

Unilateral Myelinated Retinal Nerve Fibers Associated with Ipsilateral Myopia, Amblyopia and Exotropia

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DOI: [10.36347/sjmcr.2024.v12i01.013](https://doi.org/10.36347/sjmcr.2024.v12i01.013)

| Received: 07.12.2023 | Accepted: 10.01.2024 | Published: 12.01.2024

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Abstract

Case Report

This report describes a case of unilateral myelinated retinal nerve fibers associated with ipsilateral myopia, amblyopia and exotropia. A 3-year-old boy presented with the complaint of diminished vision in his right eye. Visual acuity testing with a picture at a distance of 2.5 m revealed 1.5 in the left eye, but right visual acuity was not available. Near visual acuity with dot cards was 0.2 in the right eye and 0.6 in the left eye. The refractive error was -7.25D and +1.50D in the right and left eyes, respectively. Fundus examination revealed extensive myelinated retinal nerve fibers in the right eye, but no abnormal findings in the left eye. The axial length was 24.10 mm and 22.90 mm in the right and left eyes, respectively. Cycloplegic retinoscopy revealed a refractive error of a -7.00D in the right eye and plane in the left eye. Glasses with full optical correction were prescribed, and the patient was evaluated one month later, presenting a best-corrected visual acuity of 0.6 in the right eye and 2.0 in the left eye with a picture at a distance of 2.5 m. Three months later, his visual acuity was 0.7 in the right eye. At this point, amblyopia treatment was started with two hours daily of left eye occlusion. One year following the initial presentation, his visual acuity improved 0.8 in the right eye. The type of fixation, the initial visual acuity, the high myopia and the high anisometropia indicated a most likely bad prognosis. However, the age of diagnosis and wearing status of glasses were advantage for treatment effect in this case. Even though this condition is associated with poor visual prognosis, intensive occlusion therapy should always be attempted, along with the prescription of a cycloplegic refraction, due to unexpected and variable responses to treatment.

Keywords: Myelinated retinal nerve fiber, myopia, amblyopia, strabismus.

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INTRODUCTION

Myelinated retinal nerve fibers (MRNF) are congenital anomalies that appear as grey-white patches with feathery borders at the nerve fiber layer [1]. In a series of 3,968 consecutive autopsy cases, Straatsma *et al.*, [2] reported that MRNF were present in 0.98% of patients and in 0.54% of eyes examined, with bilateral involvement in 7.7% of patients. In a series of 7,856 patients for health screening, Makino [3] described the overall prevalence of MRNF was found to be 0.47% (37/7,856). Patients with MRNF may be completely asymptomatic or may show significant visual defects, especially those with marked axial myopia and amblyopia in the affected eye [1, 4-11]. In addition, patients with MRNF should receive corrective lenses for their myopia and astigmatism, and should undergo aggressive amblyopia therapy. However, good visual acuity can be achieved in some cases.

Herein, we report a case showing unilateral MRNF associated with ipsilateral myopia, amblyopia and exotropia.

CASE REPORT

A 3-year-old boy presented with the complaint of diminished vision in his right eye. There was no significant ocular or medical history and his birth history was normal. The refractive error was -7.25D and +1.50D in the right and left eyes, respectively. Visual acuity testing with a picture at a distance of 2.5 m revealed 1.5 in the left eye, but right visual acuity was not available. Near visual acuity with dot cards was 0.2 in the right eye and 0.6 in the left eye. The patient had right exotropia of 10 prism diopters with full extraocular motility. Slit-lamp examination yielded normal results and the findings of the dilated fundus examination were unremarkable in the left eye, but revealed MRNF in the right eye (Figure 1). MRNF were contiguous with the optic disc and prominently extended along the superotemporal vascular arcade. A-scan ultrasound

biometry showed an axial length of 24.10 mm and 22.90 mm in the right and left eyes, respectively.



Fig 1: Funduscopy image of the right eye

Note the extensive myelinated retinal nerve fibers in the right eye.

The patient was diagnosed with unilateral MRNF associated with axial myopia, amblyopia and exotropia in his right eye. Cycloplegic retinoscopy revealed a refractive error of a $-7.00D$ in the right eye and plane in the left eye. Glasses with full optical correction were prescribed, and the patient was evaluated one month later, presenting a best-corrected visual acuity (BCVA) of 0.6 in the right eye and 2.0 in the left eye with a picture at a distance of 2.5 m. Three months later, his visual acuity was 0.7 in the right eye. At this point, amblyopia treatment was started with two hours daily of left eye occlusion. One year following the initial presentation, his visual acuity improved 0.8 in the right eye.

DISCUSSION

This is a case report of unilateral myelinated retinal nerve fibers associated with ipsilateral myopia, amblyopia and exotropia with an unexpected good outcome. The type of fixation, the initial visual acuity, the high myopia and the high anisometropia indicated a most likely bad prognosis. However, the age of diagnosis and wearing status of glasses were advantage for treatment effect. Generally, it is well known that good visual prognosis in patients with MRNF due to whether macular appearance is normal or abnormal [5-11]. Hittner *et al.*, [5] reported 12 patients with unilateral peripapillary MRNF associated with myopia and/or amblyopia. According to their report, 7 patients had myopia with a mean of $-13.00D$ of anisometropia and abnormal macula. These patients had final visual acuities of 20/200 or less following conventional amblyopia therapy. In contrast, 5 patients had myopia with a mean of $-3.75D$ of anisometropia and normal macula. These patients had final visual acuities of 20/30 or greater with identical therapy. Kee and Hwang [9] studied 5 of 12

children with MRNF and anisometropic amblyopia had an improvement in visual acuity to 20/30 or better. According to their report, significant prognostic indicators were the amount of initial anisometropia, the area of myelination, and the appearance of the macula. Mean anisometropia in patients with a visual acuity of 20/30 or better was $-6.38D$. In patients with poor visual outcome, defined as final visual acuity of 20/ 200 or worse, the mean measured anisometropia was $-11.08D$. In addition, the macula appeared normal in all five patients who exhibited significant recovery. All patients with visual acuity of 20/200 or worse on final visit had an abnormal appearance to the macula. Ellis *et al.*, [6] had no significant improvement in the visual acuities of the six patients treated with occlusion therapy in their series.

Although we should be aware of the limited prognosis in order to set realistic expectations for the recovery of vision, appropriate counseling for patients and their families is important. Even though this condition is associated with poor visual prognosis, intensive occlusion therapy should always be attempted, along with the prescription of a cycloplegic refraction, due to unexpected and variable responses to treatment. Amblyopia treatment is long and requires full cooperation from the patient and the family members. We believe that this case could have presented better outcomes with strict adherence to therapy.

Disclosure: The authors have no conflicts of interest to disclose.

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