Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) masquerading as a quiet optic neuropathy: A case of bilateral vision loss and hearing deficit

Shim Ni Lim[1,2], Wan Hazabbah Wan Hitam[1], Jaya Vani Ettikan[2], Jack Son Wee[3], Hayati Abdul Aziz[2]

[1]Department of Ophthalmology & Visual Science, School of Medical Sciences, Health Campus, Universiti Sains Malaysia, Kubang Kerian, Kelantan, Malaysia, [2]Department of Ophthalmology, Hospital Sultanah Aminah, Johor Bahru, Johor, Malaysia, [3]Department of Neurology, Hospital Sultanah Aminah, Johor Bahru, Johor, Malaysia

SUMMARY

Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) is a demyelinating disorder causing oligodendropathy, often presenting with severe optic disc swelling. We describe a diagnostically challenging case lacking this hallmark feature, instead presenting as a quiet bilateral optic neuropathy with rare concurrent auditory involvement. A 38-year-old man presented with profound, sequential, painless bilateral vision loss over two weeks, accompanied by hearing impairment. Visual acuity was reduced to hand movements in the right eye and vague light perception in the left eye. No relative afferent pupillary defect was detected. Anterior segment examination was unremarkable. Fundus examination appeared deceptively normal, showing only subtle temporal pallor. Pure tone audiometry confirmed bilateral sensorineural hearing loss. Serum and cerebrospinal fluid tested positive for anti-MOG antibodies. Optical coherence tomography was initially normal, while visual evoked potentials showed delayed P100 latencies. Magnetic resonance imaging revealed extensive T2/FLAIR hyperintensities involving the posterior visual pathway, brainstem and cranial nerve. The patient received high-dose intravenous methylprednisolone followed by plasma exchange and intravenous immunoglobulin due to inadequate response. He was discharged on tapering oral corticosteroids and commenced on mycophenolate mofetil for long-term immunosuppression. At discharge, vision remained poor. At eight months, vision improved to 6/45 (right eye) and 6/60 (left eye), with optic nerve atrophy and partial recovery of hearing. This case highlights an atypical MOGAD presentation without optic disc oedema, complicated by rare auditory involvement. Early recognition and timely treatment are vital to preserve neurological and visual function.

INTRODUCTION

Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) is a recently defined demyelinating disorder of central nervous system, increasingly recognized for its broad clinical and radiological spectrum. The estimated global incidence ranges from 1.6 to 4.8 per million per year, with a prevalence of 1.3 to 2.5 per 100,000 population.[1] In Malaysia, the prevalence is reported as 0.12 per 100,000 persons.[2]

MOGAD typically presents with optic neuritis (ON), transverse myelitis, and acute disseminated encephalomyelitis (ADEM). MOGAD-associated optic neuritis (MOGAD-ON) commonly manifest as severe, often bilateral vision loss with prominent optic disc oedema and significant periorbital pain. Ancillary testing such as optical coherence tomography (OCT) often demonstrates marked retinal nerve fibre layer (RNFL) thickening in the acute phase.[1,3]

The expanding recognition of atypical presentations underscores significant diagnostic challenges. Atypical cases lacking characteristic features may lead to delayed diagnosis and suboptimal outcomes. A rare case of MOGAD is described herein presenting as a bilateral, painless optic neuropathy without disc oedema, accompanied by bilateral sensorineural hearing loss — an association seldom reported.

CASE PRESENTATION

A 38-year-old previously healthy man presented with progressive, painless bilateral visual loss over two weeks. The symptoms began with blurring of vision in the left eye, which he initially noticed as peripheral visual loss while typing a report at work and progressed to generalized visual loss. He did not seek immediate medical attention and self-medicated with over-the-counter eye drops. One week later, blurring of vision developed in the right eye. Concurrently, he developed bilateral hearing loss, worse on the left, without other otologic symptoms. There was no photopsia, heat-induced worsening of vision, ocular pain, or neurological deficits. There was no history of recent illness, vaccination, or high-risk behaviours.

