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Original Research Article

# Unilateral myelinated retinal nerve fibers associated with ipsilateral myopia and amblyopia

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**Abstract:** The objective is to evaluate the clinical features of unilateral myelinated retinal nerve fibers (MRNF) associated with ipsilateral myopia and amblyopia.We retrospectively analyzed three patients with unilateral MRNF associated with ipsilateral myopia and amblyopia.The ages at presentation range from 4 to 10 years. Refractive error in affected eye ranged from S-6.25DC-2.00D toS-9.75DC-1.25D40.Anisometropia ranged from 6.75D to 10.50D. Best corrected visual acuity in affected eye ranged from 0.04to 0.2. Strabismus was documented in 2 of 3 patients. All patients underwent amblyopic treatment, which included prescription of full cycloplegic refraction and patching of the no amblyopic eye. However, all patients had a uniformly poor visual outcome.We should beware of the limited prognosis in patients with unilateral MRNF associated with ipsilateral myopia and amblyopia.
**Keywords:**myelinated retinal nerve fiber, myopia, amblyopia, anisometropia.

## 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, we described the overall prevalence of MRNF was found to be 0.47% (37/7,856)[3].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, 4-7]. 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 evaluate the clinical features of unilateral MRNF associated with ipsilateral myopia and amblyopia.

## MATERIALS AND METHODS

We retrospectively analyzed three patients with unilateral MRNF associated with ipsilateral myopia and amblyopia.

## RESULTS

All cases are listed in Table 1 and funduscopic images in Figure 1.The ages at presentation range from 4 to 10 years. Refractive error in affected eye ranged from S-6.25DC-2.00D toS-9.75DC-1.25D40.Anisometropia ranged from 6.75D to 10.50D. Best corrected visual acuity in affected eye ranged from 0.04to 0.2. Strabismus was documented in 2 of 3 patients. Axial length in affected eye ranged from 27.21 mm to 27.50 mm.All patients underwent amblyopic treatment, which included prescription of full cycloplegicrefraction and patching of the no amblyopic eye. However, all patients had a uniformly poor visual outcome.

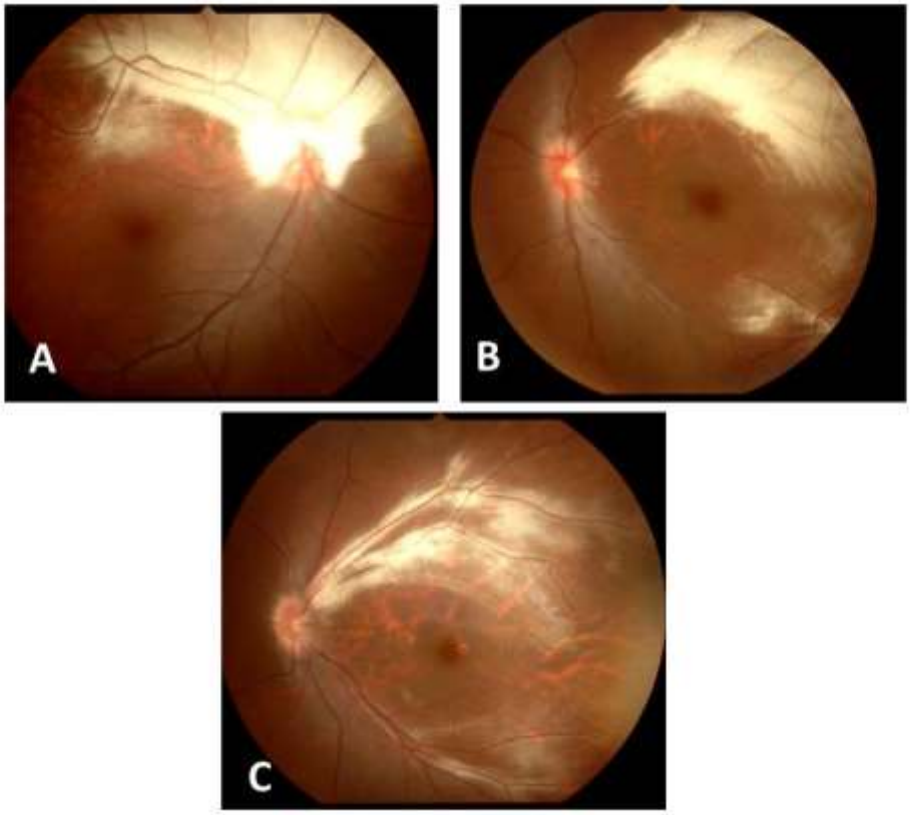


**Fig.1 Funduscopic images of case 1 (A), case 2 (B), and case 3 (C).**

**Table 1: Cases of this study**

| Case | Age Sex | Affected Eye | Refraction | Anisometropia | Axial length (mm) | Strabismus | Initial visual acuity |
|---|---|---|---|---|---|---|---|
| 1 | 4y M | R | R: S-9.50DC-2.00D180 L: 0.00D | 10.50D | R: 27.50 L: 23.50 | exotropia | R:0.04 L: 1.2 |
| 2 | 9y M | L | R: S-0.50D L: S-6.25DC-2.00D15 | 6.75D | N/A | exophoria | R:1.2 L: 0.2 |
| 3 | 10y M | L | R:S-0.50D L: S-9.75DC-1.25D40 | 9.875D | R: 23.30 L: 27.21 | Orthophoria | R:1.2 L: 0.06 |

## DISCUSSION

Generally, it is well known that good visual prognosis in patients with MRNFdue to whether macular appearance is normal or abnormal. Hittner *et al.;*[5]reported 12 patients with unilateral peripapillaryMRNF 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. In a recent study by Kee and Hwang[8], 5 of 12children studied with MRNF and anisometropicamblyopia had an improvement in visual acuity to20/30 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 outcomes, 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.

Recently, Gharai *et al.;*[9] first reported optical coherence tomography (OCT) findings of two patients with extensive peripapillary MRNF. According to their report, decreased retinal thickness is noted in both the

inner and outer rings of the macula in affected eyes in comparison with fellow eyes. In addition, Arda *et al.;*[10] first reported electrophysiological findings of three patients with extensive peripapillary MRNF. According to their report, pattern electroretinogram (PERG) results of affected eyes showed decreased amplitudes and increased latencies of P50 and N95 waves. The P100 amplitudes of the pattern and flash visual evoked response (PVER, FVER) were lower, and the latencies were higher in all affected eyes. Swaminathan *et al.;*[11] first reported multifocal electroretinogram (mfERG) findings with extensive peripapillary MRNF involving the macula. Takahashi *et al.;*[12] reported a case with reduced amplitude at the macular region without macular involvement on fundus photograph. Thus, it is not clear whether the reduced amplitude detected by mfERG occurs irrespective of macular involvement of myelinated retinal nerve fibers. Further studies with additional cases are necessary to answer this question.

Although our findings were based on small cases of unilateral MRNF associated with ipsilateral myopia and amblyopia, we should beware of the limited prognosis in order to set realistic expectations for the recovery of vision. Furthermore, appropriate counseling for patients and their families is important.

## DISCLOSURE

The authors have no conflicts of interest to disclose.

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