Section: Ophthalmology

Case Report

A CASE OF MYELIN OLIGODENDROCYTE GLYCOPROTEIN ANTIBODY ASSOCIATED ATYPICAL OPTIC NEURITIS IN A GIRL

Ragul VR[1], Prem Sai T[2], Kalavathy CM[3], Krithika M[2]

[1]Post Graduate Resident, Department of Ophthalmology, Tagore Medical College and Hospital, Rathinamangalam, Melakottaiyur, Chennai, Tamil Nadu, India
[2]Assistant professor, Department of Ophthalmology, Tagore Medical College and Hospital, Rathinamangalam, Melakottaiyur, Chennai, Tamil Nadu, India
[3]Professor and Head, Department of Ophthalmology, Tagore Medical College and Hospital, Rathinamangalam, Melakottaiyur, Chennai, Tamil Nadu, India

Received : 11/06/2024
Received in revised form : 05/08/2024
Accepted : 21/08/2024

Keywords:
Myelin Oligodendrocyte Glycoprotein Antibody Associated Atypical Optic Neuritis.

Corresponding Author:
Dr. Krithika M,
Email: krits7@gmail.com

DOI: 10.47009/jamp.2024.6.4.128

Source of Support: Nil,
Conflict of Interest: None declared

Int J Acad Med Pharm
2024; 6 (4); 645-648

Abstract

Optic neuritis is a demyelinating disorder, it is divided into typical and atypical types based on etiology and peculiar presentation. Myelin oligodendrocyte glycoprotein antibody associated disease (MOGAD) is an atypical form of optic neuritis. It accounts for 15% of atypical optic neuritis and 50% of acute demyelinating syndromes in children under 11 years of age1. The entity differs from multiple sclerosis and neuromyelitis optica spectrum disorder in terms of clinical presentation, radio diagnostic features, and prognosis. Pathogenesis is antibody against myelin oligodendrocytes glycoprotein that results in damage to myelin insulation around CNS axon. Here, we present a case of 7 year old girl with symptoms and signs suggesting MOGAD and tested positive for MOG-IgG antibody. This report highlights the importance of early diagnosis and management of MOGAD.

INTRODUCTION

MOGAD refers to a demyelinating syndrome in association with IgG antibodies targeting myelin oligodendrocyte glycoprotein, a minor transmembrane protein found on outermost lamellae of CNS myelin and oligodendrocyte.[1-3]

Earlier all atypical optic neuritis were classified as neuromyelitis optica spectrum disorder. MOGAD is an idiopathic, inflammatory, demyelinating disease of CNS, clinically and prognostically distinct from multiple sclerosis and neuromeylitica spectrum dsiorder.[4]

Previous method to diagnose MOGAD was western blot and ELISA, recently we use live cell-based assays which has enabled native human MOG to be transduced or transfected into mammalian cell line for surface expression, incubated with sera and secondary anti-human IgG antibody, analyzed for MOG-specific antibody binding, this helped in detection of clinically relevant MOG-IgG and accurate identification of children and adults.[5-7]

Most common relapsing syndrome both in children and adult is MOGAD.

CASE REPORT

A 7 year old girl came with complaints of decreased visual acuity, associated with pain around right eye more compared to left eye. On evaluation visual acuity was 3/60 and 6/60 in right and left eye respectively with normal intraocular pressure and rest of the anterior segment were within normal limits. However sluggish papillary reaction was noted in both eye and colour vision was defective in right eye > left eye. On fundus examination blurred disc margins was noted in both the eyes [Figure 1]. No signs of raised intracranial tension, alteration in consciousness or limb weakness. Routine blood investigations were normal including ESR, vitamin B12, homocysteine and tested negative for syphilis, antinuclear antibodies.

MRI brain and orbit revealed minimal increased signal intensity in the retro orbital portion of right optic nerve with loss of perioptic normal fluid signals on both sides – suggestive of bilateral chronic optic neuritis with acute changes in right side [Figure 2]. Visual evoked potential showed prolonged P100 latency in both eye [Figure 3].