The patient was alert with no meningism or focal neurological deficits. Visual acuity was markedly reduced to hand movement in the right eye and vague light perception in two quadrants in the left eye. Pupillary reactions were sluggish bilaterally, without a relative afferent pupillary

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Corresponding Author: Wan Hazabbah Wan Hitam
Email: hazabbah@usm.my

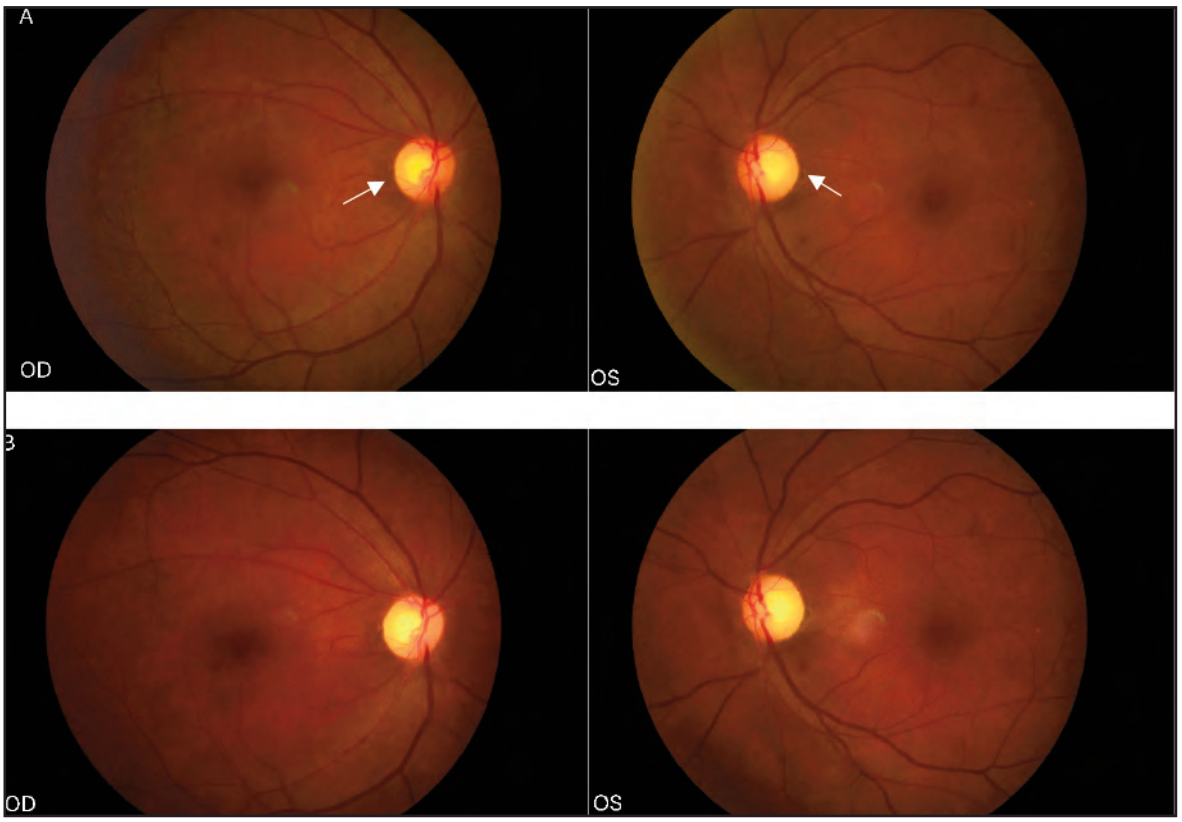

Fig. 1: Fundus photographs at initial presentation and eight-months follow-up. (A) Temporal optic disc pallor (white arrow) at presentation. (B) Partial optic nerve atrophy at eight months

