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Optical coherence tomography and multifocal electro retinogram analysis of unilateral myelinated retinal nerve fibers associated with ipsilateral myopia and amblyopia

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Abstract: This report describes a case of unilateral myelinated retinal nerve fibers associated with ipsilateral myopia and amblyopia. A 9-year-old girl presented with the complaint of diminished vision in her left eye for several years. Her best-corrected visual acuity was 1.2 in the right eye and 0.06 in the left eye. The refractive error was -0.25D and -4.00D in the right and left eyes, respectively. Fundus examination revealed extensive myelinated retinal nerve fibers in the left eye, but no abnormal findings in the right eye. The axial length was 23.30 mm and 27.18 mm in the right and left eyes, respectively. Optical coherence tomography revealed decreased retinal thickness in the left eye. A multifocal electro retinogram showed reduced amplitude in the left eye. Our findings may contribute to a better understanding of this rare condition.
Keywords: myelinated retinal nerve fiber, myopia, amblyopia, optical coherence tomography, multifocal electro retinogram

INTRODUCTION

Myelinated retinal nerve fibers are congenital anomalies that appear as grey-white patches with feathery borders at the nerve fiber layer [1]. They may be located on the disc or elsewhere on the retina. In a series of 3,968 consecutive autopsy cases, Straatsma *et al.;* [2] reported that myelinated retinal nerve fibers were present in 0.98% of patients and in 0.54% of eyes examined, with bilateral involvement in 7.7% of patients. Patients with myelinated retinal nerve fibers may be completely asymptomatic or may show significant visual defects, especially those with marked axial myopia and amblyopia in the affected eye [1, 3-6]. To our knowledge, few reports have investigated the utility of optical coherence tomography (OCT) and multifocal electro retinogram(mf ERG) in patients with this rare condition [7-9].Herein, we report a case showing unilateral myelinated retinal nerve fibers associated with ipsilateral myopia and amblyopia.

CASE REPORT

A 9-year-old girl presented with the complaint of diminished vision in her left eye for several years. There was no significant ocular or medical history and her birth history was normal. Her best-corrected visual acuity was 1.2 and 0.06in the right and left eyes with optical correction of −0.25D and −4.00 D/−1.25 D 40°, respectively. Her eye position was orthophoria. Slit-lamp examination yielded normal results and the findings of the dilated fundus examination were unremarkable in the right eye, but revealed myelinated retinal nerve fibers in the left eye (Figure 1A and B). The myelinated retinal nerve fibers were contiguous with the optic disc and prominently extended along the supero temporal vascular arcade. In addition, a tilted and slightly hypoplastic optic disc was also observed in the left eye.

A-scan ultrasound biometry showed an axial length of 23.30 mm and 27.18mm in the right and left eyes, respectively. The patient was diagnosed with unilateral myelinated retinal nerve fibers associated with axial myopia and amblyopia in her left eye.OCT (RS-3000, NIDEK, Japan) showed normal foveal architecture in both eyes(Figure 2), and a hyper reflectivity corresponding to the myelinated retinal nerve fibers in the left eye(Figure 2B arrows).

Furthermore, OCT clearly demonstrated the difference between the right and left retinal thickness. The thickness of the center (central fovea) was 268 µm and 230 µm in the right and left eye, respectively (Figure 3A and B).In addition, Early Treatment Diabetic Retinopathy Study (ETDRS) sectors map revealed that the thicknesses at the superior, inferior, nasal, and temporal quadrants were 348 µm, 342 µm, 346 µm, and 334 µm, respectively, at the inner ring(1–3 mm from the central fovea) in the right eye; in the left

eye, these values were 207 µm, 252 µm, 257 µm, and 200 µm, respectively (Figure 3A and B). Furthermore, the measurements at the outer ring(3–6 mm from the central fovea) were 304 µm, 291 µm, 314 µm, and 299 µm, respectively, in the right eye; in the left eye, these values were 195 µm, 260 µm, 279 µm, and 177 µm, respectively (Figure 3A and B). The combined retinal nerve fiber layer (RNFL), ganglion cell layer(GCL), and inner plexiform layer(IPL) map revealed that the thicknesses at the super temporal, infer temporal, super nasal, and infer nasal quadrants were 116 µm, 115 µm, 127 µm, and 123 µm, respectively, at the inner ring(1.5–4.5 mm from the central fovea) in the right eye; in the left eye, these values were 44 µm, 65 µm, 69 µm, and 104 µm, respectively (Figure 3C and D). The measurements at the outer ring(4.5–9.0 mm from the central fovea) were 75 µm, 78 µm, 121 µm, and 123 µm, respectively, in the right eye; in the left eye, the respective values were 40 µm, 51 µm, 59 µm, and 58 µm (Figure 3C and D). Mf ERG (LE-4100; TOMEY, Japan) showed reduced amplitude in the left eye (Figure 4).

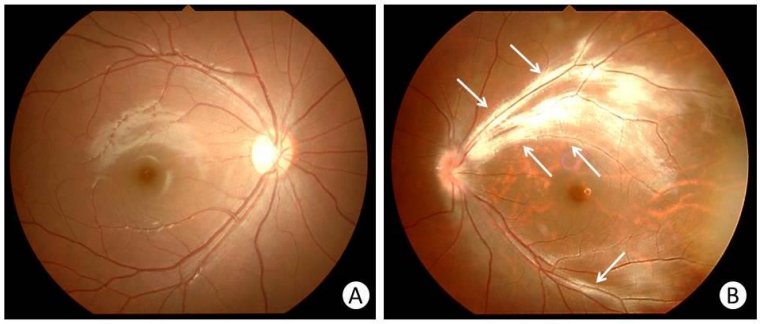

Fig.1: Funduscopic images of the right(A) and left(B) eyes.
No abnormal findings were detected in the right eye (A). Note the extensive myelinated retinal nerve fibers in the left eye (B arrows).