MOG IgG antibody was positive and aquaporin-4 antibody negative. Neurologist opinion sorted and started on intravenous methylprednisolone 450mg IV OD for 3 days, followed by tapering dose of oral prednisolone 10mg thrice a day for 11 days, and her visual acuity improved to 6/24, 6/18 in right and left eye respectively. Patient was advised to follow-up every 3 monthly intervals. On 6thmonth follow-up visual acuity in Right eye: 6/24 Left eye: 6/18, on fundus examination both eye temporal optic disc pallor present [Figure 4]. MRI showed thinning of

ISSN (O): 2687-5365; ISSN (P): 2753-6556

optic nerve suggestive of chronic optic neuritis, Visual evoked potential P100 latency was normal [Figure 6] and OCT RNFL revealed temporal thinning [Figure 5]. Neurologist opinion sorted, started on Azathioprine 25mg OD as maintenance and advised to follow-up 3monthly.

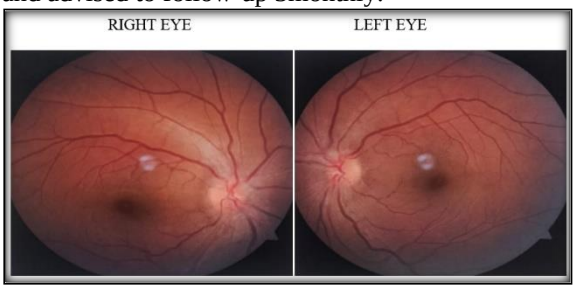

Figure 1: Blurred disc margins in both eye

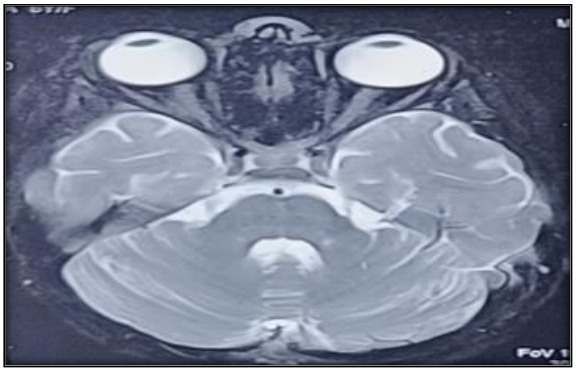

Figure 2: Bilateral optic neuritis with acute changes in right eye

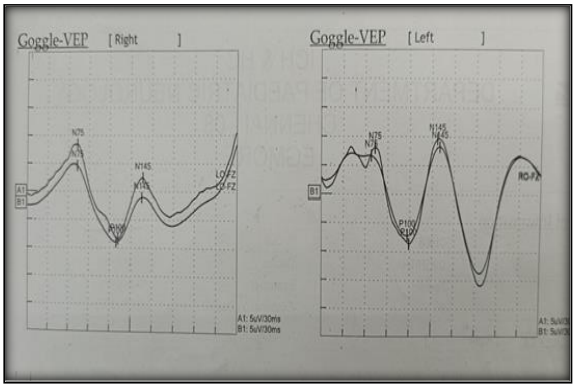

Figure 3: long latency of P100 in both eyes

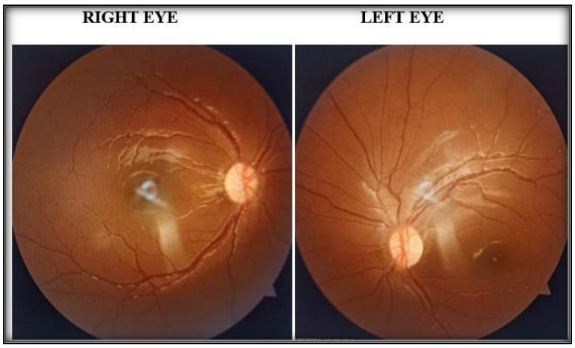

Fig 4: Both eyes showing temporal disc pallor

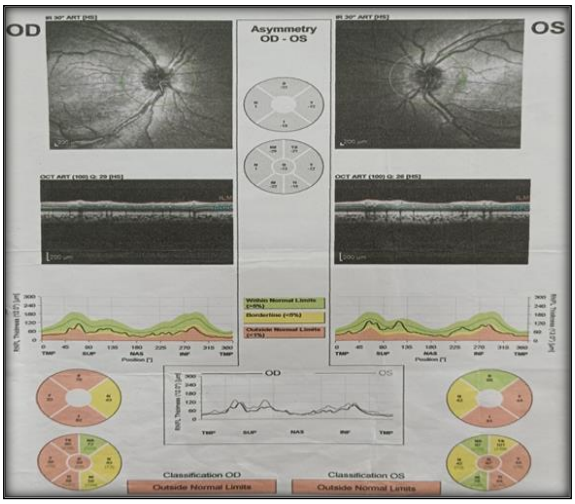

Figure 5: Both eye OCT RNFL showed temporal thinning

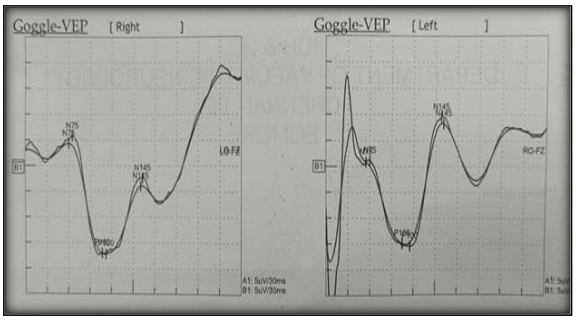

Figure 6: Visual evoked potential showed normal P100 latency

Table 1: Right Eye.

Wave	Electrode	N75	P100	N145	Stimulus	Average
A1	LO-FZ	73.8ms	132.0m	166.8m	Right	200
B1	LO-FZ	73.8ms	129.3m	165.3m	Right	200

LEFT EYE

Wave	Electrode	N75	P100	N145	Stimulus	Average
A1	RO-FZ	72.9ms	119.1m	157.2m	Left	200
B1	RO-FZ	67.8ms	117.0m	160.5m	Left	190

Table 2: RIGHT EYE

Wave	Electrode	N75	P100	N145	Stimulus	Average
A1	LO-FZ	65.1ms	107.1m	162.6m	Right	200
B1	LO-FZ	60.3ms	112.2m	160.2m	Right	200

LEFT EYE

Wave	Electrode	N75	P100	N145	Stimulus	Average
A1	RO-FZ	60.3ms	115.2m	157.8m	Left	200

ISSN (O): 2687-5365; ISSN (P): 2753-6556

B1	RO-FZ	54.9ms	104.7m	160.8m	Left	200

DISCUSSION

Myelin oligodendrocyte antibody associated disease is demyelinating disorder distinct from multiple sclerosis and neuromyelitis optica. Annual incidence of MOG-AD worldwide is approximately 1.6- 4.8 per million people with prevalence estimated at 1.3-2.5 per 100,000 people8. This disease is usually a biphasic distribution with age of onset varying from 5-10 years and 20-45 years. Optic neuritis is most common manifestation of MOGAD in adults (30-60%), followed by transverse myelitis (10-25%), second most common manifestation in children after acute disseminated encephalomyelitis.[9-11]