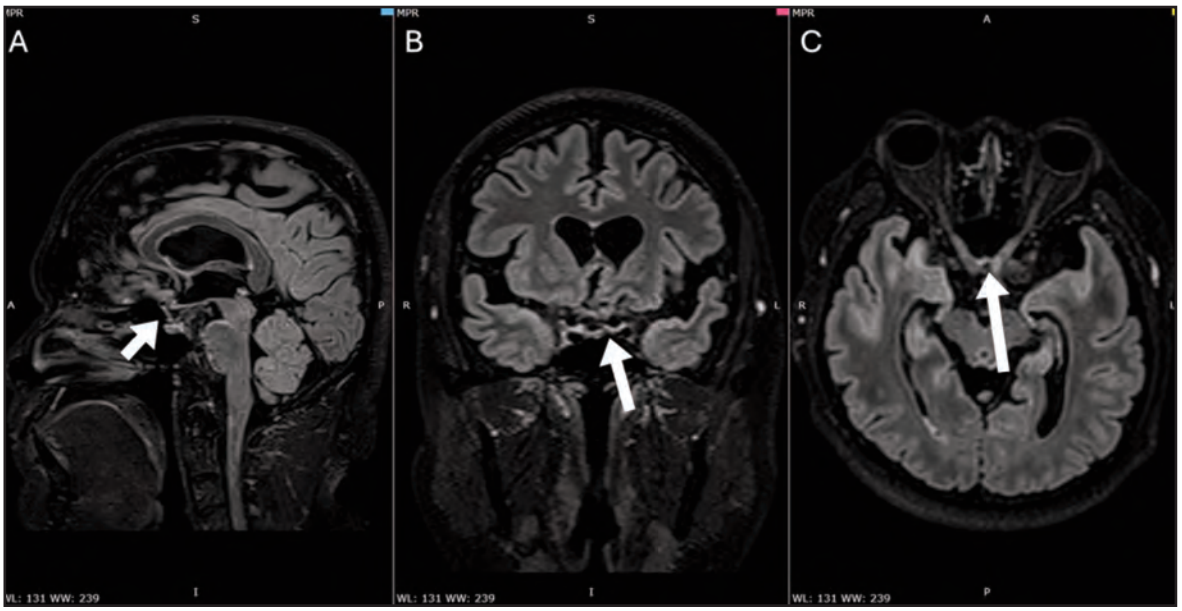

Fig. 2: T2/FLAIR MRI brain and orbit images showing hyperintensity in the optic chiasm (arrow)

defect. Anterior segment examination was unremarkable. Fundus examination showed subtle bilateral temporal optic disc pallor without disc oedema, haemorrhages, or signs of intraocular inflammation (Figure 1A). Extraocular movements were full. Colour vision and visual field could not be assessed due to profound vision loss. Pure tone audiometry confirmed left mild-to-profound and right mild-to-moderate high frequency sensorineural hearing loss.

Given the subacute, progressive bilateral visual loss, inflammatory optic neuropathy — including optic neuritis — was considered. Other differential diagnoses included compressive, infiltrative, and hereditary causes (e.g., Leber hereditary optic neuropathy). In view of the "quiet" fundus appearance despite profound vision loss, extensive investigations were performed.

Serum anti-MOG antibodies were positive as detected by the Euroimmun indirect immunofluorescence assay. Cerebrospinal fluid (CSF) analysis showed anti-MOG antibodies at a titre of 1:40, with normal opening pressure and no pleocytosis. Serum aquaporin-4 antibodies and genetic testing for LHON were negative. Infective and autoimmune screens were unremarkable. Cerebrospinal fluid (CSF) polymerase chain reaction (PCR) testing was performed to exclude infectious aetiologies of optic neuropathy and brainstem involvement. Pathogens tested included herpes simplex virus, varicella-zoster virus, cytomegalovirus, enterovirus, human parechovirus, Streptococcus agalactiae, Streptococcus pneumoniae, Haemophilus influenzae, Neisseria meningitidis, Listeria monocytogenes, Escherichia coli, Cryptococcus neoformans, and Mycobacterium tuberculosis. All CSF PCR assays and cultures were negative. Oligoclonal bands could not be tested due to insufficient sample volume.

