

Electrophysiological abnormalities associated with extensive myelinated retinal nerve fibers

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An observational case report of electrophysiological abnormalities in a patient with anisomyopic amblyopia as a result of unilateral extensive myelinated retinal nerve fibers (MNFs) is illustrated. The electrophysiological readings revealed an abnormal pattern electroretinogram (PERG) but normal full-field electroretinogram readings in the affected eye. The visual-evoked potential was also undetectable in that eye. Our findings suggest that extensive MNFs can be associated with electrophysiological abnormalities, in particular the PERG, which can aid in diagnosing the cause of impaired vision when associated with amblyopia.

Key words: Amblyopia, electrophysiology, myelinated retinal nerve fibers

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Myelinated retinal nerve fibers (MNF) are congenital anomalies that appear at the level of the retinal nerve fiber layer as whitish patches with feathery edges. They occur when myelination of the optic nerve continues past the lamina cribosa. It has an estimated prevalence of approximately 1% and is most commonly an incidental finding during ophthalmic examination.^[1]

Extensive MNF have been reported to be associated with many ocular conditions, including asymmetric myopia, amblyopia and strabismus.^[1,2] In a case series report by Straatsma *et al.*, a classic description of MNF was described in 32 patients with corresponding histopathological findings of a thickened retinal nerve fiber layer associated with myelination and a collection of dark cells with features similar to that of oligodendrocytes.^[3] Based on this evidence, it has been subsequently suggested that MNF may be considered an oligodendrocytic choristoma, of which pathology lies in the inner retina.^[4]

Electrophysiology tests in three patients with unilateral extensive MNFs showed a normal electroretinogram (ERG) in both the eyes. However, the pattern electroretinogram (PERG) of the affected eyes showed both a decrease in amplitude and an increase in latency of the P50 and N95 waves.^[5] This could be due to either a functional disturbance of the central retina or the blockage of stimulus from the MNF. However, in theory, these changes seen on PERG can be expected to be a direct result of MNF rather than as a result of a secondary effect as they involve the inner retina. We herein report a case of electrophysiological findings in a patient with diffuse MNFs and high myopia associated with amblyopia.

Case Report

A 37-year-old Chinese male presented to the Department of Ophthalmology and Visual Sciences, Alexandra Hospital, Singapore, with a history of poor vision in the left eye since childhood. Visual acuity was light perception in the left eye and 20/20 in the right eye. There was also a relative afferent pupillary defect in the left eye. Fundus examination in the left eye revealed MNF extending from the optic disc up to the equator [Fig. 1]. Examination of the right eye was otherwise normal. There was also a manifest left esotropia of 30 prism diopters.

Manifest refraction was performed with a best-corrected visual acuity (BCVA) of 20/20 in the right eye and of light perception in the left eye, with a spherical equivalent of -3.50 diopter sphere (DS) and of -12.50 DS, respectively.

To further investigate the cause of poor visual acuity, the patient was subjected to electrophysiology testing, which included a pattern PERG, pattern visual-evoked potential (VEP)

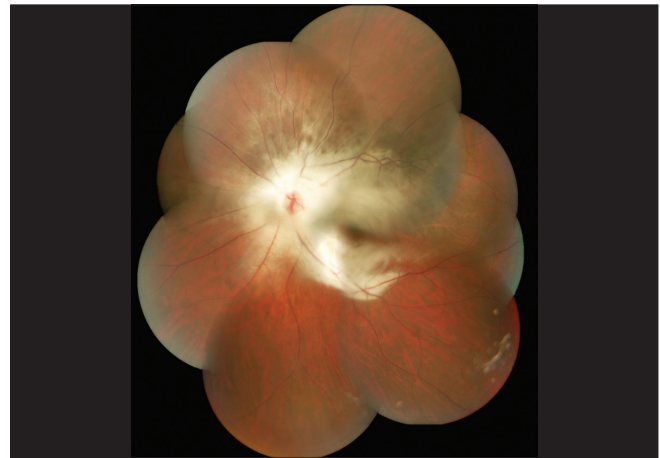


Figure 1: Montage fundal photograph of the left eye showing extensive retinal myelination

and a full-field ERG, all conducted under the International Society for Clinical Electrophysiology of Vision (ISCEV) protocol.^[6-8]

In the PERG findings, P50 and N95 amplitudes were normal in the right eye (P50 = 6.2, N95 = 9.5), suggesting normal macula and retinal ganglion cell function, but undetectable in the left eye (no observable response). Pattern reversal VEP was also undetected in the left eye (no observable response). These findings are suggestive of poor visual function in the left eye.

In the full-field ERG findings, the scotopic b-wave (rod response, a result of the depolarization of the on-bipolar neurons)^[9] was within normal limits in amplitude, although higher in the right eye compared with the left. The maximal and photopic responses were also within normal limits for both eyes [Table 1]. These findings are suggestive of a normal photoreceptor function in both the eyes.