MOG-AD classically differ from NMO (neuromyelitis optica) and Multiple sclerosis ON (optic neuritis) as following demographically biphasic age group,[11] equal female: male, course can be monophasic or relapsing, bilateral presentation is more common in MOGAD,[12] with decreased visual acuity, defective colour vision and retro orbital pain. On examination moderate to severe optic disc swelling may be associated with hemorrhage and MOGAD can cause papilledema secondary to raised intracranial pressure in association with an aseptic meningoencepalitis.[13,14] Radiologically optic nerve head swelling, longitudinal extensive optic nerve involvement and least common presentation with chiasmal involvement are seen. Serum MOG-Ab positivity marks definitive diagnosis. On ancillary investigations visual field defects shows variable field loss, delayed P100 latency in visual evoked potential. VEP testing in MOGAD is to confirm subtle optic nerve injury in case where clinical examination findings may be uncertain and in discriminating organic visual loss from functional visual loss. On OCT (ocular coherence tomography) acute phase pRNFL (peripapillary retinal nerve fiber layer) thickening and follow up pRNFL thinning is classical, thus OCT can be used as an early diagnostic tool to differentiate MOG-ON and MS-ON15. Therapeutically it is rapid steroid responsive and shows steroid dependence, Long term visual recovery is favorable in absence of subsequent relapses.

Other ophthalmic associations of MOGAD include uveitis, peripheral ulcerative keratitis, acute macular neuropathy, serous retinal detachment, venous stasis retinopathy, pre-retinal macular hemorrhage and orbital apex syndrome.[15] There are various studies showing atypical presentation of MOG-ON, it has presented in association with ulcerative colitis, a rare involvement of chiasma and optic tract can be a variable presentation. Children with a polysymptomatic disease and higher antibody titers are at greater risk for relapse.[17]

CONCLUSION

This classical case report highlights the need for testing for MOG-IgG antibodies, especially in children with bilateral optic neuritis. It is essential to differentiate the entity from other demyelinating disorders with classical presentation. The prognosis depends on early diagnosis, appropriate and timely management with regular follow-up.

