

## Association of extensive myelinated nerve fibers and high degree myopia: Case report

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Unilateral extensive myelination of the peripapillary nerve fibers may be associated with anisometropic myopia, strabismus, and reduced vision. Myelination of optic nerve fibers terminate at lamina cribrosa. Yet in some patients, myelination progresses into the peripapillary retinal nerve fibers and may affect the visual acuity. In this report, we described 4 patients. All patients presented extensive peripapillary myelinated nerve fibers associated with myopic anisometropia. After routine ophthalmic and orthoptic examinations, all patients underwent treatment for amblyopia through correction with spectacles, contact lenses, and the occlusion of the good eye. Corrected visual acuity improved in 1 patient, but 3 patients had no increase in visual acuity despite treatment with full cycloplegic refraction and appropriate patching. Probably because of structural abnormalities of the macula, visual results are often disappointing with appropriate correction of the refractive error and occlusion.

**Key words:** High myopia, myelination, reduced vision

Myelination of the optic nerve begins at the 32<sup>nd</sup> gestational week from the lateral geniculate nucleus and completes at term.<sup>[1]</sup> However, the termination mechanism has not been sufficiently explained. Peripapillary myelination can be congenital or acquired. Congenital retinal myelination is a developmental anomaly and is seen in 0.3-0.6% of population. Ipsilateral high myopia and amblyopia may sometimes accompany myelinated retinal nerve fibers.<sup>[2]</sup> In our report, clinical properties of 4 patients with ipsilateral myopia, amblyopia, and strabismus with accompanying myelinated retinal nerve fibers were discussed.

### Case Reports

#### Case 1

A 9-year-old girl was presented with strabismus. Best corrected visual acuity (BCVA) was 20/300 and 20/20 in the right and left eyes. Cycloplegic autorefractometric

measures revealed refractive error of  $-16.00$  ( $-2.25 \alpha 175^\circ$ ) dioptry (D) and  $+1.50$  ( $-0.75 \alpha 5^\circ$ ) D in the right and left eyes. Biomicroscopic examination was normal, dilated fundus examination was unremarkable in the left eye. Extensive peripapillary myelinated retinal nerve fibers were present in the right eye [Fig. 1]. Cover-uncover test revealed constant 40-prism dioptry (PD) right esotropia. A-scan ultrasound biometry showed an axial length of 24.84 mm and 21.03 mm in the right and left eyes. Contact lenses were prescribed, and occlusion therapy 4 hr/day was initiated. The visits were scheduled for 3<sup>rd</sup>, 6<sup>th</sup>, and 9<sup>th</sup> months for monitoring the visual acuity. During the 3 visits, no increase in right BCVA could be achieved [Fig. 1].

#### Case 2

A 5-year-old girl was referred to our clinic for strabismus and low vision. BCVA was 20/200 and 20/20 in the right and left eyes. Cycloplegic autorefractometric measures revealed refractive error of  $-18.00$  ( $-1.25 \alpha 165^\circ$ ) D,  $-0.25$  ( $-0.50 \alpha 15^\circ$ ) D in the right and left eyes. Cover-uncover test revealed 10 PD right esotropia. Anterior segment examination was normal. In fundus examination, extensive myelinated nerve fibers were detected in the right eye, left eye was normal. The patient was prescribed  $-15.50$  ( $-1.00 \alpha 165^\circ$ ) D and  $-0.50$  ( $-0.50 \alpha 15^\circ$ ) D spectacles, and 4 hr/day occlusion therapy was initiated. There was no increase in BCVA during the 3<sup>rd</sup>, 6<sup>th</sup>, and 9<sup>th</sup> months' visits.

#### Case 3

A 4-year-old girl was brought to the clinic for preschool screening. BCVA was 20/160 and 20/25 in the right and left eyes. Sciascopic measures with cycloplegia were  $-4.00/-4.00$  D and  $+2.00/+2.00$  D in the right and left eyes. Biomicroscopic examination was normal. No manifest squint was detected. Extensive myelinated nerve fibers were detected in the right eye. Left eye was normal. The patient was prescribed with  $-3.00$  D and vertical plane spectacles and 4 hr/day occlusion. During the 3<sup>rd</sup>, 6<sup>th</sup>, and 9<sup>th</sup> months follow-up, minimal increase in visual acuity was recorded. The BCVA was 20/125. She is still in follow-up.