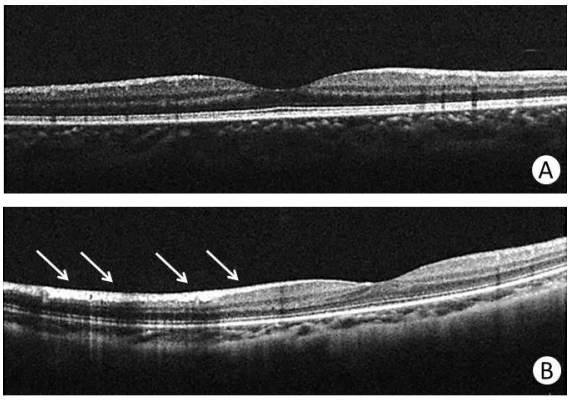

Fig 2: Vertical OCT images of the right (A) and left (B) eyes
No abnormal findings were detected in the right eye (A). Note the hyper reflectivity corresponding to the myelinated retinal nerve fibers in the left eye (B arrows).

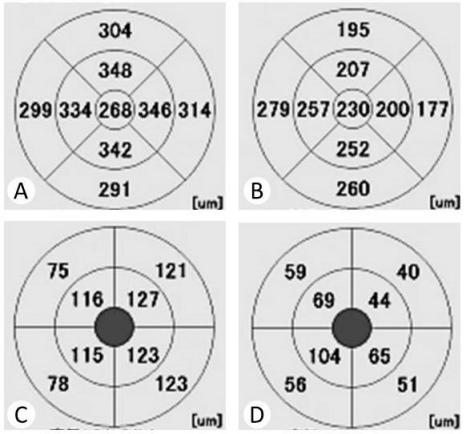

Fig3: Retinal thickness map of the right(A and C) and left(B and D) eyes.
A and B: ETDRS sectors map, C and D: RNFL + GCL + IPL map (1.5/4.5/9.0 mm).

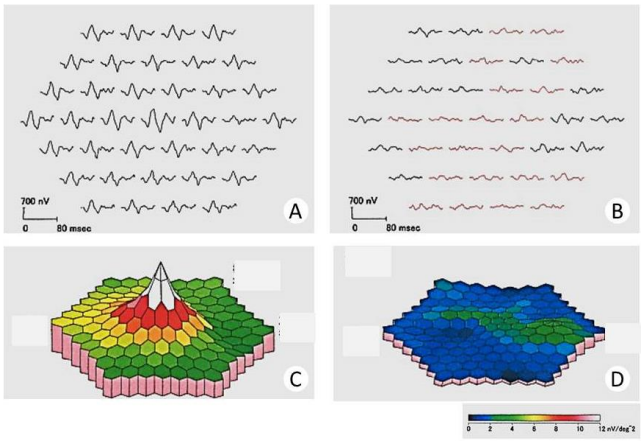

Fig.4: Multifocal electro retinogram response arrays (A and B) and 3-dimentional plots (C and D) of the right (A and C) and left(B and D) eyes.

DISCUSSION

OCT and mf ERG analyses of unilateral myelinated retinal nerve fibers associated with ipsilateral myopia and amblyopia are extremely rare [7-9]. Gharai *et al.;* [8] first reported OCT findings of two patients with extensive peripapillary myelinated retinal nerve fibers. 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 and the myopic control eye. Their results were similar to our present findings.

Lam *et al.;* [10] reported that the central macular thickness is significantly greater and the outer ring macular (3–6 mm) thickness significantly lower in highly myopic eyes than in low to moderately myopic and non-myopic eyes. In our cases, decreased retinal thickness is noted in both inner and outer rings of the macula in affected eyes compared to fellow eyes. Moreover, macular thickness is reduced following the distribution of myelination, i.e., the retinal thickness at the superior rings is lower than those at the inferior rings. However, it is not clear whether the decreased retinal thickness is due to myelinated retinal nerve fibers or myopia. Further studies with additional cases are necessary.

Arda *et al.;* [9] first reported electrophysiological findings of three patients with extensive peripapillary myelinated retinal nerve fibers. According to their report, pattern electro retinogram (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. ERG is known to be a specific test that is used to evaluate the functions of the central retina. PVEP and FVEP are also known to be specific tests used to evaluate the functions of the optic nerve. Arda *et al.;* [9] suggested that the above results may be due to the functional disturbances of the central retina and the optic nerve due to the myelinated retinal nerve fibers. In addition, Swaminathan *et al.;* [7] first reported mf ERG findings with extensive peripapillary myelinated retinal nerve fibers involving the macula. Their result was similar to our present findings. Interestingly, myelinated retinal nerve fibers did not involve the macula in our present case. Thus, it is not clear whether the reduced amplitude detected by mf ERG 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 a single case of unilateral myelinated retinal nerve fibers associated with ipsilateral myopia and amblyopia, they may contribute to a better understanding of this rare condition.

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