing for aquaporin-4 (AQP4) is important to differentiate between NMOSD. The presence of MOG antibodies and negative AQP-4-IgG support the diagnosis of MOGAD.[1,5]

The treatment of MOG antibody disease involves a combination of pharmacological

The treatment of MOG antibody disease involves a combination of pharmacological therapy to eliminate MOG antibodies. The specific treatment approach varies based on individual patient characteristics, such as age. For adults, treatment typically involves intravenous methylprednisolone (IVMP) at doses of 1-2g/kg/day over 3-5 days.[8] First-line treatment for pediatric patients include IVMP with a dose of 20-30 mg/kg/day over 3-5 days.[6,8] Acute treatment in the pediatric population includes high dose IVIG or plasma exchange. Additionally, maintenance options include azathioprine with mycophenolate mofetil which takes 3-6 months before being fully effective, and rituximab.[8] Prompt intervention is pivotal in managing MOGAD as 50% achieve nearly complete recovery, while 44% experience partial improvement.[6]

Clinical Pearls

1. Early recognition and prompt treatment with intravenous methylprednisolone are essential for improving visual and neurological outcomes in MOGAD.
2. Long-term follow-up and monitoring are necessary due to the risk of relapses, even after initial symptom resolution.
3. Diagnosis of MOGAD should include testing for MOG antibodies and differentiating it from NMOSD with aquaporin-4 testing.

References

1. Ambrosius W, Michalak S, Kozubski W, Kalinowska A. Myelin Oligodendrocyte Glycoprotein Antibody-Associated Disease: Current Insights into the Disease Pathophysiology, Diagnosis and Management. Int J Mol Sci. 2020;22(1):100. Published 2020 Dec 24. doi:10.3390/ijms22010100

2. Wynford-Thomas R, Jacob A, Tomassini V. Neurological update: Mog Antibody disease. Journal of Neurology. 2018;266(5):1280-1286. doi:10.1007/s00415-018-9122-2

3. Al-Ani A, Chen JJ, Costello F. Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD): current understanding and challenges. J Neurol. 2023;270(8):4132-4150. doi:10.1007/s00415-023-11737-8

4. Hacohen Y, Banwell B. Treatment Approaches for Mog-Ab-Associated Demyelination in Children. Current Treatment Options in Neurology. 2019;21(1). doi:10.1007/s11940-019-0541-x

5. Santoro JD, Beukelman T, Hemingway C, Hokkanen SRK, Tennigkeit F, Chitnis T. Attack phenotypes and disease course in pediatric MOGAD. Ann Clin Transl Neurol. 2023;10(5):672-685. doi:10.1002/acn3.51759

6. Marignier R, Hacohen Y, Cobo-Calvo A, et al. Myelin-oligodendrocyte glycoprotein antibody-associated disease [published correction appears in Lancet Neurol. 2021 Oct;20(10):e6] [published correction appears in Lancet Neurol. 2022 Jan;21(1):e1]. Lancet Neurol. 2021;20(9):762-772. doi:10.1016/S1474-4422(21)00218-0

7. Nurul-Ain M, Khairul Kamal ZN, Wan Hitam WH, Abd Munaaim M, Mohd Zaki F. Myelin Oligodendrocyte Glycoprotein (MOG) Optic Neuritis: A Case Series. Cureus. 2021;13(4):e14452. Published 2021 Apr 13. doi:10.7759/cureus.14452

8. Autoantibody-Binding Sites: A Review. International Journal of Molecular Sciences. 2023; 24(17):13368. https://doi.org/10.3390/ijms241713368

9. Olbert E, Brunner C, Alhani N, Našel C, Struhal W. Mog Antibody Associated Disease (mogad) presenting with extensive brain stem encephalitis: A case report. eNeurologicalSci. 2022;29:100432. doi:10.1016/j.ensci.2022.10043

Case Report Authors
Adam Dawer, BS. Jenny Lemus, BS

Medical Student Editor
Shaleen Arora, BS

Resident Editors
William Foos, MD
Julie Thomasian, MD

Attending Reviewer
David Belyea, MD MBA
Marlet Bazemore, MD MPH

Editors in Chief
Jason Dossantos, BS
Yusuf Bade, BA
Hannah Becker, BFA