

IDIOPATHIC BILATERAL OPTIC PERINEURITIS IN A YOUNG MALAY MALE

Chang Zhen Lim¹, Khairy Shamel Sonny Teo¹

¹Department of Ophthalmology, School of Medical Sciences, Health Campus, Universiti Sains Malaysia, Kubang Kerian, Kelantan, Malaysia

**Corresponding author: Khairy Shamel Sonny Teo, Department of Ophthalmology, School of Medical Sciences, Health Campus, Universiti Sains Malaysia, Kubang Kerian, Kelantan, Malaysia, E-mail: khairyshamel@usm.my*

<https://doi.org/10.32827/ijphcs.7.1.148>

ABSTRACT

Background: Optic perineuritis (OPN) is a rare inflammation involving the meningeal sheath surrounding the optic nerve. Clinically it is difficult to differentiate OPN with optic neuritis, but it is critical to have an accurate diagnosis as both the diseases have different regime of treatment. Here we report a case of bilateral optic perineuritis in a young Malay male.

Case Report: A healthy 15 years old young Malay boy presented with subacute, painless and severe blurring of vision in his both eyes, worse on the right. Relative afferent pupillary defect was negative in both eyes. Patient had bilateral generalised optic disc swelling, which is more severe on the right eye. Typical feature of optic perineuritis (OPN) 'doughnut sign' was observed on magnetic resonance imaging. Connective tissue and infective screening were negative. He was diagnosed with bilateral idiopathic optic perineuritis. He was treated with high dose intravenous corticosteroids followed by a three-month course slow tapering of oral steroids. His vision and optic nerve function recovered.

Conclusion: Magnetic Resonance Imaging (MRI) is a vital diagnostic tool to diagnose optic perineuritis. Longer steroid treatment has to be given to prevent relapse of the disease and further damage to the optic nerve.

Keywords: Optic perineuritis, young male, distinguish between optic neuritis

1.0 Introduction

Optic perineuritis (OPN) is a rare inflammatory disease involving the meningeal sheath surrounding the optic nerve. Idiopathic is a term to describe a disease where the cause is unknown. Postulated mechanism of visual loss in OPN is due to mechanical compression leading to compromise of vascular supply in the optic disc which subsequently causing optic nerve ischemia. (Margo et al., 1989) It is difficult to distinguish OPN and optic neuritis due to their similarities in clinical presentation. However, it is crucial to differentiate OPN with optic neuritis, as the prognosis of the OPN is poorer compared with optic neuritis and require longer treatment. (Tatsugawa et al. 2010) We would like to report a case of bilateral idiopathic OPN with severe bilateral visual loss in a young Malay boy.

2.0 Case Report

A previously healthy 15 years old boy presented with gradual worsening of vision in both eyes over the course of 20 days. The blurring of vision was worse on the right eye compared to the left eye. It was not associated with pain on eye movement. He denied any history associated with connective tissue disease or infection.

On examination, the visual acuity of right eye was 6/120, while that of the left eye was 6/60, both did not improve with pinhole. Relative afferent pupillary defect was negative, light brightness and red saturation were equal in both eyes. Visual field test showed central scotoma in both eyes worse on the right. [Figure 1] The anterior segment examination and intraocular pressure were normal. Funduscopy showed bilateral generalised swollen and hyperaemic optic disc, worse on the right eye. [Figure 2] However, the vessels were not tortuous nor dilated and both eyes macula were normal. Systemic examination did not reveal any abnormalities, with normal vital signs and no central or peripheral neurological deficits. Patient was worked up for connective tissue disease and infection, but the laboratory tests were all negative.

Urgent computed tomography of the brain and orbit was performed, but no abnormality was detected. He later proceeded with magnetic resonance imaging of the brain and orbit which showed hyperintensity of perineural sheath in both optic nerves, seen as characteristic “doughnut sign” on T2-weighted coronal view. [Figure 3]

Patient was treated with intravenous methylprednisolone 250 mg QID for three days. Subsequently the patient was given oral prednisolone 1 mg/kg/day, which was tapered slowly over the following 3 months. During follow-up three months post treatment, the best-corrected vision improved to 6/24 on the right eye and 6/9 on left eye. There was regression of the optic disc swelling bilaterally, and patient claimed both eyes central scotoma were reduced.