#### Case 4

A 32-year-old woman referred to our clinic for low vision in the left eye. Her BCVA levels were 20/20 and 20/200 in the

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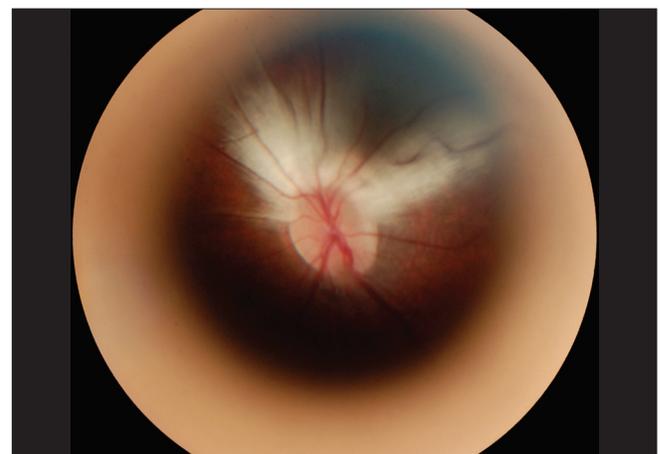


Figure 1: Myelinization of nerve fibers on retina of the right eye (Case 1)

right and left eyes. Cycloplegic autorefractometric measures revealed refractive error of +1.00 (+0.25  $\alpha$  105°) D and -6.75 D in the right and left eyes. Biomicroscopic examination was normal. Extensive myelinated nerve fibers were detected in the left eye. Right eye was normal. A-scan ultrasound biometry showed an axial length of 22.92 mm and 25.83 mm in the right and left eyes. -6.00 D contact lens was prescribed for the left eye. No increase in BCVA was recorded in the 3<sup>rd</sup>, 6<sup>th</sup>, and 9<sup>th</sup> months visits.

## Discussion

Myelination of optic nerve fibers terminate at lamina cribrosa. The control mechanism acting here has not been completely explained. However, there are several theories about this. The hypothesis, that is accepted at most is, lamina cribrosa acting as a barrier and preventing passage of oligodendrocytes to the retina. Second hypothesis is local blood-brain barrier controlling passage of various substances. The third hypothesis is the termination of retinal ganglion cells in lamina cribrosa and the proteoglycan-controlled continuation of only the axonal parts. By this way, myelination is seen around the axons and cannot pass lamina cribrosa.<sup>[3,4]</sup>

Some animals in nature, especially the rabbits, do not have lamina cribrosa and cannot show barrier function. For this reason, the retinal nerve fibers of rabbits are myelinated.<sup>[5]</sup>

Myelinated retinal nerve fibers do not always cause visual problems and can be diagnosed by chance. But, it may be associated with high axial myopia, strabismus, and amblyopia.<sup>[4,6,7]</sup>

Ellis<sup>[6]</sup> found that 83% of patients with myelinated retinal nerve fibers had myopia greater than 6 diopters. Then, why do myelinated fibers cause myopia? It remains unknown whether myelination of retinal nerve fibers is the reason for or the result of myopia. It is possible that myelinated fibers may blur retinal images and induce visual deprivation. Such deprivation at a critical stage of ocular development may contribute to myopia by including an axial enlargement. On the other hand, it is also possible that axial elongation predisposes to retinal nerve fiber myelination. Straatsma<sup>[4]</sup> found that 10% of patients with myelinated nerve fibers have myopia, amblyopia, and strabismus.

Ellis<sup>[6]</sup> suggested that low vision in these patients has an organic etiology in addition to functional amblyopia. He also suggested that myelination around the macula to be the most likely cause of poor vision and proposed that glial cells or myelin impede transmission of light through the retina or impulses from the retina to the lateral geniculate body.

If this has occurred during eye development, amblyopia is inevitable. In infancy and early childhood, anisometropia and amblyopia cause esotropia rather than exotropia.<sup>[8]</sup> We think that myelination of our cases has occurred in infancy period, and squinting was due to the deep anisometropia and amblyopia. Except for one case, deep amblyopia has been found to be resistant to therapy in our patients. In two cases with strabismus, there was no improvement in vision. Despite no manifest squint, visual acuity did not also improve in 32-year-old woman. It may probably due to the age factor. The most important reason for resistant amblyopia was high anisometropia, strabismus, and the myelinated fibers affecting macula that cause deprivation. It remains unclear which one plays the major role and which are complementary. Although there were no improvement in vision in 3 cases, Summers<sup>[8]</sup> reviewed good visual results; therefore, the first step in treatment of amblyopia in these patients must be full optical correction based on a cycloplegic refraction.

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