REFERENCES

1. Waters P, Fadda G, Woodhall M, O'Mahony J, Brown RA, Castro DA, et al. Serial Anti-Myelin Oligodendrocyte Glycoprotein Antibody Analyses and Outcomes in Children With Demyelinating Syndromes. JAMA Neurol. 2020;77:82–93.
2. 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.
3. Linnington C, Webb M, Woodhams PL. A novel myelin-associated glycoprotein defned by a mouse monoclonal antibody. J Neuroimmunol. 1984;6:387–96.
4. Rostasy K, Mader S, Schanda K, Huppke P, Gärtner J, Kraus V, et al. Anti–myelin oligodendrocyte glycoprotein antibodies in pediatric patients with optic neuritis. Arch Neurol 2012;69. doi: 10.1001/archneurol. 2011.2956
5. Brilot F, Dale RC, Selter RC, Grummel V, Kalluri SR, Aslam M, et al. Antibodies to native myelin oligodendrocyte glycoprotein in children with infammatory demyelinating central nervous system disease. Ann Neurol. 2009;66:833–42.
6. Kitley J, Woodhall M, Waters P, Leite MI, Devenney E, Craig J, et al. Myelinoligodendrocyte glycoprotein antibodies in adults with a neuromyelitis optica phenotype. Neurology. 2012;79:1273–7.
7. Sato DK, Callegaro D, Lana-Peixoto MA, Waters PJ, de Haidar Jorge FM, Takahashi T, et al. Distinction between MOG antibody-positive and AQP4 antibodypositive NMO spectrum disorders. Neurology. 2014;82:474–81.
8. Hor JY, Fujihara K. Epidemiology of myelin oligodendrocyte glycoprotein antibody-associated disease: a review of prevalence and incidence worldwide. Front Neurol. 2023;14:1260358.
9. O'Connell K, Hamilton-Shield A, Woodhall M, Messina S, Mariano R, Waters P, et al. Prevalence and incidence of neuromyelitis optica spectrum disorder, aquaporin-4 antibody-positive NMOSD and MOG antibody-positive disease in Oxfordshire, UK. J Neurol Neurosurg Psychiatry. 2020;91:1126–8.
10. Hor JY, Fujihara K. Epidemiology of myelin oligodendrocyte glycoprotein antibody-associated disease: a review of prevalence and incidence worldwide. Front Neurol. 2023;14:1260358.
11. 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;10:672–85
12. Bennett JL, Costello F, Chen JJ, Petzold A, Biousse V, Newman NJ, et al. Optic neuritis and autoimmune optic neuropathies: advances in diagnosis and treatment. Lancet Neurol. 2023;22:89–100.
13. 54. Narayan RN, Wang C, Sguigna P, Husari K, Greenberg B. Atypical Anti-MOG syndrome with aseptic meningoencephalitis and pseudotumor cerebri-like presentations. Mult Scler Relat Disord. 2019;27:30–3.
14. Wong WK, Troedson C, Peacock K, Brilot-Turville F, Menezes MP, Dale RC, et al. Steroid-responsive aseptic meningitis with raised intracranial pressure syndrome

ISSN (O): 2687-5365; ISSN (P): 2753-6556

associated with myelin oligodendrocyte glycoprotein autoantibodies. J Paediatr Child Health. 2022;58:2322–6.
15. Chen JJ, Sotirchos ES, Henderson AD, Vasileiou ES, Flanagan EP, Bhatti MT, et al. OCT retinal nerve fber 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.
16. Jeyakumar,N.,Lerch, M., Dale, R.C. et al. MOG antibody-associated optic neuritis. Eye 38, 2289-2301 (2024). https://doi.org/10.1038/s41433-024-03108-y
17. Raghavan SL, Prabu R, Mohan S, Nagarajan S. Bilateral optic neuritis as a presentation of myelin oligodendrocyte glycoprotein antibody associated disease in a child. Indian J Ophthalmol Case Rep 2023;3:118-20.

ISSN (O): 2687-5365; ISSN (P): 2753-6556