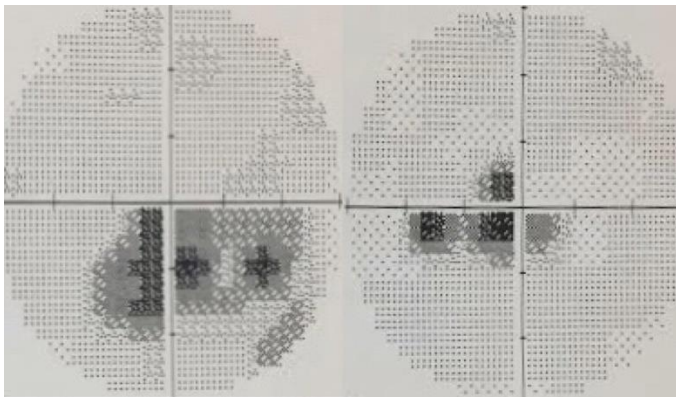


Figure 1: Central scotoma visual field defect.

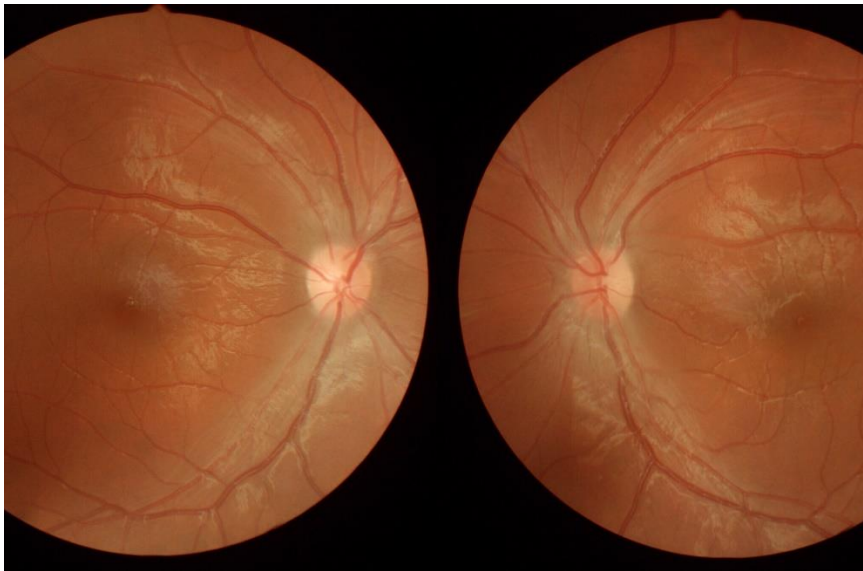


Figure 2: Bilateral funduscopy showed optic disc swelling and mild hyperaemic.

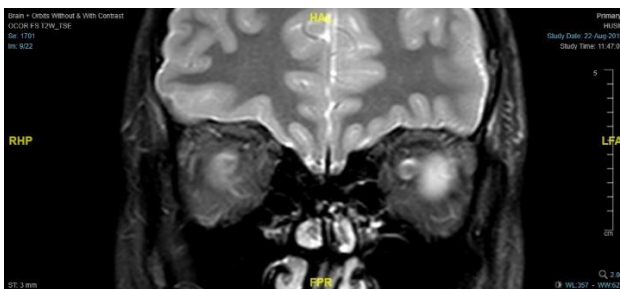


Figure 3: MR images showed ‘doughnut sign’ on coronal view

3.0 Discussion

OPN describes a condition where there is an inflammation of the perineural sheath surrounding the optic nerve. Its aetiology varies, but mostly are idiopathic as in our patient, others include connective tissue disease and infection. (Bergman et al., 2017 ;Yu-Wai-Man et al., 2007; Raghobi et al., 2012)

Clinical presentations of OPN has slight different if compare with optic neuritis. Patients with OPN usually complain of reduction of vision which progress over a period of few weeks but in optic neuritis where patient has acute loss of vision in days. (Purvin et al., 2001) OPN patient usually has central sparring visual field defect, however they may present with variable visual field defect, as in our patient shown central scotoma. (Purvin et al., 2001) Typically, OPN patient comes with unilateral blurring of vision, but case has been reported where there is bilateral eyes involvement. (Tevaraj et al., 2016)

MRI plays an invaluable role in differentiating between OPN and optic neuritis. Typical MRI features of OPN are the “doughnut sign” on coronal views and the “tram track sign” of the optic nerve sheath on axial views. (Purvin et al., 2001) These features are seen in MRI with fat suppression but not in computed topography as occur in our patient. Hence, in case of atypical optic neuritis, MRI is especially useful as a diagnostic aid to give a final diagnosis.

