Thilakavathy Tharmalingam[1,3], Shahidatul A.M[1,3], Husin M.H[2,3], Wan-Hazabbah W.H[1,3]

[1]Department of Ophthalmology and Visual Science, School of Medical Sciences, Health Campus, Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan, Malaysia

[2]Department of Radiology, School of Medical Sciences, Health Campus, Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan, Malaysia

[3]Hospital Pakar Universiti Sains Malaysia, Health Campus, Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan, Malaysia

Received 10 July 2025
Revised 13 Nov 2025
Accepted 19 May 2026
Published Online
5 June 2026

*Corresponding author
Wan Hazabbah Wan Hitam
hazabbah@usm.my

Idiopathic Optic Perineuritis Presenting as Acute Hemifield Loss: Importance of Early MRI Diagnosis and Steroid Therapy

Abstract – Optic perineuritis (OPN) is a rare inflammatory condition involving the sheath of the optic nerve. Although it may present similarly to optic neuritis, it requires distinct management approaches. A 27 years old man, came to the eye clinic with left eye visual field defect and pain on eye movement for three days. His left eye vision was counting finger and 6/6 in right eye. Relative afferent pupillary defect (RAPD) present in the left eye. He had some difficulty perceiving colours. The intraocular pressures were within normal limits bilaterally. Anterior and posterior segments examination on both eyes were unremarkable. Tests for antibodies against the water channel protein 4 (AQP4) and the oligodendrocyte myelin glycoprotein (MOG) were negative, effectively ruling out neuromyelitis optica spectrum disorder (NMOSD) and MOG antibody-associated disorder (MOGAD). Screening for infectious and autoimmune diseases were also unremarkable. Orbital MRI showed marked contrast uptake along the left optic nerve with extension into the canal of optic nerve, confirming the diagnosis of optic perineuritis. He was treated with intravenous high-dose methylprednisolone, followed by a gradually reducing course of oral steroids, resulting in full recovery of vision and colour perception within one week. This case underscores the importance of early suspicion of optic perineuritis. Urgent referral to tertiary centres with MRI facilities can help ensure accurate diagnosis and timely treatment. Early initiation of corticosteroid therapy plays a key role in preserving vision and preventing permanent visual damage.

Keywords*: Optic perineuritis, optic neuritis, corticosteroid, MRI, AQP4*

1 INTRODUCTION

Optic perineuritis (OPN) is an uncommon inflammatory condition in which the optic nerve sheath is the principal site of involvement, sparing the nerve fibres themselves. The condition leads to noticeable thickening of the optic nerve sheath due to unspecified fibrotic remodelling, in contrast to optic neuritis, where inflammation primarily affects the optic nerve axons (1,3). Clinically, it closely mimics optic neuritis, with overlapping symptoms such as periocular pain and acute visual loss. However, OPN differs in pathophysiology, radiological features, and treatment response. It's important to identify OPN promptly and correctly, as it usually responds very well to corticosteroid treatment, giving patients a good chance of regaining full vision (2,6). Delayed diagnosis may lead to irreversible visual impairment. This case highlights the importance of considering OPN in the differential diagnosis of acute vision loss, even in young patients with a normal ocular examination.

2. CASE REPORT

A previously well 27-year-old man sought medical attention after three days of sudden hemifield visual impairment in the left eye, accompanied by a dull retro-orbital ache that intensified with extraocular movement. He denied any ocular redness, discharge, trauma, constitutional symptoms, or previous ocular surgery.

Clinical assessment showed normal visual acuity of 6/6 in the right eye and counting finger in left eye. RAPD was present on left eye, with markedly reduced brightness perception and red desaturation. Intraocular pressures were 18 mmHg bilaterally. Examination of anterior and posterior of ocular structures revealed no abnormalities. Colour vision testing could not be reliably performed in the left eye due to the severity

of vision loss. Visual field testing with Humphrey perimetry of the left eye demonstrated a defect affecting half of the visual field (Figure 1).

Laboratory testing for potential systemic inflammatory, infectious, immune-mediated conditions, including vasculitis, sarcoidosis, and syphilis were negative. Gadolinium-enhanced orbital MRI revealed findings consistent with optic perineuritis. No demyelinating lesions or other intracranial abnormalities were observed (Figure 2).

The patient was started on high-dose intravenous methylprednisolone at 250 mg every six hours for five days. This was subsequently followed by a tapering course of oral corticosteroids. Vision improved rapidly during treatment, and by discharge, left eye visual acuity had fully recovered to 6/6. Follow-up confirmed normal visual fields and resolution of optic nerve function deficits.