Magnetic resonance imaging (MRI) demonstrated confluent T2/FLAIR hyperintensities involving the optic nerves, optic chiasm, and post-chiasmatic tracts (Figure 2) with additional lesions in the brainstem and cranial nerve III, IV, and V. Contrast-enhanced CT was normal. Visual evoked potentials showed prolonged P100 latency. OCT revealed normal RNFL thickness (right: 91 µm; left: 100 µm).

Intravenous methylprednisolone (1 g/day for five days) was initiated two weeks after symptom onset. Owing to an inadequate response, the patient underwent five cycles of plasma exchange followed by intravenous immunoglobulin (IVIG) at 0.4 g/kg/day for five days. Visual acuity remained at counting fingers in the right eye and hand movement in the left eye at the time of discharge. High-dose oral prednisolone was continued, with gradual tapering commencing after one month and commenced on mycophenolate mofetil for relapse prevention.

At eight months, visual acuity improved to 6/45 (right eye) and 6/60 (left). Fundus examination showed optic atrophy (Figure 1B). RNFL thinned to 72 µm (right) and 74 µm (left). Repeat audiometry showed residual bilateral mild-to-moderate sensorineural hearing loss with subjective improvement.

DISCUSSION

MOGAD is mediated by autoantibodies against the MOG protein on oligodendrocytes, leading to demyelination via complement activation and antibody-dependent cellular cytotoxicity. Histopathologically, $CD4^+$ T-cell–predominant infiltration with macrophages and complement deposition is typically observed.[1,4] The precise trigger remains uncertain, with infection or vaccine-related molecular mimicry being the most widely proposed mechanism.[1]

This case demonstrates a rare presentation of MOGAD-ON with severe bilateral vision loss without disc oedema or pain. Normal acute-phase RNFL thickness may mislead clinicians toward non-inflammatory aetiologies. Although RNFL >118 µm may help distinguish MOGAD-ON from MS-related optic neuritis5; normal RNFL measurements do not exclude MOGAD, as highlighted here.

MOGAD-ON commonly presents with marked optic disc oedema; however, this feature is not invariably present. The absence of optic disc oedema in the patient likely attributable to predominant retrobulbar involvement affecting the posterior optic nerve, optic chiasm and post-chiasmatic pathways, as demonstrated on MRI. Furthermore, delayed presentation may have allowed acute inflammatory oedema to subside before clinical evaluation, resulting in early optic disc pallor.

Sensorineural hearing loss in MOGAD is exceedingly rare; a Korean cohort reported a 4.2% prevalence.[6] Auditory dysfunction may reflect demyelination of central auditory pathways or involvement of the vestibulocochlear nerve.[7-8] MRI in this patient supported both mechanisms.

MOGAD-ON typically shows longitudinally extensive optic nerve lesions.[1,3-4] Cranial nerve involvement and brainstem lesions, as seen here, indicate multifocal disease. According to the 2023 international MOGAD diagnostic criteria,[9] low-positive MOG-IgG titres require compatible clinical and radiologic findings — all of which were fulfilled in this case.

High-dose corticosteroids are first-line therapy.[1] Steroid-refractory cases may benefit from plasma exchange or intravenous immunoglobulin (IVIG).[1,3] Early treatment improves outcomes; conversely, delays worsen RNFL loss and visual prognosis.[3] Despite timely therapy, recovery was limited, likely due to posterior pathway involvement.

Long-term, slow steroid tapering (>6 months at ≥20 mg/day) reduces relapse risk.[3] Steroid-sparing agents, including mycophenolate mofetil, further reduce relapse frequency when used in conjunction with low-dose corticosteroids.[3-4] Our patient remained relapse-free on dual therapy.