Treatment of OPN consists of high dose steroid followed by its slow tapering over long-term period. (Tatsugawa et al. 2010) In most patients it usually gives good recovery to patient visual acuity, which was demonstrated in our patient. This treatment regime can prevent the relapse of the disease, and avoid further insult to the optic nerve. (Tatsugawa et al. 2010) However, there has been reported poor response to steroid treatment, these are usually due to treatment delay. (Purvin et al., 2001)

Prolonged systemic steroid leads to multiple side effects including weight gain, reduction in bone density, prone to infection obesity and so on. (Liu et al., 2013)As to prevent recurrence of the disease, patient needs to be compliance to the long-term steroid treatment, so proper counselling and monitor has to be done to avoid treatment complication.

4.0 Conclusion

In summary, clinically it is difficult to differentiate OPN from optic neuritis. MRI can be used as a vital diagnostic tool to achieve it. An accurate diagnosis of OPN is needed and a longer steroid treatment has to be commenced to prevent relapse of the disease and further damage of optic nerve.

Acknowledgement

Ophthalmologists and staffs of Ophthalmology Department, Hospital Universiti Sains Malaysia, Kubang Kerian, Kelantan

Declaration

The authors declare no conflict of interests. The authors are responsible for the content and writing of the paper.

Authors contribution

Author 1: Wrote the manuscript

Author 2: Provided critical feedback and supervised the whole process

References

- Bergman, O., Andersson, T., & Zetterberg, M. (2017). Optic perineuritis: a retrospective case series. *Int Med Case Rep J*, 10, 181-188. doi: 10.2147/imcrj.s125972
- Liu, D., Ahmet, A., Ward, L., Krishnamoorthy, P., Mandelcorn, E. D., Leigh, R. Kim, H. (2013). A practical guide to the monitoring and management of the complications of systemic corticosteroid therapy. *Allergy Asthma Clin Immunol*, 9(1), 30. doi: 10.1186/1710-1492-9-30
- Margo, C. E., Levy, M. H., & Beck, R. W. (1989). Bilateral idiopathic inflammation of the optic nerve sheaths. Light and electron microscopic findings. *Ophthalmology*, 96(2), 200-206. doi: 10.1016/s0161-6420(89)32913-7
- Purvin, V., Kawasaki, A., & Jacobson, D. M. (2001). Optic Perineuritis: Clinical and Radiographic Features. *Archives of Ophthalmology*, 119(9), 1299-1306. doi: 10.1001/archophth.119.9.1299
- Raghibi, A., Wan Hitam, W. H., Noor, R. A. M., & Embong, Z. (2012). Optic perineuritis secondary to tuberculosis: A rare case presentation. *Asian Pacific Journal of Tropical Biomedicine*, 2(2, Supplement), S1206-S1208. doi: [https://doi.org/10.1016/S2221-1691\(12\)60385-2](https://doi.org/10.1016/S2221-1691(12)60385-2)

- Tatsugawa, M., Noma, H., Mimura, T., & Funatsu, H. (2010). High-dose steroid therapy for idiopathic optic perineuritis: a case series. *J Med Case Rep*, 4, 404. doi: 10.1186/1752-1947-4-404
- Tevaraj, J. M. P., Tai Li Min, E., Mohd-Noor, R.-A., Thavaratnam, L. K., Salmah, W. M., & Wan Hitam, W. H. (2016). Atypical Presentation of Idiopathic Bilateral Optic Perineuritis in a Young Patient. *Case Rep Ophthalmol Med*, 2016, 6741925-6741925. doi: 10.1155/2016/6741925
- Yu-Wai-Man, P., Crompton, D. E., Graham, J. Y., Black, F. M., & Dayan, M. R. (2007). Optic perineuritis as a rare initial presentation of sarcoidosis. *Clin Exp Ophthalmol*, 35(7), 682-684. doi: 10.1111/j.1442-9071.2007.01539.x