Table 1. Pathophysiological comparison of Optic Perineuritis (OPN) and Optic Neuritis (ON)

Feature	Optic Perineuritis (OPN)	Optic Neuritis (ON)
Inflammation site	Meninges surrounding the optic nerve (dura, arachnoid, pia)	Optic nerve axons (intraneural)
Mechanism	Perineural inflammation → sheath thickening → secondary compression → ischemia of optic fibers	Demyelination of optic nerve axons → impaired signal conduction
Effect on nerve fibers	Compression and secondary functional compromise	Direct axonal dysfunction due to myelin loss
MRI hallmark	Circumferential enhancement of optic nerve sheath	Intraneural enhancement of optic nerve
Clinical impact	Slower, subacute visual loss; periocular pain; atypical visual field defects (altitudinal/hemifield)	Acute visual loss; central scotoma; retro-orbital pain

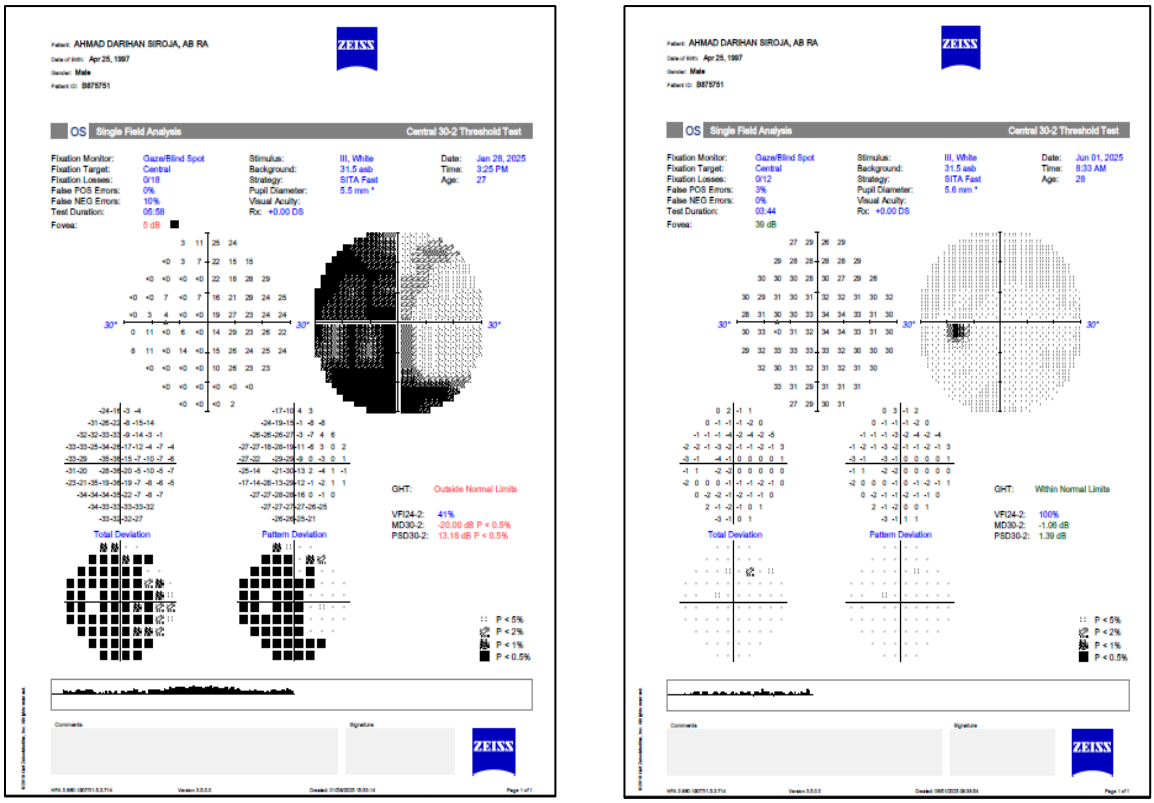

Figure 1. Visual field in the left eye on presentation showed left hemifield loss that recovered post treatment.