Discussion

The PERG has been shown to be important in clinical and research applications, as it is useful in providing information about retinal ganglion cell function, including macular function. Therefore, by using a similar stimulus to that for the VEP, the PERG can aid in its interpretation when the VEP is abnormal.^[6]

In this case, an undetectable PERG in the left eye is consistent with abnormal ganglion cell function, which correlates with amblyopia in that eye. However, in addition to studies that suggest that MNF may be considered an oligodendrocytic choristoma, further studies have also shown the disappearance of MNF in patients who had later developed branch retinal artery occlusion due to ischemic atrophy of the ganglion cell and nerve fiber layer.^[10] There has also been an implication of the loss of ganglion cells post-central retinal artery occlusion (CRAO), where the photopic negative response in cases of CRAO have been shown to be severely affected despite preservation of the cone b-wave.^[11] This makes it possible, although not easy to establish due to the lack of previous ophthalmic records, that the abnormal retinal ganglion cell function in this patient may be directly associated with the extensive MNF in the retinal nerve fiber layer rather than a

Table 1: International Society for Clinical Electrophysiology of Vision standard full-field ERG findings

	Right eye	Left eye	Normal limit
Scotopic			
b-Amplitude	230.4	161.0	127.9
b-Implicit	107	103	94
Maximal			
a-Amplitude	233.7	211.4	162.2
a-Implicit	12	14	15
b-Amplitude	287.0	255.6	204.8
b-Implicit	48	51	49
Photopic			
a-Amplitude	29	19.3	15.2
a-Implicit	16	18	16
b-Amplitude	94.0	70.5	62.2
b-Implicit	32	34	33
30 Hz flicker			
b-Amplitude	89.1	50.4	60.5
b-Implicit	27	31	33

result of the secondary amblyopia that had developed as a result of the extensive myelination. This suggestion may be reinforced from findings in a recent case report where two out of the three patients studied were not only amblyopic but also demonstrated consistent changes in the PERG.^[5] Compared with our patient, the P50 and N95 waves in the PERG were recordable but decreased amplitude and increased in latency although the MNFs described were only mainly peri-papillary and not extensive.

In conclusion, changes in the PERG may be a direct result of MNFs. Therefore, electrophysiological testing can play an important role in the assessment of visual function in a patient

with poor visual acuity associated with extensive myelination, and this diagnostic modality may be able to provide a guide to visual prognosis in such cases.

References

1. Tarabishy A, Alexandrou T, Traboulsi E. Syndrome of Myelinated Retinal Nerve Fibers, Myopia, and Amblyopia: A Review. *Surv Ophthalmol* 2007;52:588-96.
2. Straatsma BR, Heckenlively JR, Foos RY, Shahinian JK. Myelinated retinal nerve fibers associated with ipsilateral myopia, amblyopia and strabismus. *Am J Ophthalmol* 1979;88:506-10.
3. Straatsma BR, Foos BY, Heckenlively JR, Taylor GN. Myelinated retinal nerve fibers. *Am J Ophthalmol* 1981;91:25-38.
4. Rosen B, Barry C, Constable JJ. Progression of myelinated retinal nerve fibers. *Am J Ophthalmol* 1999;127:471-3.
5. Arda H, Oner A, Gumus K, Karakucuk S, Mirza E. The results of electrophysiological tests in unilateral extensive peri-papillary myelinated nerve fibers: A report of 3 cases. *Doc Ophthalmol* 2009;119:225-8.
6. Holder GE, Brigell MG, Hawlina M, Meigen T, Vaegan, Bach M. International Society for Clinical Electrophysiology of Vision. ISCEV standard for clinical pattern electroretinography -2007 update. *Doc Ophthalmol* 2007;114:111-6.
7. Marmor MF, Fulton AB, Holder GE, Miyake Y, Brigell M. International Society for Clinical Electrophysiology of Vision. ISCEV Standard for full-field clinical electroretinography -2008 update. *Doc Ophthalmol* 2009;118:69-77.
8. Odom JV, Bach M, Brigell M, Holder GE, McCulloch DL, Tormene AP, *et al.* ISCEV standard for clinical visual evoked potentials -2009 update. *Doc Ophthalmol* 2010;120:111-9.
9. Stockton RA, Slaughter MM. B-Wave of the Electroretinogram- A reflection of ON bipolar cell activity. *J Gen Physiol* 1989;93:101-22.
10. Teich SA. Disappearance of myelinated retinal nerve fibers after a branch retinal artery occlusion. *Am J Ophthalmol* 1987;103:835-7.
11. Machida S, Gotoh Y, Tanaka M, Tazawa Y. Predominant loss of the photopic negative response in the central retinal artery occlusion. *Am J Ophthalmol* 2004;137:938-40.