Visual prognosis in MOGAD is typically better than in NMOSD-ON.[10] Poor prognostic factors include bilateral onset, delayed treatment and brainstem involvement.[3,10] Persistent antibody positivity may indicate an elevated relapse risk, but is not a reliable predictor of prognosis.[1,4]

The prognosis for auditory recovery in MOGAD remains uncertain, owing to the rarity of reported cases. Available

data suggest that auditory recovery in MOGAD is often incomplete, even with prompt immunotherapy, unlike that of visual recovery.[6]

This case highlights several important diagnostic challenges. Profound bilateral visual loss without optic disc oedema with concurrent auditory symptoms are atypical for classic MOGAD-ON. The relatively normal fundus appearance and normal acute-phase RNFL thickness broadened the differential diagnoses and necessitated extensive investigations before a definite diagnosis was established. Furthermore, delayed presentation contributed to the late initiation of high dose immunotherapy. Collectively, these atypical clinical features and delayed presentation likely contributed to delayed diagnosis and late initiation of treatment and may explain the limited visual recovery observed in the patient despite aggressive immunotherapy.

CONCLUSION

This case expands the ophthalmic spectrum of MOGAD, demonstrating that it may present as severe bilateral painless optic neuropathy without disc oedema, alongside rare auditory involvement. A high index of clinical suspicion for MOGAD is essential in atypical optic neuropathy presentations, even when fundus and OCT findings are unremarkable. Early neuroimaging and serological testing are crucial for accurate diagnosis and timely treatment, thereby preventing irreversible neurological and visual disability.

REFERENCES

1. Jeyakumar N, Lerch M, Dale RC, Ramanathan S. MOG antibody-associated optic neuritis. Eye 2024; 38: 2289-301.
2. Ong ZM, Arip M, Ching YM, Kumar L, Terumalay S, Sim SH, et al. The prevalence, demographics, clinical features, neuroimaging, and inter-ethnic differences of MOGAD in Malaysia with global perspectives. Mult Scler Relat Disord 2022; 67: 104168.
3. Nowacka B, Lubiński W, Kaźmierczak B. Myelin oligodendrocyte glycoprotein (MOG) antibody-associated optic neuritis: A case report and literature review. Int Med Case Rep J 2024; 17: 391-9.
4. Sechi E, Cacciaguerra L, Chen JJ, Mariotto S, Fadda G, Dinoto A, et al. Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD): A review of clinical and MRI features, diagnosis, and management. Front Neurol. 2022; 13: 885218.
5. Chen JJ, Sotirchos ES, Henderson AD, Vasileiou ES, Flanagan EP, Bhatti MT, et al. OCT retinal nerve fiber layer thickness differentiates acute optic neuritis from MOG antibody-associated disease and multiple sclerosis: RNFL thickening in acute optic neuritis from MOGAD vs MS. Mult Scler Relat Disord 2022; 58: 103525.
6. Kwon S, Choi S, Chung YH, Min JH. Sensorineural hearing loss in seropositive neuromyelitis optica spectrum disorder and myelin oligodendrocyte glycoprotein antibody-associated disorder. Acta Neurol Scand 2024; 6724448.
7. Jarius S, Kleiter I, Ruprecht K, Asgari N, Pitarokoili K, Borisow N, et al. MOG-IgG in NMO and related disorders: A multicenter study of 50 patients. Part 3: Brainstem involvement—frequency, presentation and outcome. J Neuroinflammation 2016; 13: 281.
8. Cobo-Calvo A, Ayrignac X, Kerschen P, Horellou P, Cotton F, Labauge P, et al. Cranial nerve involvement in patients with MOG antibody-associated disease. Neurol Neuroimmunol Neuroinflamm 2019; 6(2): e543.
9. Banwell B, Bennett JL, Marignier R, Kim HJ, Brilot F, Flanagan EP, et al. Diagnosis of myelin oligodendrocyte glycoprotein antibody-associated disease: International MOGAD panel proposed criteria. Lancet Neurol 2023; 22: 268-82.
10. Akaishi T, Himori N, Takeshita T, Misu T, Takahashi T, Takai Y, et al. Five-year visual outcomes after optic neuritis in anti-MOG antibody-associated disease. Mult Scler Relat Disord 2021; 56: 103222.