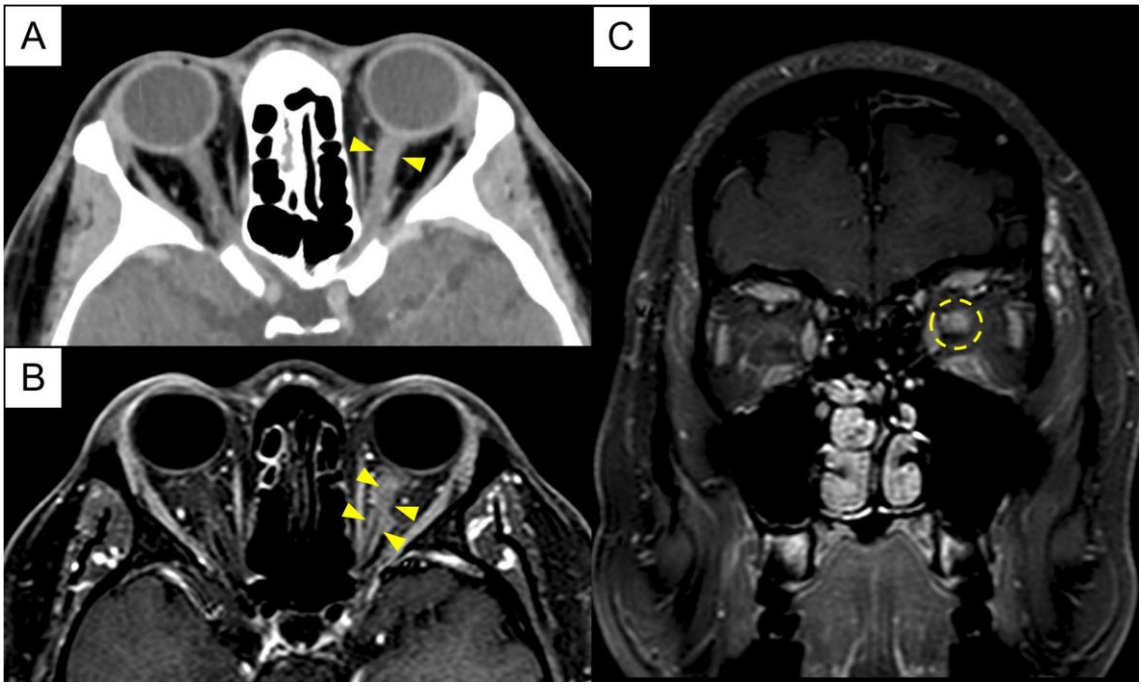

Figure 2. Contrasted CT in axial view (A) shows relative thickening of the left optic nerve with subtle enhancement (yellow arrowheads). The enhancement of the optic nerve sheath, resulting in the tram-track sign, is demonstrated in the T1-weighted with fat suppression (T1FS) post-contrast MRI B). On T1FS post-contrast MRI in coronal view (B), the circumferential enhancement results in the doughnut sign (yellow circle).

3. DISCUSSION

Optic perineuritis (OPN) is often associated with multiple sclerosis. The true prevalence of optic perineuritis is unclear, owing to its rarity, the scarcity of population-based studies, and its frequent misdiagnosis as optic neuritis. OPN can occur across a wide adult age range and does not exhibit a consistent sex predilection.

It primarily affects the perineural sheath of the optic nerve, sparing the nerve fibres themselves. While most cases are idiopathic, optic perineuritis has been linked to systemic inflammatory and infectious conditions, including syphilis, sarcoidosis, Crohn's disease, and granulomatosis with polyangiitis (1,2,4,5). Its true incidence remains uncertain due to its rarity and frequent misdiagnosis as optic neuritis

Pathologically, OPN involves inflammation of the meninges surrounding the optic nerve rather than the nerve fibres themselves (Table 1) (3). This perineural inflammation may result in swelling and heightened signal along the nerve, which can secondarily compress the optic nerve fibres Distinguishing OPN from optic neuritis is challenging because of overlapping clinical features; however, the two differ in pathogenesis, and OPN generally demonstrates a more favourable response to corticosteroid therapy (2,3).

Clinically, patients often present with subacute visual loss, periocular pain—particularly with eye movement—and impaired colour vision, with visual deficits progressing more slowly than in typical optic neuritis (1,6). Fundoscopic examination may initially appear normal. Purvin et al. reported that atypical visual field defects, such as altitudinal or hemifield loss, can arise from compression or ischemia caused by inflammation of the optic nerve sheath affecting the surrounding nerve fibres (7). A magnetic resonance scan is crucial for diagnosis, with circumferential optic nerve sheath enhancement distinguishing OPN from intraneural optic neuritis (1,6). Testing for AQP4 and MOG antibodies helps rule out neuromyelitis optica spectrum disorder and MOG-associated optic neuritis.

