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Unilateral Myelinated Retinal Nerve Fibers Associated with Ipsilateral Myopia and Amblyopia in A 2-Year-Old Boy

Medical Science

KEYWORDS : Myelinated retinal nerve fiber, Myopia, Amblyopia, Anisometropia

Shinji Makino

Department of Ophthalmology, Jichi Medical University, Shimotsuke, Tochigi, Japan

ABSTRACT

A 2-year-old boy presented with exotropia and diminished vision in his right eye. His best-corrected visual acuity was 20/1400 and 20/94 in the right and left eyes, respectively. Cycloplegic refraction was S-10.75D C-1.25D100 and S+1.00D C+1.00D80 in the right and left eye, respectively. Fundus examination revealed extensive myelinated retinal nerve fibers in the right eye. In addition, a hypoplastic optic disc was also observed in the right eye. The axial length was 25.22 mm and 21.30 mm in the right and left eyes, respectively. The patient was started amblyopic treatment, which included prescription of full cycloplegic refraction. However, it was difficult for him to wear the refractive corrected spectacle glasses. We should be aware of the limited prognosis and appropriate counseling for patients and their families is important in this rare condition.

INTRODUCTION

Myelinated retinal nerve fibers (MRNF) are congenital anomalies that appear as grey-white patches with feathery borders at the nerve fiber layer.[1] Generally, patients with MRNF may be completely asymptomatic. However, they may show significant visual defects, especially those with marked axial myopia and amblyopia in the affected eye.[1-7] Although patients with MRNF should receive corrective lenses, and should undergo aggressive amblyopia therapy, good visual acuity can be achieved in some limited cases. Herein, we present unilateral MRNF associated with ipsilateral myopia and amblyopia in a 2-year-old boy.

CASE PRESENTATION

A 2-year-old boy presented with exotorpia and diminished vision in his right eye. There was no significant ocular or medical history and his birth history was normal. His best-corrected visual acuity was 20/1400 and 20/94 in the right and left eyes, respectively. Cycloplegic refraction was S-10.75D C-1.25D100 and S+1.00D C+1.00D80 in the right and left eye, respectively. Anisometropia was 12.88D. His eye position was right exotropia. 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 1A and B). The MRNF distributed around contiguous with the optic disc and extending along vascular arcades in the right eye. In addition, a tilted and hypoplastic optic disc was also observed in the right eye. A-scan ultrasound biometry showed an axial length of 25.22 mm and 21.30 mm in the right and left eyes, respectively. The patient was diagnosed with unilateral MRNF associated with axial myopia and amblyopia in his right eye.

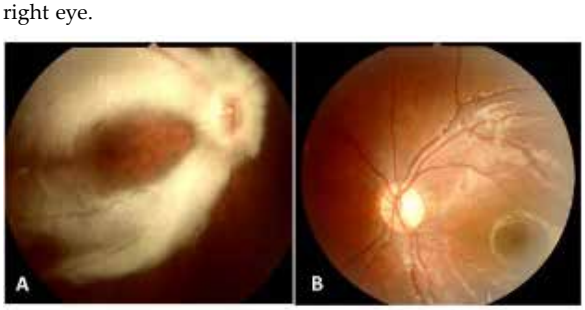

Fig. 1 Funduscopic images of the right (A) and left (B) eyes

Note the extensive MRNF distributed around and contiguous with the hypoplastic optic disc and extending along vascular arcades in the right eye (A). No abnormal findings were detected in the left eye (B).

The patient was started amblyopic treatment, which included prescription of full cycloplegic refraction. However, it was difficult for him to wear the refractive corrected spectacle glasses.

DISCUSSION

Tarabishy et al. [1] summarized 39 patients with MRNF associated with axial myopia and amblyopia described their case series along with other series in the literatures. [3, 5, 6] We selected the patients (aged ≤ 2 years) from their review article. Refractive error in affected eye ranged from S-1.50DC+3.00D to S-17.50D. Anisometropia ranged from 0.75D to 18.50D. Initial visual acuity in affected eye ranged from hand movement to 20/200. Strabismus was documented in 10 of all 13 patients. In this present case, refractive error and anisometropia were relatively large, and initial visual acuity was severely deteriorated.

The first step in treatment of amblyopia in these patients is full optical correction based on a cycloplegic refraction. However, good visual acuity can be achieved in some limited cases. Generally, it is well known that good visual prognosis in patients with MRNF due to whether macular appearance is normal or abnormal. Hittner et al. [3] 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. In a recent study by Kee and Hwang [6], 5 of 12 children studied 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 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. In addition, hypoplastic optic disc in the affected eye was detected in this present case. Visual acuity in eyes with optic disc hypoplasia is variable, and ranges from normal to no light perception. [1] Therefore, we should be aware of the limited prognosis in patients with unilateral MRNF associated with ipsilateral myopia and amblyopia.

CONCLUSIONS

Although our findings were based on single case of unilateral MRNF associated with ipsilateral myopia and amblyopia, we should be aware 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.

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