Unilateral Extensive Myelinated Retinal Nerve Fiber Layer in a Child

Rafidah Saleh and Maimunah A. Muna'aim

Abstract — We are reporting a case of an incidental finding of an extensive Myelinated Retinal Nerve Fiber Layers (MRNFL) in a healthy 12-year-old Malay boy. The child did not complain of any blurring of vision until he accidentally closed his seeing eye. On examination, the right visual acuity was 6/6 while the left visual acuity was 6/150. There was no Relative Afferent Pupillary Defect (RAPD). The anterior segment was unremarkable. The fundus of the right eye was normal with a pink optic disc with CDR of 0.4, however, the left eye showed extensive MRNFL involving the whole fundus but sparing the macula. The subjective refraction of the right eye was Plano with 6/6 vision, while the left was $-5.50/-1.50 \times 50$ with a vision of 6/150. Bjerrum of the right eye was normal but the left eye showed tunnel field at around 20 degrees with 2mm target size. His Humphrey Visual Field (HVF) 30-2, for the right eye, was normal with MD -0.90 while the left eye showed a generalized reduction of the field with MD of -20.23 . Optical Coherence Tomography (OCT) of the right eye was normal while the affected eye showed thickening of the RNFL at the peripheral. The other layers of the retina and the foveal region of anatomy were somehow preserved. The axial length of the right eye was 24.10mm while the left eye was 28.06mm. MRNFL is a benign condition. It is commonly seen as a streak of whitish patch starting from the optic disc extending to the retina following the arcuate nerve fiber layer pattern, however, extensive myelinated retinal nerve fiber layer involving the whole retina was not very common and usually associated with amblyopia, axial myopia, and squint. Even though mostly benign and solitary, MNFL can be associated with other systemic condition, therefore, clinicians must rule out other systemic diseases.

Index Terms — Amblyopia, Anisometropia, child, myelin sheath, Optical Coherence Tomography.

I. CASE REPORT

We are reporting a case of an incidental finding of an extensive myelinated Retinal Nerve Fiber Layers (RNFL) in a healthy 12-year-old Malay boy. The child did not complain of any blurring of vision until this year, he accidentally closed his seeing eye. The right visual acuity was 6/6 while the left visual acuity was 6/150 or roughly Finger Counting at 1 foot. Both Intraocular pressure (IOP) was normal. The eyes were orthophoric and there was no Relative Afferent Pupillary Defect (RAPD). On examination the anterior segment was unremarkable. The fundus of the right eye was normal with a pink optic disc with CDR of 0.4, however, in the left eye, the Optic Disc was pink but severely obscured by the extensive whitish pale fibers starting from the disc, extending to the peripheral fundus, following the arcuate RNFL pattern but sparing the macula. There were no other abnormalities seen at the retina (Fig.1).

Fig. 1. Colored photo of the right eye showed the fundus of the right eye was normal with a pink optic disc with CDR of 0.4, however, in the left eye, the Optic Disc was pink but severely obscured by the extensive whitish pale fibers starting from the disc, extending to the peripheral fundus, following the arcuate RNFL pattern but sparing the macula.

The subjective refraction of the right eye was Plano with 6/6 vision, while the left was $-5.50/-1.50 \times 50$ with a vision of 6/150. Bjerrum of the right eye was normal but the left eye showed small tunnel field at around 20 degrees with 2mm target size (Fig. 2).

His Humphrey Visual Field (HVF) 30-2, for the right eye, was normal with MD -0.90 while the left eye showed a generalized reduction of the field with MD of -20.23 . Spectral-Domain Optical Coherence Tomography (OCT) of the right eye was normal while the affected eye showed thickening of the RNFL at the peripheral, so thick that it cast a long shadow obscuring the retinal layers below it. The foveal region anatomy was somehow preserved (Fig. 3).

SD-OCT RNFL of both Optic Nerve Head showed normal thickness over the right side while severely thickened over the left side (Fig. 4).

The axial length of the right eye was 24.10mm while the left eye was 28.06mm showed a severely anisometropic eye

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with dense amblyopia.

Fig. 2: The right Bjerrum Chart was normal while the left Bjerrum showed small tunnel vision about 20 degrees central.

Fig. 3. SD-OCT Retina Map OU report showed the right eye with normal retinal anatomy while the left eye showed thickening of the peripheral retina, so thick that it casts a long shadow obscuring the layers below it.

Fig. 4. SD-OCT RNFL of both Optic Nerve Head showed normal thickness over the right side while severely thickened over the left side.

II. DISCUSSION

In the case discussed above, the child was found to have an extensive myelinated RNFL associated with severe anisometropia and dense amblyopia. This condition is congenital and benign. It is found in 0.57 to 1% of the population. [1] On funduscopy, it usually appeared as white patches following the arcuate pattern of RNFL. [2] In terms of patterns, there was a lot of variety in its locations. It can either present in all quadrant of the retina or mainly in the superior and inferior quadrant. The common location was juxtapapillary with the mean largest base diameter was 2.7 mm with the mean of three clock hours in size [3]. Extensive myelinated RNFL involving the whole fundus is not common and usually associated with amblyopia, axial myopia, and squint. On imaging such as red-free, it is seen as white. The whitish opaque appearance is due to its high lipid content inside the myelin sheath. On Optical Coherence Tomography (OCT) the RNFL appears thickened at the periphery. On the Fundus Autofluorescence (FAF) it appears dark and also causes blockage on Fluorescein Angiography. Even though it is usually solitary and non-progressive, it may be associated with a lot of systemic conditions such as optic disc drusen [4], optic nerve hypoplasia [5], keratoconus [6], Leber's hereditary optic neuropathy [7] and optic nerve glioma as well as myopia, anisometropia, and amblyopia syndrome [8]. Therefore, one must think of its associated features before simply discharging the patient. In our patient, the main cause of poor vision was the densely myelinated fibers and the severe anisometropia. If he would have presented earlier, patching therapy with full optical cycloplegic correction using contact lens might be possible since there is a little bit of tunnel vision left in that affected eye, however, the prognosis is still very poor in this type of case in view of the extensive myelination.

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