Intensive steroid therapy continues to be the primary treatment and usually lead to rapid recovery of vision (10). Early recognition is critical, as delayed diagnosis may lead to irreversible visual deficits. Patients with underlying systemic disease require careful follow-up due to the risk of relapse.

4 CONCLUSION

Prompt recognition of optic perineuritis (OPN) is essential in patients presenting with acute unilateral visual loss, especially when retro-orbital pain is present despite a normal fundus examination. In contrast to optic neuritis, optic perineuritis (OPN) is typically not self-limiting and, if treatment is delayed, may result in permanent visual impairment. This case highlights the important role of primary care physicians in identifying suspicious features and arranging urgent referral to tertiary centres with MRI facilities, as early diagnosis and treatment can greatly improve visual and clinical outcomes.

ACKNOWLEDGEMENTS

The authors wish to express their sincere appreciation to the staffs of Hospital Pakar Universiti Sains Malaysia for their invaluable contributions to the clinical assessment, management, and follow-up of this patient. We are also grateful to the hospital administration for facilitating access to clinical facilities and resources. Finally, we extend our gratitude to the patient for his cooperation and for granting permission to share his case for educational and research purposes.

AUTHOR CONTRIBUTIONS

Thilakvathy Tharmalingam: Conceptualization, patient management, data collection, literature review, manuscript drafting and manuscript revision.

Mohd Hafizuddin Husin: Radiological interpretation, data acquisition, critical review of the manuscript, and supervision

Wan Hazabbah Wan Hitam, Shahidatul Adha Mohamad: Clinical supervision, manuscript review and final approval of the version to be published

AUTHORSHIP CLARIFICATION

All authors meet the JBCS criteria for authorship and approved the final manuscript.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Ethics approval was not required for this case report. Written informed consent was obtained from the patient.

CONSENT FOR PUBLICATION

Not applicable

CONFLICTS OF INTEREST

The authors declare that they have no competing interests.

REFERENCES

[1] 1. Saitakis G, Chwalisz BK. Optic perineuritis. Current Opinion in Ophthalmology. 2022;33(6):519 524. doi:10.1097/ICU.0000000000000900.

[2] Li H, Zhou H, Sun J, Wang H, Wang Y, Wang Z, Li J. Optic perineuritis and its association with autoimmune diseases. Frontiers in Neurology. 2021; 12:627077. doi:10.3389/fneur.2020.627077.

[3] Neuro Ophthalmology Group, Ophthalmology Branch of Chinese Medical Association. Chinese expert consensus on the diagnosis and treatment of infiltrative optic neuropathy (including OPN). Chinese J Ocul Fundus Dis. 2022;38(12):955 962. doi:10.3760/cma.j.cn511434 20221107 00588.

[4] Aladawi M, Crespo D, Xu R, Zabad R, Vuppala A A. Bilateral longitudinally extensive optic perineuritis post COVID 19 presenting as “idiopathic” intracranial hypertension. Clin Exp Neuroimmunol. 2023;14(3):128 132. doi:10.1111/cen3.12749.

[5] Yokogawa N, Shimada K, et al. Optic perineuritis in eosinophilic granulomatosis with polyangiitis (EGPA). Rheumatology. 2024;63(7) e195. doi:10.1093/rheumatology/kead687

[6] Miller C, Vu N H, Carozza R B. Idiopathic optic perineuritis in a pediatric patient. Journal of Child Neurology. 2025;40(10):906 909. doi:10.1177/08830738251341771.

[7] Escamilla Ocañas C E, Morales Cardona N C, Jamal F. Optic perineuritis and orbital inflammation secondary to zoledronate: A case report. J Neurol Ther. 2024;3(1):1 3. doi:10.14312/2397 1304.2024 1.

[8] Tawengi M M, Fael M, Hourani R F, Alyaarabi T, Tawengi A M, Alfitori G. Optic perineuritis presenting with transient monocular vision loss (TMVL): Case report. Int Med Case Rep J. 2024;17:665 669. doi:10.2147/IMCRJ.S460611.

[9] Jung E, Choi J H, Choi K D, Choi S Y. Unilateral optic perineuritis as the initial presentation of multiple myeloma. Frontiers in Ophthalmology. 2025;5:1616532. doi:10.3389/fopht.2025.1616532.

[10] Osman O, Gammack J, Toledo Franco L, et al. Acute unilateral vision loss and optic perineuritis (OPN) in geriatric patients: A series of three cases. Mathews Journal of Ophthalmology. 2024;9(1). doi:10.30654/MJOP.10028